

SECOND EDITION

PEDIATRIC EMERGENCY MEDICINE



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Second Edition

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CONTENTS

<i>Contributors</i>	xiii
<i>Preface</i>	xxi

Section 1

CARDINAL PRESENTATIONS	1
1 Approach to the Child in the Emergency Department <i>Valerie McDougall Kestner</i>	1
2 The Febrile or Septic Appearing Neonate <i>Robert A. Felter and Ron D. Waldrop</i>	4
3 The Febrile- or Septic-Appearing Infant or Child <i>Ron D. Waldrop and Robert A. Felter</i>	9
4 Respiratory Distress <i>Joanna Cohen and Kathleen M. Brown</i>	13
5 Sudden Infant Death Syndrome and Apparent Life- Threatening Event <i>Collin S. Goto and Sing-Yi Feng</i>	16
6 Altered Mental Status and Coma <i>Susan Fuchs</i>	21
7 Seizures <i>Susan Fuchs</i>	28
8 Chest Pain <i>Wendy C. Matsuno</i>	38
9 Acute Abdominal Conditions <i>Jonathan Singer</i>	42
10 Vomiting, Diarrhea, and Gastroenteritis <i>William R. Ahrens</i>	49
11 Feeding Disorders <i>William R. Ahrens</i>	53
12 Jaundice <i>Anjali Singh and William R. Ahrens</i>	56
13 Crying Infant <i>Joan M. Mavrinac</i>	64
14 Limping Child <i>Isabel A. Barata</i>	68
15 Mild Head Injury in Children <i>Eustacia (Jo) Su</i>	74
16 Approach to the Patient with Rash <i>Daniel McManus and Gregory Garra</i>	79
17 Neck Masses <i>Raemma Paredes Luck and Robert W. Schafermeyer</i>	83
18 Neonatal Emergencies <i>Jenice Forde-Baker</i>	91
19 The Transplant Patient in the Emergency Department <i>Susan M. Scott and Neil Evans</i>	96

Section 2**SEDATION, ANALGESIA, AND IMAGING 99**

- 20 Procedural Sedation and Analgesia *Amy L. Baxter* 99
- 21 Pain Management *Amy L. Baxter* 107
- 22 Imaging *Wendy C. Matsuno* 112

Section 3**RESUSCITATION 119**

- 23 Airway Management *Loren G. Yamamoto* 119
- 24 Respiratory Failure *Lynette L. Young* 127
- 25 Shock *Jonathan Marr* 130
- 26 Cardiopulmonary Resuscitation *Alson Inaba* 136
- 27 Neonatal Resuscitation *Paul J. Eakin* 144

Section 4**TRAUMA 153**

- 28 Evaluation and Management of the Multiple-Trauma Patient *Michael Gerardi* 153
- 29 Head Trauma *Kimberly S. Quayle* 168
- 30 Pediatric Cervical Spine Injury *Julie C. Leonard and Jeffrey R. Leonard* 174
- 31 Thoracic Trauma *Karen O'Connell, Wendy Ann Lucid, and Todd Brian Taylor* 182
- 32 Abdominal Trauma *Shireen M. Atabaki, Wendy Ann Lucid, and Todd Brian Taylor* 193
- 33 Genitourinary Trauma *Joyce C. Arpilleda* 202
- 34 Maxillofacial Trauma *Joanna York and Stephen A. Colucciello* 208
- 35 Orthopedic Injuries *Greg Canty* 213
- 36 Pediatric Sports Injuries in the Ed *Greg Canty* 220
- 37 Injuries of the Upper Extremities *Jim R. Harley* 222
- 38 Injuries of the Pelvis and Lower Extremities *Greg Canty* 233
- 39 Soft Tissue Injury and Wound Repair *D. Matthew Sullivan* 241

Section 5**RESPIRATORY EMERGENCIES 253**

- 40 Upper Airway Emergencies *Richard M. Cantor and Linnea Wittick* 253
- 41 Asthma *Kathleen M. Brown* 257
- 42 Bronchiolitis *Kathleen M. Brown* 269
- 43 Pneumonia *Sharon Mace* 275

44	Pertussis	<i>Sharon Mace</i>	282
45	Bronchopulmonary Dysplasia	<i>Madeline Matar Joseph</i>	287
46	Cystic Fibrosis	<i>Sabah F. Iqbal, Dinesh K. Pillai, Kathleen M. Brown, and Bruce L. Klein</i>	291

Section 6**CARDIOVASCULAR EMERGENCIES** 297

47	Congenital Heart Disease	<i>Timothy Horeczko and Kelly D. Young</i>	297
48	Congestive Heart Failure	<i>Donna M. Moro-Sutherland, William C. Toepper, and Joilo Barbosa</i>	309
49	Inflammatory and Infectious Heart Disease	<i>William T. Tsai</i>	316
50	Dysrhythmias in Children	<i>Ghazala Q. Sharieff</i>	322
51	Pediatric Hypertension	<i>Emily C. MacNeill</i>	328
52	Thromboembolic Disease	<i>Lee S. Benjamin</i>	335

Section 7**NEUROLOGIC EMERGENCIES** 341

53	Syncope	<i>Susan Fuchs</i>	341
54	Ataxia	<i>Susan Fuchs</i>	347
55	Weakness	<i>Susan Fuchs</i>	353
56	Headache	<i>Susan Fuchs</i>	361
57	Hydrocephalus	<i>Susan Fuchs</i>	368
58	Cerebral Palsy	<i>Susan Fuchs</i>	369
59	Cerebrovascular Syndromes	<i>Susan Fuchs</i>	372

Section 8**INFECTIOUS EMERGENCIES** 379

60	Influenza	<i>Karen C. Hayani and Arthur L. Frank</i>	379
61	Meningitis	<i>Steven Lelyveld and Gary R. Strange</i>	388
62	Toxic Shock Syndrome	<i>Eiman Abdulrahman and Shabnam Jain</i>	392
63	Kawasaki Disease	<i>Anthony Cooley and Shabnam Jain</i>	396
64	The Pediatric HIV Patient in the ED	<i>John F. Marcinak</i>	400
65	Tick-Borne Infections	<i>Scott A. Heinrich</i>	407
66	Common Parasitic Infestations	<i>Steven Lelyveld and Gary R. Strange</i>	411
67	Imported Diseases/Diseases in the Traveling Child	<i>Thomas L. Hurt</i>	417
68	Bioterrorism—A Pediatric Perspective	<i>Janet Lin and Timothy B. Erickson</i>	425

Section 9**IMMUNOLOGIC EMERGENCIES** 431

- 69 Common Allergic Presentations: Allergic
Conjunctivitis/Rhinitis *William R. Ahrens* 431
- 70 Anaphylaxis *E. Bradshaw Bunney* 434

Section 10**GASTROINTESTINAL EMERGENCIES** 437

- 71 Abdominal Pain *Philip H. Ewing* 437
- 72 Gastrointestinal Bleeding *Cristina M. Estrada* 440
- 73 Gastroesophageal Reflux *Jamie N. Deis and
Thomas J. Abramo* 446
- 74 Gastrointestinal Foreign Bodies *Philip H. Ewing* 450
- 75 Liver and Gall Bladder Disease *Susan M. Scott
and Ashley Kumar* 452

Section 11**ENDOCRINE EMERGENCIES** 457

- 76 Disorders of Glucose Metabolism *Nicholas Furtado* 457
- 77 Adrenal Insufficiency *Nicholas Furtado* 462
- 78 Hyperthyroidism *Nicholas Furtado* 465
- 79 Rickets *Carla Minutti* 468
- 80 Fluid and Electrolyte Disorders *Susan A. Kecskes* 475
- 81 Inborn Errors of Metabolism *George E. Hoganson* 486

Section 12**GENITOURINARY EMERGENCIES** 493

- 82 Male Genitourinary Problems *John W. Williams* 493
- 83 Urinary Tract Diseases *John W. Williams* 498
- 84 Specific Renal Syndromes *Roger M. Barkin* 500

Section 13**DERMATOLOGIC EMERGENCIES** 507

- 85 Petechiae and Purpura *Malee V. Shah and
Robert A. Wiebe* 507
- 86 Pruritic Rashes *Malee V. Shah and Robert A. Wiebe* 511
- 87 Superficial Skin Infections *Malee V. Shah
and Robert A. Wiebe* 514
- 88 Exanthems *Robert A. Wiebe and Malee V. Shah* 516
- 89 Infant Rashes *Robert A. Wiebe and Malee V. Shah* 523

Section 14**OTOLARYNGOLOGIC EMERGENCIES 527**

90 Ear and Nose Emergencies *Evan J. Weiner* 527

91 Emergencies of the Oral Cavity and Neck
Erica Katz and Gregory Garra 531

Section 15**OPHTHALMOLOGIC EMERGENCIES 539**

92 Eye Trauma *Jeremiah J. Johnson*
and Stephen A. Colucciello 539

93 Eye Emergencies in Childhood *Lauren P. Ortega,*
Katherine M. Konzen, and Ghazala Q. Shariieff 544

Section 16**GYNECOLOGIC EMERGENCIES 553**

94 The Adolescent Pregnant Patient *Pamela J. Okada,*
Adriana M. Rodriguez, and Jeanne S. Sheffield 553

95 Gynecologic Disorders of Infancy, Childhood,
and Adolescence *Maria Stephan* 560

96 Vaginitis *Maria Stephan* 565

97 Sexually Transmitted Diseases *Maria Stephan* 567

98 Dysmenorrhea and Dysfunctional Uterine Bleeding
Pamela J. Okada and Mercedes Uribe 572

Section 17**HEMATOLOGIC AND ONCOLOGIC
EMERGENCIES 581**

99 Anemia *Audra L. McCreight* 581

100 Sickle Cell Disease *Audra L. McCreight*
and Jonathan E. Wickiser 587

101 Bleeding Disorders *Audra L. McCreight and*
Jonathan E. Wickiser 592

102 Blood Component Therapy *Audra L. McCreight*
and Jonathan E. Wickiser 599

103 Oncologic Emergencies *Audra L. McCreight*
and Jonathan E. Wickiser 602

Section 18**NONTRAUMATIC BONE AND JOINT DISORDERS 613**

- 104** Infectious Musculoskeletal Disorders
Kemedy K. McQuillen 613
- 105** Inflammatory Musculoskeletal Disorders
Kemedy K. McQuillen 619
- 106** Nonmalignant Tumors of Bone
Kemedy K. McQuillen 628

Section 19**TOXICOLOGIC EMERGENCIES 635**

- 107** General Approach to the Poisoned Pediatric Patient
Timothy B. Erickson 635
- Part 1 Antipyretic Analgesics** 643
- 108** Acetaminophen *Leon Gussow* 643
- 109** Aspirin *Michele Zell-Kanter* 646
- 110** Nonsteroidal Anti-inflammatory Drugs
Michele Zell-Kanter 649
- Part 2 Household Chemicals** 652
- 111** Toxic Alcohols *Timothy B. Erickson* 652
- 112** Organophosphates and Carbamates *Leon Gussow* 657
- 113** Caustics *Jenny J. Lu and Trevonne M. Thompson* 660
- 114** Hydrocarbons *Trevonne M. Thompson* 662
- 115** Rodenticides *Arthur Kubic, Anthony M. Burda,
and Michael S. Wahl* 664
- Part 3 Prescription Drugs** 671
- 116** Cardiotoxins *Allan R. Mottram and Jerrold Leikin* 671
- 117** Prescription Drugs: Antidepressants
Michael R. Christian 677
- 118** Neuroleptics *Timothy B. Erickson* 681
- 119** Isoniazid Toxicity *Jenny J. Lu and Theodore Toerne* 685
- 120** Carbon Monoxide Poisoning *Sean M. Bryant* 688
- Part 4 Drugs of Abuse** 692
- 121** Opioids *Timothy B. Erickson* 692
- 122** Cocaine Toxicity *Michael R. Christian
and Steven E. Aks* 695
- 123** Phencyclidine & Ketamine *Matthew Valento* 698
- 124** Amphetamines *James Rhee* 700
- 125** Gamma-Hydroxybutyrate *Jenny J. Lu and
Timothy B. Erickson* 702
- 126** Lead Poisoning *Mark B. Mycyk* 705
- 127** Iron *Michael R. Christian and Steven E. Aks* 707

Part 5 Environmental Poisons	710
128 Cyanide Poisoning <i>Mark B. Mycyk</i>	710
129 Mushroom Poisoning <i>Matthew Valento</i>	711
130 Poisonous Plants <i>Ejaaz A. Kalimullah and Andrea G. Carlson</i>	714
131 Lethal Toxins in Small Doses <i>Leon Gussow</i>	721
132 Chemical Weapons: Nerve Agents and Vesicants <i>Leon Gussow</i>	724

Section 20

ENVIRONMENTAL EMERGENCIES

133 Human and Animal Bites <i>David A. Townes</i>	727
134 Snake Envenomations <i>Timothy B. Erickson, Andrew Zinkel, and Valerie Dobiesz</i>	730
135 Spider and Arthropod Bites <i>Timothy B. Erickson, Renee King, and Valerie Dobiesz</i>	736
136 Marine Envenomations <i>Timothy B. Erickson, Armando Márquez, and Valerie Dobiesz</i>	742
137 Drowning <i>Julie Martino and Mark Mackey</i>	747
138 Burns <i>Kavitha P. Reddy</i>	749
139 Electrical and Lightning Injuries <i>Mary Ann Cooper</i>	755
140 Heat and Cold Illness <i>Heather M. Prendergast</i>	762
141 High Altitude Illness <i>Janis P. Tupesis and Ira J. Blumen</i>	767
142 Dysbaric Injuries <i>Ira J. Blumen and Lisa Rapoport</i>	770
143 Radiation Emergencies <i>Ira J. Blumen, Eric Beck, and James Rhee</i>	778

Section 21

PSYCHOSOCIAL EMERGENCIES

144 Sexual Abuse <i>Sara L. Beers and Matthew Cox</i>	789
145 Abuse and Neglect <i>Robert A. Wiebe, Matthew Cox, and Sara L. Beers</i>	793
146 Psychiatric Emergencies <i>Catherine P. Moore</i>	799
147 Death of a Child in the Emergency Department <i>William R. Ahrens</i>	804

Section 22
**EMERGENCY MEDICAL SERVICES
AND MASS CASUALTY INCIDENTS** 809

148 Pediatric Prehospital Care *Craig J. Huang and
Maeve Sheehan* 809

149 Interfacility Transport *Maeve Sheehan and
Craig J. Huang* 812

150 Mass Casualty Management *Janet Lin* 815

Section 23
MEDICOLEGAL AND ADMINISTRATIVE ISSUES 821

151 Medico-Legal Considerations *William R. Ahrens* 821

152 Ethical Considerations *Alan Johnson* 825

153 Withholding or Terminating Resuscitation and
Brain Death *Howard Hast* 832

Index 835

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PREFACE

Pediatric Emergency Medicine: Just the Facts, 2nd Edition, is a distillation of the material provided in *Pediatric Emergency Medicine, 3rd Edition*, which was published in 2009. *Just the Facts* has been developed as a resource for pediatric emergency physicians, general emergency physicians, pediatricians, and others who regularly provide pediatric emergency care or who only occasionally are called upon to care for sick or injured children. Following the same topical organization as the larger text, this work provides the essential information needed in the emergency care of children in a readily accessible manner. In order to do this, we have omitted much information that is of a supporting or enhancing nature. The current work is not intended to be comprehensive but is designed to be a quick reference, an overview for the first-time user, or a refresher course for the experienced clinician. By virtue of its smaller size, the work is more affordable and more portable than its larger companion. We have included new multiple choice questions with each chapter to facilitate the use of the material as a self-test and as a review for those preparing for in-training, board certification, or recertification examinations.

We would like to express our gratitude to the chapter authors and to Anne Sydor and Brian Kearns at McGraw-Hill for their wonderful support and encouragement. Likewise, we would like to express our gratitude to our families, friends, and colleagues who have tolerated our withdrawal from them while we worked to produce this work.

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Section 1

CARDINAL PRESENTATIONS

1 APPROACH TO THE CHILD IN THE EMERGENCY DEPARTMENT

Valerie McDougall Kestner

HIGH-YIELD FACTS

- The game plan for approaching children in the emergency department (ED) is completely different than for the adult. The physician gets one attempt to engage the patient, greet the parent, perform the examination, and formulate a treatment plan.
- The ED itself must be prepared for the pediatric patient. The American Academy of Pediatrics and the American College of Emergency Physicians have established a list of recommended pediatric resuscitation equipment and emergency medications.
- The Pediatric Assessment Triangle (PAT) can establish a quick initial assessment based on appearance, effort of breathing, and circulation to skin.
- Several tools are available to help a provider with weight-based dosing, including the length-based Broselow tape, the PEMSOFIT calculator software package, Pediatric Advanced Life Support (PALS), or regional children's hospital code cards.
- It is helpful to have a clinical pharmacologist or pharmacist present at pediatric codes.

PREPARING FOR THE EXAMINATION

- There is a high element of anxiety for parents and children in the ED.
- Discern whether the child has a chronic medical condition.
- Listen to the parents. Figure out what is the parents' main concern.

- Expect to relay information to multiple concerned family members.
- Make sure the right equipment is available in the room.
- Anticipate the developmental stage of the patient.

DEVELOPMENTAL STAGES AND ACTIVITY LEVEL

- Have a mental image of the child before entering the room.
- A neonate has a vigorously suck, opens eyes briefly, moves all extremities, and exhibits the Moro (startle) reflex.
- A 3-month-old smiles, tracks, and lifts the head.
- A 6-month-old sits up with support, holds the head up, and babbles.
- A 9-month-old explores, pulls to stand, and grabs by raking with the hands.
- A 1-year-old crawls, takes independent steps, may be walking, and says one-two words.
- A 2-year-old has a large vocabulary and puts two words together.
- The child is generally cooperative and can start giving some history at age 3.
- The child's maturity, anxiety, intelligence, and the parental vibe play a large role.

THE HISTORY AND PHYSICAL

- Be flexible when conducting the pediatric examination. Observe the child during play. Let the parent hold the child.
- If the patient has a respiratory or cardiac complaint, examine the child's lungs and heart first before the history.

- Examine the painful part last. Use distraction techniques to help facilitate examination.
- Use Child Life specialists during procedures. Gauge the parents' attitude—will the parent be a help or a hindrance? Let the school-aged and adolescent children talk. Get siblings out of the room if possible.

ASSESSMENT AND PLAN

- The need for blood work is impacted by the child's age, immunization status, and past medical history.
- The "as low as reasonably achievable" (ALARA) radiation concept is important in pediatrics. Judicious use of CT scan is important in pediatrics.
- Create feasible treatment plans for the child and parent. Consider dosing medication as few times per day as possible.
- Visual illustration, written instructions, and verbal review of the plan all increase the chance of compliance.
- Discharge instructions should be clear and written out, including follow-up phone numbers, names of subspecialists, and a time frame for follow-up. Provide specific symptoms to look for as reasons to return to the ED.

SUMMARY

- Pediatric patients in the ED present a wonderful, yet challenging opportunity.
- The physician will face several challenges during history-taking, physical exam, and assessment and plan.
- Enlist the role of the parent, decrease anxiety of all parties, and educate with clear instructions.

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QUESTIONS

1. A 2-month-old girl presents to the ED pale and limp with shallow respirations. Which of the following provides a length-based, color-coded reference during resuscitation?
 - A. PEMSOFTE calculator
 - B. PALS code card
 - C. Broselow tape and carts
 - D. Pediatric Assessment Triangle
 - E. American Academy of Pediatrics website
2. A 3-year-old girl with Syndrome Y presents with her parents to your emergency department with the chief complaint of cough. You remember learning about this syndrome in medical school 5 years ago. The parents say the child once required a bronchoscopy when she had a cough like this. The most appropriate next step in management of this child is:
 - A. Order the cough protocol, which is the standard in your ED
 - B. Conduct a bronchoscopy
 - C. Review her past records online
 - D. Consult a subspecialist to assess the need for a bronchoscopy
 - E. C&D

3. The nurse calls you back into a room as she is going through discharge instructions with a family. During your visit you reassured them that their daughter didn't have asthma, pneumonia, or croup. They are upset that no prescription was given for their daughter's cold. What could you have done to avoid this communication breakdown?
 - A. Talked to the parents and figured out what the parents' main concern was
 - B. Patiently relayed information to the mom in the room and the dad on the phone
 - C. Prescribed a cough medication
 - D. Ordered a chest x-ray
 - E. Checked a pulse oximetry reading
4. The baby you are examining is sitting on his mom's lap with his head tilted to the side, intermittently smiling at you, and watches your penlight move. This child is most likely how old?
 - A. Neonate
 - B. 3 months
 - C. 6 months
 - D. 9 months
 - E. 12 months
5. There is a 4-year-old in the next exam room with his mother. He cut his forehead on the coffee table tonight and requires stitches. The biggest reason to use the papoose board to restrain the child for the procedure is:
 - A. That is what you used for the last 4-year-old who needed stitches
 - B. The child is crying
 - C. The child's mother is crying
 - D. The child edges away and kicks when you approach him to evaluate and clean the wound
 - E. Every 4-year-old who needs stitches requires a papoose board for restraint
6. A 2-year-old boy presents for limp. He is crying for the entire examination. There are no obvious deformities. What is a good rule of thumb for examining this child?
 - A. The child should not play during the exam
 - B. TV should be turned off during the exam
 - C. Have the child lie down on the exam table
 - D. An x-ray is indicated
 - E. Examine the injured or painful leg last
7. A 9-year-old child with autism presents with forehead laceration. He is calm with the examination but asks several questions regarding the procedure. The best strategy to approach his laceration repair:
 - A. Consult child life specialist to provide distraction techniques
 - B. Refer child to operating room for repair under anesthesia
 - C. Give oral midazolam to ease anxiety prior to repair
 - D. Sedate the child with ketamine for procedure
 - E. Consult plastic surgery for repair
8. A 13-year-old female presents with her mother for 1-day history of abdominal pain, vomiting, and fever. The mother is worried that the child may have appendicitis. A mandatory component of the evaluation is:
 - A. A CT scan of the abdomen and pelvis
 - B. Obtaining further history with the mother out of the room
 - C. A pelvic exam in any female adolescent with abdominal pain
 - D. Performing complete blood count and liver function tests
 - E. Referring the child to a pediatric hospital for further evaluation
9. An 18-month-old child presents to the ED after falling off the kitchen counter onto the wood floor. He cried immediately and has taken a bottle since. He has a 3-cm hematoma on the right side of the forehead. The best explanation to provide parents with regarding to obtaining imaging studies is:
 - A. A fall from this height requires a head CT
 - B. This large hematoma on the forehead likely indicates the presence of a skull fracture
 - C. Skull x-rays are often helpful in children with head injuries
 - D. It is important to expose children to radiation "as low as reasonably achievable"
 - E. There is a high likelihood of intracranial hemorrhage

ANSWERS

1. C. The Broselow tape and carts provides a length-based, color-coded standard reference during resuscitation. The Pediatric Assessment Triangle provides a quick framework for initial impression of sick children.
2. E. Listen to the parents. They likely know more about their children's rare conditions than the doctor knows. It is extremely helpful to review electronic past medical history or previous ED encounters prior to formulating a treatment plan. Likewise, a good compromise in this scenario is to acknowledge their feeling that the patient needs a bronchoscopy by consulting a subspecialist instead of proceeding with a test that may or may not be indicated.
3. A. Good communication is a key concept in the pediatric emergency medicine encounter. The practitioner may complete a very thorough history,

physical, and diagnostic plan and still have unhappy parents. One should be sure to find out what the parents' main concern is and address it in real time.

4. B. These are developmental milestones of a 3-month-old. A neonate has poor head control and does not track. A 6-month-old will have good head control and sit with support. By 9 months the child should be pulling to stand, and by 12 months this child may be taking a few steps on his own or even walking.
5. D. A papoose board is very helpful in performing pediatric procedures. If the child is edging away and kicking when he is approached, he will most likely not cooperate for the procedure. An important concept in pediatric emergency medicine is that each age has a wide range of developmental readiness—what works for one 4-year-old, for example, may be totally opposite of what works for the next one.
6. E. Toddlers often cry in the emergency department for several reasons—anxiety, strangers, hunger, and pain are all possibilities. In the toddler, examine the painful part last to help decrease anxiety, build trust, and increase the chance of getting a decent examination. Allowing the child to play during the exam lets the practitioner observe the child's natural movement and state of well-being. Distractions such as the TV can facilitate an accurate examination. One should be flexible and examine the child in a position of his or her choice for parts of the exam; mandating the child to lie on the table may hinder the examination efforts. An x-ray may be indicated depending on examination findings.
7. A. Child life specialists are invaluable for pediatric procedures. While the initial thought in this child with autism may be to sedate him, the fact that he is quite verbal with several questions suggests that he would be very amenable to distraction techniques that child life could provide.
8. B. The fine balance between autonomy and parents as historians is a unique issue in pediatric emergency medicine. Especially in the adolescent with abdominal pain and vomiting obtaining further history with the mother out of the room is important. The adolescent may have other vague complaints, and a good rule of thumb is to interview the patient alone if the story is not adding up. Pelvic exam, CT scan, and blood work can be ordered to confirm or rule out diagnoses as guided by a proper history and physical. Referring the child to a pediatric hospital may be indicated but not as a first step.
9. D. The “as low as reasonably achievable” movement in radiology encourages practitioners to limit

amount of radiation exposure to young children to that which is imperative to diagnosis. This child has a normal neurologic examination and thus a low likelihood of intracranial hemorrhage. This type of injury is quite common in pediatrics and is not usually associated with fracture. Skull x-rays are not widely used, but in the right setting they may be helpful.

2

THE FEBRILE OR SEPTIC APPEARING NEONATE

Robert A. Felter

Ron D. Waldrop

EPIDEMIOLOGY

- Neonates in the first 28 days of life have the greatest risk for serious bacterial infection (SBI).
- Premature infants should have the amount of prematurity subtracted from their chronological age for SBI evaluation considerations.
- There is currently no accepted laboratory evaluation that can separate neonates at low risk for sepsis, although some studies say a child over seven days with a completely normal workup and physical examination may be admitted for observation without antibiotics.
- The most frequent bacterial pathogens are group B *Streptococcus*, *Escherichia coli*, and *Listeria monocytogenes*; infection is usually spread hematogenously.
- 50% of newborns with mothers who have Group B streptococcal infection are colonized but only 1% develops infection.
- *L monocytogenes* infection has up to 45% mortality and that is why ampicillin is used in empiric therapy.
- Both group B streptococcal and *L monocytogenes* infections have distinct early and late presentations.
- *E. coli* sepsis frequently has an associated urinary tract infection.
- Among viruses, herpes simplex and nonpolio enterovirus are the most common causes of serious illness.

FEVER AND SEPSIS

- Neonates with a documented fever or septic appearance should have a sepsis evaluation (Table 2-1) and be admitted to the hospital for 48–72 hours of antibiotic coverage.

TABLE 2-1 Potential Evaluation of the Febrile Neonate

Complete blood count (CBC)
Blood culture
Urinalysis and culture (regard less of U/A results)—catheterized specimen
Lumbar puncture with cell count and differential, culture, protein, glucose
CSF-PCR for herpes and enterovirus
Chest radiograph (when indicated)
Complete metabolic panel (electrolytes, glucose, liver function tests)
Stool culture and leukocyte count (with diarrhea)
C-reactive protein
Procalcitonin

- Fever is a temperature of $\geq 38^{\circ}\text{C}$ or 100.4°F taken rectally.
- Septic neonates may present with hypothermia, which is a rectal temperature less than 36°C or 96.8°F .
- Persistent tachycardia or tachypnea can be a sign of sepsis.
- Irritability, lethargy, or decreased feeding may be the only signs of early sepsis.

EVALUATION

- Initial evaluation of the neonate includes noting the general condition and reviewing vital signs.
- Nonspecific symptoms of sepsis include decreased interest in feeding, poor muscle tone, decreased interactivity, and irritability.
- Even if a specific focus of infection is found (eg, otitis media), the same evaluation and treatment is indicated.
- Skin findings are important in evaluating the neonate. Vesicular rashes suggest herpes infection; purpuric or petechial rashes are ominous; look for areas of cellulitis.
- Skin mottling can indicate more advanced sepsis.

SEPSIS WORKUP

- Sepsis workup includes blood for CBC and blood culture, urine for analysis and culture, and lumbar puncture (Table 2-1).
- CSF PCR for herpes and enterovirus should be ordered with the appropriate clinical presentation.
- In the unstable child, attention to hemodynamic stability is the first priority.
- Always check immediately for hypoglycemia and treat if identified; hypoglycemia may mimic sepsis.
- Chest radiograph is indicated if there are respiratory signs or symptoms.

TABLE 2-2 Antibiotic Therapy for the Febrile- or Septic-Appearing Neonate

	BW 1200–2000 g	BW >2000 g
Infants Younger Than 1 wk (mg/kg)		
Gentamicin IV, IM	2.5 q 12 h	2.5 q 12 h
Cefotaxime IV, IM	50 q 12 h	50 q 8 or 12 h
Ceftriaxone IV, IM	50 q 24 h	50 q 24 h
Ampicillin IV, IM	25–50 q 12 h	25–50 q 8 h
Vancomycin IV	10–15 q 12–18h	10–15 q 8–12 h
Clindamycin IV, IM, PO	5 q 12 h	5 q 8 h
Erythromycin PO	10 q 12 h	10 q 12 h
Infants Aged 7 to 28 d (mg/kg)		
Gentamicin IV, IM	2.5 q 8 or 12 h	2.5 q 8
Cefotaxime IV, IM	50 q 8 h	50 q 8 or 12 h
Ceftriaxone IV, IM	50 q 24 h	50–75 q 24 h
Ampicillin IV, IM	25–50 q 8 h	25–50 q 6 h
Vancomycin IV	10–15 q 8–12 h	10–15 q 6–8 h
Clindamycin IV, IM, PO	5 q 8 h	5–7.5 q 6 h
Erythromycin PO	10 q 8 h	10 q 8 h

TREATMENT

- Ensure hemodynamic stability following the ABCs of resuscitation.
- After sepsis work up is performed, start antibiotics.
- Antibiotic choices are listed in Table 2-2.
- HSV infection should be considered with patients who have had seizures, have blood in the CSF (especially with a mononuclear pleocytosis), afebrile septic appearing infants, those with elevated transaminases, or those with herpetic rashes.
- Enteroviral infection can present almost any way but is considered during the appropriate season and with post or peripartum fever. When suspected, add acyclovir to empiric therapy.
- If the child looks well, is over 7 days of age, and the sepsis workup is completely normal, admission for observation only has been recommended.

SPECIFIC CONDITIONS THAT MAY LOOK LIKE NEONATAL SEPSIS

- Neonates have the highest risk of morbidity and mortality from pertussis.
- Pertussis is difficult to diagnose in the neonate because the catarrhal phase is brief, pleocytosis is often absent, and “whoop” may be absent.
- Patients may present with hypoxia, apnea, poor feeding, seizures, or dehydration.
- PCR of nasopharyngeal specimen is more likely to be positive than cultures.
- Erythromycin must be added to therapy if pertussis is suspected.

- Congenital heart disease may present at any time after birth and should always be considered when faced with the septic appearing neonate.
- There are over 400 known inborn errors of metabolism many of which can present in the neonatal period and mimic sepsis.

OTHER NEONATAL CONCERNS

- Omphalitis is an infected area around the umbilicus in the neonate; because of neonatal inability to contain infection, this infection can extend into the vasculature or abdomen; treatment should be directed against *Staphylococcus aureus*.
- Conjunctivitis may be caused by several different organisms; a large purulent discharge within the first week of life may be due to *Neisseria gonorrhoeae* and must be treated with IV antibiotics.
- Chlamydia infection can present initially with an eye discharge; at this stage, topical erythromycin ointment is adequate treatment.

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QUESTIONS

1. A 21-day-old infant is brought to the ED for poor feeding. The child was a term infant with an

uncomplicated vaginal delivery. At the second week check-up the child was doing well and gaining weight appropriately. The baby is breast- and bottle-fed. For the last 24 hours, the infant has not been interested in feeding. The child has the following vital signs: Temp: 100.0°R; HR 190; RR 35; capillary refill: 2 seconds. The physical examination is unremarkable. The next appropriate step is:

- A. The child has a normal examination and can be sent home
 - B. Immediate sepsis workup, admit, and start antibiotics
 - C. Perform urinalysis and if normal may send home
 - D. Observe vital signs and observe feeding
 - E. Obtain a CBC and blood culture; if WBC is over 15,000 admit for observation.
2. An 18-day-old infant had a rectal temperature at home of 101°F. Mother gave an appropriate dose of Tylenol. She was advised by her pediatrician to bring the child to the ED. In the ED his vital signs are: Temp: 99.9°R; HR 130; RR 32; capillary refill <2 seconds. Physical examination is unremarkable and the infant is active and feeds well. The next appropriate step is:
 - A. Observe the infant for 4 hours. If his fever does not return and he remains asymptomatic he can be discharged and sent to his pediatrician within 24 hours.
 - B. Complete sepsis workup and admit for 48 hours of antibiotic therapy.
 - C. Draw CBC and blood culture and give 50 mg/kg ceftriaxone IM and arrange for follow up in 24 hours.
 - D. Draw CBC and obtain urinalysis. If both are normal, send for cultures and discharge child to follow up with primary care physician.
 - E. Perform complete sepsis workup and if normal admit for observation.
 3. A 5-week-old child comes to the ED for a temperature of 101°F rectally. The child has otherwise been acting normally. The child was born at 36 weeks but went home in 2 days. The child had mild bilirubin elevation but did not require phototherapy. On physical examination, vital signs were normal except for a temperature of 100.5°F. There is a right otitis media but no other findings. What do you do next?
 - A. Perform a complete sepsis work up and admit for IV antibiotics.
 - B. Prescribe amoxicillin (90 mg/kg per day in two equal doses) for 10 days.

- C. Give 50 mg/kg ceftriaxone and arrange follow up in 24 hours.
- D. Obtain a CBC and blood culture; if WBC are elevated admit for IV antibiotics.
- E. Obtain CBC and blood culture, U/A and urine culture and send home on oral amoxicillin (90 mg/kg/day); follow up cultures with primary care physician.
4. A 10-day-old child is brought to the ED because of decreased activity. The child was the product of a term spontaneous vaginal delivery and was doing well until 24 hours ago. Mother said he stopped feeding and has become progressively more lethargic. Initial vital signs are: Temp: 98.0°F rectally; pulse 210; RR 50; capillary refill 4 seconds. Physical examination reveals a lethargic infant with no specific physical findings. The next step in evaluation of this patient is:
- A. Perform a complete sepsis workup and admit for IV antibiotics
- B. Put under warmer and observe in ED
- C. Obtain blood for CBC and culture and start IV antibiotics
- D. Start oxygen by mask, start IV and give bolus of 20 cc/kg of NS, put in warmer
- E. Get a portable chest x-ray
5. A 10-day-old infant is brought to the ED with a history of “not looking right” and an episode when the child “stiffened and eyes seemed to be looking to the right without moving.” The child’s vital signs on admission are: Temp: 98.0°F rectal, HR 190, RR 15, capillary refill 3 seconds. Bedside glucose after the IV is started is 25. The appropriate management for this child is:
- A. Perform a complete sepsis workup and start antibiotic administration as soon as possible.
- B. Perform a complete sepsis work up, give an appropriate dose of D10W and start antibiotics.
- C. Perform a sepsis workup, give D10W and phenytoin.
- D. Establish IV line and immediately give antibiotics.
- E. Establish IV line, give push of D10W, start antibiotics and acyclovir.
6. A 3-week-old child had rhinorrhea that started 3 days ago. One day ago he started to have coughing fits, which usually ended with emesis. He has had decreased feeding for the last 24 hours. The mother brings the child to the ED. On admission, his vital signs are: Temp: 99.4°F rectal; HR 175, RR 50, capillary refill <2 seconds; pulse oximetry 92%. During your examination the baby has a period of apnea, which requires stimulation to correct. Which of the following statements is true about this infant?
- A. CBC will probably show a leukocytosis with a lymphocyte predominance
- B. If the child has pertussis, he will recover more quickly than older children
- C. If the infant has pertussis, the nasopharyngeal culture is the best way to make the diagnosis
- D. Recommended antibiotic therapy includes the addition of erythromycin
- E. Chest radiograph will probably show a lobar infiltrate
7. A 3-week-old child is brought to the ED by parents because they noted redness and swelling around the umbilicus for the last 24 hours. They said it appeared to be painful. On admission the vital signs were: Temp: 99.8°F rectal; HR 150; RR 35; capillary refill <2 seconds. The examination is negative except for an area around the umbilicus about 1 cm, which is red and indurated. The appropriate treatment for this infant is:
- A. Obtain CBC, blood cultures, umbilical cultures and admit and treat with IV vancomycin.
- B. Advise parents about appropriate umbilical hygiene and have follow up with their primary physician the next day.
- C. Start the child on oral clindamycin and arrange for follow up in 24 hours.
- D. Perform a complete sepsis workup and admit for observation if normal.
- E. Advise the parents to put warm soaks on the area every 4 hours, keep the diaper below the umbilicus and return if the redness gets worse.
8. A 6-day-old infant presents to the ED with a rectal temperature of 97.0°F, pulse 190, RR 40, capillary refill 2 seconds. The child was born at term and had an uncomplicated vaginal delivery. The mother was cultured and was positive for group B *Streptococcus* and was treated appropriately. You suspect sepsis. Your initial work up shows a normal CBC and a normal urinalysis. The lumbar puncture was bloody; which of the following is true about this patient?
- A. Because the mother was treated for Group B *Streptococcus*, it is unlikely the infection is due to that organism.
- B. Even though the mother was treated, Group B *Streptococcus* is still the most likely organism.
- C. *Chlamydia trachomatis* is most likely because the child is afebrile.

- D. If the urinalysis is normal there is no UTI.
 E. Because the tap was bloody the cause is most likely herpes simplex.
9. A 2-week-old is brought to the ED by extremely anxious parents because the baby does not want to take the breast. The mother has a history of genital herpes but said there were no obvious lesions prior to birth. She was febrile 24 hours prior to delivery. The patient's vital signs are: Temp: 100.9°F rectally, HR 190, RR 50; capillary refill less than 2 seconds. The examination is normal except for the vital signs. A sepsis workup is performed and is normal with the exception of a WBC count in the CSF of 250 cells, mostly lymphocytes. The mother is very upset and asks what is wrong with the baby and what is going to happen to her baby. A correct response is:
- A. It is probably a viral illness and the baby will do fine.
 B. The baby probably does not have a herpes infection and it is probably not bacterial but the antibiotics will take care of it if it is.
 C. It is serious but because the baby has an early meningitis there should be no major issues after treatment.
 D. The child probably has a herpes infection as well as meningitis and may have some neurological problems.
 E. It is probably not a herpes infection because the child has no rash.
10. A 21-day-old infant presents to the ED in critical condition. He is mottled and lethargic. Vital Signs are temperature 96.8°F rectal, HR 220, RR 50, capillary refill 4 seconds, pulse ox 90%. The first steps in management are:
- A. Establish IV and start cefotaxime and ampicillin
 B. Immediately start assisted ventilation with 100% oxygen
 C. Immediately perform an LP, blood culture, and urine culture and start IV antibiotics
 D. Put under a warmer, start IV, and begin maintenance fluids
 E. Place under warmer, start oxygen by mask, obtain IV access and dextrostick and give 20 cc/kg NS push.
- not feed well, then he should be admitted for observation if all else is normal. A is incorrect because the child has abnormal vital signs and a concerning history. More information is needed. B is wrong. Although the child has a few signs of early sepsis, he is stable enough to observe to determine if the vital signs remain abnormal. C is wrong for a child this age. While, if it is positive, it would lead to a sepsis work up and admission, if it is negative, it could miss other causes of sepsis in a neonate. E is wrong because the CBC is not an adequate screening test to rule out sepsis.
2. B. The infant is under 28 days and has a documented fever. Even though the infant responded to acetaminophen, the complete sepsis workup is indicated followed by admission for IV antibiotics. A is wrong because an infant may only have a fever briefly and still have an SBI. C is wrong because the antibiotic coverage is not appropriate and the patient needs inpatient treatment and observation. D is wrong because both CBC and U/A can be normal in a child with sepsis. E is wrong because with our current state of knowledge, there is no laboratory parameter that can determine which neonates can be observed and which require antibiotics.
3. A. Even though the infant is 5 weeks old, he was 4 weeks premature, so has to be considered to be under 28 days. Also, finding a source in an infant does not change the management of the febrile neonate. B is incorrect; child is clinically under 28 days. C is incorrect; inappropriate antibiotic and requires IV antibiotics. D is incorrect; child is clinically under 28 days. E is incorrect; child is clinically under 28 days.
4. D. The first priority in management of the septic child is establishing hemodynamic stability. A is incorrect; the child is hemodynamically unstable. B is incorrect; the child has no history of cold exposure, so vital signs are more likely due to sepsis. C is incorrect; we must address ABC's first. E is incorrect; the increased RR may be due to pneumonia but the child's overall condition must be addressed first.
5. E. The most critical issue is to correct hypoglycemia. Since this child has had a seizure, in addition to antibiotics, acyclovir is given and herpes simplex must be included in the differential diagnosis. A is incorrect. The child is too unstable to do a complete sepsis work up. The hypoglycemia must be corrected. Antibiotics and acyclovir should be started. B is incorrect. The child is too unstable to perform a complete sepsis work up. In addition to the D10W and antibiotics, acyclovir is indicated.

ANSWERS

1. D. The infant has an elevated heart rate, which can be an early sign of sepsis. If the heart rate remains elevated, a sepsis evaluation is indicated. Also, the infant should be observed feeding; if the child does

C is incorrect. The child is too unstable to perform a complete sepsis workup. The hypoglycemia must be corrected. Anticonvulsants are not indicated unless the child has repeated seizures or is in status epilepticus. Antibiotics and acyclovir should be given as soon as possible. D is incorrect. First priority is to correct hypoglycemia, then give antibiotics and acyclovir.

6. D. Because pertussis is in the differential, erythromycin is indicated. A is incorrect. A leukocytosis is often absent in neonates. B is incorrect. Neonates have the highest risk of morbidity and mortality with pertussis. C is incorrect. Due to the brief catarrhal phase, when the cultures are performed it is often late and they are usually negative. PCR tests are more accurate. E is incorrect. Chest radiographs are variable.
7. A. This is probably omphalitis and the neonate may not be able to contain the infection. B is incorrect. The exam is suggestive of a cellulitis not irritation. C is incorrect. Oral antibiotics are not adequate in the neonate. D is incorrect. The child is neither febrile nor septic appearing. The child needs IV antibiotics against *S aureus*. E is incorrect. Omphalitis must be treated aggressively. A watch and wait is not adequate.
8. B. The most common infection in the newborn period remains group B *Streptococcus*. A is incorrect; even though treatment has reduced the risk of infection with group B *Streptococcus* in the newborn, it has not eliminated it. C is incorrect. *C trachomatis* usually causes ophthalmia neonatorum and sometimes an afebrile pneumonia later in the newborn period. D is incorrect. Neonates can have a normal urinalysis and still have a UTI. E is incorrect. A bloody tap by itself does not indicate herpes simplex.
9. D. This child may have a *Herpes* infection and has a significant risk of mortality and morbidity. A is incorrect. This viral illness may be *Herpes* and the morbidity and mortality is high. B is incorrect. This child may have herpes infection. C is incorrect. Infants with *Herpes* meningitis have 50% mortality and most have neurological sequelae. E is incorrect. Up to 40% of infants with *Herpes* infections have no skin lesions.
10. E. Multiple things need to be done at the same time; as the patient is in shock, 20 cc per kilogram NS push is imperative. A is incorrect; antibiotics are secondary to cardiovascular stabilization. B is incorrect; while the patient has a low pulse oximetry, he is breathing well. He does need oxygen but not assisted ventilation. C is incorrect; the patient

is too unstable to perform a sepsis workup. D is incorrect; the child is in shock and needs aggressive fluid management.

3

THE FEBRILE- OR SEPTIC-APPEARING INFANT OR CHILD

Ron D. Waldrop

Robert A. Felter

INTRODUCTION

- *Fever* is the most common complaint for children presenting to the emergency department (ED), representing more than 20% of all presenting complaints annually.

THERMOREGULATION

- Maintenance of body temperature involves a complex interplay of physiologic heat loss by radiation, evaporation, convection, and conduction; heat gain by metabolic and physiologic activity; and variations in ambient temperature.
- Examples of heat exchange mechanisms include sweating and hyperventilation for losses, shivering, and catabolism for heat gain and swaddling for ambient temperature changes.
- The gold standard for measuring core body temperature in the noncritical care setting is rectally, by either glass mercury thermometer or electronic probe. Other sites, such as axillary, oral, tympanic, or skin, and other devices, such as infrared or liquid crystal, may serve as effective screening tools, but are less sensitive in the young infant and child.

FEVER

- Fever in young infants and children is defined as a body temperature greater than 100.4°F (38°C).
- Fever may cause any child to appear ill due to increased metabolic rate, decreased peripheral vascular resistance, and increased tissue demand leading to tachypnea, tachycardia, diaphoresis, and chills.
- There is no reliable relationship between fever height and the clinical response to antipyretics in predicting serious illness.
- Hyperpyrexia is defined as an extreme body temperature elevation above 106°F (41.1°C). Hyperpyrexia is

associated with a significant increased risk of serious infection, but is also associated with central nervous system abnormalities, medication toxicity, as with psychotropic drugs and neuroleptic malignant syndrome from anesthetics, and heat stroke.

- Hyperpyrexia is associated with an increased incidence of febrile seizures and rhabdomyolysis.
- Significant parental coaching is often warranted to avoid over- or under-evaluation and treatment.

SEPSIS

- Sepsis is more correctly termed systemic immune response syndrome (SIRS) and is defined by temperature instability, age inappropriate tachycardia, or tachypnea, as well as an abnormal leukocyte count. SIRS represents one step in the continuum from infection to occult bacteremia, SIRS, severe sepsis, septic shock, and finally multiorgan dysfunction syndrome (MODS).

BACTEREMIA

- In recent studies, the risk of bacteremia appears to have been modified dramatically by the use of more advanced broad-spectrum antibiotics, advanced diagnostic testing and immunization first against *H influenzae* and subsequently against *S pneumoniae*.
- Unfortunately, new and more resistant bacteria, such as methicillin-resistant *Staphylococcus aureus* (with skin and soft tissue infections) and *Escherichia coli* (with urinary infection) are now the most common causes of bacteremia and are equally capable of being invasive or leading to fulminant infection and sepsis.
- Given the extremely low risk of bacteremia and its sequelae, the best expectant therapy in the well-appearing child is close observation and follow-up pending culture results.

PRESENTATION AND HISTORY

- Careful history is one of the keys to accurate diagnosis in young infants and children. The caretaker's perception of illness is vitally important as a background description of normal and abnormal behavior in the patient. Specific observations include activity level, feeding behavior, and interactive behavior.
- Because of physiologic reserve in young infants and children, symptoms described by the parents may be intermittent with intervening periods of relatively normal behavior.

TABLE 3-1 Febrile Illnesses in Children Younger than 36 Months

Viral	Bacterial
Croup	Pharyngitis (Strep)
Stomatitis	Otitis media
Roseola	Sinusitis
Influenza	Lymphadenitis
Bronchiolitis	UTI
Gastroenteritis	Abscess
Influenza	Consolidated pneumonia
Pharyngitis (viral)	Meningitis
Upper respiratory infection	Osteomyelitis
Interstitial pneumonia	Septic arthritis
Varicella	

- Proper immunization significantly reduces the likelihood of serious bacterial infection (SBI) caused by *H influenzae* and *S pneumoniae*, as well as the classic childhood illnesses of diphtheria, pertussis, polio, and varicella (Table 3-1).

PHYSICAL EXAMINATION

- *The physical examination* is of paramount importance in evaluating a febrile infant or child and remains the most useful tool in determining risk of SBI and the need for antibiotic therapy. Often, the first clues to significant illness are abnormal vital signs. Vital signs should be compared with published age-appropriate norms for heart and respiratory rate.
- In the unstable child, signs of clinical toxicity include altered or decreased mental status; significantly abnormal vital signs; dyspnea; color changes, such as cyanosis and pallor; and hypoxia as measured by pulse oximetry.
- Examination of the clinically stable child should include an assessment of mental status, including activity level and interaction with parents, as well as interaction with the health care provider. In the young infant, poor feeding behavior and poor consolability may be important clues to serious illness.
- Decreased activity or lethargy is always a sign of serious illness.
- Character of the cry may be informative, with a lusty loud cry suggesting a healthy child and a weak, high-pitched cry or absent cry suggesting a child who is seriously ill.
- Normal peripheral perfusion is indicated by capillary refill of less than 3 seconds in a warm extremity. In the presence of dehydration or septic shock, perfusion will be delayed
- Reassessment of perfusion and mental status is mandatory as clinical condition may wax and wane

TABLE 3-2 Pediatric Patients Independently at High Risk for SBI

Unimmunized
Immunocompromised
Acquired or congenital neutropenia
HIV
Debilitated state
Splenuctomy
Sickle cell disease
Congenital heart disease
Lung disease
Bronchpulmonary dysplasia
Cystic fibrosis
Central nervous system abnormality
Trauma
Cerebrospinal fluid shunts

TABLE 3-3 Common Seasonal Viral Illnesses in Infants and Children Causing Fever in the United States

ORGANISM	SYMPTOMS	SEASON
Adenovirus	Pharyngitis, conjunctivitis, croup, bronchiolitis, gastroenteritis	Winter–early spring
Influenza A/B	Croup, bronchiolitis, pneumonia, myalgias	Winter
Parainfluenza type 1–3	Pharyngitis, croup, bronchiolitis	Summer–autumn
Respiratory syncytial virus	URI, croup, bronchiolitis, pneumonia	Fall–winter
Rotavirus	Gastroenteritis	Winter–spring
Rhinovirus	URI, bronchiolitis	Fall–spring

in young infants and children due to cardiovascular reserve, but when the reserve is exhausted, sudden vascular collapse can occur.

- Petechiae and purpura should always alert the practitioner to the possibility of meningococcal disease or sepsis, especially in the ill-appearing child. Petechiae distributed on the face and chest may also suggest intense vasalva maneuvers associated with vomiting or severe cough (Table 3-2).

RISK OF SBI IN PATIENTS WITH VIRAL INFECTIONS

- The risk of SBI in the presence of these identifiable viral infections is significantly less; however, a small but significant group of febrile infants will still have either a concomitant or secondary SBI (Table 3-3).

MANAGEMENT (FIG. 3-1)

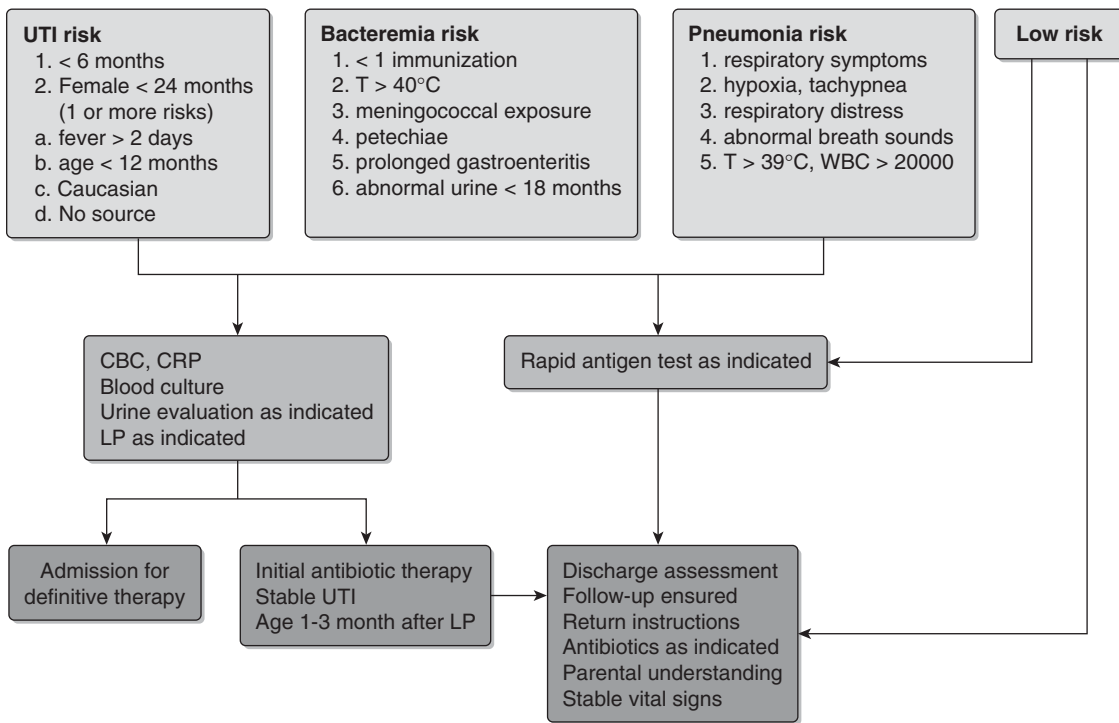


FIG. 3-1. Management of nontoxic febrile child (29 days to 36 months).

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QUESTIONS

1. A 6-month-old is brought to the ED for evaluation of fever. The mother states that the family has just returned from a camping trip where the outdoor temperature was colder than expected. She reports wrapping the child in several blankets. She also admits not taking the infant's temperature, but insists she felt "very warm." A tympanic temperature of 100.2°F is obtained in triage. Which of the following statements is true?
 - A. Shivering is a mechanism for heat loss
 - B. A fever is defined as a temperature of 100.4°F
 - C. Children are not susceptible to ambient temperature changes
 - D. Tympanic thermometers are as accurate as rectal
 - E. Hyperventilation results in an insignificant amount of heat loss.
2. A 1-year-old girl is brought by her parents for evaluation of fevers for 3 days. The parents report high fevers of 104°F at home. The fevers have been transiently responsive to acetaminophen and ibuprofen. As a result, the parents delayed MD evaluation. Which of the following statements is true?
 - A. Children rarely appear ill due to fever alone.
 - B. Higher fever and response to antipyretics predict risk of serious bacterial infection (SBI).
 - C. Hyperpyrexia (>41°C) is not associated with a risk of SBI.
 - D. Fever, inappropriate tachycardia, and tachypnea for age and elevated WBC indicate SIRS (systemic inflammatory response syndrome).
 - E. SIRS includes the entities of sepsis and septic shock.
3. A 4-year-old girl is brought to the ED by her grandmother. As per the grandmother, the child was less playful and had a decreased appetite. The grandmother believes the child has another urinary tract infection. The child appears well hydrated and nontoxic. You suspect a viral illness. Which of the following statements is true?
 - A. Methicillin-resistant Staph aureus (MRSA) and *E coli* associated with UTI are self limited infections.
 - B. Lack of septic appearance in the ED rules out SBI.
 - C. Activity level and feeding behavior are important observations.
 - D. The presence of a usual childhood viral infection rules out SBI.
 - E. Patients with documented viral illness have an inconsequential incidence of concomitant SBI.
4. You are listening to a medical student presentation of a 4-month-old infant boy brought in for a persistent fever by his mother. It appears the mother has been underdosing antipyretics for the infant. The infant has normal vital signs and normal appearance. The medical student concludes that the risk of SBI in this infant is low. Which of the following statements is true?
 - A. The most common cause of SBI today is *Hemophilus influenzae*.
 - B. Workup for SBI includes a thorough skin examination and an appropriately obtained urine specimen for evaluation and culture.
 - C. Once a child becomes ill appearing, they will remain so until treated.
 - D. Stable vital signs rule out the presence of a SBI.
 - E. Physical examination findings are of minimal use in the evaluation of a child with possible SBI.
5. A 3-week-old is brought to the ED for increased irritability and refusing to breastfeed. The infant appears ill. Which of the following would confirm your suspicions?
 - A. Re-establishment of perfusion with IV fluids rules out ongoing shock.
 - B. A weak high-pitched cry may be an indicator of SBI.
 - C. Hypoxia is easily determined clinically.
 - D. Meningococcal vaccine is available for the young child.
 - E. A loud forceful cry may be an indicator of SBI.

ANSWERS

1. B. Fever is defined as a temperature of 100.4°F. Shivering is a mechanism for heat gain not heat loss. Children are susceptible to ambient temperature changes. Tympanic thermometers are not as accurate as rectal.

2. D. Fever, inappropriate tachycardia, and tachypnea for age and elevated WBC indicate SIRS (systemic inflammatory response syndrome). Higher fever and response to antipyretics do not predict risk of serious bacterial infection (SBI). Hyperpyrexia ($>41^{\circ}\text{C}$) is associated with a risk of SBI. SIRS represents one step in the continuum from infection to occult bacteremia, SIRS, severe sepsis and septic shock.
 3. C. Activity level and feeding behavior are important observations. Methicillin resistant *Staph aureus* (MRSA) and *E coli* associated with UTI are not self-limited infections and require antibiotics. The lack of septic appearance in the ED does not rule out SBI. Patients with documented viral illness can have a significant incidence of concomitant SBI.
 4. B. Workup for SBI includes a thorough skin examination and an appropriately obtained urine specimen for evaluation and culture. Stable vital signs do not rule out the presence of a SBI. Physical examination findings are of significant value in the evaluation of a child with possible SBI.
 5. B. A weak high-pitched cry may be an indicator of SBI. Reestablishment of perfusion with IV fluids does not rule out ongoing shock.
- The PNS provides innervation to the muscles of respiration and can be disrupted in diseases of the peripheral motor nerve, neuromuscular junction, or the respiratory muscles.
 - Any process that compromises the delivery of oxygen to the alveoli or blood to the capillaries will cause a ventilation/perfusion (V/Q) mismatch and lead to respiratory distress.
 - Increased metabolic demands or states that affect the blood's ability to deliver oxygen can also produce respiratory distress.

4 RESPIRATORY DISTRESS

Joanna Cohen

Kathleen M. Brown

INTRODUCTION

- Respiratory distress is characterized by increased respiratory effort, rate or work of breathing as manifested by tachypnea, hyperpnea, nasal flaring, and inspiratory retractions.
- Respiratory distress can progress to respiratory failure, the most common precipitating cause of in-hospital cardiopulmonary arrest.

PATHOPHYSIOLOGY

- Control of gas exchange is maintained through the respiratory system, the central and peripheral nervous systems (CNS, PNS), the diaphragm, the chest wall, and the circulatory system.
- CNS control of respiration lies in the respiratory centers of the medulla.

CLINICAL PRESENTATION

- Quiet tachypnea occurs from nonpulmonary diseases such as DKA or shock.
- Tachypnea with grunting, stridor or wheezing represents airway obstruction.
- Seesaw respiration or abdominal breathing occurs with upper airway obstruction.
- Stridor results from extrathoracic airway obstruction.
- Wheezing results from intrathoracic airway obstruction.
- Grunting occurs with pulmonary edema, pneumonia, or atelectasis.
- Decreased breath sounds occur with airflow obstruction, parenchymal lung disease, or poor respiratory effort.
- Slow breathing or irregular respiratory rate occurs with fatigue, CNS depression, or hypothermia.
- A child in respiratory distress whose respiratory rate goes from rapid to normal may be improving; however, if the child's level of consciousness is waning, a decreasing respiratory rate may actually indicate a declining status.

LABORATORY AND RADIOGRAPHIC FINDINGS

- Pulse oximetry measures oxygen tissue saturation.
- End-tidal carbon dioxide monitoring assesses ventilation.
- Capnography shows a graphic display of exhaled carbon dioxide via a probe in the nostril of a spontaneously breathing patient or in line with an endotracheal tube.
- Arterial blood gas (ABG) assesses blood gas exchange in the lungs, the acid-base balance of the body and electrolyte levels.
- High anion gap metabolic acidosis results from diarrheal dehydration, DKA, an inborn error of metabolism, sepsis, or toxin ingestion.

- Normal anion gap metabolic acidosis is associated with hypernatremic dehydration, renal tubular acidosis, or rapid volume expansion.
- Chest radiographs may show a radiopaque foreign body, lobar pneumonia, pneumothorax, atelectasis, hyperinflation, or cardiac enlargement.
- Lateral neck radiographs are used to assess for retropharyngeal abscess or aspirated radiopaque foreign body.

DIFFERENTIAL DIAGNOSIS (TABLES 4-1 AND 4-2)

TABLE 4-1 Major Causes of Respiratory Distress in Newborns

Upper airway	Lower airway
Craniofacial anomalies	Neonatal respiratory distress syndrome
Laryngomalacia	Bronchopulmonary dysplasia
Laryngeal webs	Gastroesophageal reflux
Vascular rings	Meconium aspiration
	Diaphragmatic hernia
Cardiac	Nervous system
Congenital heart disease	Congenital CNS anomalies
	Neonatal apnea
Metabolic	Seizure
Inborn errors of metabolism	

TABLE 4-2 Major Causes of Respiratory Distress in Infants and Children

Upper airway	Lower airway
Craniofacial anomalies	Asthma
Foreign body	Bronchiolitis
Facial or neck trauma	Aspiration of emesis
Tonsillitis/peritonsillar abscess	Foreign body
Ludwig's angina	Near-drowning
Burns	Gastroesophageal reflux
Epiglottitis	Pneumonia
Croup	Cystic fibrosis
Retropharyngeal abscess	Pertussis
Tracheitis	Pulmonary edema
Anaphylaxis	Pulmonary contusion
Angioedema	Pulmonary embolism
	Smoke inhalation
	Pneumothorax/Tension pneumothorax
	Pleural effusion/empyema
	Hemothorax
	Rib fractures/flail chest
Cardiac	Nervous system
Myocarditis	Intoxication
Acute decompensated heart failure	Seizure
	Tetanus
Metabolic	Myelitis
Hyperventilation	Guillain-Barré syndrome
Diabetic ketoacidosis	Botulism
Other causes of acidosis	Tick paralysis

TREATMENT

- Immediate lifesaving interventions may be needed for upper airway obstruction, tension pneumothorax, cardiac tamponade, or respiratory failure.
- Alert child: minimize agitation.
- Obtunded child: relieve soft tissue obstruction with a chin-lift or jaw thrush, suction the mouth and nose, remove visualized foreign body.
- Maintain airway patency: oropharyngeal airway, nasopharyngeal airway, endotracheal tube, and surgical airway.
- Trauma patient: consider blunt or penetrating injury to the trachea and tension pneumothorax.
- Croup: nebulized racemic epinephrine and dexamethasone.
- Anaphylaxis: IM epinephrine, diphenhydramine, H₂-blockers, methylprednisolone, and an isotonic crystalloid fluid bolus.
- Asthma: albuterol and steroids.
- For patients with severe respiratory distress or impending failure: high-concentration humidified oxygen should be delivered to maintain cerebral and myocardial oxygenation.
- Effective bag-valve-mask (BVM) ventilation is still the single most important intervention in managing a patient with respiratory failure.

DISPOSITION/OUTCOME

- Likely discharge: patients with asthma that respond to steroids and inhaled bronchodilators, foreign bodies that have been removed, intoxication with recovery.
- Indications for hospital admission: persistent increased work of breathing, hypoxia, concern for rebound of symptoms
- Indications for ICU admission: endotracheal intubation, severe respiratory distress, and impending respiratory failure.

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QUESTIONS

1. A 2-year-old boy presents with inspiratory stridor and retractions. He has no medical problems and no known allergies. He attends a preschool program and had been previously well. Likely diagnoses include which of the following?
 - A. Foreign body ingestion
 - B. Asthma
 - C. Croup
 - D. A and B
 - E. A and C
2. A 12-year-old female is brought to the ED by her mother for altered mental status. On examination, the child appears lethargic with quiet tachypnea. She has no known medical history. There is no history of trauma. Her parents report an outbreak of a gastrointestinal illness at her school. Which of the following is the most likely diagnosis?
 - A. Asthma
 - B. Croup
 - C. Anaphylaxis
 - D. DKA
 - E. Retropharyngeal Abscess
3. A 6-month-old boy is rescued from a house fire. He is obtunded and tachypneic. His carboxyhemoglobin level at the scene is 25%. Which of the following is correct regarding his condition?
 - A. He is at the same risk of central nervous system disturbances as an adult.
 - B. His V/Q mismatch is secondary to an increased metabolic demand for oxygen.
 - C. He has decreased blood flow to the lungs.
 - D. He should be treated with 100% or greater oxygen delivery.
 - E. He is likely to present with a cherry red skin color.

4. A 7-year-old female ate a pine nut for the first time and developed lip and tongue swelling associated with wheezing and hives. She is treated in the ED with IM Epinephrine, steroids, and antihistamines and is feeling much better one hour after her arrival. Which of the following is correct regarding her condition?
 - A. She can be safely discharged home now that she is feeling better.
 - B. She requires ICU admission immediately.
 - C. She will require an EpiPen with instruction on use upon discharge.
 - D. The placement of an IV can safely be avoided in this patient.
 - E. The presence of hives in this patient is diagnostic for anaphylaxis.

ANSWERS

1. E. Stridor is a high-pitched inspiratory noise that suggests an extrathoracic airway obstruction mechanism, such as foreign body or croup, while wheezing during exhalation suggests an intrathoracic airway obstruction etiology such as asthma.
2. D. Quiet tachypnea is usually an attempt to increase minute ventilation to blow off carbon dioxide from nonpulmonary diseases, such as DKA or shock, whereas, tachypnea with grunting, stridor, or wheezing suggest airway obstruction.
3. D. Anemia and abnormal hemoglobin states, such as methemoglobinemia or carboxyhemoglobinemia, affect the blood's ability to deliver oxygen to the tissues. The ensuing V/Q mismatch leads to respiratory distress. Most patients with carboxyhemoglobinemia will look pale, rather than cherry red. Treatment is with 100% FiO₂ or hyperbaric oxygen.
4. C. Anaphylaxis can be treated with IM epinephrine, diphenhydramine, H₂ blocker, methylprednisolone, and an isotonic crystalloid fluid bolus. Patients can have a recurrence of symptoms with no further exposure to the allergen and should therefore be observed for a period of time prior to discharge. After observation, this patient can be discharged with instructions for use of the EpiPen should another similar event occur.

5 SUDDEN INFANT DEATH SYNDROME AND APPARENT LIFE-THREATENING EVENT

*Collin S. Goto
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SUDDEN INFANT DEATH SYNDROME

DEFINITION OF SUDDEN INFANT DEATH SYNDROME (SIDS)

- The sudden death of an infant younger than 1 year which remains unexplained after a thorough case investigation, including a complete autopsy, examination of the death scene, and review of the clinical history.

EPIDEMIOLOGY AND PATHOPHYSIOLOGY

- SIDS remains the most common cause of death for children aged 1 month to 1 year in developed countries.
- Approximately 2500 infants die from SIDS every year in the United States.
- The peak incidence is between 2 and 4 months of age and 90% of SIDS deaths occur in the first 6 months of life.
- Boys are more likely to die than girls at a ratio of 60:40.
- Younger maternal age, lack of prenatal care, low birth weight, prone sleeping position, overheating, and preterm birth are all risk factors for SIDS (Table 5-1).
- In the United States, African-Americans and Native Americans have SIDS rates that are two to three times the national average irrespective of socioeconomic status.

TABLE 5-1 Factors Associated with Sudden Infant Death Syndrome

MEDICAL/GENETIC	ENVIRONMENTAL/BEHAVIORAL/ SOCIOCULTURAL
Congenital defects	Bed sharing
Low birth weight	Head covering
Preterm infant	Higher ambient temperature
Polymorphisms causing impaired autonomic regulation and arousal	Low socioeconomic status
	Maternal smoking
	Multiple layers of clothing
	Soft sleeping surfaces
	Smoke exposure
	Supine sleep position

- The pathophysiology of SIDS is polygenic and multifactorial with medical, genetic, environmental, and behavioral/sociocultural factors.
- Prone sleeping is associated with SIDS and has been shown to increase the time infants spend in a state of reduced spontaneous arousability possibly due to the trapped carbon dioxide around the infant’s face.
- The SIDS rate in the United States has decreased since the American Academy of Pediatrics first published its recommendation that infants should sleep in a nonprone position in 1992 and started the “back to sleep” campaign in 1994.

EVALUATION AND MANAGEMENT

- Resuscitation efforts should be initiated immediately on the unresponsive infant (Fig. 5-1).
- Past medical history, present illnesses, current medications, and any history of trauma should be ascertained and the child should be thoroughly examined for any congenital abnormalities, signs of concurrent illness, or evidence of physical abuse.
- SIDS is a diagnosis of exclusion.

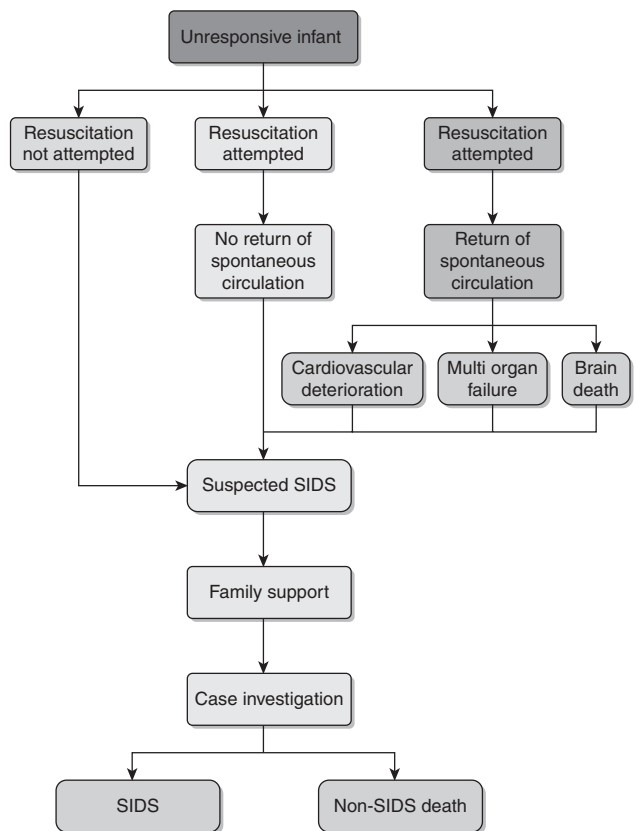


FIG. 5-1. Management of sudden infant death syndrome.

- The differential diagnosis of SIDS includes sepsis, pneumonia, myocarditis, congenital heart defect, cardiomyopathy, arrhythmia, prolonged QT syndrome, accidental or nonaccidental trauma, suffocation, adrenal hyperplasia, and inherited metabolic disorders.
- Autopsies and death scene investigations are warranted to help determine the cause of death and provide valuable information and closure for the family.
- Prevention is the key to reducing mortality secondary to SIDS and risk reduction is the most important measure in preventing SIDS.
- Risk reduction strategies include nonprone sleeping, avoiding maternal smoking in pregnancy, decreasing environmental smoke exposure, maintaining comfortable ambient temperature, providing a safe sleep environment, and fully immunizing the child.

DISPOSITION

- The loss of a child is a devastating event. The parents should be allowed to see and hold the baby and details of the resuscitation should be explained.
- Immediate social work and pastoral support will help the family to cope with the difficult and confusing situation.
- Surviving siblings and other family members need age-appropriate support.

THE APPARENT LIFE-THREATENING EVENT

DEFINITION

- Definition of ALTE: an episode that is frightening to the observer and is characterized by some combination of apnea (central or obstructive), color change (cyanosis, pallor, erythema, or plethora), marked change in muscle tone (rigidity or limpness), or unexplained choking or gagging.
- The majority of patients will appear well and the challenge for the emergency physician is to determine whether a true life-threatening event has occurred.

EPIDEMIOLOGY

- ALTE is estimated to occur in 0.5% to 6% of all children during the first year of life and
- It is more common in boys and premature infants.
- The peak incidence is in infants younger than 2 months.
- The exact relationship between ALTE and SIDS is not clear.

ETIOLOGY

- ALTE is primarily a historic description of the event rather than a single, unifying pathophysiologic process.
- Apnea is a common presentation but is also the final common pathway for many disease processes seen in infants.
- A definitive diagnosis of the ALTE is found in only approximately 50% of patients.

INITIAL ASSESSMENT AND STABILIZATION

- The initial evaluation of the unstable or ill-appearing infant is directed at identifying and stabilizing immediate life-threatening conditions (Fig. 5-2).
- A more thorough secondary survey is performed after stabilization to identify any physical findings that may elucidate the etiology of the ALTE.

HISTORY

- The history often provides the most important information in the evaluation of an ALTE.
- Information about the details of the event should be ascertained, including the infant's respiratory effort, skin color, mental status, muscle tone, the duration of the event, and the degree of resuscitation required prior to evaluation in the emergency department.

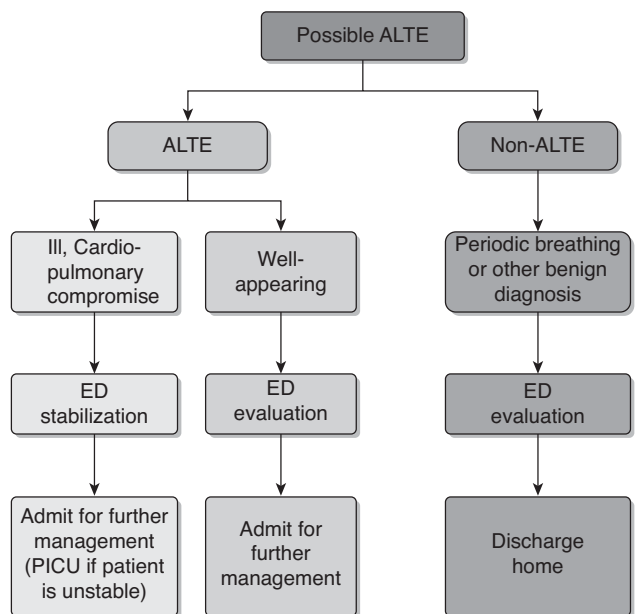


FIG. 5-2. Management of apparent life-threatening event.

- Central apnea is characterized by an absence of respiratory effort while obstructive apnea is usually associated with choking, gasping, or gagging.
- An infant who remains awake and alert during an event is unlikely to have suffered prolonged hypoxia or an acute neurologic event.
- Hypotonia associated with apnea or color change implies significant hypoxia or decreased cerebral perfusion, while hypertonicity is characteristic of seizures.
- A history of apnea that required vigorous physical stimulation or cardiopulmonary resuscitation implies a true life-threatening event.
- Information should be gathered concerning past medical history and any recent illness that may have contributed to the ALTE.
- A history of a sibling with SIDS is a recognized risk factor for sudden death.
- The possibility of factitious ALTE because of Munchausen's syndrome by proxy must be considered in the infant who repeatedly presents with an ALTE or other unexplained illnesses.

PHYSICAL EXAMINATION

- A thorough head-to-toe examination of the infant may provide clues to the etiology of the ALTE.
- Particular attention should be paid to the respiratory, cardiovascular, and neurologic systems.
- Continuous monitoring in the emergency department may provide the opportunity to observe events such as gastroesophageal reflux, choking, cyanosis, or apnea.

EVALUATION AND MANAGEMENT

- The many diagnostic studies to be considered in the evaluation of an ALTE reflect the diverse differential diagnosis (Table 5-2).
- For suspected serious bacterial infection, a complete blood count, blood culture, urinalysis with urine culture, and lumbar puncture should be performed.
- Nasopharyngeal swabs for viral identification should be considered when a viral respiratory infection is suspected.
- A chest radiograph should be obtained for any infant with respiratory or cardiac abnormalities.
- Electrocardiogram is useful to assess for cardiac pathology, including prolonged QT syndrome, Wolff-Parkinson-White syndrome, myocarditis, or anomalous left coronary artery with myocardial ischemia.
- Serum electrolytes, glucose, blood urea nitrogen, serum creatinine, calcium, magnesium, and phosphorus are obtained to evaluate seizures.

TABLE 5-2 Differential Diagnosis of an Apparent Life-Threatening Event

Cardiovascular System
Anemia
Cardiomyopathy
Congenital heart disease
Dysrhythmia (prolonged QT syndrome, Wolff-Parkinson-White syndrome)
Hemorrhage (child abuse)
Myocarditis
Vascular rings and slings
Central Nervous System
Apnea of prematurity
Congenital brain malformation
Head trauma (child abuse)
Idiopathic central apnea
Increased intracranial pressure (congenital hydrocephalus, tumor)
Meningitis/encephalitis
Seizure
Respiratory System
Breath-holding spell
Bronchiolitis (respiratory syncytial virus)
Congenital malformation (choanal atresia, laryngeal cleft, tracheoesophageal fistula)
Foreign body
Laryngomalacia/tracheomalacia
Laryngospasm (choking spell, gastroesophageal reflux)
Obstructive sleep apnea
Periodic breathing of infancy
Pertussis
Pneumonia
Smothering (intentional or unintentional)
Upper airway obstruction (nasal congestion)
Systemic/Metabolic/Other
Dehydration
Electrolyte abnormality (hyponatremia, hypocalcemia, congenital adrenal hyperplasia)
Factitious (Munchausen's syndrome by proxy)
Hypoglycemia
Hypothermia
Inborn errors of metabolism
Sepsis
Toxins/drugs

- Computed axial tomography scan of the head, skeletal survey, and drug screen should be obtained if evaluating child abuse.
- Further evaluation after hospitalization may include sleep studies, pH probe testing, electroencephalography, and other studies that are beyond the scope of the emergency department.

DISPOSITION

- An infant may be discharged from the emergency department if a detailed history and physical examination do not indicate that a true ALTE has occurred.
- A reasonable period of observation in the emergency department is important, but does not prove that the prior event was insignificant and does not rule out recurrence.

- After parental education, the infant without true ALTE may be discharged home with specific instructions for follow-up in 24 hours with a primary care provider or return to the emergency department sooner if any problems occur.
- Any infant with a history of apnea, pallor, cyanosis, limpness, or unresponsiveness requiring vigorous physical stimulation, or cardiopulmonary resuscitation is excluded from outpatient consideration.
- It is best to admit the infant for observation and monitoring if there is any question about the nature of the event, the parents' ability to care for the infant at home, or the adequacy of follow-up.
- Any infant who is unstable should be admitted to the pediatric intensive care unit.

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QUESTIONS

1. You are peer reviewing a case of SIDS, which was seen in the emergency department several weeks ago. The child was a two-month-old female infant, full term, small for gestational age with a birth weight of 2 kg. The 30-year-old mother had placed the infant on her back in the crib the night before. The next morning the infant was found unresponsive and apneic and she was brought to the emergency department where she was declared dead about an hour and a half later. Which of the following characteristics of this patient is a known risk factor for SIDS?
 - A. Female
 - B. Full term birth
 - C. Supine sleeping position
 - D. 30-year-old mother
 - E. 2 kg birth weight
2. You have been asked to be an expert discussant on SIDS at a conference. You have been asked specifically to give a statement about the recommended infant sleeping position. Which of the following statements is true regarding sleeping positions and SIDS?
 - A. The American Academy of Pediatrics advocates the prone sleeping position to prevent SIDS.
 - B. Prone sleeping has been shown to increase the time infants spend in a state of reduced spontaneous arousability.
 - C. Prone sleeping is believed to cause nitrogen to be trapped around the infant's nose and mouth.
 - D. Prone sleeping position has not been found to be associated with SIDS.
 - E. Supine sleeping position has been strongly associated with SIDS.
3. During a recent small group discussion, a medical student asked the question of what exactly defines SIDS in children. Which of the following characteristics is needed to qualify a death as SIDS?
 - A. Age greater than 1 year of age
 - B. Cause of death unexplained despite investigation
 - C. Male infant
 - D. Patient with diagnosed chronic illness
 - E. Previous history of an apparent life-threatening event (ALTE)
4. You are educating new parents about SIDS. One of the parents asks you for examples of risk-reducing strategies. You respond with one of the following answers:
 - A. Avoiding maternal smoking during pregnancy
 - B. Increased temperature of the sleeping environment
 - C. Increasing environmental smoke exposure
 - D. Limiting immunizations in children
 - E. Prone sleeping position

5. You are helping to develop a multidisciplinary team to help families grieve when a child dies of SIDS. Your team includes social workers, chaplains, and child life specialists. Which of the following is an appropriate grieving method?
 - A. Age-appropriate support for family members
 - B. Antidepressant therapy for surviving family members
 - C. Counseling the family as a group
 - D. Encouraging the temporary use of substances to help in grieving
 - E. Use of stimulant medication to enhance mental focus
6. A 2-month-old female infant is brought to the emergency department by her parents after a choking spell at home. Which of the following statements best fits the definition of an ALTE?
 - A. The infant's skin was pink and warm during the choking spell.
 - B. The muscle tone was normal throughout the event.
 - C. The parents appear unconcerned about the event.
 - D. The patient was breathing well throughout the episode.
 - E. The patient was found choking and gagging during a nap.
7. The ALTE is a common reason for families to bring children to the emergency department for evaluation. However when considering the epidemiology of ALTE, which of the following statements is true?
 - A. An ALTE is a reliable predictor of SIDS
 - B. An ALTE occurs in 50% of all infants
 - C. ALTE is more common in female infants rather than male infants
 - D. ALTE is more common in full term infants rather than premature infants
 - E. The peak incidence is in children under 2 months of age
8. A 2-month-old male infant is being evaluated in the emergency department for a possible ALTE. Which of the following statements is true regarding the evaluation of an infant's breathing during an ALTE?
 - A. A history of apnea that required cardiopulmonary resuscitation suggests a true life-threatening event.
 - B. An infant who remains awake and alert during an event has likely suffered severe, prolonged hypoxia.
 - C. During central apnea the infant appears to be distressed and struggling to breathe.
 - D. During obstructive apnea the infant appears to be limp with absence of respiratory effort.
 - E. Normal periodic breathing is often associated with cyanosis.
9. A 4-month-old infant is being evaluated for a possible apneic episode. Which of the following scenarios is most compatible with safe discharge from the emergency department and close out-patient follow-up?
 - A. A stable infant with no pediatrician and a single 16-year-old mother with no private transportation.
 - B. A well-appearing infant with periodic breathing whose detailed history and physical examination do not indicate that a true ALTE has occurred.
 - C. An infant with apnea to whom the parents administered mouth-to-mouth resuscitation.
 - D. An infant with limpness that required vigorous stimulation.
 - E. An infant with prolonged apnea associated with cyanosis.
10. When evaluating an infant for ALTE, which of the following tests is most important to perform in the emergency department prior to hospital admission?
 - A. An electroencephalogram to evaluate for seizures.
 - B. Esophageal pH probe study to evaluate for gastroesophageal reflux.
 - C. Serum electrolytes and glucose to evaluate for acute life-threatening metabolic derangements.
 - D. Sleep study or polysomnogram to determine the type of apnea that is occurring.
 - E. Urine organic acid and serum amino acid profile to evaluate for inborn errors of metabolism.

ANSWERS

1. E. The best choice for the question would be the low birth weight of 2 kg. Other known risk factors for SIDS are young maternal age, lack of prenatal care, prone sleeping position, overheating, and preterm birth. Also, boys are more likely to die from SIDS than girls at a ratio of 60:40.
2. B. Prone sleeping is associated with SIDS and has been shown to increase the time infants spend in a state of reduced spontaneous arousability. The mechanism is not completely understood but it is believed that carbon dioxide becomes trapped around the infant's nose and mouth. The American

Academy of Pediatrics advocates a supine sleeping position as a SIDS risk reduction strategy.

3. B. The definition of SIDS is the sudden death of an infant younger than 1 year of age, which remains unexplained after a thorough case investigation, including a complete autopsy, examination of the death scene, and review of the clinical history. Although male infants seem to have greater incidence of SIDS, the sex of the infant is not needed to diagnose SIDS. Also, chronic illness and previous ALTE are not necessary for the diagnosis of SIDS.
4. A. The risk reduction strategies include avoiding maternal smoking in pregnancy, nonprone sleeping, decreasing environmental smoke exposure, maintaining comfortable ambient temperature, providing a safe sleep environment, and fully immunizing the child.
5. A. Age-appropriate therapy should be provided to siblings and family members by the members of your multidisciplinary team. During the initial phase of grieving, medications and substances should not be encouraged. Although counseling the family as a group may be helpful, it does not meet all the needs of each individual family member.
6. E. An ALTE is defined as an episode that is frightening to the observer and is characterized by some combination of apnea (central or obstructive), color change (cyanosis, pallor, erythema, or plethora), marked change in muscle tone (rigidity or limpness), or unexplained choking or gagging.
7. E. The peak incidence is in infants younger than 2 months. Although the true incidence of ALTE is unknown, it is estimated to occur in 0.5% to 6% of all children during the first year of life and is more common in boys and premature infants. Although there is some overlap of epidemiologic risk factors, the exact relationship between ALTE and SIDS is not clear.
8. A. A history of apnea that required vigorous physical stimulation or cardiopulmonary resuscitation is ominous and implies a true life-threatening event. Normal periodic breathing is not associated with skin color changes. Central apnea is characterized by an absence of respiratory effort, whereas during obstructive apnea, the infant typically appears to be struggling to breathe with choking, gasping, or gagging. The infant's mental status during the event is also important. An infant who remains awake and alert during an event is unlikely to have suffered prolonged hypoxia or an acute neurologic event such as a seizure.
9. B. An infant may be discharged from the emergency department if a detailed history and physical examination do not indicate that a true ALTE has occurred, provided the infant continues to do well and the parents are comfortable with the situation and capable of observing the infant at home. Examples of such a situation include periodic breathing mistaken for apnea or a minor coughing or gagging episode. Any infant with a history of apnea, pallor, cyanosis, limpness, or unresponsiveness requiring vigorous physical stimulation or cardiopulmonary resuscitation is excluded from outpatient consideration. If there is any question about the nature of the event, the parents' ability to care for the infant at home or the adequacy of follow-up, it is best to err on the side of caution and admit the infant for observation and monitoring.
10. C. Of the available choices, the serum electrolytes and glucose are most likely to impact immediate decision making, including determining the severity of the event and stability of the infant. For example, severe hyponatremia, hypoglycemia, or metabolic acidosis may be diagnosed and treated. The studies should also help to determine the disposition of the patient (ie, admit to monitored ward versus intensive care setting). Further studies can be performed after admission including sleep studies, pH probe testing, electroencephalography, and other studies that are beyond the scope of the emergency department. Results of urine organic acid and serum amino acid testing will not return for several days and will not affect emergency department decision making.

6

ALTERED MENTAL STATUS AND COMA

Susan Fuchs

INTRODUCTION

- *Altered mental status* refers to an aberration in a patient's level of consciousness. It always implies serious pathology and mandates an aggressive search for the underlying disorder.
- More precise terminology describes the degree of altered mental status and has important implications for differential diagnosis and management:
 - *Lethargy* is a state of reduced wakefulness in which the patient displays disinterest in the environment

TABLE 6-1 Glasgow and Children's Coma Scale

GLASGOW	CHILDREN'S	SCORE
Eye opening		
Spontaneous	Spontaneous	4
To command	To speech	3
To pain	To pain	2
None	None	1
Motor response		
Follows command	Spontaneous	6
Localizes pain	Withdraws to touch	5
Withdraws to pain	Withdraws to pain	4
Abnormal flexion (decorticate)	Abnormal flexion	3
Abnormal extension (decerebrate)	Abnormal extension	2
No response	No response	1
Verbal response		
Oriented	Coos, babbles, age-appropriate verbalizations	5
Confused	Irritable cry	4
Inappropriate words	Cries to pain	3
Incomprehensible	Moans, grunts	2
No response	No response	1

Modified from Taylor, APLS.

and is easily distracted but is easily arouseable and can communicate.

- *Delirium* is characterized by disorientation, delusions, hallucinations, fearful responses, irritability, and sensory misperception.
- Obtundation is severe blunting of alertness with a decreased response to stimuli.
- *Stupor* exists when the patient can only be aroused by extremely vigorous and repeated stimulation.
- *Coma* occurs when a profound reduction in neuronal function results in unresponsiveness to sensory stimuli. It constitutes the most severe manifestation of altered mental status.
- Coma scoring systems
 - Scoring systems permit objective and reproducible assessment of the degree of altered mental status and allow effective communication among health care providers.
 - Glasgow coma scale (GCS), scores three responses with a range from 3 to 15. The GCS has been modified so that it can be applied to infants and children. The main difference is the verbal response (Table 6-1).

PATHOPHYSIOLOGY

- In general, patients with altered mental status have suffered a diffuse insult to the brain. The more severe the insult, the greater the alteration in mental status.

- For coma to occur, the underlying abnormality must involve damage to either both cerebral hemispheres or to the ascending reticular activating system, which traverses the brain stem through the upper pons, mid-brain, and diencephalon, and plays a fundamental role in arousal.
- Metabolic, infectious, and toxic etiologies tend to produce diffuse but symmetric deficits.
- Structural lesions result in focal deficits that progress in a predictable pattern.
 - Supratentorial lesions produce focal findings that progress in a rostral-caudal fashion, whereas sub-tentorial lesions result in brain stem dysfunction followed by a sudden onset of coma, cranial nerve palsies, and respiratory disturbances.
- The causes of coma are listed in Tables 6-2 and 6-3.

HISTORY

- Focuses on identifying the underlying abnormality.
 - Events prior to the onset of mental status changes such as headache, febrile illness, trauma, and drug ingestion.
 - Associated symptoms such as vomiting, diarrhea, or respiratory difficulties are important clues.
 - Past medical history including diabetes, seizure disorder, or underlying heart or kidney disease is elicited. A prior history of similar episodes may imply an underlying metabolic abnormality, such as an inborn error of metabolism.

PHYSICAL EXAMINATION

- Focuses on assessing the degree of neurologic impairment and localizing the lesion responsible for the patient's altered mental status.
 - Vital signs, including temperature.
 - Airway, breathing, and circulation must be evaluated and managed prior to completing the examination.
 - Important parameters of the general physical examination in patients with altered mental status are outlined in Table 6-4.
 - The neurologic evaluation focuses on an exact description of the patient's mental status, which provides a baseline for comparison during the course of illness.
 - Cranial nerves and motor function are assessed for potentially localizing findings,
 - Evaluate the response of the extremities to a painful stimulus.
 - The biceps, triceps, patellar, and Achilles reflexes are tested for strength and symmetry and the

TABLE 6-2 Etiology of Altered Mental Status Based on the Mnemonic “Tips From the Vowels”

MNEMONIC DEVICE	CATEGORY	CAUSE
A	Abuse	Head trauma Shock
E	Epilepsy (and other causes of seizures)	Hypernatremia Hypocalcemia Hypoglycemia Hyponatremia Postictal state Status epilepticus
	Endocrine	Addison’s disease Hyperthyroidism Hypothyroidism Inborn errors of metabolism
	Electrolyte disorders	Hypercalcemia Hypernatremia Hyponatremia
I	Infection	Brain abscess Encephalitis Meningitis Sepsis Subdural empyema
	Intussusception	Neurologic presentation
O	Overdose	Alcohol Carbon monoxide Lead Opiates Salicylates Sedatives
U	Uremia (and other metabolic causes)	Hemolytic uremic syndrome Hepatic encephalopathy Hypoxia Renal failure Reye’s syndrome
T	Trauma	Child abuse Head trauma Hemorrhage
	Tumor	
I	Insulin-related problems	Diabetic ketoacidosis (DKA) Hyperglycemia Hypoglycemia Ketotic hypoglycemia Nonketotic hypoglycemia
P	Psychogenic	Diagnosis of exclusion
S	Shock	Anaphylactic Cardiogenic Hemorrhagic Hypovolemic Neurogenic Septic
	Stroke (and other CNS lesions)	Arteriovenous malformations Hemorrhage
	Shunt-related problems	Hydrocephalus Shunt dysfunction

patient is evaluated for the presence of a Babinski response, which indicates an upper motor neuron lesion.

- Decorticate posturing signifies dysfunction of the cerebral hemispheres with an intact brain stem.

TABLE 6-3 Mnemonic for Coma Using Childhood Immunizations

DPT Dehydration Poisoning Trauma	HIB Hypoxia, hypothermia Intussusception Brain tumor
OPV Occult trauma (child abuse), overdose Postictal Ventriculoperitoneal shunt problem	MMR Meningitis Metabolic Reye’s syndrome
IPV Infection Postanoxia Viral encephalitis	Hep B Hyperthermia Electrolyte, endocrine Psychogenic Bleeding

- Decerebrate posturing signifies a lesion of the midbrain
- For patients in coma, the area of the brain involved can be localized by considering physical examination findings (Table 6-5).

DIAGNOSTIC TESTING

- All patients with altered mental status should have a bedside glucose determination.
- Other helpful studies include complete blood count with differential and platelets, electrolytes, calcium, renal functions, and urinalysis. In some cases, arterial blood gas and serum ammonia are indicated.
- Patients who may have ingested a toxin require a urine drug screen, as well as blood levels for suspected toxins such as aspirin or alcohol.
- Infants suspected of suffering from inborn errors of metabolism require testing for urine and serum, amino and organic acids, ammonia, liver function, thyroid function, plasma free fatty acids, and serum carnitine.
- If infection is suspected, cultures are obtained from the blood, urine, and cerebrospinal fluid. Lumbar puncture is withheld until increased intracranial pressure (ICP) is excluded (Fig. 6-1).
- Radiographic examination of the cervical spine is performed if there is any suspicion of trauma.
- If physical examination findings suggest a structural lesion, herniation, or increased ICP, a computed tomography (CT) scan should be performed.

THERAPY

- Intubation is required for patients with altered mental status who have lost protective airway reflexes and who are at risk for aspiration. Intubation is also indicated for patients with evidence of critically increased ICP.

TABLE 6-4 Important Parameters of the General Physical Examination of Patients with Altered Mental Status

BODY AREA	TECHNIQUE	FINDINGS
Head	Palpation Fontanelle palpation (in infants)	Hematoma, fracture Fullness, depression, pulsations (reflects ICP)
Eyes	Eye position Reactivity of the pupils Funduscopy examination	Midposition, deviated Constricted, dilated Papilledema, retinal hemorrhages
Ears	Inspection	Bleeding, CSF drainage
Nose	Inspection	Bleeding, CSF drainage
Neck	Palpation Auscultation	Tenderness, spasm, stepoff Bruit
Breath	Check for odor	DKA: fruity smell Hepatic coma: musty smell Uremia: urine smell
Skin	Inspection	Jaundice, petichiae, purpura
Chest	Auscultation	Signs of respiratory pathology
Abdomen	Palpation	Hepatosplenomegaly, masses, evidence of intussusception

TABLE 6-5 Localization of the Area of Brain Dysfunction Using Physical Findings

FINDING	DESCRIPTION	SIGNIFICANCE
Patient Posture	Decorticate: arms flexed, legs extended Decerebrate: extension and internal rotation of the upper and lower extremities (spontaneous or in response to examination or pain) Unilateral decerebrate posturing Flaccid paralysis	Dysfunction of the cerebral hemispheres with intact brain stem Lesion of the midbrain, toxic, or metabolic etiology
Respiratory Pattern	Consistent hyperventilation Cheyne-Stokes (periods of tachypnea followed by apnea) Ataxic breathing (irregular rate and depth) Irregular breathing	Uncal herniation Diffuse lesions of both hemispheres and brain stem Compensation for metabolic acidosis, e.g., DKA, or salicylate intoxication; lesions of the midbrain or low pons Bilateral hemispheric abnormality with intact brain stem; impending temporal lobe herniation
Pupils	Small, reactive pupils Pinpoint, nonreactive pupils Midposition, fixed pupils Unilateral dilated pupil (in the presence of coma) Bilateral fixed pupils	Lesions of the pons or upper medulla Lesion at the level of the medulla Lesions affecting the cerebral hemispheres or intoxication Metabolic derangement Lesions of the midbrain or upper pons Third nerve compression, as with uncal herniation; in the late phase the pupil is nonreactive
Reflex Eye Movements	Oculocephalic (doll's eye) reflex—contraindicated with suspicion of neck trauma Oculovestibular response—contraindicated with suspicion of neck trauma Corneal reflex (touch the cornea with a small piece of cotton)	Tectal herniation; severe hypothermia; may imply severe permanent brain damage Passively move head from side to side: if eyes move together toward the side opposite that to which the head is turned (+), brain stem function is intact. Impairment indicates a lesion at the midbrain or upper pons, sedation, toxic or metabolic encephalopathy Absence indicates lesion at the lower pons or medulla Elevate the head to 30 degrees and irrigate one or both ear canals with ice water. Normal: slow deviation of the eyes toward the irrigated side with lateral nystagmus away from the irrigated ear Asymmetric response indicates brain stem lesion Bilateral loss indicates metabolic or structural brain stem lesion In the unconscious patient, only slow deviation to the irrigated ear is seen If warm water is used, the quick phase is toward the irrigated ear. COWS = cold, opposite, warm, same Normal: close/blink both eyes simultaneously Bilateral absence: pontine lesion, intoxication, metabolic disorder, or paralysis

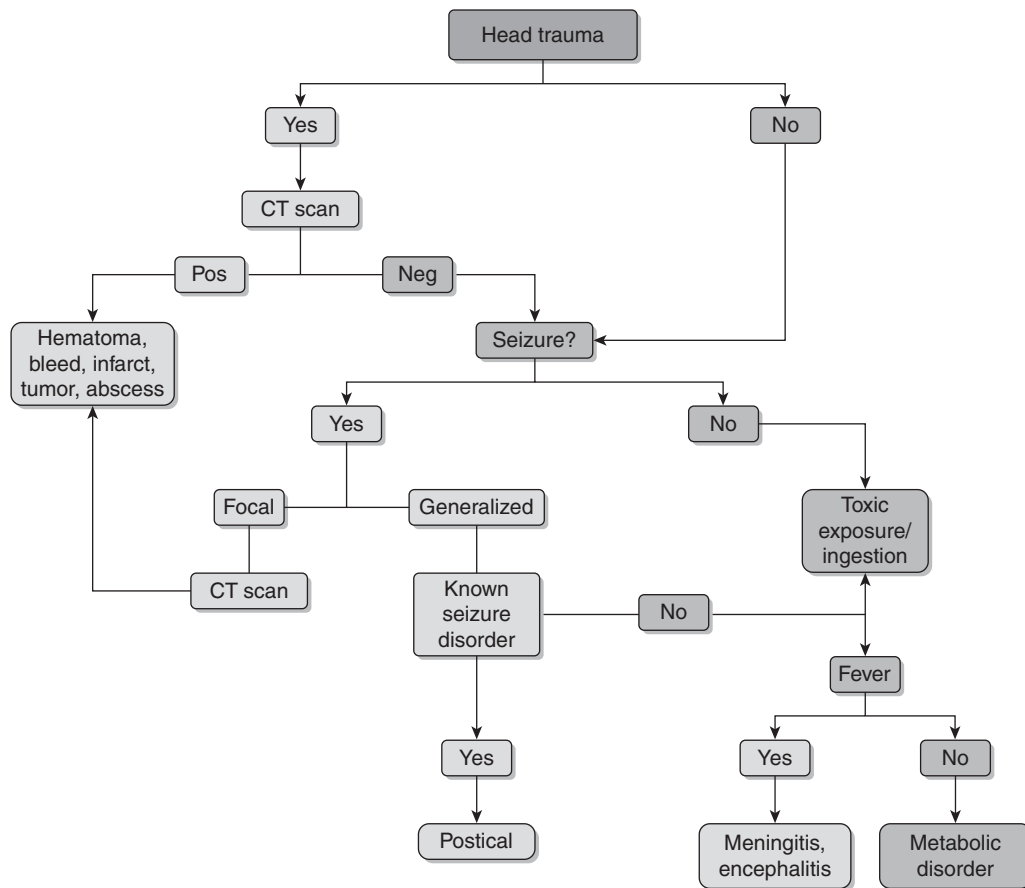


FIG. 6-1. Etiology of altered mental status.

- All patients should receive oxygen and, if hypoglycemia is suspected, 0.5 to 1.0 g/kg of glucose.
- If an ingestion is suspected, a trial of naloxone may prove useful.
- Patients who are hypotensive are resuscitated with crystalloids. Fluids are titrated carefully for patients who may have increased ICP, in whom overaggressive hydration can precipitate herniation. Yet, hypotension is avoided, since it can result in cerebral hypoperfusion and ischemia.
- Hypertension can be secondary to increased ICP, but can also be due to hypertensive encephalopathy. If the etiology is the latter, the blood pressure should be lowered slowly. The goal is to lower the diastolic blood pressure to 100 to 110 mm Hg or a maximum of 25% over 2 to 6 hours.
- Hyperventilation produces vasoconstriction of the cerebral arteries and has been used as the treatment for impending herniation due to elevated ICP. If there are no signs of impending herniation, it is best not to

reduce the P_{CO_2} below 35 torr because severe vasoconstriction and cerebral ischemia can result.

- Elevating the head of the bed to 30° may be beneficial.
- Mannitol or furosemide may be useful adjuncts for patients with severely increased ICP, but hypotension must be avoided and a neurosurgeon should be consulted.
- Additional therapy is directed at maintaining normal body temperature, controlling seizures and correcting acid-base or electrolyte abnormalities

DISPOSITION

- Patients with significant alteration in mental status are best managed in an intensive care unit.
- For patients with milder disease, the decision to admit to the hospital or discharge from the emergency department largely depends on the etiology of the problem.

SPECIAL CONSIDERATIONS

- Several causes of altered mental status and coma are characteristic of the pediatric population and deserve special mention. None are common, but all represent serious problems that confront the emergency physician.

LEAD ENCEPHALOPATHY

- Lead toxicity is a consideration in the differential diagnosis of any child with profoundly altered mental status or coma.
- Lead encephalopathy can be associated with increased ICP and seizures.
- Patients often have a history of pica and parents may have noted abdominal pain, constipation, or vomiting prior to the development of encephalopathy.
- The evaluation and management of lead encephalopathy is discussed in Chapter 113.

INTUSSUSCEPTION

- Intussusception is a fairly common gastrointestinal emergency in children younger than 3 years.
- Although it commonly presents with episodes of intermittent abdominal pain and vomiting, there is a “neurologic presentation” in which the child manifests a depressed level of consciousness that can range from lethargy to obtundation.
- The overall appearance of the patient can mimic shock, with fulminant sepsis a consideration.
- In some cases the abdominal examination may reveal a mass and rectal examination shows heme-positive or “currant jelly” stools.
- Intussusception is discussed in detail in Chapter 9.

REYE’S SYNDROME

- Reye’s syndrome is a disorder characterized by the acute onset of encephalopathy, often developing approximately 2 weeks following a viral infection.
- The diagnosis is based on the presence of encephalopathy, elevated liver enzymes, and the presence of microvesicular fatty changes in the liver.
- The pathophysiology is unknown, but it may involve the interaction of salicylates and certain viruses, especially influenza and varicella.
- The syndrome begins with unremitting vomiting and can progress from lethargy to disorientation, combativeness, and coma.
- The encephalopathy is characterized by increased ICP with elevated liver function tests and serum ammonia

is generally three times normal. Serum bilirubin is only slightly elevated and jaundice is absent.

- Hypoglycemia is common in infants and in patients with severe encephalopathy.
- Fatty microvesicular metamorphosis of the liver can be confirmed by biopsy.
- If the diagnosis of Reye’s syndrome is entertained, aggressive management is indicated.
 - Intravenous fluids are administered at or slightly below maintenance requirements.
 - Hypoglycemic patients may require 10% or 15% dextrose.
 - Patients who are not arousable to voice or light pain are candidates for elective intubation and ventilation.
 - Mannitol may be required for control of ICP. Reye’s syndrome is further discussed in Chapter 6.

INBORN ERRORS OF METABOLISM

- Numerous inborn errors of metabolism can present early in life with vomiting, seizures, and altered mental status. These disorders are discussed in Chapter 81.

HYPOGLYCEMIA

- In any patient with altered mental status, hypoglycemia is a consideration. See Chapter 76 for further discussion.

CONGENITAL ADRENAL HYPERPLASIA

- In a child with congenital adrenal hyperplasia, hypoglycemia may result from the absence of cortisol.
- A constellation of symptoms such as lethargy, vomiting, dehydration, and altered mental status should suggest this disorder.
- See Chapter 77 for further discussion.

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QUESTIONS

1. A 4-year-old boy is brought by his parents for evaluation of altered mental status. The parents report difficulty arousing him after an afternoon nap. Mother reports the child has complained of abdominal pain and has several episodes of vomiting over the last 24 hours. The child was recently diagnosed with pica. Which of the following management principles would apply to this patient?
 - A. The absence of seizures in this child is reassuring for a benign course.
 - B. A lumbar puncture should be withheld until increased ICP can be excluded.
 - C. A trial of naloxone is contraindicated.
 - D. Intravenous glucose at 0.5 to 1.0 g/kg.
 - E. Fluid resuscitation at 20 cc/kg of normal saline.
2. A 2 ½-year-old girl previously healthy with no past medical history is rushed to the emergency department by EMS after a frantic call to 911 for an unresponsive child. The parents report that the child has had intermittent waves of abdominal pain however since she appeared “normal” in between episodes and they deferred seeking immediate medical attention. On arrival to the ED, the child appears in shock and is immediately resuscitated. Physical examination reveals an abdomen mass with heme positive stools. Notably, there is an absence of “currant jelly stools” on exam or by history. Which of the following is true regarding the diagnosis of intussusception in this patient?
 - A. The diagnosis is less likely because a neurologic presentation excludes the diagnosis of intussusception.

- B. The diagnosis is more likely because of the absence of significant past medical history.
 - C. The diagnosis is less likely because of the presentation of shock. Sepsis is a more likely diagnosis
 - D. The diagnosis is likely because of the physical examination findings.
 - E. Intussusception is unusual below the age of 3 years.
3. A 1 ½-year-old boy is brought to the ED for evaluation of altered mental status. The father reports that 1 ½ weeks ago the child was diagnosed with a viral infection. He reports alternating antipyretics for intermittent fevers. He also reports the child received aspirin a “couple of times.”.The father states the child had unremitting vomiting until he “passed out”. Which of the following is contraindicated in the immediate management of this patient?
 - A. Concentrated dosages of dextrose, typically 10–15%
 - B. Mannitol for increased ICP
 - C. Intubation and ventilation
 - D. Lowering the diastolic blood pressure by 25%
 - E. IV fluids at maintenance levels.

ANSWERS

1. B. Lead toxicity is a consideration in the differential diagnosis of any child with profoundly altered mental status or coma. Lead encephalopathy can be associated with increased ICP and seizures. Lumbar puncture is withheld until increased ICP is excluded. Patients often have a history of pica and parents may have noted abdominal pain, constipation, or vomiting prior to the development of encephalopathy. Glucose is only indicated for hypoglycemia. Likewise, patients who are hypotensive are resuscitated with crystalloids. Fluids are titrated carefully for patients who may have increased ICP, in whom overaggressive hydration can precipitate herniation.
2. D. Intussusception is a fairly common gastrointestinal emergency in children younger than 3 years. Although it commonly presents with episodes of intermittent abdominal pain and vomiting, there is a “neurologic presentation” in which the child manifests a depressed level of consciousness that can range from lethargy to obtundation. The overall appearance of the patient can mimic shock, with fulminant sepsis a consideration. In some cases the abdominal examination may reveal a mass and rectal examination shows heme-positive or currant-jelly stools.
3. D. This patient has a fairly typical presentation of Reye's syndrome. If the diagnosis of Reye's

syndrome is entertained, aggressive management is indicated. This would include giving hypoglycemic patients 10% or 15% dextrose. Patients who are not arousable to voice or light pain are candidates for elective intubation and ventilation. Mannitol may be required for control of ICP. Hypertension can be secondary to increased ICP, but can also be due to hypertensive encephalopathy. If the etiology is the latter, the blood pressure should be lowered slowly. The goal is to lower the diastolic blood pressure to 100 to 110 mm Hg or a maximum of 25% over 2 to 6 hours.

7 SEIZURES

Susan Fuchs

INTRODUCTION

- A seizure results from abnormal, excessive, paroxysmal electrical neuronal discharge within the brain.
- Epilepsy is defined as seizures that occur over a period of time without an obvious precipitant.
- In the US, childhood epilepsy has an incidence of 4 to 9 cases per 1000. One percent of children will experience a febrile seizure by the age of 14 and 5% of children will experience a febrile seizure by the age of 6.

CLASSIFICATION

- Seizures are classified as location-related (partial, focal, and local), generalized, undetermined, and special syndromes and then further classified as shown in Table 7-1.
- Generalized seizures can be convulsant or nonconvulsive and involve both cerebral hemispheres,

impair consciousness and have bilateral motor symptoms.

- Absence (petit mal) seizures are nonconvulsive characterized by abrupt and brief loss of awareness (<15 seconds), which may include staring or eye blinking, without postictal confusion. These can often be induced by hyperventilation or photic stimulation.
- Myoclonic seizures are brief muscle contractions of one or several muscles characterized by jerking and flexor muscle spasms that can be irregular and asymmetric. Tonic seizures are due to sustained muscle contraction resulting in rigidity, and tonic-clonic seizures (grand mal) combine the tonic, clonic movements, and a postictal phase.
- Atonic seizures (drop attacks) involve a loss of muscle tone, causing a fall to the floor.
- Benign childhood epilepsy with centrotemporal spikes, also known as benign rolandic epilepsy, with onset between 3 and 13 years of age is the most common partial epilepsy syndrome in children, and often occurs upon awakening, and consists of facial movements, grimacing, drooling, and vocalizations. It can also occur during sleep as tonic or clonic muscle activity.
- Juvenile myoclonic epilepsy (Janz syndrome) is characterized by myoclonic jerks of the arm that occur after awakening, but can also include some generalized tonic-clonic seizures and absence seizures. It begins between 8 and 18 years of age, and there is a strong family history of seizures. Sleep deprivation, hyperventilation, photosensitivity, and alcohol can trigger a seizure.
- West syndrome (infantile spasms) is characterized by sudden symmetric bilateral tonic contractions of the extremities, head, and trunk. The onset is at 5 to 12 months of life, with spasms occurring upon falling asleep or after awakening, with occurrences a few times to hundreds of times a day. Tuberous sclerosis is the most common cause. The classic EEG finding is hypsarrhythmia.

TABLE 7-1 Classification of Seizures

	GENERALIZED	PARTIAL, FOCAL OR LOCAL	FEATURES
Idiopathic or Primary	X	X	No identifiable cause and normal development
Symptomatic or Secondary	X	X	Identifiable brain lesion or systemic disease
Simple		X	No impairment in consciousness
Complex		X	Consciousness impaired; both hemispheres of brain involved
Motor		X	Motor manifestations such as jerks, spasms, twitches present
Somatosensory		X	Numbness, tingling, parathesia or visual phenomena present
Autonomic		X	Changes in sweating, heart rate, pupil size; also psychic symptoms such as aphasia
Psychomotor		X	Repetitive behaviors such as chewing or swallowing, gestures such as clapping, or repetitive verbalizations that are not remembered after the seizure

TABLE 7-2 Etiology of Childhood Seizures

Infections	Hypnatremia
Meningitis	Hypocalcemia
Meningoencephalitis	Hypomagnesemia
Brain abscess	Pyridoxine deficiency
Parasites	Inborn errors of metabolism
Trauma	Vascular
Hemorrhage: epidural, subdural	Intracranial hematoma
Posttraumatic	Embolism
Intoxication	Infarction
Lead	Hypertensive encephalopathy
Cocaine	Tumor
PCP	Psychological
Amphetamine	Hyperventilation
Aspirin	Breath-holding spells
Carbon monoxide	Congenital
Isoniazid	Malformations
Organophosphates	Birth asphyxia
Theophylline	Neurocutaneous syndromes
Lidocaine	Other
Lindane	S/P DPT immunization
Drug withdrawal (anticonvulsants)	Seizure disorder
Metabolic	Noncompliance
Hypoglycemia	Inadequate drug level
Hyponatremias	

Data from Isaacman DJ, Trainor JL, Rothrock SG. Central nervous system. In: Gausche-Hill M, Fuchs S, Yamamoto L, eds. *APLS: The Pediatric Emergency Medicine Course*. Sudbury, MA: Jones and Bartlett Publishers; 2004:168–182; Chiang VW. Seizures. In: Fleisher GR, Ludwig S, Henretig FM, eds. *Textbook of Pediatric Emergency Medicine*. 5th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2006:629–636.

- Lennox–Gastaut syndrome has its onset at 1 to 8 years of age and consists of multiple seizure types. These children often have seizures every day, and there is an associated deterioration in intelligence, as well as behavior disorders.

THE FIRST AFEBRILE SEIZURE

- Fever is the most common cause, followed by infections, trauma, toxic exposures, metabolic disorders, and failure to take prescribed anticonvulsants. In addition, childhood seizures are often idiopathic. A more thorough list is included in Table 7-2.

HISTORY

- Elicit detailed information regarding the episode itself, including preceding events such as aura, hallucinations (auditory or visual) and the duration of the episode.
- Obtain a clear description of the patient's level of consciousness during the episode, memory of the event, duration, and any postictal phenomena.
- Note and characterize abnormal motor movements as localized or generalized.

- Elicit information about abnormal eye movements, facial grimacing, lip movements, and urinary or fecal incontinence.
- If there have been recurrent episodes, a pattern may be evident, such as a predilection for the event to occur upon waking or when the patient is fatigued.
- Obtain a history of fever, trauma, prior seizures, drug use or withdrawal, underlying medical disorders, perinatal problems, developmental milestones, and a family history of seizures.

DIFFERENTIAL DIAGNOSIS

- Differential diagnosis includes syncope, gastroesophageal reflux, breath-holding spells, migraines, sleep disorders, behavioral disorders, movement disorders, and psychogenic causes.
- Syncopal episodes are usually preceded by blurred vision, dizziness, and pallor.
- Gastroesophageal reflux usually results in an arched back position with crying, no loss of consciousness, and is usually associated with feeding.
- Cyanotic breath-holding spells usually occur after a crying episode, and result in limpness and loss of consciousness, occasionally with posturing. With a pallid breath-holding spell, the infant often sustains minor head trauma, loses consciousness, stops breathing, becomes pale, and limp. They may develop generalized increased muscle tone with incontinence, and have a postictal period.
- A child who is daydreaming maintains his or her posture and head control, and can be interrupted by name calling or touch.
- During a pseudoseizure, the patient often keeps his or her eyes tightly closed, resists eye opening, and avoids painful stimuli, and afterwards there is rapid return to a normal level of consciousness.

PHYSICAL EXAMINATION

- If the child is actively seizing, stabilize airway, breathing, and circulation first.
- Obtain vital signs including temperature, heart rate, respiratory rate, and blood pressure.
- Determine the child's level of consciousness.
- Measure the head circumference in young infants to detect micro- or macrocephaly, and palpate the head in any child with trauma to detect hematomas or skull fractures.
- Examine the eyes, assessing pupillary reactivity, whether gaze is conjugate or disconjugate, and perform a fundoscopic examination for papilledema or retinal hemorrhages.

- Document the presence or absence of meningismus and photophobia.
- Examine skin for petechiae and signs of neurocutaneous syndromes.
- Perform a thorough neurologic examination, which may reveal Todd's paresis, a transient paralysis that can follow a seizure. It is usually unilateral and may involve both the face and extremities.

LABORATORY EVALUATION

- Perform a bedside glucose check on all patients.
- Other laboratory studies are based on the type of seizure, history, and likely etiologies, and should be individualized. If the child has been on antiseizure medication, a drug level is obtained, if available.
- Perform a lumbar puncture in any patient suspected of having a central nervous system infection. Delay the lumbar puncture for CT if there are focal findings on physical examination or suspicion of a mass lesion or increased intracranial pressure. In infants younger than 6 months with a first afebrile seizure, with no etiology, and persistent decreased level of consciousness, a lumbar puncture should be performed.

RADIOLOGIC EVALUATION

- There is little use for neuroimaging in most patients with a generalized seizure, no focal findings on physical examination, and no history of trauma.
- Obtain a head CT or MRI if there is a postictal focal deficit (Todd's paralysis) that does not quickly resolve, the level of consciousness remains decreased, or if mental status does not return to baseline within several hours.
- If trauma is suspected, a CT scan is preferred as an acute hemorrhage can be detected, but MRI is preferred for brain damage and old hemorrhage detection.

ELECTROENCEPHALOGRAM

- EEG should not be performed until a few days to weeks after the seizure.

DISPOSITION

- Consider admission for any child with abnormal neurologic examination.

- For patients with a first afebrile seizure, a nonfocal neurologic examination, a negative ED workup, discharge with instructions to follow up with neurology. See Fig. 7-1 for seizure algorithm.
- Recurrence is not predictable. The majority of recurrences occur within 1 to 2 years. The overall 3-to-5-year recurrence risk is 40% to 50%. Withholding treatment does not change prognosis.
- Reassurance and addressing safety concerns are the most important pieces of information to provide the patient and parents.
- In some cases, discussion of rescue medications such as rectal diazepam may be helpful. Rectal diazepam comes in a premeasured gel formulation in a prefilled special tip syringe that is useful for children with prolonged seizures and those far from medical care. The dose is 0.2 to 0.5 mg/kg with a maximum dose of 5 mg for those <5 years, and 10 mg for those ≥5 years.

ANTIPILEPTIC DRUGS

- Many anticonvulsants are available, some of which have efficacy for certain types of seizures (Tables 7-3 and 7-4).
- Consult a pediatric neurologist before initiating therapy and see Table 7-5 for dosing, side effects, and drug interactions.
- Therapeutic drug levels are available for some AEDs, but serious side effects tend to be idiosyncratic (Table 7-6).

NEONATAL SEIZURES

- These are seizures that occur during the first 28 days of life, mostly soon after birth.
- Because the cerebral cortex is immature, seizures in neonates can be convulsive (asymmetric or bilateral posturing of the trunk or extremities and eye deviation), or nonconvulsive (motor automatisms such as of lip smacking, tongue movements, random eye movement, and pedaling movements of legs).
- Neonatal seizures are commonly related to perinatal asphyxia; intracranial hemorrhage; central nervous system infections; cerebral infarction; metabolic abnormalities, especially hypoglycemia and hypocalcemia; and congenital abnormalities of the brain.
- Less commonly, seizures are related to inherited metabolic abnormalities, including urea cycle defects and abnormalities in amino acid metabolism. These defects often become apparent after the infant begins

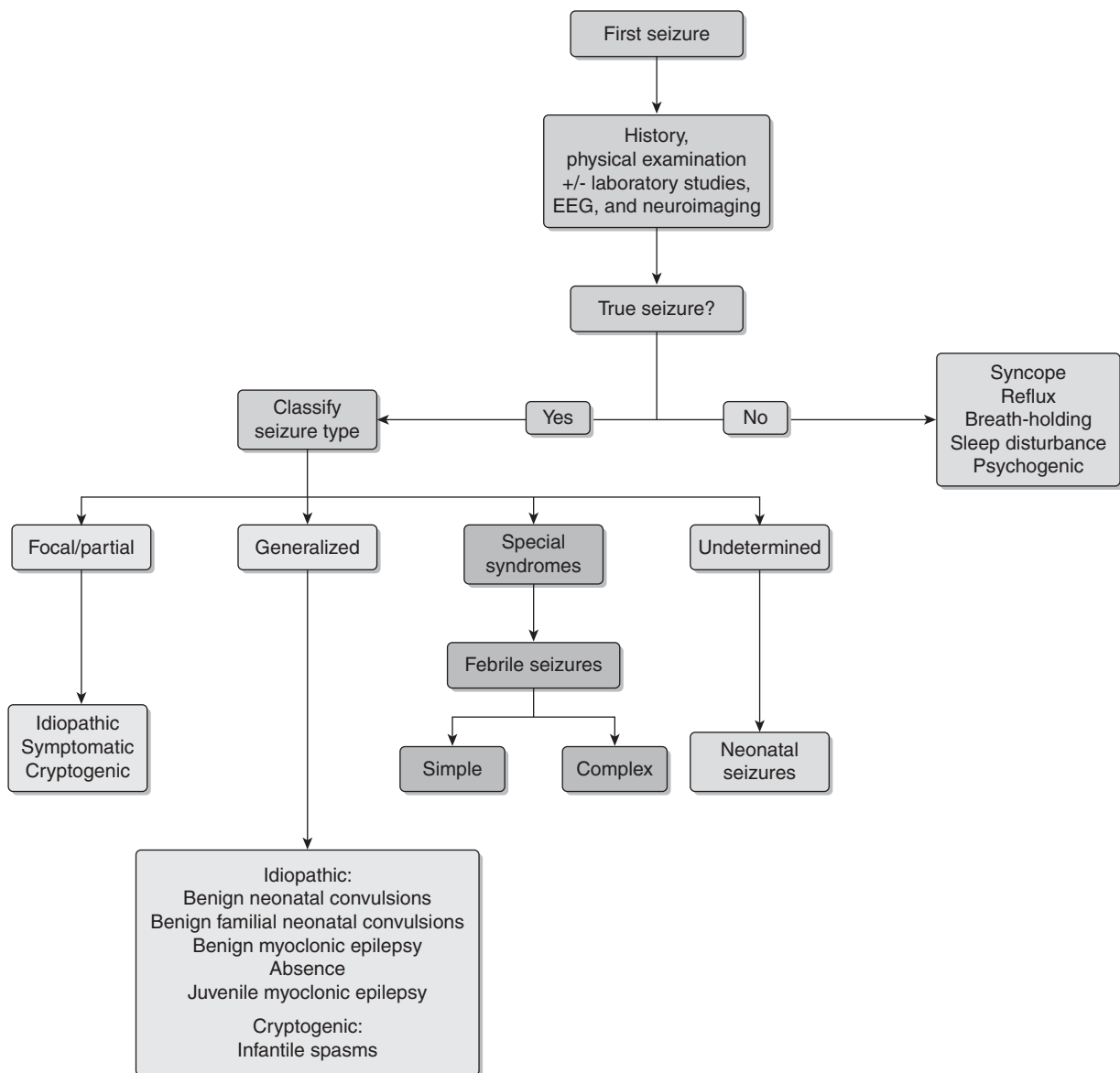


FIG. 7-1. Seizure algorithm. (Data from Major P, Thiele EA: Seizures in children: determining the variation. *Pediatr Rev.*28:363–371, 2007; LaRoche SM, Helmers SL: The new antiepileptic drugs: clinical applications. *JAMA* 291:615–620, 2004.)

feeding and usually cause lethargy, vomiting, and poor feeding as well as seizures.

- A rare cause of refractory seizures in neonates is inherited pyridoxine deficiency, which is inherited as an autosomal recessive trait. Specific neonatal epileptic syndromes are listed in Table 7-7. Important aspects of the history for patients with neonatal seizures are listed in Table 7-8.

LABORATORY EVALUATION

- Obtain bedside glucose, serum glucose, electrolytes, calcium, and magnesium.
- Unless trauma is suspected and CT needed, perform a lumbar puncture for bacterial and viral cultures, cell count, protein, glucose, and Gram stain as soon as possible.

TABLE 7-3 Antiepilepsy Drugs (AEDs) Used for Specific Seizure Types

PARTIAL/FOCAL SEIZURE	FOCAL OR GENERALIZED	SYNDROME SPECIFIC
Carbamazepine	Valproic acid	Ethosuximide (absence)
Fos/Phenytoin	Felbamate	Viagbatrin (infantile spasms) ^a
Primidone	Primidone	Valproic acid (Lennox-Gastaut)
Gabapentin	Levetiracetam	
Tiagabine	Tiagabine	
Oxcarbazepine		
Pregabalin		

^aNot available in the United States.

Data from Major P, Thiele EA. Seizures in children: Laboratory diagnosis and management. *Pediatr Rev.* 2007;28:405–414; Wilfong A. Treatment of seizures and epileptic syndromes in children. In: Rose BD, ed. *UpToDate*. Waltham, MA: UpToDate; 2007; Chapman K. *Controversies in the Treatment of Epilepsy. Seizure Medications, the Old and the New*. San Francisco, CA: American Academy of Pediatrics, National Conference and Exhibition; 2007.

TABLE 7-4 Anticonvulsant Choice—Daily Oral Medications

SEIZURE TYPE	DRUG OF CHOICE
Absence	Ethosuximide (Zarontin) 15–30 mg/kg/d Valproic acid (Depakene) or divalproex (Depakote) 15–60 mg/kg/d Lamotrigine (Lamictal) 10–12 mg/kg/d if given alone; 2–5 mg/kg/d when given with valproic acid
Atonic	Benzodiazepines, Valproic acid, clonazepam (Klonopin) 0.2 mg/kg/d, ethosuximide
Myoclonic	Valproic acid, clonazepam, lamotrigine
Partial	Carbamazepine (Tegretol/Carbatrol) 10–40 mg/kg/d bid or qid Felbamate 45–60 mg/kg/d Levetiracetam (Keppra) 10–60 mg/kg/d Phenytoin/fosphenytoin 4–8 mg/kg/d Valproic acid Phenobarbital 4–6 mg/kg/d Primidone (Mysoline) 10–20 mg/kg/d Gabapentin (Neurontin) 40–80 mg/kg/d Oxcarbazepine (Trileptal) 20–40 mg/kg/d Tiagabine (Gabatril) <12 y 0.5 mg/kg/d, ≥12 y 4 mg/kg/d
Generalized, tonic-clonic	Carbamazepine, phenytoin, phenobarbital, primidone, valproic acid, lamotrigine, topiramate (Topamax) ^a 4–10 mg/kg/d
Infantile spasms	ACTH, benzodiazepines, valproic acid, topiramate, ketogenic diet

^aNot available in the United States.

Data from Wilfong A. Treatment of seizures and epileptic syndromes in children. In: Rose BD, ed. *UpToDate*. Waltham, MA: *UpToDate*; 2007; Friedman MJ, Shariief GQ. Seizures in children. *Pediatr Clin North Am.* 2006;53:257–277.

TABLE 7-5 Adverse Effects of AEDs

AED	ADVERSE EVENTS
Carbamazepine	Blood dyscrasia, elevated liver functions, rash
Ethosuximide	Blood dyscrasia, somnolence, rash
Felbamate	Aplastic anemia, severe hepatic toxicity
Gabapentin	Fatigue, ataxia, weight gain
Lamotrigine	Rash, TEN, SJS
Levetiracetam	Ataxia, behavioral changes
Oxcarbazepine	Hypo Na, hepatic or blood dyscrasia
Phenobarbital/primidone	Somnolence, cognitive impairment, rash
Phenytoin/fosphenytoin	Ataxia, rash, somnolence
Pregabalin	Ataxia, fatigue
Tiagabine	Spike wave stupor, weakness
Topiramate	Renal calculi, glaucoma, weight loss, metabolic acidosis
Valproic Acid	Drowsiness, thrombocytopenia, hepatic necrosis
Vigabatrin ^a	Dizziness, retinal degeneration
Zonisamide	Rash, renal calculi, photosensitivity

SJS, Steven's Johnson Syndrome; TEN, Toxic epidermal necrolysis.

^aNot available in the United States.

Data from Major P, Thiele EA. Seizures in children: laboratory diagnosis and management. *Pediatr Rev.* 2007;28:405–414; Wilfong A. Treatment of seizures and epileptic syndromes in children. In: Rose BD, ed. *UpToDate*. Waltham, MA: UpToDate; 2007; Chapman K. *Controversies in the Treatment of Epilepsy. Seizure Medications, the Old and the New*. San Francisco, CA: American Academy of Pediatrics, National Conference and Exhibition; 2007; LaRoche SM, Helmers SL. The new antiepileptic drugs: scientific review. *JAMA.* 2004;291:605–614.

TABLE 7-6 Therapeutic Monitoring

AED	MONITORING
Carbamazepine	4–12 µg/mL
Ethosuximide	40–100 µg/mL
Felbamate	40–100 µg/mL
Phenobarbital	10–40 µg/mL
Phenytoin	10–20 µg/mL
Valproic acid	50–100 µg/mL
Gabapentin (Neurontin)	None
Lamotrigine (Lamictal)	None
Levetiracetam (Keppra)	None
Oxcarbazepine (Trileptal)	None
Pregabalin (Lyrica)	None
Tiagabine (Gabatril)	None
Topiramate (Topamax)	None
Zonisamide (Zonegran)	None

Data from Wilfong A. Treatment of seizures and epileptic syndromes in children. In: Rose BD, ed. *UpToDate*. Waltham, MA: *UpToDate*; 2007; Conway JM, Kriel RL, Birnbaum AK. Antiepileptic drug therapy in children. In: Swaiman KF, Ashwal S, Ferriero DM, eds. *Pediatric Neurology: Principles and Practice*. 4th ed. Philadelphia, PA: Mosby/Elsevier; 2006:1105–1130; Chapman K. *Controversies in the Treatment of Epilepsy. Seizure Medications, the Old and the New*. San Francisco, CA: American Academy of Pediatrics, National Conference and Exhibition; 2007.

TABLE 7-7 Causes of Neonatal Seizures

Hypoxia/anoxia (intrauterine or perinatal)
Cerebral ischemia (secondary to hypoxia/anoxia)
Hemorrhage
Subarachnoid (birth trauma)
Subdural (birth trauma)
Intraventricular/intracerebral (prematurity)
Infection
Meningitis: group B streptococci, <i>Escherichia coli</i>
Meningoencephalitis: herpes, cytomegalovirus, toxoplasmosis
Metabolic
Hypoglycemia (especially first day of life)
Hypocalcemia (days 3–14)
Pyridoxine (vitamin B ₆) deficiency
Inborn errors of metabolism (days 4–7)
Aminoacidurias
Maple syrup urine disease
Phenylketonuria
Urea cycle defects: citrullinemia
Organic acidurias: propionic academia
Structural anomalies
Lissencephaly
Hereditary disorders
Tuberous sclerosis
Drug withdrawal
Narcotics
Neonatal epileptic syndromes

TABLE 7-8 Important Aspects of the History for Patients with Neonatal Seizures

Gestational age of the infant
History of maternal infections
Maternal drug use during pregnancy
Maternal fever during labor
Premature rupture of membranes
Duration of labor
Method of delivery
Complications during delivery
Need for the newborn to be aggressively resuscitated, which may indicate perinatal asphyxia
Feeding pattern and the type of formula (important for a child who has a seizure after 3 d of age, when inherited metabolic defects become more likely)

- If an inherited metabolic defect is considered, serum ammonia, serum, and urine amino acids, arterial blood gas and, if possible, serum lactate are indicated.
- Cranial ultrasound or CT scan can be useful to diagnose hemorrhage.

TREATMENT

- Secure an adequate airway, ensure oxygenation, and obtain vascular access.
- If hypoglycemia is present administer 2 mL/kg of D₁₀W intravenously, followed by an infusion of D₁₀W.

- Use phenobarbital (20 mg/kg intravenously) or phenytoin (20 mg/kg) to stop seizures.
- If seizures are refractory, give pyridoxine (100 mg intravenously).
- Correct other metabolic abnormalities such as hypocalcemia (<7 mg/dL) and hypomagnesemia.

FEBRILE SEIZURES

- Febrile seizures are accompanied by a fever without evidence of intracranial infection, intracranial abnormality, metabolic abnormality, toxins, or an endotoxin.
- Febrile seizures usually occur between 6 months and 5 years of age. Most are self-limited, generalized, last for less than 15 minutes, and occur once in a 24-hour period, in which case they are classified as simple.
- A complex febrile seizure lasts more than 15 minutes, occurs more than once in a 24-hour period, or has a focal component.
- Following a febrile seizure, children will usually have a postictal period during which they are lethargic, irritable, or confused, and may have a brief period of hemiparesis.
- Approximately 2% to 5% of all children will have a febrile seizure. They occur most commonly in children younger than 2 years.
- Twenty-five to thirty percent of children who have one febrile seizure will have a recurrence.
- Risk factors that correlate with an increased risk of subsequent epilepsy include a prolonged or unilateral seizure, a prior neurologic deficit, and a family history of epilepsy.
- Commonly implicated etiologies include upper respiratory tract infections, viral syndrome, pharyngitis, otitis media, pneumonia, gastroenteritis, urinary tract infections, and roseola. Febrile seizures can also occur after immunizations.
- Take a history focused on any preceding febrile illness and a description of the seizure and its duration
- Perform a complete physical with attention to the cause of fever and ruling out any central nervous system infection.
- In a febrile seizure, the neurologic examination is normal. If a neurologic deficit exists, consider another etiology for the fever.

LABORATORY EVALUATION

- Obtain a bedside glucose in all patients.
- For the first simple febrile seizure, there are no other required laboratory studies.

- The greatest controversy is whether lumbar puncture is required.
- Patients >18 months of age who are nontoxic, with normal mental status, and no evidence of neck pain or stiffness do not require a lumbar puncture.
- In a child who is still postictal or noncommunicative (younger than 12 months) or who has received prior antibiotics, detecting meningismus may be difficult. In this situation, a lumbar puncture should be strongly considered.
- Other studies such as skull radiographs, CT scan, and even EEG are rarely helpful and not warranted after the first simple febrile seizure, unless history or physical findings suggests some underlying pathology.

THERAPY

- Stabilize the airway and ensure adequate oxygenation if the child is still seizing.
- Give anticonvulsants if the seizure persists for more than 5 minutes (see Status Epilepticus).
- Administer acetaminophen, 15 mg/kg po or pr or ibuprofen, 10 mg/kg po.
- Treatment with anticonvulsants is not indicated after a first febrile seizure.

DISPOSITION

- Discharge patients who have had a febrile seizure with instructions to follow up with primary care unless an underlying infection precludes discharge. Treat bacterial infection with appropriate antibiotics.
- Parental reassurance and education regarding the benign nature of febrile seizures, the low risk of recurrence, and the low incidence of subsequent epilepsy are part of the discharge instructions.

STATUS EPILEPTICUS

- Status epilepticus is a seizure lasting 30 minutes or longer or two or more seizures without recovery of consciousness in between.
- Status can present in several forms, including generalized (convulsive and nonconvulsive) and partial seizures (convulsive and nonconvulsive).
- Etiologies overlap those for a first seizure and include non-CNS fever or infection, central nervous system infection, medication change or non-compliance in children on anticonvulsant therapy, head trauma, hypoxia, metabolic disorders, toxic

- ingestions, ethanol, tumor, vascular lesions, and progressive neurologic disorders.
- Stabilize the airway and ensure adequacy of oxygenation first. Secure venous access as soon as possible.
- Use the chin lift or jaw thrust to open the airway. An oral or nasal airway can be inserted as needed. Oral suctioning may be required, so this should be available.
- Administer high-flow oxygen (15 L) is administered to all patients via nonrebreather mask or bag-mask ventilation.
- Monitor cardiac status and pulse oximetry continuously.
- Intubation may be necessary to oxygenate and ventilate the patient adequately.
- Determine bedside glucose and draw blood for complete blood count, electrolytes, BUN, glucose, calcium, and magnesium.
- For patients on anticonvulsant therapy, drug levels are obtained, and in some patients a toxicology screen may be indicated.
- If the glucose is <60 mg/dL, give 0.5 to 1.0 g/kg of dextrose as D₂₅W, 2 to 4 mL/kg or D₅₀W 1 to 2 mL/kg. In adolescents, also give thiamine 100 mg.
- If vascular access cannot be obtained, intraosseous access is acceptable, and some initial AED medications can be given rectally, intranasally or intramuscular route.
- Begin medications within 5 to 10 minutes of status onset, as there is a risk of mortality in 1% to 3% of prolonged seizures.
- Benzodiazepines are effective for treatment of an actively seizing patient.
- Lorazepam (Ativan) has an onset of action of 2 to 3 minutes and a relatively long half-life of 12 to 24 hours. Side effects include respiratory depression and sedation. The dose is 0.10 mg/kg, up to a maximum of 4 mg per dose.
- Diazepam (Valium) is useful for control of seizures. It has an onset of action of 1 to 3 minutes, but its half-life of 15 to 20 minutes means that repeated doses are often required. The dose is 0.1 to 0.3 mg/kg, slowly by intravenous push. Side effects include respiratory depression, hypotension, sedation, and bradycardia.
- Diazepam can also be given rectally, using the intravenous formulation in a dose of 0.5 mg/kg for the first dose and 0.25 mg/kg for any subsequent dose, to a maximum of 20 mg. With rectal administration, the onset of action is usually within 5 to 10 minutes. There is also a rectal gel form of diazepam (Diasat) available, with the dose via this formulation of 0.2 to 0.5 mg/kg with a maximum dose of 5 mg for those <5 years, and 10 mg for those ≥5 years. It is

available in several premeasured sizes: 2.5, 5.0, and 10 mg.

- Midazolam (Versed) is a benzodiazepine that is rapidly absorbed after intramuscular injection and is an alternative to other benzodiazepines when it is impossible to obtain intravenous or intraosseous access.
- The intravenous dose is 0.1 mg/kg, and the intramuscular dose is 0.2 mg/kg with an onset of action in approximately 2 to 3 minutes IV and 15 minutes IM.
- Benzodiazepines are not useful for long-term seizure control, thus providing a long-acting antiseizure medication is also required
- Fosphenytoin is a water-soluble prodrug of phenytoin. The conversion is 1.5 mg fosphenytoin = 1 mg phenytoin = 1 mg phenytoin equivalents (PE). (Therefore, 150 mg fosphenytoin = 100 mg phenytoin = 100 mg PE, and all doses listed below are in PE.) The intravenous loading dose is 20 mg PE/kg, which can be given at a rate of 3 mg PE/kg/min up to 150 mg PE/min. (It can also be given IM.) The only side effects are pruritus and paresthesias (groin). Because it is in a neutral solution and does not contain propylene glycol, it can also be given intramuscularly (same dose as intravenously); however, peak levels are reached in 3 hours.
- Phenytoin, when given intravenously, has rapid brain deposition but takes longer to control the seizure than a benzodiazepine (10 to 30 minutes). The loading dose is 20 mg/kg, which must be given slowly, 50 mg/min in adults or 1 mg/kg/min in children <50 kg. Side effects include hypotension and cardiac conduction disturbances (widened QT interval and arrhythmias), which if they occur should prompt a slower infusion or stopping the medication. Phenytoin will precipitate in glucose solutions, so it should be given directly into the vein, or in saline.
- Phenobarbital is still a useful drug for treating status epilepticus, and it remains the drug of choice for neonatal seizures. Peak brain levels are reached in 10 to 20 minutes, and its duration of action is >48 hours. The loading dose is 20 mg/kg IV given slowly at 100 mg/min. Side effects include respiratory depression (additive with benzodiazepines), sedation, and occasionally hypotension. If seizures stop before the entire loading dose is given, the remainder can be given intravenously or even orally within 1 to 2 hours
- If status persists after giving one dose of a benzodiazepine followed by phenytoin or phenobarbital, an additional dose of benzodiazepine can be given. If the seizure persists after phenytoin or phenobarbital is given, the alternate drug of the two can be administered. For refractory status, midazolam, pentobarbital,

propofol or valproic acid may be given as continuous infusions.

- Pentobarbital is given as a loading dose of 5 to 15 mg/kg intravenously followed by an infusion of 0.5 to 5 mg/kg/h, to keep the level between 20 and 50 μ g/mL and to produce burst suppression on the EEG, or cessation of epileptic activity. Vasopressors are often needed with pentobarbital coma.
- Midazolam is given as a 0.2 mg/kg load followed by an infusion of 1 μ g/kg/min, and titrated upward (up to 4 μ g/kg/min) over 60 minutes until there is cessation of seizures or burst suppression on the EEG
- Another option is the use of intravenous propofol (1–3 mg/kg load followed by 2–10 mg/kg/h). However, side effects include hypotension with rapid infusion, as well as fatal acidosis and rhabdomyolysis with maintenance infusions. It is also contraindicated in children on a ketogenic diet.
- Valproic acid can also be given IV (Depacon) as a 15 to 20 mg/kg load, and repeated every 10 to 15 minutes to a maximum of 40 mg/kg, followed by an infusion of 5 mg/kg/h.
- Therapy of nonconvulsive status epilepticus is similar to that of convulsive status, using a benzodiazepine, fosphenytoin, or phenytoin. For absence status, a benzodiazepine can be followed by oral or nasogastric ethosuximide.

DISPOSITION

- Admit all patients given lorazepam or long-acting anticonvulsants.
- Since diazepam is short acting, if no other drugs have been given, individualize the admission decision.
- Admit all patients in status or receiving/needing assisted ventilation to the ICU.

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QUESTIONS

1. A 14-year-old female is brought to the ED by her parents after a loss of consciousness. The patient describes feeling everything getting very small beforehand. She denies a headache or a history of head trauma. This best describes which of the following?
 - A. Simple partial seizure
 - B. Complex partial seizure
 - C. Syncope
 - D. Pseudoseizure
 - E. Generalized seizure
2. A 7-year-old male is brought to the ED for crying and drooling upon awakening a few times in the last week. Parents deny any muscle activity or trauma. There is no family history of seizures. This is an example of which of the following?
 - A. West syndrome
 - B. Janz syndrome
 - C. Febrile seizure
 - D. Rolandic epilepsy
 - E. Lennon Gastaut syndrome
3. A 2-year-old male is brought to the ED after losing consciousness. The parents state he was crying beforehand, then went limp and turned blue. In the ED, he is very active and back to normal according to the parents. This best describes:
 - A. Breath holding spell
 - B. Afebrile seizure
 - C. Febrile seizure
 - D. A tic
 - E. A pseudoseizure
4. A 5-year-old child presents to the ED seizing for the last 5 minutes. The first step of management is which of the following?
 - A. IV access
 - B. Rectal diazepam
 - C. Open the airway
 - D. Intubation
 - E. Watch until seizure stops
5. A 6-year-old had an afebrile seizure at home. In the ED, he is still lethargic and his neurologic examination shows no focal findings. Which of the following should be performed first?
 - A. Lumbar puncture
 - B. CT scan of his head
 - C. Bedside glucose
 - D. MRI of his brain
 - E. Complete blood count
6. A 2-year-old had a generalized seizure at home lasting 30 seconds. When seen in the ED he is awake and playful, but has a fever of 39.5°C. He is found to have otitis media. Which of the following is needed prior to discharge?
 - A. Parental reassurance
 - B. CT of brain
 - C. CBC
 - D. Urinalysis
 - E. Lumbar puncture
7. A 5-day-old infant comes to the ED with lip smacking and blue lips. She was full term, had no perinatal problems and was discharged 3 days ago with her mother. After establishing an airway, providing oxygen and establishing vascular access, the most appropriate medication is which of the following?
 - A. Prostaglandin A
 - B. Calcium
 - C. Pyridoxine
 - D. Phenobarbital
 - E. Glucose
8. A 9-year-old comes to the ED after the teacher says he seems to be daydreaming in school. In the ED the child says he feels funny when he works on the computer, but does not fall out of his chair. You suspect absence seizures. Which of the following would be most beneficial?
 - A. CT of the head
 - B. Outpatient EEG
 - C. See if they occur after hyperventilation
 - D. ECG
 - E. Diazepam for home use
9. A 13-year-old female comes to the ED in status epilepticus. There is no history of trauma or seizure disorder. Jaw thrust and oxygen maintains her airway, and vascular access is obtained. The seizure continues after a dose of lorazepam. The next medication to provide is
 - A. Diazepam
 - B. Phenobarbital

- C. Propofol
 - D. Valproic acid
 - E. Fosphenytoin
10. A 3-year-old with complex partial seizures comes to the ED with increasing seizures. The child is on gabapentin, levetiracetam, carbamazepine, topiramate, and carnitine. Which of these drugs levels can be checked in the ED?
- A. Gabapentin
 - B. Levetiracetam
 - C. Carbamazepine
 - D. Topiramate
 - E. Carnitine

ANSWERS

1. B. Complex partial seizure. Complex partial seizures result in impaired consciousness and involve both sides of the brain. Motor involvement is not always present, complex somatosensory symptoms such as numbness, tingling, or paresthesias of an extremity, or visual phenomena are always present. Autonomic symptoms include sweating, change in heart rate, pupil size, and piloerection; psychic symptoms include aphasia, déjà vu or jamais vu experiences, and illusions of perception (size and sound); and affective symptoms include such things as fear, anger or depression, and even hallucinations. A simple partial seizure results in no impairment in consciousness. Syncopal episodes are often preceded by blurred vision, followed by a loss of consciousness. A pseudoseizure tends to be a diagnosis of exclusion, but the patient often avoids painful stimuli and returns to a normal level of consciousness after the seizure. A generalized seizure results in impaired consciousness, and bilateral motor symptoms.
2. D. Rolandic epilepsy. Benign childhood epilepsy with centrotemporal spikes, also known as benign rolandic epilepsy, has an onset between 3 and 13 years of age and is the most common partial epilepsy syndrome in children. It often occurs upon awakening, and consists of facial movements, grimacing, drooling, and vocalizations. It can also occur during sleep as tonic or clonic muscle activity. West syndrome (infantile spasms), is characterized by sudden symmetric bilateral tonic contractions of the extremities, head, and trunk. The onset is at 5 to 12 months of life, with spasms occurring upon falling asleep or after awakening, with occurrences a few times to hundreds of times a day. Juvenile myoclonic epilepsy (Janz syndrome) is characterized by myoclonic jerks of the arm after awakening, but can also include some generalized tonic-clonic seizures and absence seizures. It begins between 8 and 18 years of age, and there is a strong family history of seizures. Sleep deprivation, hyperventilation, photosensitivity, and alcohol can trigger a seizure. Lennox–Gastaut syndrome has its onset at 1 to 8 years of age and consists of multiple seizure types. These children often have seizures every day, and there is an associated deterioration in intelligence, as well as behavior disorders.
3. A. Breath holding spell. Cyanotic breath-holding spells usually occur after a crying episode, and result in limpness and loss of consciousness, occasionally with posturing. While this could be an afebrile seizure, the rapid return to normal level of consciousness makes this less likely. Without a documented fever, this cannot be a febrile seizure. A tic is a movement disorder without loss of consciousness. A pseudoseizure tends to be a diagnosis of exclusion, but the patient often avoids painful stimuli and returns to a normal level of consciousness after the seizure. It would be uncommon for a 2-year-old to have pseudoseizures.
4. C. Open the airway. Initial therapy consists of meticulous attention to maintaining patency of the airway and adequacy of oxygenation and ventilation. Oral suctioning may be required, so this should be available. High-flow oxygen (15 L) is administered to all patients via non-rebreather mask or bag-mask ventilation. Venous access is secured as soon as possible. Treatment of the seizure via the rectal route, or IV route (once secured) can follow. Intubation is usually not necessary.
5. C. Bedside Glucose. A bedside glucose check should be performed on all patients to detect hypoglycemia. Other laboratory studies are based on the type of seizure, history, and likely etiologies, and should be individualized. A lumbar puncture should be performed in any patient suspected of having a central nervous system infection. For most patients there is little use for a CT scan or MRI. Neuroimaging should be reserved for those with a postictal focal deficit (Todd's paralysis) that does not quickly resolve, a child whose level of consciousness remains decreased, or who has not returned to baseline mental status within several hours.

6. A. Parental reassurance. Parental reassurance and education regarding the benign nature of febrile seizures, the low risk of recurrence, and the low incidence of subsequent epilepsy are part of the discharge instructions. Patients who have had a febrile seizure may be discharged, with follow-up by their primary care provider, unless an underlying infection precludes discharge. If a bacterial infection is the etiology of the fever, it is treated with appropriate antibiotics. There is no need for a CBC, urinalysis, CT or lumbar puncture based on history and physical examination.
7. D. Phenobarbital. Phenobarbital (20 mg/kg intravenously) is the drug of choice for neonatal seizures, with phenytoin (20 mg/kg) the second choice. In refractory seizures, pyridoxine (100 mg intravenously) is indicated to treat the potential for pyridoxine-dependent seizures. If the infant is hypoglycemic (<40 mg/dL), 2 mL/kg of D₁₀ W is administered intravenously, followed by an infusion of D₁₀ W. If a metabolic abnormality such as hypocalcemia (<7 mg/dL) is found, it should be corrected, but hypomagnesemia may be made worse by giving calcium. Prostaglandin A is the treatment for a cyanotic infant with a ductal dependent lesion of the heart.
8. B. Outpatient EEG. An EEG is the study of choice in the evaluation of childhood afebrile seizures, including absence seizures, but it should be performed a few days to weeks after the seizure. The EEG helps to confirm if the event was a seizure, classify the seizure type or epilepsy syndrome, predicts recurrence, and guides therapy. While hyperventilation may prompt a seizure, it cannot truly be diagnosed unless EEG recordings are characteristic for absence seizures. A CT of the head will not diagnose the seizure. If the concern is syncope versus seizure, ECG may be helpful. Diazepam for home use is not appropriate in this case.
9. E. Fosphenytoin. Fosphenytoin is the next drug to be given during status epilepticus. Diazepam is another benzodiazepine, and is unlikely to terminate the seizure if lorazepam did not work. Phenobarbital is usually a third line drug. Propofol and valproic acid are used for refractory status.
10. C. Carbamazepine. Carbamazepine (Tegretol) is the only one of these drugs in which a therapeutic level can be obtained in the emergency department. Gabapentin, levetiracetam, topiramate, and carnitine levels are unavailable for management of this patient.

8

CHEST PAIN

Wendy C. Matsuno

Chest pain is a worrisome symptom that often causes parents to bring their child to the emergency department (ED) for evaluation. The rate of pediatric patients presenting to the ED with a complaint of chest pain is 3 to 6 for every 1000 patient visits.

In the majority of cases, the etiology of chest pain is benign, but symptoms are distressing enough to cause 27% to 30% of children to miss school.

CLINICAL PRESENTATION

- Average age of presentation is 10 to 12 years.
- Equal distribution between sexes.
- Younger children usually have a cardiorespiratory source, whereas adolescent patients usually have chest pain of psychogenic origin.
- Patients that present with a complaint of chronic chest pain (>6 months duration) usually have idiopathic or psychogenic chest pain.

DIFFERENTIAL DIAGNOSIS

CARDIAC

- A cardiac cause is found in 4% to 5% of cases presenting to the ED for evaluation of chest pain.
- Myocardial infarction is rare in the pediatric population.
- These patients usually present with the classic severe, substernal chest pain with radiation to the left arm or jaw.
- Patients are at greater risk for myocardial ischemia if they have a history of congenital heart disease, acquired heart disease (eg, Kawasaki disease), or drug abuse (eg, cocaine).
- Pericarditis usually presents with sharp, substernal chest pain, which may be alleviated by leaning forward. On physical examination, the patient classically has distant heart sounds, a friction rub, and signs of congestive heart failure.
- Myocarditis patients often have vague symptoms including chest pain, dyspnea, dizziness, nausea, vomiting, and fatigue. Physical examination usually reveals a gallop, signs of congestive heart failure, and tachycardia unresponsive to fluids.
- Hypertrophic cardiomyopathy patients usually give a history of increased chest pain with exertion.

- Aortic stenosis, pulmonary stenosis, abnormal coronary arteries, and mitral valve prolapse, if severe, can lead to ischemia of the heart and papillary muscles.
- Ventricular tachycardia can present as a fleeting, sharp pain, or palpations.
- Supraventricular tachycardia (SVT) is usually described as a rapid heartbeat.

RESPIRATORY

- Pediatric chest pain attributed to a pulmonary etiology was found in 12.5% to 19% of cases.
- An asthma exacerbation can cause chest tightness and pain.
- Bronchospasm and persistent coughing can lead to excessive use of the chest wall muscles and result in chest pain.
- Pneumonia can present with fever, tachypnea, and upper respiratory symptoms. Physical examination may reveal decreased breath sounds or rales.
- Acute pain and subsequent respiratory distress could be a spontaneous pneumothorax or pneumomediastinum.
 - Patients with asthma, Marfan's syndrome, or cystic fibrosis are at increased risk.
 - Physical examination may reveal decreased breath sounds and crepitus.
- Pulmonary embolism is rare in pediatrics, but should be considered in patients with dyspnea, pleuritic chest pain, hemoptysis, and low-grade fever.
 - Risk factors for a pulmonary embolism are the use of birth control pills, recent abortion, prolonged immobility, inherited hypercoagulable disorders, indwelling central lines, and major trauma, particularly to the lower extremities.

GASTROINTESTINAL

- Gastrointestinal causes make up 3% to 4% of ED pediatric chest pain visits.
- Gastroesophageal reflux disease causes a burning, substernal type of pain due to resulting gastritis and esophagitis. Epigastric tenderness and the association of pain with eating food are suggestive of a gastrointestinal origin.
- A foreign body lodged in the esophagus can cause chest pain.

MUSCULOSKELETAL

- A musculoskeletal etiology is found in 32.5% to 43% of visits to the ED for chest pain.

- Trauma can cause fractures and contusions that may result in chest pain.
- Overuse or overexertion of the chest wall muscles may cause muscle strain.
- Costochondritis has reproducible chest pain elicited by palpating the costochondral joints. The etiology of costochondritis is unknown, but it is considered to be a benign, inflammatory condition.
- Tietze's syndrome, also occurs at the costochondral junctions, but has the associated findings of swelling, redness, and warmth. This is a self-limited inflammatory condition.
- Slipping rib syndrome usually occurs at false or floating ribs with a sharp, intermittent pain that lasts a few minutes and settles to a dull ache. The pain is thought to result from the anterior end of the rib, slipping out of place and aggravating the adjacent intercostal nerve. The "hooking maneuver," where the patient is instructed to lie on the unaffected side and the practitioner reaches under the lower costal margin and pulls the rib anteriorly, results in the reproduction of the patient's pain and a click sensation.
- Precordial catch syndrome, or Texidor's twinge, is a benign condition that causes a brief, sharp pain to the left chest without radiation. The etiology is unclear, but is thought to occur from the parietal pleura, intercostal nerves, or from the stretching of the supporting ligaments of the heart.

PSYCHOGENIC

- A psychogenic source accounts for 5% to 9% of ED chest pain visits.
- Pediatric patients experiencing anxiety, depression, or stress can have symptoms manifesting as chest pain.

IDIOPATHIC

- Idiopathic chest pain is diagnosed in 12% to 45% of chest pain cases seen in the ED.

DIAGNOSTIC EVALUATION

- Chest x-rays can reveal a cardiac process (eg, congestive heart failure), a pulmonary etiology (eg, pneumonia, pneumothorax, hemothorax, pneumomediastinum), a musculoskeletal cause (eg, fracture), or a gastrointestinal finding (eg, foreign body).

- Electrocardiograms (ECGs) can help to diagnose conduction abnormalities, structural defects, myocardial, and pericardial disease.
 - Common ECG findings with childhood myocardial infarction include Q waves greater than 35 ms, ST segment elevation greater than or equal to 2 mm, and prolonged calculated QT interval greater than 440 ms.
- CK can be released by skeletal muscle as well as heart muscle, thus a more specific CK-MB fraction is a better indicator of myocardial damage.
- Troponin I is found only in the myocardium and can be detected within 3 to 6 hours after the onset of ischemia.
- Troponin T is also found in the cardiac myocytes and can be present in the serum within 3 hours.
- Troponin I and T are more sensitive and specific than CK and CK-MB, and are the preferred method of detecting myocardial injury.

MANAGEMENT

- Appropriate therapy should be initiated in the ED dependent upon the underlying cause, with specialty consultation and referrals as needed.
- Cardiac causes of chest pain generally require cardiology consultation.
- Pulmonary causes such as asthma and pneumonia should be treated with standard medications.
- Pneumothorax and hemothorax may require emergent intervention with needle decompression or chest tube placement, but would depend on the size of the lesion and patient status.
- Patients with a pulmonary embolism require admission and anticoagulation therapy similar to adults (see Chapter 52)
- Patients with gastroesophageal reflux disease can have medication therapy initiated and subsequent follow-up with either a gastroenterologist or pediatrician.
- Patients with an esophageal foreign body will usually be able to pass it through the gastrointestinal tract, but if concerned because of the size or other factors, a consultation to remove the foreign body can be made (see Chapter 74).
- Musculoskeletal causes of chest pain can generally be treated with rest and nonsteroidal antiinflammatory agents.
- The slipping rib syndrome can be treated with education and avoidance of the offending movements. Local nerve blocks, corticosteroid injections and surgery are sometimes necessary.

- Precordial catch syndrome is a self-limiting condition that requires only education and supportive care.
- Psychogenic causes of chest pain usually require outpatient follow-up by the patient's pediatrician or psychiatrist unless the anxiety, depression, or stress is severe.
- In cases of idiopathic chest pain, no specific therapy is needed, but follow-up is essential.

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QUESTIONS

1. A 12-year-old boy is brought to the ED for evaluation of chest pain. The mother reports that he recently started an after school activity program. He has no significant medical history and there have been no sick contacts. Which of the following is true regarding his chest pain?
 - A. Males are more often affected than females
 - B. The usual age of presentation is 2–5 years of age

- C. A gastrointestinal etiology is the most common
- D. Chest pain is a chief complaint for 3 to 6 of every 1000 pediatric patient visits
- E. Patients with chronic chest pain usually have a cardiac etiology
2. A 7-year old girl is brought to the ED by her father for chest pain. You learn that the child has history of “heart problems” as a child. Dad is unable to recall the exact diagnosis. Which of the following conditions would place this patient at a higher risk for a more benign etiology for her chest pain?
- A. Supraventricular Tachycardia
- B. Cocaine use
- C. Kawasaki’s disease
- D. Congenital heart disease
- E. Tietze’s syndrome
3. A 15-year-old star basketball athlete is brought to ED. During the championship finals, he began complaining of chest pain. You are told he has a history of cardiomyopathy. You suspect he may be having a myocardial infarction. Which of the following would be the most sensitive and specific test to detect myocardial injury?
- A. Creatine kinase
- B. CK-MB
- C. Troponin
- D. Lactate
- E. Hematocrit
4. You suspect your pediatric patient is having a myocardial infarction. In reviewing the electrocardiogram, which of the following ECG findings would you expect to see to confirm your suspicion?
- A. Q waves less than 35 ms
- B. ST segment elevation greater than or equal to 2 mm
- C. Shortened QT interval
- D. Absence of a p wave
- E. Presence of a delta wave
5. A 12-year-old female presents with chest pain and difficulty breathing. She has a history of asthma, but has not used her inhaler for the last 2 years. There is no history of foreign body ingestion or choking episodes. There is no history of fever, but she has had a runny nose and cough. On examination, there are no wheezes, but she has decreased breath sounds, tachypnea, and oxygen saturations of 92% on room air. What would be the next step in management?
- A. Bronchodilator
- B. Chest imaging
- C. EKG
- D. Troponin
- E. Echocardiogram
6. A 14-year-old male presents to the ED with chest pain. On examination, he is found to have reproducible chest pain elicited by palpating the costochondral joints. What is the likely diagnosis?
- A. Pneumothorax
- B. Pneumonia
- C. Costochondritis
- D. gastroesophageal reflux
- E. myocarditis
7. An 11-year-old female was at band practice when she experienced the acute onset of right sided chest pain. She denies trauma. On exam, she is nontoxic, has symmetric chest rise, breath sounds slightly decreased on the right, trachea is midline. What would be the next best course of action?
- A. CBC
- B. EKG
- C. Bronchodilator
- D. Chest x-ray
- E. Antibiotics

ANSWERS

1. D. Chest pain is a chief complaint for 3 to 6 for every 1000 patient visits. Chest pain has an equal distribution between genders. Patients are usually 10 to 12 years of age. Gastrointestinal causes for chest pain are found in 3–4 % of pediatric patients with chest pain. Patients with chronic chest pain usually have a psychogenic or idiopathic etiology.
2. E. Tietze’s syndrome is an inflammatory condition of the costochondral junctions, and not related to heart disease. All other causes put the patient at risk for myocardial ischemia.
3. C. Troponin. Troponin I and T are more sensitive than CK and CK-MB to detect myocardial injury. Levels are usually elevated in 3–6 hours after injury. The other tests would not be specific for myocardial injury.
4. B. ST segment elevation greater than or equal to 2 mm. Common ECG findings with childhood myocardial infarction include Q waves greater than 35 ms, ST segment elevation greater than or equal to 2 mm, and prolonged calculated QT interval greater than 440 ms. The absence of a p wave is seen in SVT. Presence of a delta wave is seen in WPW.

5. A. Bronchodilator. Patient has a history of asthma, and is likely suffering from an asthma exacerbation. She has no wheezes, but decreased breath sounds. A chest x-ray would be reasonable, but would not be the next step in management with his current respiratory distress. EKG, troponin and echocardiogram would likely not assist the management of this patient.
 6. C. Costochondritis. Costochondritis is a benign condition and patients have reproducible pain when the costochondral joints are palpated. Pneumothorax, pneumonia, gastroesophageal reflux, and myocarditis do not classically have this finding.
 7. D. Chest x-ray. The patient likely has a spontaneous pneumothorax. She is stable so immediate needle decompression or chest tube placement is not needed and a chest film is reasonable to evaluate the extent of the pneumothorax. A cbc, ekg, bronchodilator, and antibiotics would not be appropriate.
 - Distal complete obstruction—dilated loops of bowel and air-fluid levels.
 - Incomplete obstruction—gas pattern may appear normal.
 - Ultrasound may reveal “whirlpool” sign, bowel wall edema, or intraluminal fluid.
- Upper gastrointestinal series preferred
 - Descending duodenum obstruction just over the right of the spine is pathognomonic.
 - Intestine distal to obstruction may have “corkscrew” appearance.
 - Treatment
 - Intestinal intubation, decompression, fluid resuscitation, occasional blood replacement, and antibiotics if toxic. Laparotomy is mandatory.

9 ACUTE ABDOMINAL CONDITIONS

Jonathan Singer

THE OBSTRUCTIONS

MALROTATION WITH MIDGUT VOLVULUS

- Occurs due to arrest of embryonic rotation of intestinal tract.
- Midgut volvulus includes duodenum, small bowel and colon up to midtransverse portion
- Symptoms are vague
 - Failure to thrive, chronic recurrent abdominal distension, pain-free episodic vomiting, persistent unexplained diarrhea.
 - Bilious vomiting, abdominal discomfort or episodic crying, failure to pass stools or constipated stools common; gangrenous bowel is rare.
- Stool may be positive for occult blood.
- Males are affected twice as often as females.
- Nearly half present within first week of life, two-thirds within first month, and >90% within first year of life.
- 90% infants appear well initially but with time will become ill appearing.
- Plain radiographs may be normal or exhibit obstructive pattern
 - Duodenal obstruction—double bubble sign.

PYLORIC STENOSIS

- Most common cause of obstruction after first month of life.
 - Range is 1 week-3 months of age; usual is 2–6 weeks of life.
- Males more common than females; first born males most common.
- Presents as nonbilious, postprandial projectile vomiting; anorexia is absent.
- Early on infant appears well, later presents with dehydration.
- Palpable “olive” mass is seen in 25–50% of cases.
- Ultrasound is preferred study—may see antral nipple sign.
- Laboratory may reveal hypokalemia, hypochloremia, and metabolic alkalosis
- Treatment
 - Correct dehydration, acidosis, and electrolyte imbalance
 - Surgical intervention

INTUSSUSCEPTION

- Most frequent cause of intestinal obstruction between 3 months and 5 years old
 - >60% occur in first year of life with most between 5 and 9 months of age
 - Males more common
- Symptoms: colicky pain, vomiting, blood stools
 - Sudden onset severe abdominal pain for several minutes followed by period of normal appearance
 - Vomiting and loose stools or diarrhea may occur
 - Variable amounts of blood in stool
 - “current jelly” stools appear late and are not common

- Anorexia is usual
- Apathy or listlessness, mental status change may occur
- Abdominal exam is variable with guarding or distention uncommon
 - Sausage shaped mass may be found in any location.
 - Stools may be grossly bloody or positive only for occult blood.
- Plain abdominal radiographs recommended
 - Rule out contraindications for contrast enema and to review findings of obstruction.
 - Films may be normal up to 30% of time.
- Ultrasonography is useful
 - Findings may be large sonographic target, bull's eye or doughnut sign, and a sleeve or pseudokidney sign.
- Spiral CT may be useful in equivocal cases
- Treatment
 - NPO, intravenous fluid hydration, electrolyte replacement, Nasogastric tube
 - Surgical consultation with radiologist consultation mandatory
 - Nonsurgical air insufflation or barium enema may be recommended
 - Operative therapy is needed if nonsurgical failure occurs

INCARCERATED HERNIAS

- It is important that all *children must be fully undressed* for exam.
- Hernia types—most common is umbilical followed by inguinal and scrotal regions, midline epigastrium and lateral border of rectus sheath.
- More common in males.
- Greatest risk for incarceration during first 6 months of life.
- Hallmark sign—asymptomatic bulge that becomes more prominent with increased abdominal pressure.
- Irritability is first symptom of incarceration in infant; older children may have cramping abdominal pain that does not necessarily localize to hernia site.
- Nonbilious vomiting may become bilious or even feculent over time.
- Radiographs rarely needed; ultrasonography may be useful in premature infants.
- Nonoperative reduction of a strangulated hernia can be attempted by emergency physician. If unsuccessful, surgical consultation mandated.

INTRAABDOMINAL SEPSIS

ACUTE APPENDICITIS WITHOUT PERFORATION

- Most commonly seen in late elementary school age children; <2% in children under 2 years of age.
- Classically presents first as abdominal pain then nonbilious vomiting, low-grade fever.
- Abdominal pain starts in epigastric or periumbilical area and then localizes to right lower quadrant at McBurney's point over 1–12 hours.
- Dysuria and diarrhea may occur and may lead to misdiagnosis.
- Iliopsoas and/or obturator sign may be positive if appendix is retrocecal.
- CBC is useful if diagnosis is less secure; usual wbc >15,000 cells/mm³.
- Radiographs generally not useful.
- Ultrasound may be useful—target lesion and thickened appendix seen.
- CT abdomen may be useful if diagnosis is uncertain.
- Early surgical consultation should be obtained.

ACUTE APPENDICITIS WITH PERFORATION

- High likelihood of perforation increased in children <2 years of age because symptoms may be vague and misleading.
- Rapid progression to perforation within 6–12 hours from onset of symptoms.
- Pain may lessen or cease once perforation occurs.
- Older children may have vague abdominal symptoms for days to weeks.
- Acute perforation often appear acutely ill with tachycardia, high fever, rebound, guarding.
- 75% have elevated white blood count count—often with bandemia.
- Radiographs, ultrasound or CT may be helpful.
- Surgical consultation is mandatory.

SPONTANEOUS PERITONITIS

- Most commonly associated conditions in descending order:
 - perforated appendix
 - intestinal obstruction
 - incarcerated hernia
 - inflammatory bowel disease
 - Hirschsprung's disease
 - posttraumatic (including instrumentation and foreign body)

- spontaneously ruptured viscus (including Meckel’s, bile duct, colon, ileum)
- necrotizing enterocolitis (NEC)
- Increased risk in children with VP shunt, immunodeficiency, ascites, or nephrosis
- More often in females, 5–10 years of age
- Acute to insidious onset, poorly differentiated pain
 - Occurs over hours to days followed by nonbilious vomiting, diarrhea and fever
- Appearance—ill with fever, tachycardia. May have grunting respirations.
- Abdomen is distended, tender, and guarded with rebound.
- Radiographs often reveal distended large and small intestines with air fluid levels.
- Paracentesis may prevent exploratory laparotomy; however, if diagnosis is unclear, laparotomy is necessary.
- Abdominal exam may reveal soft, nontender fecal mass in left lower quadrant
- Rectal exam may reveal empty vault with “squirt”—explosive release of stool when finger is withdrawn
- Progression to enterocolitis occurs more commonly with newborns
 - Sudden abdominal distention, generalized abdominal discomfort and explosive diarrhea that rapidly becomes bloody
 - Fever, dehydration and altered mental status common
 - May lead to colonic perforation, peritonitis, and gram-negative septicemia
- Abdominal radiographs may be normal or show dilated colon
- Barium enema may be confirmatory

NECROTIZING ENTEROCOLITIS

- Most common in premature who have undergone blood loss, transient hypotension, and birth asphyxia or who required central line.
- May be self-limited or progress to a fatal disease.
- Anorexia, abdominal distention, nonbilious vomiting, or diarrhea.
- Hemochezia common—grossly bloody stools or guaiac positive seedy stool.
- Infants often septic appearing.
- Radiographs may reveal bowel distention or intraluminal air (pneumatosis intestinalis).
 - If radiographs are ambiguous, barium enema may be helpful.
- Treatment
 - NPO, parenteral nutrition, Nasogastric tube, parenteral, and intraluminal antibiotics
 - Surgical consult—surgery is withheld in the absence of perforation, obvious peritonitis, and gangrenous bowel.

HIRSCHSPRUNG’S DISEASE

- Characterized by absence of intramural ganglion cells usually in rectosigmoid area
- More common in males than females
- Presentation variable
 - Newborns may have delayed passage of first meconium stool
 - If undiagnosed may have increased fecal retention, obstipation, constipation
 - Poor appetite and extended periods of failure to thrive are common

GASTROINTESTINAL FOREIGN BODIES

- Size, configuration, consistency, or chemistry of ingested object is important
 - Small <15–20 mm round oval or cuboid without sharp edges—rare problem
 - Rigid, elongated, slender objects—increased complications
 - Single magnet not problematic but multiple magnets may cause pressure necrosis or bowel perforation
 - Bezoars from repeated ingestion of hair or vegetable matter can lead to obstruction
 - Ingested batteries are serious and must be removed
- Children at risk <1 year of age or have underlying congenital, anastomotic, or inflammatory disease of mediastinal structures.
- Obstruction, perforation, peritonitis, or abscess may be immediate or delayed for months.
 - Prolonged esophageal foreign bodies may cause airway obstruction, mediastinitis, and erosion into major vessels.
 - Obstructions often occur at hypopharynx, thoracic inlet, or cardioesophageal junction.
 - Hypopharyngeal—persistent gagging and pooling of oral secretions, superior neck pain or inability to swallow or speak.
 - Aortic arch—localize pain to area of sternal notch; also dysphagia and drooling but lack dysphonia.
 - Distal esophagus- vague chest discomfort, dysphagia and odynophagia.
 - Lower intestinal tract—intermittent abdominal pain +/- vomiting.

- Radiograph of chest and abdomen or handheld metal detector useful for locating metallic foreign body.
- Single coin lodged for <24 hours in esophagus—high likelihood to pass.
 - Treatment—NPO, IV hydration and observe × 12 hours; may need sedation
 - Balloon extraction and esophagoscopy sometimes employed
- Corrosive batteries or pointed/elongated objects must be *immediately* removed
- Parents should be advised at discharge of potential for delayed complications

MEGACOLON

- Occurs in late childhood or early adolescence.
- Caused by ulcerative colitis or Crohn's disease often seen as persistent diarrhea followed by the appearance of mucus and bloody stools.
- Results from inflammation of transverse colon leading to dilatation, hemorrhage, peritonitis, and overwhelming sepsis.
- May progress over hours to days as fever, malaise, anorexia, abdominal pain, and distension, grossly bloody stools and lethargy.
- Radiographs reveal transverse colon dilatation of >6–7 cm in diameter; free air seen with perforation.
- CT scan of abdomen diagnostic.
- Treatment:
 - IV fluid resuscitation; may need albumin or blood transfusion
 - High dose steroids
 - Surgical consultation
 - Nasogastric tube
 - Parenteral antibiotics

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QUESTIONS

1. 8-month-old healthy male is brought by parents to emergency department (ED) for constipation and excessive crying. Parents relate a single episode of green colored emesis which occurred 1 hour prior to presentation. Stools are reportedly normal in appearance. At your exam, the infant is afebrile, consolable, and well appearing. His abdomen is soft and nontender to exam. Plain radiographs of the abdomen are ordered and air fluid levels in a dilated stomach and duodenum with no gas noted in the remainder of the bowel. Which of the below represents the most appropriate management:
 - A. Nasogastric tube insertion followed IV hydration, antibiotics and admit for observation.
 - B. A barium/air enema should be performed to attempt reduction of obstruction.
 - C. Immediate surgical consultation for prompt laparotomy should be arranged.
 - D. Discharge home with parents and instructions to administer a vegetable based laxative.
 - E. Discharge home with dietary instructions of bananas, applesauce, toast, and arrange for follow-up with pediatrician in 24 hours.
2. 6-week-old healthy male is brought by parents to ED for a complaint of vomiting intermittently × 3 days. Parents state child occasionally vomits milk forcefully after eating. On examination the child is well appearing, has a social smile and is eagerly taking his bottle. Physical exam reveals an afebrile infant in no distress, abdomen is soft and nondistended with a small round mass palpable in the epigastrium, capillary refill is 3 seconds. Your recommendation to the parents for further care would be
 - A. Discharge home with instructions to continue feeds and follow-up with pediatrician next week.
 - B. Obtain ultrasound of abdomen.
 - C. Nasogastric tube insertion followed by IV hydration, antibiotics, and admission for observation.

- D. Admit for immediate laparotomy.
E. Obtain CT scan to identify mass and obtain diagnosis.
3. A 3-year-old is brought by parents for vomiting and abdominal pain, which is intermittent in nature. The parents have also noted three loose stools. Physical exam reveals a listless afebrile child who currently denies abdominal pain. The abdominal exam is soft and nontender. Rectal examination is strongly positive for occult blood. The next appropriate management step should be
- Begin oral hydration using the WHO formulation and discharge home if child appearance improves and tolerates oral liquids.
 - Obtain stool cultures and begin antibiotics and discharge home.
 - Obtain plain radiographs and if normal, discharge home after IV fluid hydration.
 - Perform ultrasonography and evaluate for a target or bull's eye on transverse or cross section of intestines.
 - Discharge home with instructions to collect stool for culture.
4. A 6-month-old presents with colicky abdominal pain which results in bouts of inconsolable crying followed by periods of playfulness (Fig. 9-1). Physical exam reveals a happy playful child. Abdominal exam is soft with a tender sausage-shaped mass in the right lower quadrant. The parents also relate two episodes of vomiting. Plain films are done which reveal air fluid levels but no free air. Your next step in the evaluation should be
- Insert a nasogastric tube, begin IV hydration, and admit for observation.
 - Consult surgery for an immediate laparotomy for appendicitis.
 - Consult surgery and discuss with radiologist whether an air/barium enema should be employed.
 - Obtain a CBC and urinalysis. Give oral liquid challenge to child. If lab work is normal and oral liquids are well tolerated, discharge home with specific instructions to return should child resume vomiting or abdominal pain returns.
 - Discharge home with laxative and instructions to follow-up with pediatrician if not improved in 24 hours.
5. A 2-month-old male from a full term delivery, is brought to the ED for inconsolable crying and refusal to feed for six hours. On examination, the child is afebrile but irritable. The abdominal exam reveals a soft abdomen with a 2 cm mass noted in the right scrotum. While in the ED, the child vomits a bilious colored emesis. Further ED evaluation to be considered should be:
- Keep child NPO, apply ice to the mass, and following sedation attempt to relocate the mass using continuous bimanual pressure.
 - Obtain immediate CT scan.
 - Consult radiology for barium enema.
 - Administer antibiotics, perform lumbar puncture, and admit.
 - Obtain a testicular ultrasound and consult urology.
6. 4-year-old female is brought to the ED for a 1-day history of nonbilious vomiting and abdominal pain and temperature of 38°C. On examination, the

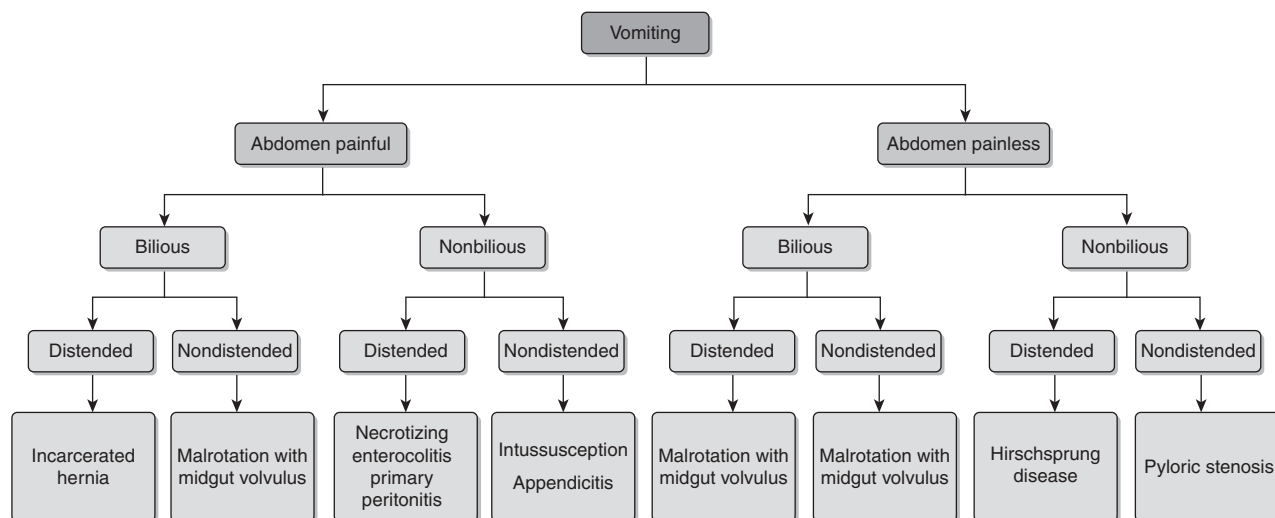


FIG. 9-1. Approach to selected abdominal emergencies in childhood.

abdomen is soft, flat with guarding, and rebound tenderness in the right lower quadrant. Further workup should include which of the following mandatory tests:

- A. CBC
 - B. Air/barium enema
 - C. CT abdomen with contrast
 - D. Plain radiographs of the abdomen
 - E. Surgical consultation
7. A 10-day-old female, delivered at 36 weeks, is brought to the ED by her parents with a complaint of poor feeding, nonbilious vomiting, and diarrhea. On exam you find an afebrile, pale, septic appearing neonate with a distended abdomen, hypoactive bowel sounds. Rectal exam reveals grossly bloody stools. Which of the following radiographic findings are expected?
- A. Normal gas pattern.
 - B. Ultrasonography detailing a bull's eye or target lesion.
 - C. Fecalith in the right lower quadrant.
 - D. Plain radiographs of the abdomen reveal bowel distention with intraluminal air.
 - E. CT abdomen reveals a right lower quadrant sausage shaped mass.
8. A 1-year-old infant is brought to the ED for chronic constipation and sporadic abdominal distention. Parents relate that they have unsuccessfully tried many remedies and laxatives to relieve the constipation and have seen numerous physicians in an attempt to find a cure. A diagnosis of Hirschsprung's disease is suspected. Which of the following symptom is supportive of this diagnosis?
- A. The child's underwear is stained by stool
 - B. The abdomen is tender and tensely distended
 - C. Insertion of a finger in the rectum may result in an explosive "squirt" of stool
 - D. Rectal exam reveals a dilated empty vault
 - E. The disease is most common in females
9. An 11-month-old female is brought to the ED for a possible "shiny" foreign body ingestion. The parents state that they saw the child put something in their mouth. On examination, you find a well appearing child in no distress and without stridor or drooling. Your next action should be
- A. CT scan of the neck.
 - B. Reassure the parents, discharge home, and ask the parents to evaluate the fecal contents for the foreign object and to return if not found or child becomes ill.
 - C. Arrange for an immediate esophagoscopy.

- D. Obtain a anteroposterior film of chest and abdomen.
- E. Administer glucagon and observe for foreign body passage to stomach.

10. Parents bring a 4-year-old female to the ED for evaluation of abdominal pain. On examination, you find a child who appears lethargic with a significant distended abdomen. Rectal examination reveals a grossly bloody stool. Which of the following findings would support a diagnosis of toxic megacolon?
- A. Radiographic findings of a dilatation of the transverse colon to 6 cm
 - B. Chronic constipation with explosive nonbloody diarrhea
 - C. Examination of the abdomen reveals soft, mobile masses in the left lower quadrant
 - D. Bilious vomiting without diarrhea
 - E. CT scan reveals a dilated appendix

ANSWERS

1. C. Bilious emesis is a concern for malrotation, which is highly suspected following the plain radiographs which demonstrate a "double bubble" sign. Barium/air enema reductions are often attempted as nonoperative treatment of intussusception. Children with bilious emesis should not be discharged home without surgical consultation.
2. B. The initial symptom of pyloric stenosis is unpredictable infrequent projectile vomiting which is nonbilious and postprandial. Unless significantly electrolyte and volume depleted, infants will appear healthy with a good suck, and if fed, will swallow without difficulty. An epigastric, rounded mass, described as an "olive" is found in a quarter to a half of all cases. Ultrasound allows for direct visualization and measurement of the pylorus muscle to obtain the diagnosis. Treatment is IV fluid hydration, nasogastric tube insertion, and surgical consultation for pyloromyotomy. Antibiotics are not needed. CT scan is not recommended due to radiation risk.
3. D. Intussusception classically presents with colicky abdominal pain, vomiting and bloody stools. The abdominal pain is described as intermittent and the child may appear well between episodes of pain. At the same time as the vomiting, the child may experience several bowel movements, which vary from formed stools to thin liquid. The presence of blood in the stool may be trace to grossly bloody. Often the child will present with mental status changes,

which may vary from apathy or listlessness to obtundation. Plain radiographs may be normal up to 30% of the time but may also show localized air-fluid levels, dilated small bowel loops, reduced intestinal air, minimal fecal content in the colon or inability to visualize the liver tip or mass lesion. Sonographic findings of intussusception may include a large sonographic target, bull's eye or doughnut sign on the transverse or cross section of the colon, and a sleeve or pseudokidney sign on the longitudinal section.

4. C. Nonoperative attempts to reduce an intussusception may be performed if no contraindications such as perforated bowel exist or if child is toxic in appearance. If pneumatic or hydrostatic pressure techniques fail to reduce the intussusception, an additional repeat effort at nonoperative reduction may be done. Should reduction techniques fail, or if child toxic in appearance, operative therapy is performed. Abdominal pain with intussusception is described as severe, colicky, and intermittent followed by periods of quiescence. A tender sausage shaped mass may be palpable in any quadrant.
5. A. Nonoperative reduction of a strangulated hernia can be attempted by the emergency physician. Greatest success follows a period of NPO, application of ice to the hernia sac and sedation followed by continuous bimanual pressure. Surgical consultation is mandated for suspected strangulation or unsuccessful manual reduction. The diagnosis of incarcerated hernia is not difficult if the child is completely undressed. All children with incarceration appear uncomfortable. Radiography confirmation is rarely necessary.
6. E. A complete blood count is unnecessary in the patient with obvious appendicitis. Flat plate and upright abdominal radiographs are suggestive but not pathognomic for appendicitis. Conventional CT scan utilizing intravenous and oral contrast may be used for patients in whom the diagnosis of appendicitis is uncertain but the risk of radiation makes this a test to use on a selective basis. Barium/air enema is used to diagnose and reduce intussusception. For patients with classic symptoms of appendicitis—fever, right lower quadrant pain and vomiting, surgery consultation is mandatory.
7. D. Necrotizing enterocolitis (NEC) is characterized by gastric dilatation, functional ileus and erosive mucosal injury. Premature infants who have sustained multiple stresses to their cardiovascular system such as acute blood loss, transient hypotension, and birth asphyxia or who require central vascular instrumentation are at increased risk. Affected infants are often pale and septic appearing. Abdominal distention may be generalized or a single dilated colonic segment may occur. Bowel sounds are diminished. Rectal examination reveals grossly bloody stool or seedy stool that is guaiac positive. On an abdominal flat plate, bowel distention is the most common finding. Intraluminal air (pneumatosis intestinalis) may be limited to scattered colonic segments or be generalized. Ultrasonography in NEC has only limited benefit. It may be used to detect and track the passage of air through the portal vein system. The finding of a bull's eye or target lesion identified by ultrasonography is associated with intussusception. A sausage shaped mass in the abdomen may be palpated in any quadrant of the abdomen with intussusception. Fecaliths in the right lower quadrant may be seen with acute appendicitis.
8. C. Patients with Hirschsprung's disease may present with increasing fecal retention, obstipation, constipation and sporadic abdominal distention. Parents frustrated by repeated therapeutic failures for chronic constipation may bring the child to the ED for a second opinion. The abdomen may have mild to moderate distention but is usually soft and nontender. The child's underwear is not soiled from overflow and the rectal exam reveals an empty vault that is not dilated. An examining finger may cause an explosive "squirt" upon withdrawal. The disease is four time more common in males than females.
9. D. It is important to do a thorough evaluation for the foreign body and attempt to locate the size, shape, and type of object ingested. Of concern is the ingestion of batteries, magnets, and coins. The primary mode to locate a metallic foreign body is the AP film of chest and abdomen. A handheld metal detector may also be used. CT scan is of benefit only if the patient has a perforated organ. Even a single retained battery can lead to corrosive damage and tragedy. Esophagoscopy or balloon extraction must be employed if the child has a battery with corrosive potential regardless of level point of the esophageal impaction.
10. A. The hallmark radiologic feature of toxic megacolon seen on a supine abdominal film is the dilatation of the transverse colon to greater than 6–7 cm. In the majority of cases, the children have persistent diarrhea followed by the appearance of bloody stools with mucus. The abdomen is generally distended and tender and vomiting will be nonbilious. A CT scan is diagnostic revealing the distended transverse colon.

10 VOMITING, DIARRHEA, AND GASTROENTERITIS

William R. Ahrens

- Vomiting is the forceful expulsion of the contents of the stomach. Diarrhea is defined as frequent loose or liquid bowel movements. By far the most common cause of vomiting and/or diarrhea is acute infectious gastroenteritis.
- The history regarding the patient who is vomiting focuses on the duration of the illness, the frequency of the vomiting, the character/color of the contents vomited, and associated abdominal pain.
- In infants of 2 to 6 weeks of age with multiple episodes of vomiting pyloric stenosis should be considered.
- Bilious vomiting is rare in infants and children, and always raises the possibility of a malrotation, with or without a volvulus.
- Vomiting associated with persistent, severe, or localized abdominal pain suggests peritonitis.
- Vomiting accompanied by a headache raises the possibility of increased intracranial pressure.
- The history regarding diarrhea focuses on the duration of the problem, the frequency of the stools, and the characteristics of the stool; the presence of blood or mucus in the feces increases the possibility of a bacterial or inflammatory illness.
- Frank rectal bleeding suggests an anatomic lesion, such as an intussusception, Meckel's diverticulum, or juvenile polyps.
- Protracted diarrhea suggests a malabsorption syndrome or inflammatory bowel disease.
- Perhaps the most important aspect of the physical examination of the patient with vomiting and diarrhea is mental status.
- A happy, playful infant or child is unlikely to have a life-threatening problem. A patient who is lethargic is either significantly dehydrated or is at high risk for a significant metabolic or anatomic lesion.
- Vomiting and altered mental status can occur secondary to increased intracranial pressure; associated physical findings include splitting of the cranial sutures and/or a bulging anterior fontanelle.
- Metabolic abnormalities due to inborn errors of metabolism, diabetic ketoacidosis, uremia, and hyper or hyponatremia can also present with vomiting and altered mental status. Inborn errors of metabolism usually present in infants.
- A rare cause of vomiting and altered mental status is Reye's syndrome. A child with intussusception can present with profound lethargy, usually associated with a history of vomiting.

TABLE 10-1 Life-Threatening Causes of Vomiting

Increased intracranial pressure
Bowel obstruction (malrotation, intussusception, incarcerated hernia, adhesions)
Inborn errors of metabolism
Diabetic ketoacidosis
Toxic ingestions
Reye's syndrome

TABLE 10-2 Life-Threatening Causes of Diarrhea

Toxic megacolon (Hirschsprung disease, ulcerative colitis)
<i>E. coli</i> 0157 (hemolytic uremic syndrome)
Pseudomembranous enterocolitis (antibiotic-associated diarrhea)

- Patients with a history of diarrhea who present with altered mental status are at risk for toxic megacolon which can occur secondary to Hirschsprung disease. Physical examination may reveal palpable stool throughout the abdomen and an empty rectal vault.
- Profound diarrhea, in which rapid losses of fluid and electrolytes occur, can cause altered mental status secondary to shock.
- Diarrheal illness secondary to shigella can be associated with altered mental status.
- Bloody diarrhea can be caused by infection with *Escherichia coli* 0157; up to 10% of affected children will develop potentially life-threatening hemolytic uremic syndrome. Tables 10-1 and 10-2 list some life-threatening causes of vomiting and diarrhea.

GASTROENTERITIS

ETIOLOGY

- Rotavirus remains the most common cause of infectious diarrhea in children worldwide.
- Other viral causes of acute infectious gastroenteritis include adenovirus serotypes 40 and 41, noroviruses (Norwalk-like viruses) virus, calcivirus, and astrovirus. Noroviruses are responsible for a large number of food-borne outbreaks. Rotavirus and adenovirus predominantly infect children younger than 3 years of age.
- In the developing world, bacterial infections account for a high percentage of diarrheal illnesses. Enterotoxigenic bacteria include enterotoxigenic *E. coli*, *Clostridium perfringens*, Cholera species, and *Vibrio* species; diarrhea is caused by the secretion of toxin. Enteroinvasive bacteria include enteroinvasive and enterohemorrhagic *E. coli*, *Shigella* species,

Salmonella species, Campylobacter, Yersinia species, and Pleosiomonas species; diarrhea is associated with inflammatory changes in the small and/or large bowel.

- *Clostridium difficile*, an antibiotic-associated diarrhea, is toxin mediated. Although a fairly high percentage of infants are colonized with *Clostridium difficile*, it rarely causes disease. Noninvasive parasites that cause diarrhea include Giardia species and Cryptosporidium species. Giardia is a pathogen associated with children attending day care; it can cause acute and chronic diarrhea. An invasive parasitic cause of diarrhea is *Entameba histolytica*.

PATHOPHYSIOLOGY

- Viruses tend to directly damage the small intestinal villi, causing malabsorption of carbohydrates, which result in watery osmotic diarrhea.
- Bacteria produce diarrhea by a variety of mechanisms. Enterotoxigenic *E coli* infects the small intestine and causes noninvasive watery diarrhea via two major toxins. Enteropathogenic *E coli* are an important cause of diarrhea in infants in developing countries, especially diarrhea lasting more than 2 weeks.
- Shigella infection is associated with seizures in some children.
- Salmonella enteritis is a common cause of self-limited, noninvasive diarrhea. Infants younger than 1 year of age, patients with sickle cell disease, and immunocompromised patients are at risk for bacteremia from *S enteritis*. *Salmonella typhi* and *paratyphi* are capable of causing serious blood-borne disease, especially in infants and children.
- Campylobacter are the most common cause of bacterial gastroenteritis in developed countries. Infection causes inflammatory enteritis; diarrhea can be bloody. Campylobacter infection is associated with the Guillan–Barre syndrome.

DIAGNOSTIC EVALUATION

PHYSICAL ASSESSMENT

- All children with significant vomiting and diarrhea will have a deficit of fluids and electrolytes. How “sick” the patient is will depend not only on the fluid deficit, but also on the rate at which it occurred. Fluid losses that occur slowly are compensated for by water in the intracellular and interstitial spaces that shifts into the intravascular space.
- Fulminant losses from severe diarrhea are poorly compensated for, especially in infants. The degree

of dehydration is notoriously difficult to assess in younger infants.

- There are a variety of established, teachable, and reproducible methods for estimating the extent to which a patient is dehydrated. Currently, the emphasis is on simplifying the assessment. As it can be difficult to distinguish between mild and moderate dehydration, the two can be grouped together. Important findings indicating the presence of significant dehydration are delayed capillary refill, abnormal skin turgor, and an abnormal respiratory pattern. Other important findings include tachycardia, dry mucus membranes, and the absence of tears. The importance of assessing mental status cannot be overstated. Guidelines regarding the assessment and treatment of diarrhea in children from the World Health Organization (WHO) and UNICEF for physicians and nonphysician health care providers are available on the Internet.

DIAGNOSTIC STUDIES

- If moderate or severe dehydration is present it is reasonable to evaluate serum electrolytes. Most patients with significant diarrhea will have low serum bicarbonate.
- Hypoglycemia can complicate gastroenteritis. In patients with bloody diarrhea or protracted diarrhea, a stool sample for culture and ova/parasites should be sent. Fecal leukocytes suggest an enteroinvasive infection.
- For infants, patients with sickle cell disease, and patients who are immunocompromised in whom salmonella is suspected a blood culture is indicated.
- Shigella infection is associated with bacteremia.
- Rotavirus antigen can be detected in the stool via enzyme immunoassay and latex agglutination.

TREATMENT

ORAL REHYDRATION THERAPY

- Oral rehydration therapy (ORT) is an effective treatment for the vast majority of patients with gastroenteritis-related dehydration. The mechanism crucial to the efficacy of ORT is the coupled transport of sodium and glucose molecules at the intestinal brush border accompanied by free water.
- There are a variety of oral hydration solutions (ORS) available; current literature supports the use of formulas containing between 65–75 mg/L of sodium and 75–90 mmol/L of glucose. In the vast majority of

cases, rehydration can be accomplished without risk of causing hypo or hypernatremia.

- In infants and children with minimal dehydration, treatment is directed at maintaining nutrition and preventing dehydration. Fluid intake can be generally increased, or ORS can be administered at a volume of 10 mL/kg for each stool, or 2 mL/kg for each emesis. Diet should not be restricted.
- Patients with mild-to-moderate dehydration should receive 50 to 100 mL/kg of ORS over 2 to 4 hours. Ongoing losses are also replaced. Nasogastric administration of ORS is safe and effective. Moderately dehydrated patients who require intravenous rehydration can be rehydrated over several hours in the emergency department.
- Patients in shock or with severe dehydration require intravenous therapy. An initial bolus of 20 mL/kg of 0.9 NS or Lactated Ringers is rapidly administered; it is repeated until adequate perfusion is restored. Once perfusion is restored, hydration can be continued intravenously or ORT initiated. Intravenous therapy, if utilized, consists of replacement of the estimated volume deficit along with maintenance fluid therapy. If ongoing fluid losses are severe, they need to be added to the therapy.

TABLE 10-3 Assessment of Dehydration

Two of the following signs: Lethargic or unconscious Sunken eyes Not able to drink or drinking poorly Skin pinch goes back very slowly	Severe dehydration
Two of the following signs: Restless, irritable Sunken eyes Drinks eagerly, thirsty Skin pinch goes back slowly	Some dehydration
Not enough signs to classify as some or severe dehydration	No dehydration

ADJUNCTIVE THERAPY

- Ondansetron can facilitate oral therapy in children with persistent vomiting and decrease the number of children who require intravenous salvage therapy.
- Zinc supplementation reduces the severity and duration of diarrhea in children younger than 5 years. It also reduces the incidence of subsequent diarrheal illness. The WHO recommends zinc supplementation (10–20 mg/d for 10–14 days) in children with diarrhea.

TABLE 10-4 Treatment of Common Gastrointestinal Infections

CAUSE	ANTIBIOTIC	ALTERNATIVE TREATMENT	COMMENT
Cholera	Adults: Docycycline Tetracycline Children: Tetracycline	Erythromycin	
Shigella dysentery	Adults and children: Ciprofloxacin Levofloxacin	Trimethoprim/sulfamethoxazole Ampicillin Azithromycin Ceftriaxone Pivmecillinam	Treatment recommended for patients with dysentery; increasing resistance to commonly used antibiotics
Giardiasis	Adults and children: Metronidazole	Tinidazole Albendazole Mebendazole	Albendazole and mebendazole effective in children, also treat other parasitic infections
Amebiasis (dysentery)	Adults and children: Metronidazole	Tindazole	
Campylobacter	Adults and children: Azithromycin	Erythromycin Ciprofloxacin	Most cases self-limited and do not require therapy; treatment in severe cases shortens duration of symptoms
Enterotoxigenic <i>E. coli</i>	Levofloxacin Ciprofloxacin	Azithromycin Trimethowrim/sulfamethoxazole	Treatment is empiric; most cases self-limited
Salmonella (non-typhoid)	Ciprofloxacin	Azithromycin Trimethoprim/sulfamethoxazole Ampicillin Cefotaxime Ceftriaxone	Treatment recommended for patients at risk for invasive disease: infants younger than 3 mo of age, patients with sickle cell disease, patients with HIV or other forms of immunosuppression
<i>E. coli</i> 0157	none		Treatment with antibiotics may trigger or exacerbate hemolytic uremic syndrome (controversial)

- It is vitally important to maintain the nutritional status of children with diarrhea, especially those who are malnourished. Breast-feeding should be continued, as should an age-appropriate diet. There is insufficient evidence to support withholding lactose-containing formulas in patients with acute diarrhea. Liquids containing high sugar loads, such as colas and apple juice, are best avoided.
- Antibiotic therapy is not indicated in the majority of cases of diarrheal illnesses, and is becoming increasingly complicated by the rapid development of resistance to commonly used medications. Ideally, therapy is directed by culture and sensitivity. (Tables 10-3 and 10-4)

PREVENTION

- Prevention of gastroenteritis fundamentally depends on improving hygiene. In the developing world, access to potable water is crucial. Access to latrines is also an important public health goal in developing nations.
- Two vaccines against rotavirus have been developed and are currently in use. Salmonella typhi, which is capable of causing fatal disease, especially in young children, is especially prevalent in Asia. Two vaccines for Salmonella typhi—one oral and the other injectable—are available for children older than 2 years of age. Oral cholera vaccines have been developed that are effective in reducing incidence and severity of disease. Vaccines against shigella, campylobacter, and enterotoxigenic *E coli* are in various stages of development.

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QUESTIONS

1. A 4-month-old male presents with a one-day history of vomiting; there is no history of diarrhea. On arrival, the baby is lethargic, afebrile, with normal vital signs. This implies
 - A. The need for emergent aggressive volume resuscitation
 - B. A severe electrolyte imbalance
 - C. A high likelihood of an inborn error of metabolism
 - D. The possibility of increased intracranial pressure
2. A 2-week-old infant on cow's milk formula presents with a history of occasional vomiting, which on detailed questioning appears to be bilious. The baby is afebrile and the physical examination is normal. Appropriate intervention would be
 - A. Reassure the parents the baby has benign regurgitation and discharge home.
 - B. Order an upper gastrointestinal study.
 - C. Check serum electrolytes and if they are normal discharge home.
 - D. Order an obstructive series and if it is negative discharge home.
3. A 4-year-old girl presents with fever and lethargy. The history reveals 2 days of bloody diarrhea. Her abdominal exam is unremarkable; rectal exam reveals heme positive stool. The most likely finding is
 - A. A toxic ingestion
 - B. An intussusception diagnosed by barium emema
 - C. Enteroviral meningitis
 - D. Fecal leukocytes
4. While on a medical mission to Africa, you encounter an obviously malnourished 9-month-old girl with a 3-week history of nonbloody nonmucoid diarrhea. Her mother has tried three courses of antibiotic—she cannot recall what kind. The baby

- appears well hydrated. The best course of management would be
- Stop breast feeding and place the baby on clear liquids for a week.
 - Continue regular feeding and place the baby on a course of zinc.
 - Empiric therapy with high dose antibiotics.
 - Do nothing as no diagnostic testing is available.
5. A robust 5-month-old male presents with 2 days of vomiting and diarrhea. On physical examination, the baby appears to be moderately dehydrated. No one can start an IV. Further management to consider would be
- Start an intraosseous line
 - A nasogastric tube is contraindicated in moderate dehydration
 - There is no role for ondansetron in moderate dehydration
 - ORT has a high likelihood of success
5. D. In a robust baby with moderate dehydration an intraosseous line is very unlikely to be necessary. A nasogastric tube is an acceptable and safe way to administer oral rehydration therapy, which is likely to succeed even in moderately dehydrated patients; ondansetron is likely to be beneficial in this situation.

11 FEEDING DISORDERS

William R. Ahrens

HIGH-YIELD FACTS

COMMON FEEDING PROBLEMS IN INFANTS

- For the human infant, the first year of life, and especially the first 6 months, is a period of explosive growth. Sustaining normal weight gain and development require virtually continuous intake of a large amount of calories. For the first 4 months, this occurs almost exclusively via the intake of liquid, either as breast milk or formula. After 4 months, solid foods are gradually introduced. This chapter will focus on two feeding problems especially common in younger infants: GER and formula intolerance.

GASTROESOPHAGEAL REFLUX

- GER occurs when there is retrograde flow of gastric contents into the esophagus. Some degree of GER is so common in infants that it can be considered a normal variant in the first year of life. It is thought to be due to immaturity of lower esophageal sphincter function that results in transient lower esophageal sphincter relaxations. Less commonly, GER can be secondary to motility disorders or gastric outlet obstruction.
- Clinically, gastrointestinal reflux presents as regurgitation or vomiting of breast milk or formula. The vast majority of babies vomit during the first week of life; 60% to 70% suffer from some element GER at age 3 to 4 months. Emesis can be forceful, or of the mild variety often referred to as “spitting up.” The emesis usually occurs after feeding; it should never be bilious or bloody.
- Most cases of GER are physiologic, which implies that there is no associated underlying pathology,

ANSWERS

- D. A 1-day history of vomiting is unlikely to cause dehydration sufficient to cause mental status changes or a severe electrolyte imbalance. An inborn error of metabolism is possible, they are very rare. The combination of vomiting and lethargy must always call to mind the possibility of increased intracranial pressure; in this case aggressive fluid resuscitation could provoke cerebral herniation. This is a classical presentation of the shaken-impact syndrome.
- B. Bilious vomiting in an infant must always prompt consideration of the possibility of a malrotation, with the attendant risk of a midgut volvulus. Benign regurgitation does not cause bilious emesis. Electrolytes are unhelpful in diagnosing a malrotation, and an obstructive series can be normal. An upper gastrointestinal study is the diagnostic study of choice.
- D. A toxic ingestion is unlikely given the history. An intussusception is possible, but the fever suggests an infectious etiology. Meningitis is unlikely given the history of bloody diarrhea. Shigellosis is a classic cause of fever, lethargy, and bloody diarrhea. Fecal leukocytes are highly likely, as is a significant bacteremia.
- B. It is inadvisable to withdraw regular feeding from an already malnourished baby with diarrhea. With no diagnostic testing available, it would be impossible to select an appropriate antibiotic, especially for a baby who has already received multiple courses of medication. Zinc has been shown to be beneficial in malnourished infants with diarrheal diseases.

growth and development are normal, and there are no resultant medical complications. Physiologic reflux usually resolves by approximately 1 year of age.

- Pathologic GER is associated with multiple complications, including poor weight gain or failure to thrive, esophagitis, and respiratory complications, including reactive airway disease and recurrent pneumonia, thought to be secondary aspiration. There may be a relationship between GER and acute life-threatening events.
- Risk factors for GER include prematurity and underlying neurologic disease. CMPA may also be implicated as a cause of GER.
- The history of the infant suspected of having GER includes determining the amount and frequency of feeding, especially in formula-fed infants, as well as noting whether the formula is cow's milk or soy based. Stooling pattern is evaluated, especially for the presence of bloody diarrhea that could indicate a formula allergy.
- Infants with pathologic or symptomatic GER often have a history of excessive crying or irritability, poor appetite, and "arching" of their back that may be related to pain secondary to erosive esophagitis.
- The physical examination in the infant with GER is most likely to be unremarkable, except in the patient with an underlying neurologic disorder or with severe GER that results in failure to thrive.
- No laboratory studies available in the ED are diagnostic. An upper GI barium study may be useful in the child with a history of repetitive vomiting, since it may reveal esophageal dysmotility or anatomic lesions that cause vomiting in young infants, but the test is not sensitive for reflux. A 24-pH probe study is currently the most sensitive test to evaluate the presence of GER.

TREATMENT

- The vast majority of infants with physiologic GER respond to very conservative measures. These include smaller more frequent feeding, positioning the baby upright after feeding, and thickening the formula with cereal.
- Formula can be thickened by adding one tablespoon (15 mL) of dry rice cereal per ounce of formula. Premixed thickened formulas are also commercially available (Table 11-1).

TABLE 11-1 Thickened Formulas

United States: Enfamil AR LIPIL

Canada: Enfamil A+ Thickened with rice starch

- Prone positioning is not recommended in infants younger than 6 months due to a possible increase in the risk of sudden infant death syndrome.
- Patients with GER thought to be secondary to CMPA may improve if switched to an extensively hydrolyzed or amino acid formula. Medical therapy is predominantly directed at reducing secretion of gastric acid and reducing gastric emptying time. Medications used include H2 receptor blockers such as ranitidine and nizatidine and proton pump inhibitors such as omeprazole and lansoprazole.
- A small percentage of infants, especially those with neurologic disorders, will not improve with medical therapy, and are at risk for long-term complications of GER, especially involving the lungs. The majority of these patients will respond well to surgical intervention; the standard operation to correct GER is the Nissan Fundoplication.

FORMULA INTOLERANCE AND HYPERSENSITIVITY

- Formula intolerance is a vague term that can generally be described as any adverse reaction to ingested formula. It can be allergic or nonallergic in nature—an example of nonallergy-mediated intolerance is lactose intolerance.
- Approximately 5% to 15% of infants are in some way intolerant of cow's milk. Of these, between 2% and 7.5% have CMPA.
- Parents often change from one formula to another based on the belief that the infant is experiencing some difficulty or another. Common reasons given for changing formula were colic, "spitting up," and diarrhea or constipation. In some cases, tolerance may be associated with relatively subtle differences in the composition of two formulas.
- Food allergy affects approximately 6% to 8% of infants. It is more common in children with severe atopic dermatitis. In infants, the most common food allergens are cow's milk and soy protein. Many infants with CMPA are also soy protein intolerant.
- Breast-feeding reduces the incidence of CMPA; however, breast-fed babies can develop CMPA from antigens secreted in breast milk.
- Allergic symptoms can be IgE or non-IgE mediated. Manifestations of IgE sensitivity usually develop shortly after ingesting the offending allergen and include urticaria, angioedema, and wheezing; in its most severe form, an IgE-mediated reaction can result in anaphylaxis.
- Non-IgE-mediated hypersensitivity often presents in infants between 1 week and 3 months of age.

Vomiting and diarrhea are common complaints; bloody stools are not uncommon. Poor weight gain and failure to thrive may be noted. GER can occur.

- Specific diagnoses include allergic proctocolitis, enterocolitis, food protein–induced proctocolitis, and food protein-induced enteropathy syndromes.
- In the ED, diagnosis of formula intolerance, allergy, or specifically CMPA depends mainly on a careful history and high level of clinical suspicion.
- In infants in whom formula intolerance or allergy is suspected, the potentially offending formula is withdrawn; if symptoms resolve, the patient can be rechallenged with the formula—return of symptoms is considered diagnostic.
- The ED management of infants with suspected formula intolerance/allergy will depend on the severity of symptoms and, in the case of a likely allergy, the type of reaction involved.

If an anaphylactic reaction is suspected, the offending formula should be immediately stopped and the patient referred to an allergist; consideration should be given to prescribing an Epi-Pen. Infants with lactose intolerance should be placed on a lactose free formula.

- Current recommendations are that infants with CMPA be changed to an extensively hydrolyzed or amino acid based formula; soy formulas are avoided because of cross-sensitization with cow’s milk protein. Some infants will tolerate an extensively hydrolyzed formula. Infants who do not tolerate a hydrolyzed formula should be placed on an amino acid formula. Those infants with CMPA who have symptoms such as severe eczema, reflux esophagitis, or failure to thrive may benefit from an immediate introduction to an amino acid formula without a preceding trial of extensively hydrolyzed formula (Table 11-2).
- Breast-feeding mothers whose infants develop CMPA are advised to avoid consuming milk-based products.
- The natural history of CMPA is such that most affected infants will eventually tolerate cow’s milk.

TABLE 11-2 Lactose-Free, Extensively Hydrolyzed, and Amino Acid Formulas

Lactose-free formulas
United States: Enfamil LactoFree Lipil, Isomil DF, Similac
Lactose-free advance
Canada: Enfalac LactoFree, Similac LF
Extensively hydrolyzed formulas
United States: Nutramigin LIPIL, Pregestimil, Alimentum
Advance
Canada: Nutramigin, Pregestimil, Alimentum
Amino acid formula
Neocate

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QUESTIONS

1. A 4-month-old infant is brought to the emergency department (ED) by her parents. The parents are concerned that she is “spitting up” after feedings and is at risk of becoming dehydrated. You learn that the infant was full-term born by cesarean section with no complications. The infant has a normal vitals, is well appearing with a normal examination, The parents present you with their Google search. You suspect that the infant has gastroesophageal reflux. Which of the following statement is correct?
 - A. Is rare in full term infants
 - B. Often results in bilious emesis
 - C. Virtually always requires medication or surgery
 - D. Usually responds to conservative measures
2. A 5-month-old boy is brought to the ED for persistent diarrhea. The mother states the infant was recently switched from cow’s mild formula to a soy-based formula. Which of the following is a true statement regarding Cow’s milk protein allergy (CMPA)?
 - A. Never causes vomiting
 - B. Invariably results in anaphylaxis
 - C. Can cause bloody diarrhea
 - D. Is virtually unheard of in breast fed infants
3. You strongly suspect CMPA in a 4-month-old infant girl with colic and diarrhea. The best management strategy for this patient would include which of the following:
 - A. Can continue to consume cow’s milk formula but should be started on an antihistamine
 - B. Should be changed to a soy based formula
 - C. Should be started on an extensively hydrolyzed or amino acid based formula
 - D. Are doomed to a lifetime of no ice cream

4. A 11-month-old is brought to the ED for recurrent vomiting. You review the medical record and note that the child has had numerous visits throughout the year for similar complaints. The child has had poor weight gain and appeared malnourished. Following an ED evaluation, you decide to admit the child. Which of the following statement are true regarding pathologic Gastroesophageal reflux?
- Can cause significant pulmonary problems
 - Rarely responds to surgical treatment
 - Rarely affects overall weight gain
 - Is not associated with underlying neurologic disease

ANSWERS

- D. GER is extremely common in full term infants. It does not cause bilious emesis, which should prompt consideration of an anatomical lesion. GER usually responds to conservative treatment.
- C. CMPA can cause both vomiting and bloody diarrhea. Anaphylaxis is uncommon, but can occur. CMPA can occur in breast fed infants whose mothers consume cow's milk products.
- C. Cow's milk formula should be stopped in infants with suspected CMPA. Soy formulas are avoided because of a high incidence of cross sensitivity. An extensively hydrolyzed or amino acid based formula is recommended. Most infants "outgrow" CMPA, so fortunately the little darlings will eventually get their ice cream.
- A. Discussion: Pathologic GERD often causes pulmonary problems, such as reactive airway disease and recurrent aspiration pneumonia. GER is associated with neurologic disorders, such as cerebral palsy, and often causes failure to thrive. It usually responds to surgical correction (Nissen fundoplication).

12 JAUNDICE

Anjali Singh
William R. Ahrens

HIGH-YIELD FACTS

INTRODUCTION

- Jaundice is a yellowish-green discoloration of the skin and sclera caused by hyperbilirubinemia. It is apparent when the serum bilirubin reaches 5 mg/dl.

In newborns, some degree of hyperbilirubinemia is virtually universal.

- Although sequelae from hyperbilirubinemia are relatively rare, unconjugated bilirubin is potentially toxic to the neonatal central nervous system.
- Conjugated hyperbilirubinemia, although not neurotoxic, is often a marker for serious underlying disease.

PATHOPHYSIOLOGY

- Bilirubin is largely formed by the destruction of red blood cells and the catabolism of heme proteins. Bilirubin is transported to the liver, where it undergoes enzymatic-mediated conversion from an insoluble unconjugated form to a water-soluble conjugate. The conjugating enzyme is markedly diminished in the newborn infant.
- The insoluble form of bilirubin is indirect reacting, the water-soluble form direct reacting.
- After conjugation, bilirubin is excreted in the bile and from there into the intestinal tract. In the intestinal tract, some of the conjugated bilirubin is reabsorbed into the enterohepatic circulation.
- In the newborn, elevated unconjugated hyperbilirubinemia is potentially neurotoxic. Bilirubin enters the brain if it is unbound to albumin, unconjugated, or if the blood-brain barrier has been disrupted. The concentration of bilirubin in the brain and the duration of exposure are important determinants of neurotoxicity.
- The newborn is especially vulnerable to hyperbilirubinemia due to increased hemolysis secondary to shortened red blood cell survival time and/or hemolysis from fetal-maternal blood group incompatibility. Impaired hepatic up-take and inadequately developed enzymes delay its conjugation, and increased enterohepatic circulation results in inefficient excretion.
- Hyperbilirubinemia is especially common in newborns and young infants, and it is helpful to consider this age group separately.

UNCONJUGATED HYPERBILIRUBINEMIA

NEWBORNS AND YOUNG INFANTS

DIFFERENTIAL DIAGNOSIS

Physiologic Jaundice

- The most common cause of unconjugated hyperbilirubinemia in the neonatal period is physiologic jaundice. It is thought to result primarily from a

six-fold increase in bilirubin load, and a marked deficiency in enzyme activity. In addition, the hepatic uptake and excretion of bilirubin is transiently impaired.

- Physiologic jaundice becomes visible on the second to third day of life and peaks around the fourth day. The maximum elevation is usually <6 mg/dL. In premature infants, jaundice both peaks and resolves somewhat later, and peak levels can reach 12 mg/dL. Physiologic jaundice is a nonpathologic condition, with no neurologic sequelae.

BREAST MILK JAUNDICE

- In general, jaundice is more common in breast-fed infants than in bottle-fed infants. Early-onset jaundice is referred to as breastfeeding jaundice, which is akin to a relative starvation state, putting infants at risk for dehydration and increased enterohepatic reuptake of bilirubin. Effective early lactation is the key to its prevention. Breastmilk jaundice typically occurs after the first 3 to 5 days of life, and can persist for several weeks to a few months. Breast milk jaundice may result from substances contained in breast milk that antagonize the conjugation and excretion of bilirubin. Rarely, breast-fed infants can develop elevations of unconjugated bilirubin starting in the first week of life that can reach 15 to 27 mg per 100 mL by the second or third week. Hyperbilirubinemia resolves with the cessation of breast-feeding and does not recur when it is resumed.
- Interruption of breast-feeding is reserved for infants with bilirubin levels that place the newborn at risk for kernicterus. It is important to note that although 50% of breastfed infants develop jaundice, less than 1% develop bilirubin levels that are of concern.

INCREASED HEMOLYSIS

- Increased hemolysis in newborn infants is the most common cause of hyperbilirubinemia severe enough to warrant phototherapy or exchange transfusion. It is usually secondary to maternal-fetal blood group incompatibility in either rhesus or ABO antigens. Jaundice usually appears in the first 24 hours of life.
- Other causes of hemolysis include hereditary spherocytosis and elliptocytosis, and G6PD deficiency. Severe bruising or cephalohematoma secondary to trauma during delivery can also result in increased metabolism of heme proteins and unconjugated hyperbilirubinemia.

MISCELLANEOUS

- Unconjugated hyperbilirubinemia can result from a variety of unusual causes. These include hypothyroidism, Down's syndrome, polycythemia, and pyloric stenosis or other high intestinal obstructions.
- Bacterial infections, including those from the urinary tract, can cause unconjugated hyperbilirubinemia, although there may also be a component of conjugated bilirubin. It is generally believed that neonatal sepsis causes hyperbilirubinemia by either hemolysis as a result of oxidative stress or bacterial endotoxins reducing bile flow.

SEQUELAE

- In the immediate neonatal period, unconjugated hyperbilirubinemia is of great concern largely because of its association with kernicterus, an irreversible neurologic disorder that occurs when unbound bilirubin is deposited in the central nervous system, especially the basal ganglia and various brainstem nuclei.
- Unconjugated bilirubin has the potential to penetrate the nervous system and cause either transient bilirubin encephalopathy or the permanent sequelae of kernicterus. There have been conflicting published reports of the association between bilirubin levels and long-term neurocognitive outcomes, with most data from term neonates with bilirubin levels less than 25 mg/dL.

ACUTE BILIRUBIN ENCEPHALOPATHY

- Acute bilirubin encephalopathy is characterized clinically by lethargy that can progress stage-wise to stupor and coma. Changes in the brainstem auditory evoked response have been noted, though the long-term consequences of these are unknown.
- There have been some reports that if emergent exchange transfusion is performed during the intermediate phase, the central nervous system changes may be reversible.

CHRONIC BILIRUBIN ENCEPHALOPATHY (KERNICTERUS)

- In its full-blown form, chronic bilirubin encephalopathy ultimately results in choreoathetosis or athetoid cerebral palsy, auditory dysfunction, dental enamel

dysplasia, paralysis of upward gaze, extrapyramidal signs, and less often mental retardation. In full-term newborns, kernicterus is associated with levels of unconjugated serum bilirubin levels >20 mg/dL. In premature infants, lower levels can cause kernicterus.

EVALUATION AND MANAGEMENT

The evaluation of unconjugated hyperbilirubinemia depends on the specific age at which hyperbilirubinemia is noted, and the rate of rise of bilirubin. The American Academy of Pediatrics subcommittee on hyperbilirubinemia has put forth guidelines for the management of hyperbilirubinemia in infants of more than 35 weeks gestation or more, which largely still serve as the cornerstone of management.

HISTORY AND PHYSICAL

- Special attention should be given to whether the infant has accompanying poor feeding, lethargy, or fever. Sepsis is rare among jaundiced well-appearing infants. Other important considerations are feeding history and weight gain to help assess hydration status.
- In neonates, jaundice progresses in a cephalocaudal direction. Blanching of skin with digital pressure for visual inspection of the skin alone is not an accurate method for estimating the degree of hyperbilirubinemia, especially in darkly pigmented infants. It is also important to look for a cephalohematoma or extensive bruising.

LABORATORY EVALUATION

- Measurement of the serum bilirubin is the standard for accurate diagnosis. Various transcutaneous devices (TcB) are also available to measure bilirubin noninvasively. TcB has been shown to correlate with serum bilirubin levels less than 12–15 mg/dL; however, it is less accurate in darkly pigmented babies, babies of lower gestational age, and babies undergoing phototherapy.
- Laboratory evaluation includes a complete blood cell count, type and screen, blood smear, reticulocyte count, and Coomb's test. If a bacterial infection is a consideration, cultures of blood, cerebrospinal fluid, and urine are obtained, in addition to a urinalysis.

MANAGEMENT APPROACH

- Important considerations are rate of rise of bilirubin and identification of any risk factors. The most important risk factors are breast-feeding, gestational age less than 38 weeks, significant jaundice noted in a sibling, and jaundice noted prior to discharge. Newborns with unconjugated bilirubin >5 to 6 mg/dL after 2 to 3 days of life merit investigation. In addition, further workup is indicated if cord blood bilirubin is 4 mg/dL or greater, increasing at a rate of 0.5 mg/dL over a 4–8 hour period, or increasing greater than 5 mg/dL in the first 24 hours of life.
- Work up is also indicated if for bilirubin greater than 13 to 15 mg/dL in term infants, greater than 10 mg/dL in premature infants, and when jaundice persists beyond 10 days of life in term infants, or 21 days in premature infants. Any infant who is going to need phototherapy requires a workup to identify the cause of jaundice.
- Based on a study of healthy term and near term infants, Bhutani and colleagues established a percentile based predictive nomogram using age-in-hours specific bilirubin levels to determine infants at risk of developing severe hyperbilirubinemia. It is imperative to always interpret bilirubin levels in terms of the infant's age in hours (Fig. 12-1).
- Treatment of hyperbilirubinemia may consist of phototherapy, exchange transfusion or pharmacologic agents. The criteria for initiating phototherapy are not completely clear, but largely depend on precise gestational age and birth weight. A serum bilirubin greater than 25 mg/dl is considered a medical emergency; affected newborns should be admitted directly to the hospital rather than directed to the emergency department, so that treatment is not delayed. Guidelines published by the AAP in 2004 can help direct the decision to begin initiate therapy (Fig. 12-2).
- Phototherapy principally acts by converting bilirubin into isomers, which bypass hepatic conjugation and are excreted without further metabolism. It is the treatment of choice for most babies with moderate to severe elevation of indirect bilirubin. It is recommended that intravenous gamma globulin be administered if the bilirubin continues to rise in spite of intensive phototherapy, especially in infants with hemolytic disease.
- Infants with rapidly rising serum bilirubin who do not respond to phototherapy may require exchange transfusion. Exchange transfusion should be performed in consultation with a neonatologist (Fig. 12-3).

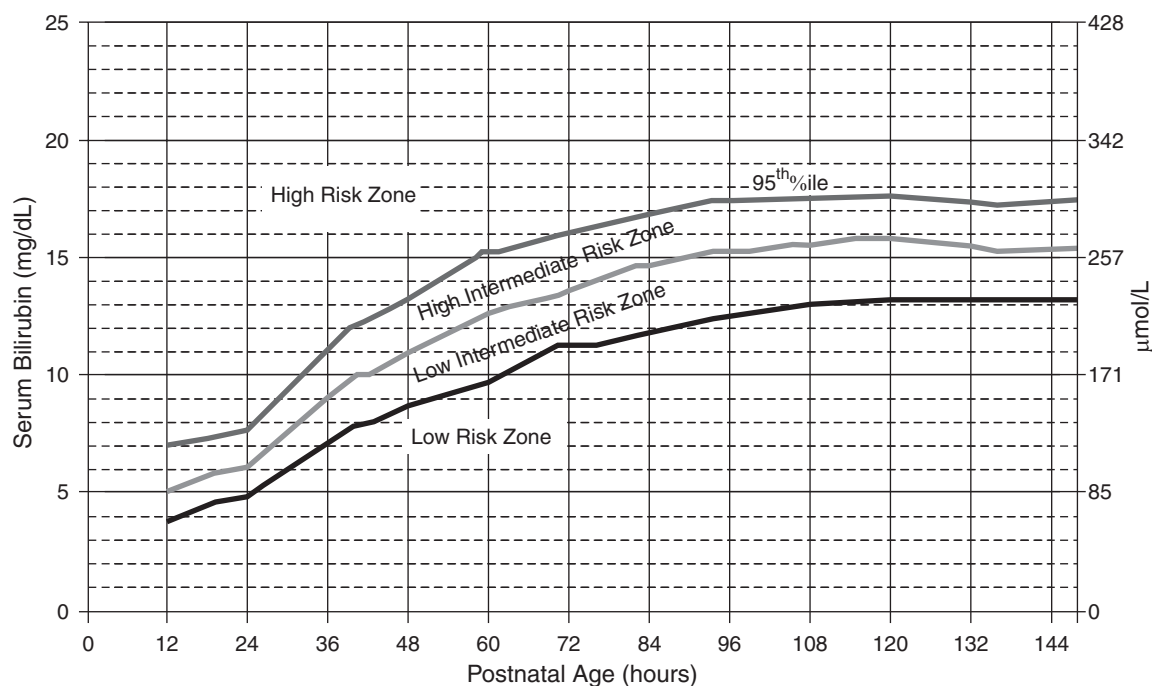


FIG. 12-1. Nomogram of bilirubin serum levels in neonates older than 35-weeks gestation. (Reproduced with permission from American Academy of Pediatrics Subcommittee on Hyperbilirubinemia: Maisels MJ, Baltz RD, Bhutani VK, Newman TB, Palmer H, Rosenfeld W, et al. Clinical practice guideline: management of hyperbilirubinemia in the newborn infant ≥ 35 weeks of gestation. *Pediatrics*. 2004;114:297–316. Copyright 2004 by the AAP.)

OLDER INFANTS AND CHILDREN

DIFFERENTIAL DIAGNOSIS

- In older children, unconjugated hyperbilirubinemia is most likely the result of a hemolytic process or an inherited defect in the conjugation of bilirubin. Hemolytic anemia can be congenital, as in sickle cell disease, thalassemia, hereditary spherocytosis, pyruvate kinase or G6PD deficiency or can be acquired, as in drug-induced hemolysis.
- Gilbert's syndrome results in mild, intermittent unconjugated hyperbilirubinemia resulting from partial deficiency of glucuronyl transferase. Symptoms are usually nonspecific abdominal pain and nausea with an elevated bilirubin up to 5 mg/dL. Crigler Najjar syndrome is characterized by a partial (type II) or complete absence (type I) of glucuronyl transferase, with type II presenting later in childhood and amenable to treatment with Phenobarbital.

HISTORY AND PHYSICAL

- Important considerations are general appearance, hepatomegaly, splenomegaly or other signs of hemolysis.

LABORATORY EVALUATION

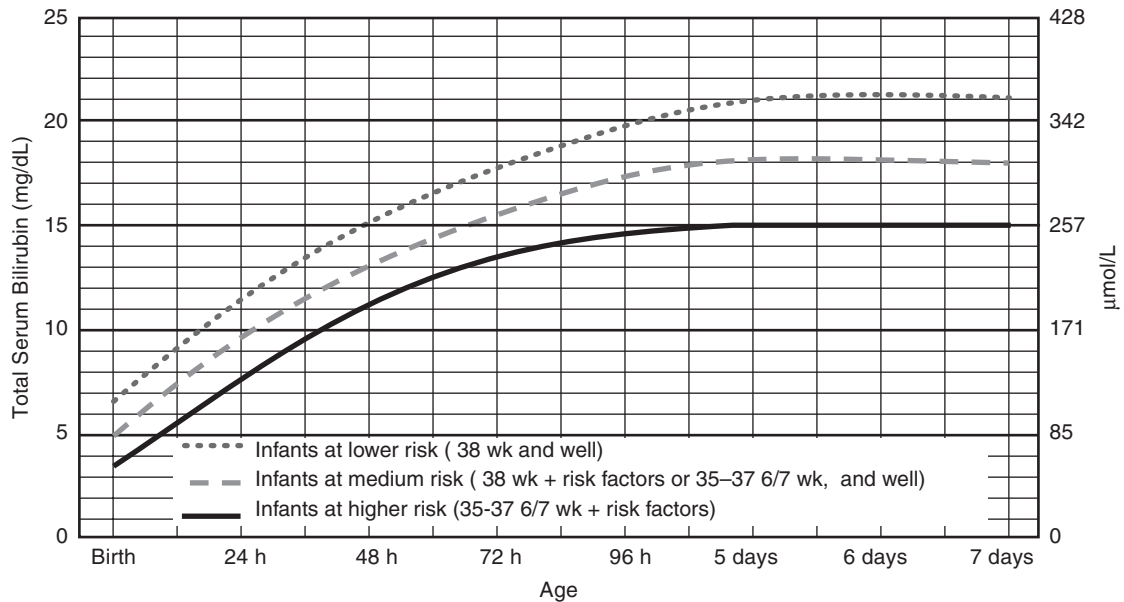
- Workup begins with the serum bilirubin level. The work-up for hemolytic anemia includes a complete blood cell and reticulocyte count and a serum haptoglobin level. If anemia is present, a peripheral smear looking for abnormal cell morphology is warranted. Additional testing includes Coombs test, G6PD level and hemoglobin electrophoresis. Liver function tests will help differentiate hepatic disease from hematologic disease.

MANAGEMENT APPROACH

- Etiology specific treatment is the mainstay of therapy in this age group.

CONJUGATED HYPERBILIRUBINEMIA NEWBORN

- Conjugated hyperbilirubinemia is present when the conjugated fraction of bilirubin exceeds 20% of the total bilirubin, or is greater than 2 mg/dl. It is far less common in newborns and young infants than is



- Use total bilirubin. Do not subtract direct reacting or conjugated bilirubin.
- Risk factors = isoimmune hemolytic disease, G6PD deficiency, asphyxia, significant lethargy, temperature instability, sepsis, acidosis, or albumin < 3.0 g/dL (if measured).
- For well infants 35–37 6/7 wk can adjust TSB levels for intervention around the medium risk line. It is an option to intervene at lower TSB levels for infants closer to 35 wks and at higher TSB levels for those closer to 37 6/7 wk.
- It is an option to provide conventional phototherapy in hospital or at home at TSB levels 2–3 mg/dL (35–50 μmol/L) below those shown but home phototherapy should not be used in any infant with risk factors.

FIG. 12-2. Guidelines for phototherapy in hospitalized infants older than 35-weeks gestational age. (Reproduced with permission from American Academy of Pediatrics Subcommittee on Hyperbilirubinemia: Maisels MJ, Baltz RD, Bhutani VK, Newman TB, Palmer H, Rosenfeld W, et al. Clinical practice guideline: management of hyperbilirubinemia in the newborn infant \geq 35 weeks of gestation. *Pediatrics*. 2004;114:297–316. Copyright 2004 by the AAP.)

unconjugated hyperbilirubinemia. It most commonly occurs secondary to intrahepatic cellular damage; less often, it is due to obstruction of biliary flow. Conjugated hyperbilirubinemia is always pathologic. Most infants with conjugated hyperbilirubinemia will present within the first month of life.

INFECTIOUS CAUSES

- Neonatal cholestasis can occur secondary to hepatic injury from a multitude of infectious causes. Cytomegalovirus, rubella, herpes simplex, varicella, coxsackie, and hepatitis B are common viral etiologies. Syphilis and toxoplasmosis are also implicated. Most of these diseases are present in-utero, and are often associated with congenital anomalies and hepatosplenomegaly.
- Bacterial sepsis can result in conjugated hyperbilirubinemia, although the unconjugated fraction is also usually increased. The urinary tract is a common site of infection, and can involve gram-negative organ-

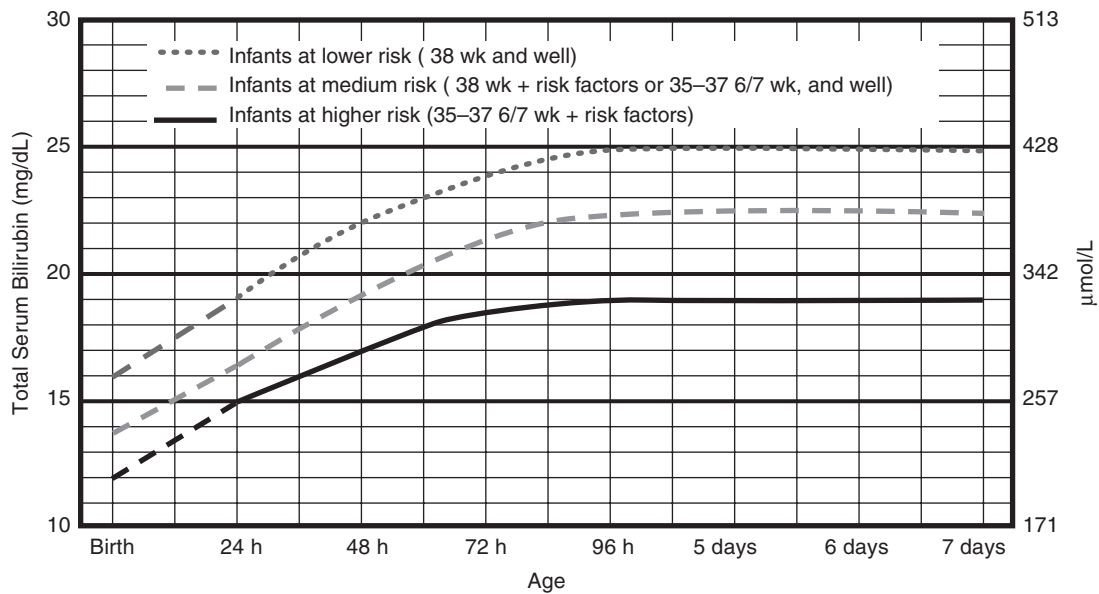
isms such as *Escherichia coli*. Jaundice often starts at 3 to 4 days of age and, in some instances, is the only manifestation of infection.

METABOLIC CAUSES

- Metabolic disorders that can cause conjugated hyperbilirubinemia include alpha₁ antitrypsin deficiency, cystic fibrosis, and galactosemia. Most metabolic disorders will have clinical manifestations other than jaundice. Another hepatocellular cause of neonatal cholestasis is idiopathic neonatal hepatitis, which is characterized by prolonged jaundice and a liver biopsy demonstrating “giant” hepatocytes.

EXTRAHEPATIC DISEASES

- The major extrahepatic cause of conjugated hyperbilirubinemia in infancy is biliary atresia,



- The dashed lines for the first 24 hours indicate uncertainty due to a wide range of clinical circumstances and a range of responses to phototherapy.
- Immediate exchange transfusion is recommended if infant shows signs of acute bilirubin encephalopathy (hypertonia, arching, retrocollis, opisthotonos, fever, high pitched cry) or if TSB is 5 mg/dL (85 µmol/L) above these lines.
- Risk factors—isoimmune hemolytic disease, G6PD deficiency, asphyxia, significant lethargy, temperature instability, sepsis, acidosis.
- Measure serum albumin and calculate B/A ratio (see legend).
- Use total bilirubin. Do not subtract direct reacting or conjugated bilirubin.
- If infant is well and 35–37 6/7 wk (median risk) can individualize TSB levels for exchange based on actual gestational age.

FIG. 12-3. Guidelines for exchange transfusion in hospitalized infants older than 35-weeks gestational age. (Reproduced with permission from American Academy of Pediatrics Subcommittee on Hyperbilirubinemia: Maisels MJ, Baltz RD, Bhutani VK, Newman TB, Palmer H, Rosenfeld W, et al. Clinical practice guideline: management of hyperbilirubinemia in the newborn infant ≥ 35 weeks of gestation. *Pediatrics*. 2004;114:297–316. Copyright 2004 by the AAP.)

a syndrome characterized by absence of the bile ducts anywhere between the duodenum and hepatic ducts. Patients present with jaundice, dark urine, and often with acholic stools. Mild hepatomegaly may be present. Depending on the location of the lesion in the biliary tree, the Kasai procedure, involving surgical anastomosis of the remaining bile ducts to the bowel may be palliative. A major complication of the Kasai procedure is ascending cholangitis.

- Another cause of extra-hepatic biliary obstruction is choledochal cyst, a congenital saccular dilatation of the common bile duct. It can present with jaundice and a right upper quadrant mass, or with symptoms of cholangitis, including fever and leukocytosis. A right upper quadrant ultrasound may facilitate the diagnosis.
- Most patients with conjugated hyperbilirubinemia require referral to a pediatric gastroenterologist for definitive evaluation.

CONJUGATED HYPERBILIRUBINEMIA IN OLDER CHILDREN

- Conjugated hyperbilirubinemia in older children most commonly results from infectious hepatitis. Hepatitis A virus infection is the most common cause of acute jaundice in young children. Infants are typically asymptomatic, while older children exhibit a prodrome of fever and malaise followed by abdominal pain, nausea, vomiting, and jaundice with elevated hepatic enzymes. Hepatitis B virus infections typically have a more protracted course, with older children being jaundiced in the acute phase and typically asymptomatic in the chronic carrier stage. Epstein Bar virus is another cause of jaundice in older children as part of the infectious mononucleosis syndrome.
- Drug-induced liver injury is fairly common. Acetaminophen overdose is one of the leading causes of fulminant hepatic failure in adolescents and young adults.

TABLE 12-1 Laboratory Evaluation of Hyperbilirubinemia in Older Children

Unconjugated Hyperbilirubinemia
Complete blood count
Reticulocyte count
Blood smear
Serum haptoglobins
Direct and indirect Coombs test
Hemoglobin electrophoresis
Red cell enzyme assay
Test for spherocytosis
Conjugated Hyperbilirubinemia
Liver function test (AST, ALT, ALP, and GGT)
Synthetic liver function (prothrombin time, total protein, albumin, glucose, cholesterol, ammonia)
Abdominal ultrasonography
HAV IgM, HBsAg, IgM-anti-HB score, anti-HCV, monisopt/EBV titers
Serum ceruloplasmin, 24-hour urinary copper excretion
Serum IgG, autoantibodies (ANA, ASMA, anti-liver-kidney-microsomal antibody)
Serum α_1 -antitrypsin level and phenotype
Liver biopsy

- Less commonly, genetic or metabolic disorders can present with jaundice and conjugated hyperbilirubinemia. Relatively common metabolic defects include α_1 antitrypsin deficiency and Wilson disease. Wilson disease is an autosomal recessive disorder of copper metabolism resulting in excess accumulation of copper in the liver, central nervous system, kidney, cornea, and heart. The liver involvement ranges from acute hepatitis with jaundice to fulminant liver failure with encephalopathy.
- Although α_1 antitrypsin deficiency commonly presents as neonatal cholestasis, new onset jaundice can occur any age. It is the most common genetic disorder requiring liver transplantation in children.
- Hepatobiliary disease is now assuming a greater role in older children with cystic fibrosis. Extrahepatic biliary tract disorders may present in older children with obstructive jaundice.
- Autoimmune hepatitis is a chronic progressive inflammatory disorder of possible autoimmune etiology. Type 1 disease affects adolescent females, causing abdominal pain, malaise, and jaundice. Type 2 causes a rapidly progressive illness in younger infants.

EVALUATION AND MANAGEMENT

- Jaundice beyond the neonatal period is usually pathologic, and requires investigation. Once a distinction is made between conjugated versus unconjugated hyperbilirubinemia, cholestatic versus noncholestatic causes should be explored. Historical clues are an

important diagnostic tool, especially when features of specific disorders or a family history of jaundice are present. A review of medications and inquiring about risk factors for viral hepatitis are important. Physical exam should focus on signs that distinguish chronic from acute liver involvement. Important laboratory evaluations to consider are outlined in the table provided but should be tailored to the individual child. A strong consideration should be given to an abdominal ultrasound in any child who presents with conjugated hyperbilirubinemia. Liver biopsy may ultimately lead to the definitive diagnosis. Treatment is directed to the specific underlying cause along with attention to nutrition and hydration Table 12-1.

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QUESTIONS

1. A 4-day-old newborn girl is brought to the emergency department (ED) by concerned parents. The parents are concerned about the yellowish discoloration of her eyes. The infant was full term with a normal vaginal delivery and discharged on day 2. The infant has normal vitals signs and is bottle-fed.

You suspect physiologic jaundice. Which of the following statements is correct regarding your working diagnosis?

- A. Is rare in full-term infants
 - B. Virtually never occurs in preterm infants
 - C. Often requires phototherapy
 - D. Usually peaks at about 4 days of age
 - E. Usually requires exchange transfusion
2. A 3-day-old infant is referred from the outpatient clinic to the ED. The infant is exclusively breast-fed and the mother reports the infant has had difficulty “latching on” and now she has noticed decreased feeding. On examination, you note yellowish-green discoloration of the skin and sclera. The infant is otherwise well appearing with normal vital signs. In addition to laboratory work, which of the following conversations with the mother would be most appropriate in this case?
- A. Depending on the bilirubin levels, advise the mother to discontinue breast-feeding permanently.
 - B. Regardless of the bilirubin levels, advise the mother to discontinue breast-feeding temporarily, but caution the mother that symptoms will recur if breast feeding is interrupted and then restarted.
 - C. Depending on the bilirubin levels, advise the mother to discontinue breast-feeding temporarily, but reassure her that the symptoms usually do not recur if breast feeding is interrupted and then restarted.
 - D. Regardless of the bilirubin levels, advise the mother of the potential risk for kernicterus associated with this condition.
 - E. Regardless of the bilirubin levels, no specific conversation is necessary with the mother as complications are extremely rare
3. A newborn is brought to the emergency department for lethargy and extreme jaundice. The infant was full term with no complications. However, there is a history of maternal-fetal blood group incompatibility. Which of the following would be correct regarding the development of kernicterus?
- A. Has little to do with gestational age.
 - B. Is inevitable once bilirubin encephalopathy develops.
 - C. Can be prevented by exchange transfusion.
 - D. Never occurs in full term infants.
 - E. Has little to do with increased hemolysis.
4. A 6-day-old African-American infant is brought to the emergency department for evaluation of jaundice. His history is significant for being born premature at 32 weeks. Which of the following is correct regarding the evaluation of the jaundiced neonate?
- A. Focuses solely on hepatic pathology.
 - B. Rarely requires laboratory testing as visual assessment is accurate.
 - C. Is accurately assessed by transcutaneous bilirubin.
 - D. Usually requires a complete blood count, reticulocyte count, and coombs test.
 - E. Is rarely necessary in the emergency department.
5. A 1-month-old infant is brought to the ED for evaluation of fever, jaundice, and decreased feeding. The infant was full term born by caesarean section secondary to an active genital herpes outbreak by her mother. On laboratory evaluation, the complete blood count is normal, the urinalysis has positive nitrates, and the conjugated fraction of bilirubin exceeds 20% of the total bilirubin. Which of the following is accurate regarding conjugated hyperbilirubinemia?
- A. Is rarely pathologic in neonates
 - B. Usually results from infectious causes in older children
 - C. Rarely results from anatomic causes in neonates
 - D. Cannot be assessed by ultrasounds in neonates
 - E. Never results from acetaminophen overdose

ANSWERS

1. D. Physiologic jaundice is common in both premature and full term babies; it is a nonpathologic condition; bilirubin level peaks at about 4 days of age; neither phototherapy nor exchange transfusion are necessary.
2. B. Some degree of jaundice is more common in breast-fed infants. Indirect bilirubin rarely reaches the level at which the baby is at risk for kernicterus or mandates cessation of breast-feeding. It rarely recurs if breast-feeding is interrupted, then restarted. Breast milk jaundice may result from substances in the milk itself.
3. C. Premature babies are at greater risk of kernicterus than healthy full term babies, although it can also occur in full term infants. Increased hemolysis is a risk factor for the development of kernicterus, even in full term infants. In full-term newborns, kernicterus is associated with levels of unconjugated serum bilirubin levels >20 mg/dL. In premature infants, lower levels can cause kernicterus. Increased hemolysis in newborn infants is the most common cause of hyperbilirubinemia

severe enough to warrant phototherapy or exchange transfusion. It is usually secondary to maternal-fetal blood group incompatibility in either rhesus or ABO antigens. Bilirubin encephalopathy may be reversible; exchange transfusion can lower unconjugated bilirubin and prevent kernicterus.

4. D. Given the danger of elevated unconjugated hyperbilirubin in a newborn, the jaundiced neonate presenting to the emergency department requires evaluation. Transcutaneous measurement is not sufficiently accurate, nor is visual inspection especially in darker pigmented infants. In addition to hepatic pathology, the patient must be assessed for increased hemolysis, which entails the studies detailed in answer.
5. B. Conjugated hyperbilirubinemia is virtually always pathologic in neonates; an ultrasound can diagnosis some congenital anatomic defects. While acetaminophen overdose can result in conjugated hyperbilirubinemia, especially in adolescents, infections causes are the most common etiology in older children.

13 CRYING INFANT

Joan M. Mavrinac

With a thorough history, a meticulous physical examination, limited diagnostic tests, and a period of observation in the emergency department (ED), most of the causes of the crying, irritable infant will be identified. Clues to the diagnosis may be subtle. Perform serial examinations and observe until the cause is found or infants return to their normal baseline behavior.

DEFINITIONS

- Normal crying:
 - Peaks in the second month of life and then gradually decreases.
 - Peak crying time may be 2 to 3 hours/day at 6 weeks of age.
 - Crying decreases to 1 hour/day by 14 weeks of age.
- Colic:
 - Chronic crying syndrome from 3 weeks to 3 months of age.
 - Unexplained, paroxysmal crying in healthy infants with normal weight gain.
- Crying episodes last >3 hours/day on >3 days/week.
- Colic is a diagnosis of exclusion.

HISTORY

“Listen to the patient, he is telling you the diagnosis” (Sir William Osler)

- Resist the urge to rush through the history.
- Elicit the following: onset, duration, pattern, exacerbating, and alleviating factors, quality of the cry (high-pitched or weak), activity and appearance before and after the crying episode (normal, listless, or lethargic), responsiveness to parents.
- Ask about related symptoms such as fever, vomiting, diarrhea, constipation, hematochezia, cough, or nasal congestion.
- Ask about trauma, possible ingestions, or exposures.
- Ask about paradoxical irritability (infant is more irritable when held).
- Oral intake: feeding history, medication history, and toxic ingestion.
- Relationship to feeding:
 - Crying only with feeding: consider oral pathology (gingivostomatitis, herpangina, oral burn, torn frenulum).
 - Crying only with bowel movement: consider anal fissure. If hematochezia: consider infectious enteritis, milk allergy.
 - Crying during or after feeding, especially with arching of neck and upper back: consider gastroesophageal reflux (GERD).
- Output: vomiting, diarrhea, constipation, hematochezia, urination.
- Past medical history:
 - Perinatal and birth history: maternal premature rupture of membranes, herpes, group B streptococcus status, drug abuse, medication use.
 - Immunization history: recent infant immunizations.
 - Growth and development: weight gain, developmental milestones.

PHYSICAL EXAMINATION

- Observe first: infant behavior (toxic or nontoxic appearing), infant–parent interaction, color, vigor of cry, pitch of cry, quality of respirations, work of breathing, symmetry of movement of extremities.
- Completely undress the infant at some point during the examination.
- Nonthreatening examinations first, preferably in parent’s arms: auscultation, abdominal palpation.

- Threatening examinations last: fundoscopy, otoscopy, oral pharynx, genital, and rectal examinations.
- General: Infant's activity (toxic or nontoxic appearing), infant–parent interaction, color, and pitch of cry.
- Vital signs:
 - Temperature (hypo or hyperthermia: infectious process).
 - Pulse (tachycardia, bradycardia: infectious process, dehydration, anemia, cardiovascular etiology).
 - Respiratory rate, oxygen saturation (respiratory or cardiovascular pathology).
 - Weight (abnormal weight gain may be the sign of an ill infant).
- HEENT:
 - Head: anterior fontanelle (bulging in increased intracranial pressure, depressed in dehydration), ecchymosis (suggestive of trauma).
 - Eyes: corneal abrasion, foreign body, and retinal hemorrhages.
 - Ears: tympanic membranes (otitis media).
 - Nose: inspect for obstruction (infants are often obligate nose breathers until 4 to 6 months).
 - Oral–pharynx: inspect for pharyngitis, gingivostomatitis, oral burns, torn frenulum.
- Respiratory: retractions suggest either respiratory or cardiovascular pathology, tachypnea may signify infection, pain, or metabolic problem (eg, diabetic ketoacidosis).
- Cardiovascular: check pulse to rule out supraventricular tachycardia (SVT); bradycardia may represent sepsis; note murmurs or gallops.
- Abdominal: palpate for masses, signs of peritoneal irritation.
- Genitourinary: examine for incarcerated hernias, testicular torsion, or hair tourniquets.
- Musculoskeletal: look for symmetry of movement (asymmetry suggests possibility of fractures or septic arthritis).
- Skin: look for hair tourniquets, bruises, rashes.
- Neurological: Paradoxical irritability is a dangerous sign (the infant cries with holding and is calm when not touched) and it may occur with meningeal irritation, peritoneal irritation, fracture, or septic arthritis.

DIFFERENTIAL DIAGNOSES

TOXIC APPEARING INFANTS

- Infections: sepsis, meningitis, encephalitis
- Trauma: head injury (nonintentional or abusive head trauma)

- Ingestions/poisons: medications, toxins, or drugs
- Abdominal pathology: intussusception (late), peritonitis (appendicitis)

NONTOXIC APPEARING INFANTS

- Infections: otitis media, pharyngitis, urinary tract infection, septic arthritis.
- Trauma: corneal abrasion, foreign body in eye, fractures (accidental or nonaccidental), hair tourniquet, burns.
- Ingestions/poisons: carbon monoxide, pseudoephedrine, antihistamines.
- Abdominal pathology: intussusception (early), GERD, milk allergy, constipation.
- Genital-urinary pathology: incarcerated hernia, testicular torsion.
- Cardiovascular : SVT, congestive heart failure.
- Other: electrolyte disturbances (hypoglycemia, hypernatremia, hyponatremia, hypercalcemia, hypocalcemia), immunization reactions.
- Diagnoses of exclusion: Colic, parental anxiety, overstimulation, sensory integration problems (autism).

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QUESTIONS

1. 2-month-old female presents with intermittent crying for 2 days. Crying is more frequent after feedings. The infant is gaining weight well. She has slight regurgitation of milk after feedings. Parents describe the infant arching her neck and upper back during the episodes of crying. She is alert and well

between feedings. Vital signs are normal. Physical examination is normal.

Crying noticed after feeding in a well infant with normal weight gain may be secondary to

- A. Colic
 - B. Gastroesophageal reflux
 - C. Pyloric stenosis
 - D. Intestinal malrotation
 - E. Clavicular fracture
2. A 2-week-old male infant presents with crying for the last 2 hours. He was born full-term, the product of a normal vaginal delivery. The mother had prolonged rupture of membranes, a fever that resolved within 12 hours of delivery, and she did not receive intrapartum antibiotics. The infant has been well since birth, with normal weight gain. The physical examination is remarkable for rectal temperature of 38.5°C, heart rate of 90 beats/minute, respiratory rate 30 breaths/minute, and oxygen saturation 98% and capillary refill time of 3 seconds. The remainder of the physical examination is unremarkable.

The most important diagnosis to consider in this infant is

- A. Pharyngitis
 - B. Viral illness
 - C. Bacterial sepsis or meningitis
 - D. Otitis media
 - E. Omphalitis
3. An 11-month-old male presents to the ED with crying for the last 6 hours. The crying is episodic, lasts for a few minutes, and recurs a few times each hour. The infant draws his legs to his abdomen during the episodes and is well appearing between the paroxysms of crying. He had one emesis on presentation to the ED.

The most important diagnosis to consider in this infant is

- A. Pharyngitis
 - B. Viral illness
 - C. Bacterial sepsis or meningitis
 - D. Otitis media
 - E. Omphalitis
4. A 3-month-old female infant presents for evaluation of crying. She has been crying for 3 hours at home and parents are unable to console her. Her past medical history is unremarkable. Appetite, urine, and stool output are normal. There is no history of trauma. Physical examination is normal. There is no evidence of corneal abrasion or hair tourniquet. While in the ED the infant stops crying and appears alert and playful.

Your next step in the management of this infant would be

- A. Intramuscular cephalosporin and discharge home.
 - B. Observation in the ED for 1–2 hours, if well appearing, contact the primary care physician and discharge home with close follow up.
 - C. Admission to hospital for monitoring after septic work-up.
 - D. Obtain a CT scan of the head.
 - E. Obtain a skeletal survey.
5. An 8-week-old female infant presents to the ED with a history of intermittent crying for 3 hours prior to arrival. She was born by normal spontaneous vaginal delivery at 36 weeks gestation; mother was positive for Group B Streptococcus. Physical examination is remarkable for an ill appearing infant with mottled appearance, weak, intermittent crying, and moaning. Vital signs: Rectal temp: 38.6°C, heart rate: 180 beats/minute, respiratory rate: 40/minute, pulse Oximetry: 98%. HEENT: Anterior fontanelle: full, bulging.

The most likely diagnosis is

- A. Group B Streptococcus meningitis
 - B. Otitis media
 - C. SVT
 - D. Congestive heart failure
 - E. Dehydration secondary to viral illness
6. An 8-month-old male infant presents with crying for 4 hours. He has paradoxical irritability; his crying intensifies when he is being held. There is no history of fever, emesis, or diarrhea. Physical examination reveals asymmetry of movement of the lower extremities, with the right lower extremity held in flexion.

The paradoxical irritability in this infant most likely represents

- A. Fracture
 - B. Peritoneal irritation
 - C. Meningitis
 - D. Sepsis
 - E. Encephalitis
7. A 6-month-old male presents with intermittent irritability and crying for 3 days and 3 episodes of emesis. He recently received immunizations. The physical examination reveals an afebrile, ill-appearing, lethargic infant, with a full fontanelle. He has bruises in various stages of healing over his extremities and a torn frenulum.

Evaluation of this infant should include

- A. Lumbar puncture
- B. CT head and skeletal survey

- C. Urinalysis and urine culture
 D. Upper GI series
 E. Chest radiograph
8. A 3-month-old female infant presents for evaluation of crying. Patient has been crying for the last 3 hours. There is no history of fever, emesis, diarrhea, or change in urinary output. Physical examination reveals an afebrile, nontoxic appearing infant with a swollen, red fourth right toe.
 The first step toward further management of this infant is
- A. Complete blood count and blood culture
 B. Foot radiography
 C. Sickie prep
 D. Examination of the digit under magnification
 E. Intravenous antibiotics
9. A 9-month-old male infant presents for evaluation of crying and emesis. Crying started 4 hours ago, initially intermittent and has progressed to continuous crying. His past medical history is negative except for hydrocele at birth. Initial physical exam is done with the infant dressed and in the mother's arms.
 After examining the patient in the mother's arms, the next step should be
- A. Give a trial of oral hydration.
 B. Insert an intravenous line and give a 20 cc/kg bolus of normal saline.
 C. Undress the infant completely and examine the genitourinary area.
 D. Administer acetaminophen and reexamine the infant.
 E. Perform fluorescein staining of the eyes.
10. A 10-month-old irritable female infant presents with intermittent crying and poor feeding for 24 hours prior to arrival. The infant wants to be held. She is fully immunized and attends daycare. Parents report an elevated temperature and have noticed a rash on her hands and feet. The infant is nontoxic appearing. What is the most likely cause of the crying in this infant?
- A. Occult bacteremia
 B. Otitis media
 C. Pharyngeal ulcers
 D. Intussusception
 E. Aseptic meningitis

ANSWERS

1. B. Gastroesophageal reflux may present with crying, especially after feedings, arching of the neck and upper back, and regurgitation of milk

in a well appearing child. Colic is a diagnosis of exclusion and it is not associated with feedings. Infants with pyloric stenosis present with projectile nonbilious emesis. Intestinal malrotation should be considered in any infant with bilious emesis. It is common for a clavicular fracture to be suspected only after a callous mass is noted over the affected clavicle at 7–10 days. However, early on, infants with a clavicular fracture may present with irritability and decreased movement of the upper extremity on the affected side.

2. C. The most important diagnosis to consider when confronted with a crying, febrile neonate is bacterial sepsis or meningitis. Historical risk factors for bacterial sepsis or meningitis include maternal prolonged rupture of membranes without intrapartum antibiotics and maternal peripartum fever. This infant's physical examination is also concerning for bacterial sepsis or meningitis as noted by rectal fever (normal range: 36.8–38°C), bradycardia (normal range: 120–160 beats/minute) and decreased perfusion as noted by the delayed capillary refill (normal range: <2 seconds). Viral illness, pharyngitis, or otitis media may present with fever. However, these are diagnoses of exclusion in a febrile neonate, which are made after a septic work-up and admission. Omphalitis or umbilical site cellulitis would be evident on physical examination.
3. D. Intussusception is the most frequent cause of intestinal obstruction in children between 3 months and 6 years. It presents with intermittent crying episodes, caused by paroxysms of abdominal pain, followed by emesis, which is sometimes bilious. Early in the course, the child appears normal between episodes of crying. Hematochezia or currant jelly appearing stools as well as lethargy may occur later (12–24 hours). While the other diagnoses listed may present with intermittent crying and emesis, intussusception is the most important entity to exclude in this case.
4. B. If the crying abates during the ED visit, a 1 to 2 hour period of observation is appropriate in the well-appearing infant with a normal physical examination. The infant may be sent home with close follow up with the primary care physician and specific instructions to return if symptoms recur. If the infant remains well appearing and playful during the observation period, septic workup and hospital admission are not indicated. Administration of antibiotics without evaluation for sepsis is inappropriate. While nonaccidental trauma should always be considered in the differential diagnosis

of the crying infant, in this scenario neither the history nor the physical examination findings are suggestive. Therefore, neither the CT of the head nor skeletal survey is indicated.

5. A. Serious bacterial illness must be considered in the differential diagnoses of the ill appearing, febrile, crying infant. Risk factors for serious bacterial illness in this infant include prematurity and maternal Group B Streptococcus. A bulging fontanelle in an infant with a weak cry, moaning, irritability, and lethargy suggests meningitis. Otitis media may cause intermittent crying, however it would not explain the bulging fontanelle and mottled appearance. SVT in infants may present with irritability and mottled appearance, however the heart rate is usually greater than 220 beats/minute. Congestive heart failure presents with diaphoresis, increased feeding duration, and tachypnea. Dehydration may present with lethargy in infants, however, the fontanelle is flat or depressed.
6. A. Paradoxical irritability, intensification of crying when the infant is held, may be an indication of meningeal irritation, peritoneal irritation, septic arthritis, or fractures. The infant in this scenario has no fever, emesis, or other indications suggestive of systemic illness. The findings on physical examination are concerning for a right lower extremity fracture.
7. B. Nonaccidental trauma must always be considered in the crying infant. Complete physical examination with the child undressed may reveal signs suggestive of trauma, as the bruises and torn frenulum described in this case. Retinal hemorrhages are also suggestive of nonaccidental trauma. CT of the head and skeletal survey are indicated to identify intracranial hemorrhages or occult fractures. Lumbar puncture is diagnostic for meningitis or encephalitis, which is unlikely in this afebrile infant. There is no indication that this infant has gastrointestinal, respiratory or urinary pathology.
8. D. In a crying infant with a swollen digit, a hair tourniquet must always be considered. Completely undressing the infant is the key to making the diagnosis. Once found, the tourniquet must be released or ischemic injury may occur. Examination under magnification is the first step in the management. The diagnoses of cellulitis or fracture may be entertained after hair tourniquet is excluded. Dactylitis secondary to sickle cell disease presents with diffuse swelling of the hands and feet.

9. C. Incarcerated inguinal hernias present with emesis and crying. The diagnosis is evident as soon as the diaper is removed. In the examination of the crying infant, completely undressing the infant cannot be overemphasized. If the diagnosis is not evident after a complete physical examination, a corneal abrasion (diagnosed by fluorescein staining of the eyes) or urinary tract infection needs to be considered. Intravenous hydration, oral rehydration or acetaminophen administration is not appropriate without a complete physical examination
10. C. It is always important to obtain a complete physical examination when evaluating a crying infant. The infant in this scenario has an enteroviral infection, coxsackie, known as hand-foot-mouth disease: a vesicular rash on the palms and soles and pharyngeal ulcers. The pharyngeal ulcers are the cause of the fever, irritability and decreased feeding. Occult bacteremia is unlikely because this infant is fully immunized. Otitis media is unlikely because the usual prodrome of a viral upper respiratory tract infection is not present. Intussusception is one of the gastrointestinal complications of Henoch–Shonlein Purpura (HSP). HSP is the most common vasculitis in children. The rash is petechial and/or vasculitic and usually involves the lower trunk, legs, buttock, and perineum. Aseptic meningitis is unlikely because the infant is nontoxic and does not have paradoxical irritability.

14 LIMPING CHILD

Isabel A. Barata

HISTORY

- Painless limp is likely to be the result of mechanical or neuromuscular disorders and seldom presents acutely. Painful limp is more likely to be infectious, traumatic, or neo-plastic in etiology.
- Limp that appears worse in the morning suggests a rheumatologic process. Night pain, especially pain that wakes a child from sleep, is a worrisome indicator of a malignant process and warrants a rapid diagnostic approach.
- Fever suggests an infection or an inflammatory condition. A recent upper respiratory infection could be the instigating event to a septic process or raise the possibility of post streptococcal reactive arthritis.
- Weight loss or malaise suggests a systemic process.

- Abdominal pain, diarrhea, or urinary symptoms may suggest nonorthopedic etiologies such as appendicitis, psoas abscess, orchitis, or testicular torsion.
- Immunization history is important since Hemophilus influenza type B may cause both septic arthritis and osteomyelitis.
- Lyme disease should be considered if the patient lives in or travels to an endemic area.
- Past medical history may be helpful since the history of endocrine problems such as hypothyroidism or delayed sexual development may be associated with slipped femoral capital epiphysis.
- A family history should also be obtained for rheumatologic or muscular diseases that may be inherited.

PHYSICAL EXAMINATION

- The child's temperature may help identify an infectious process. However, the absence of fever should not sway from the diagnosis.
- The generalized demeanor of the child, for example, a child with septic arthritis, may be quiet.
- Observe the child's gait. In an antalgic gait, the child has asymmetrical cadence due to less time spent in the stance phase of the affected leg. A child who walks stiffly may be attempting to reduce pain in the spine, such as that occurring in diskitis. In addition, a child who walks with a Trendelburg gait, the torso shifts over the pathologic limb, may be an indication of hip inflammation or hip muscle weakness, such as Perthes' disease and transient synovitis.
- Complete a thorough examination from head to toe, including examination of the abdomen and genitalia.
- Perform a focused neurologic examination by observing the child walking on the toes, walking on the heels, and hopping on one foot. Test for deep tendon reflexes and clonus.
- Complete a musculoskeletal examination of the spine, sacroiliac joint, hip, knee, ankle, and foot. All joints should be taken through their full range of motion passively and actively and be palpated for tenderness and warmth. Loss of range of motion will help localize the site of pathology.
- Examine the feet looking for ingrown toenails and any evidence of puncture wounds. Also check the feet for clawing of the toes or cavus foot deformity, which are red flags for an underlying neurologic condition, especially if either condition is unilateral.

SPINE

- Check the spine on flexion to identify an asymmetric turning of the spine, a sign of spinal cord pathology.

Diskitis causes limited spinal flexion accompanied by a stiff posture and localized tenderness.

- Spondylosis or spondylolisthesis patients experience an exacerbation of lumbosacral pain with spinal extension, so check the spine on extension as well by having the patient bend backward.

SACROILIAC JOINTS

- The flexion, abduction, and external rotation (FABER) test consisting of hip flexion, abduction, and external rotation is positive if it causes groin pain. It indicates a hip rather than a spinal problem.
- Press firmly on the flexed knee and on the opposite anterosuperior iliac crest, pain in the sacroiliac area indicates a problem with sacroiliac joints.

HIP JOINTS

- Slipped capital femoral epiphysis: there is obligate external rotation of the involved lower extremity when the hip is flexed. These rotational changes can occur when the radiographic findings are still quite minimal.
- Inflammatory conditions of the hip as well as developmental dysplasia of the hip present with asymmetry in hip abduction. Hip abduction is tested with the hips flexed and extended making certain the pelvis remains level.
- The Galeazzi test is performed by putting the child in a supine position and bringing the ankles to the buttocks with the hips and knees flexed. The test is positive when the knees are at different heights, suggesting developmental dysplasia or a leg-length discrepancy.
- Measurement of thigh and calf circumference is important since atrophy (more than 1–2 cm of difference between sides) should be seen in a patient with any hip or knee condition that has limited function for more than 1 to 2 months.
- Septic hip is usually held in a position of flexion, abduction and external rotation.
- Differentiating transient synovitis (TS) from septic arthritis is particularly challenging, because both conditions present with decreased motion of the hip. A modified log-roll test may be helpful in differentiating the degree of hip irritation. While distracting the supine child by gently holding the big toe and pretending to examine the foot from different angles, significant hip rotation may be attempted. If an arc of 30° or more of hip rotation is possible without complaints of pain, a diagnosis of TS is more likely than one of sepsis.

KNEE

- The knee examination should include ballotment of the patella to identify an effusion or hemarthrosis (blood in the joint).
- Pain at extremes of flexion or extension can signal meniscal pathology; however, in an acute injury, the amount of fluid and pain in the knee joint often makes motion limited.

ANKLE/FOOT

- Examination of the ankle should include careful palpation of the fibula and tibial physis, foot, and toes to identify possible occult fracture through the growth plate.

EVALUATION

- If appropriate from the history and physical examination order a complete blood count with differential, erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) level.
- The CRP becomes elevated earlier than the sedimentation rate and is considered more sensitive for an infectious process. CRP, as compared to ESR, has a better negative predictive value than a positive predictor. If CRP is less than 1 mg/dL, the probability that the patient does not have septic arthritis is 87%.
- A CRP level of >2.0 mg/dL (>20 mg/L) was a strong independent risk factor and a valuable tool for assessing and diagnosing children suspected of having septic arthritis of the hip.
- The probability of septic arthritis was 99.6% when all four of these clinical factors were present: history of fever, nonweight-bearing, ESR of at least 40 mg/h, and a serum white blood cell count greater than 12×10^9 .
- Multivariate regression analysis showed that body temperature >37°C, ESR >20 mm/h, CRP >1 mg/dL, WBC >11 000/mL, and an increased hip joint space, a displacement or blurring of periarticular fat pads of >2 mm were independent multivariate predictors of acute septic arthritis.
- Multivariate analysis demonstrated that when all three variables of duration of symptoms >1 and <5 days, temperature >37.0°C, and ESR >35 mm/h were present, the predicted probability of infection was 0.66, falling to 0.01 when none were present. This multivariate model enables us to rule out musculoskeletal infection with 99% certainty in

limping children with none of these three presenting variables.

- If a septic process is suspected, the first priority should be to aspirate joints with effusion and evaluate the synovial fluid. The synovial fluid needs to be sent for cell count, Gram stain, anaerobic, and aerobic cultures. The evidence is not as strong on sending the synovial fluid for protein and glucose analyses.
- The sensitivity and specificity of white blood cell count in the synovial fluid above 40 000 per mm^3 exceeded 90% in differentiating septic arthritis from other kinds of arthritis. The white blood cell count in synovial fluid greater than 50 000 per mm^3 is considered to be an indication of an infectious process.
- A septic joint partially treated with antibiotics may have a lower-than-expected white cell count. In addition, a negative culture does not rule out a septic joint; in approximately 33% of cases, the joint aspirate does not recover an organism.
- If the joint in question is the knee and the patient is sexually active, the fluid should be cultured for gonorrhea by using a special media (Thayer-Martin) for growth.
- If the patient lives in an area that is endemic for Lyme disease, blood titers should be determined because acute Lyme disease can mimic a septic process.
- Serum rheumatoid factor antinuclear antibody and HLA typing should not be considered initially, because inflammatory diseases in children tend to be seronegative, making blood work less critical than the clinical history and physical examination.

RADIOGRAPHIC ANALYSIS

- Plain films of the areas in question should be the first radiographic studies ordered since they provide an excellent means of screening for fracture, joint effusion, lytic lesions, periosteal reaction, and avascular necrosis.
- In the nonverbal patient, a screening anteroposterior film from hips to feet identifies a fracture in one-fifth of patients presenting with a limp.
- Hip films anteroposterior and frog-lateral views are necessary because the pathology is frequently best seen on lateral views.
- Acute septic arthritis plain radiographs may show a displacement or blurring of periarticular fat pads.
- The pelvic films should be examined for bone fragmentation (Perthes'), joint space widening (sepsis, Perthes'), and structural abnormalities (hip dysplasia).

- Fractures of the upper femur are rather uncommon, and if they occur in the young age group, they usually consist of a nondisplaced Salter–Harris type I fracture.
- Toddler’s fracture in young children is a spiral fracture of the tibia. However, there are other fractures that should be considered within this symptom complex: original Toddler’s fracture (type I), the hyper-extension-induced upper tibial fracture (type II); various buckle fractures of the distal tibia and fibula; the so-called “bunk-bed foot” such as type I fracture, an impaction-buckle fracture of the base of the first metatarsal, and the type II fracture, a compression fracture of the cuboid bone as well as ankle fractures; and plastic bending fractures of the fibula.
- Child abuse should be considered with multiple fractures or a fracture of a suspicious nature.

ULTRASOUND

- Ultrasonography is noninvasive but highly dependent on the operator for an accurate result.
- Ultrasonography is much more sensitive than plain films, identifying an effusion in two-thirds of a series of 500 children with normal plain films. The sonographic detection of hip effusion is better when the hip is in slight extension and abduction than in the neutral hip position.
- Power Doppler does not seem to allow the exclusion of septic arthritis and should not be used to preclude aspiration when otherwise clinically indicated.
- Ultrasound is also helpful in identifying fluid in the soft tissues consistent with a soft-tissue abscess.

BONE SCAN/CT SCAN/MRI

- When the cause of a child’s limp cannot be localized by history or physical examination, a bone scan is an excellent way to help localize pathology.
- The overall accuracy of bone scans was found to be 81%. The predictive value for a bone scan to be correct was 100% for a cold scan and 82% for a hot scan. The main reason for a false-positive scan was contiguous soft-tissue infection. A negative bone scan makes an infection, a fracture, or most tumors less likely so that observation may be more safely pursued.
- An area that is bright on bone scan can be further evaluated with plain films, computed tomographic (CT) scan with bone windows, or magnetic resonance imaging (MRI) studies.

- CT scan best delineates bone structure, whereas MRI best highlights areas of pathology in the soft tissues as well as inflammation of bone.

MICROBIOLOGY

- Microscopy of the joint fluid cannot be used to rule out septic arthritis. The fluid must be cultured.
- Parenteral antibiotics should be given only after synovial fluid has been obtained for Gram stain and culture.

CONCLUSION

- A child who limps often presents a diagnostic challenge.
- The differential diagnosis is extensive. Although the most common cause is trauma, awareness of other potential causes is important.
- The age of the child and the pattern of the gait help narrow the differential diagnosis. In most cases, a diagnosis can be made from the history and physical examination.
- If the diagnosis is not obvious after a careful clinical evaluation, plain radiographs provide an excellent means of screening for fracture, joint effusion, lytic lesions, periosteal reaction, and avascular necrosis. Other tests should be ordered only when indicated.

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QUESTIONS

1. A 20-month-old female is brought to the ED by her parents with a complaint that she will not stand on her right leg since earlier this afternoon after going down a sliding pond. There is no history of fever or prodromal symptoms, and there is no history of previous injuries. Her parents report that she is able to walk, but with a limp.
The most likely diagnosis is
A. Toddler's fracture
B. Femur fracture
C. Cuboid fracture
D. Septic joint
E. None of the above
2. A 4-year-old female child who is brought to the ED with fever and right leg pain since the previous night. Her fever and leg pain worsened to the point that she was no longer able to ambulate. She appears nontoxic with a temperature of 40°C. She prefers to keep her right hip abducted and externally rotated, with the right knee flexed. She refuses to bear any weight on the right leg. Range of motion of the right hip is severely limited, especially internal rotation, adduction, and extension. Examination of the remainder of the right lower extremity, the left lower extremity, and spine are within normal limits. Patient is most likely to have
A. Toxic Synovitis
B. Septic Arthritis
C. Lyme arthritis
D. Avascular necrosis
E. None of the above
3. Appropriate tests at this time for patient in question 2 are
A. Pelvis and Hip X-rays
B. CBC with diff
C. C-reactive protein (CRP) and sedimentation rate (ESR)
D. Only A and B
E. A, B, C
4. This is a 15-year-old female living in the Northeastern US with acute onset of left lower extremity pain and swelling of the knee after a weekend ski trip in December. On physical exam patient is afebrile, has a swollen left knee, decreased range of motion. The complete blood count shows mild leukocytosis, erythrocyte sedimentation rate is 28. Synovial white cell count is 50,000 milliliters. The following test should be sent from the synovial fluid:
A. Gram stain
B. Lyme titer
C. Aerobic and anaerobic cultures
D. Only A and C
E. A, B, and C
5. Patient in question 3 is more likely to have:
A. Septic Arthritis
B. Gonococcal Arthritis
C. Lyme Arthritis
D. Toxic Synovitis
E. None of the above
6. A 14-year-old boy comes to the ED complaining of left hip pain. He has a 4 week history of left medial thigh pain that has become increasingly severe. An AP radiograph of the hip reveals no abnormalities. What is the most likely diagnosis?
A. Femoral stress fracture
B. Anterior hip dislocation
C. Legg–Calvé–Perthes disease (LCPD)
D. Slipped capital femoral epiphysis
E. Posterior hip dislocation
7. A 3-year-old with history of anemia and is on iron supplements presents with a petechial rash that started one week ago in the buttocks, now on the upper chest, neck, around right eye and mouth. The patient was diagnosed with anemia and she also a cast on the right leg for presumed occult fracture with persistent leg pain. She had a persistent limp before the cast was placed. The most likely diagnosis for this patient's limp is:
A. Septic arthritis
B. Henoch–Schönlein Purpura
C. New onset leukemia
D. Slipped capital femoral epiphysis
E. None of the above.
8. A 4-year-old presents to the ED with history of not being able to bear weight on the right leg. Patient had a recent history of upper respiratory infection. On physical exam, he appears well and has low-grade temperature. He has pain on range of motion

of the hip but is able to tolerate some range of motion. The most likely diagnosis for this patient's limp is:

- A. Septic Arthritis
 - B. Gonococcal Arthritis
 - C. Lyme Arthritis
 - D. Transient Synovitis
 - E. LCPD
9. The flexion, abduction, and external rotation (FABER) test consisting of hip flexion, abduction, and external rotation. What does it indicate if it causes groin pain:
- A. A hip rather than a spinal problem
 - B. A spinal rather than a hip problem
 - C. Sacroiliac area problem
 - D. Both A and B
 - E. Both A and C
10. Which of the following is true about ultrasound use in the diagnosis of hip effusion?
- A. Ultrasound is highly dependent on the operator for an accurate result.
 - B. Ultrasound is much more sensitive than plain films
 - C. Power Doppler does not seem to allow the exclusion of septic arthritis and should not be used to preclude aspiration when otherwise clinically indicated.
 - D. Ultrasound is also helpful in identifying fluid in the soft tissues consistent with a soft-tissue abscess.
 - E. All of the above

ANSWERS

1. A. Toddler's fracture was first described by Dunbar in 1964 as a subtle, nondisplaced oblique fracture of the distal tibia in children, 9 months to 3 years of age. The child usually presents with an acute onset of limp or refusal to bear weight on one leg. An unsteady toddler may have fallen with a twist, or the child may have gotten his/her foot caught and fallen, twisting it while trying to free his/her foot. Many times the fall is unwitnessed and parents are unsure of what happened. The mechanism is not consistent with a femur fracture or cuboid fracture. There is no clinical evidence of septic arthritis.
2. B Septic arthritis is an inflammation of a synovial membrane with purulent effusion into the joint capsule, usually due to bacterial infection. The age range of those affected is broad, from the neonatal period to advanced age. In children, septic arthritis most commonly affects the hip joint. Septic arthritis usually is divided into gonococcal and nongonococcal arthritis, as clinical and treatment regimens differ. Treatment consists of a combined medical and surgical approach. Toxic synovitis is unlikely with the patient with an elevated temperature and significant pain on range of motion. Lyme arthritis most frequently affects the knee joint. Avascular necrosis is unlikely with such a sudden onset.
3. E Appropriate work up includes radiographs of the pelvis and hip, as well as complete blood cell count, C-reactive protein (CRP) and sedimentation rate (ESR). C-reactive protein becomes elevated earlier than the sedimentation rate and is considered more sensitive for an infectious process. CRP, as compared to ESR, has a better negative predictive value than a positive predictor. If CRP is less than 1 mg/dL, the probability that the patient does not have septic arthritis is 87%. Sedimentation rate (ESR) is usually elevated in patients with septic arthritis.
4. E. Lyme arthritis needs to be ruled out due to the patient's geographic location and the season. Lyme arthritis will usually present in the Fall early winter after exposure in the summer to the *Ixodes*, or deer tick carrying the spirochete *B. burgdorferi*.
5. C. In Lyme arthritis, joints are the exclusive target in most cases. There may be a history of skin involvement in the form of erythema migrans. This often begins as a migratory polyarticular process involving bursae, tendons, and joints, which evolves over 1-2 days into a monoarticular process involving the knee, ankle, and wrist, in decreasing frequency.
6. D. Slipped capital femoral epiphysis seen between 10 and 17 and usually gradual onset over days to months and often starts as groin pain. Radiographs are often subtle and best seen on lateral views of the hip. LCPD is the idiopathic osteonecrosis of the capital femoral epiphysis of the femoral head. It usually affects younger children aged 3–12 years, with a median age of 7 years. Anterior and posterior hip dislocation would have an acute presentation.
7. C. New onset leukemia presenting with prior history of a limp, anemia and now with a petechial rash. With Henoch–Schönlein Purpura you should not have anemia.
8. D. Transient synovitis (TS) is the most common cause of acute hip pain in children aged 3–10 years.

The disease causes arthralgia and arthritis secondary to a transient inflammation of the synovium of the hip. Recent history of an upper respiratory tract infection, pharyngitis, bronchitis, or otitis media is elicited from approximately half of patients with transient synovitis.

9. A. The flexion, abduction, and external rotation (FABER) test consisting of hip flexion, abduction, and external rotation if it causes groin pain indicates a hip rather than a spinal problem. A problem with sacroiliac joints can be diagnosed by firmly pressing on the flexed knee and on the opposite anterosuperior iliac crest and it will produce pain in the sacroiliac area.
10. E. All are true. Ultrasonography is noninvasive but highly dependent on the operator for an accurate result. Ultrasonography is much more sensitive than plain films. Power Doppler does not seem to allow the exclusion of septic arthritis and should not be used to preclude aspiration when otherwise clinically indicated. Ultrasound is also helpful in identifying fluid in the soft tissues consistent with a soft-tissue abscess.

15 MILD HEAD INJURY IN CHILDREN

Eustacia (Jo) Su

ANATOMIC CONSIDERATIONS

- Disproportionately large head (size and weight) relative to rest of body, especially in infants and toddlers.
- Short, weak, flexible neck, allowing more movement and momentum transfer to head from any impact.
- Open fontanelles (up to 8 months) and sutures (up to 2 years) provide flexibility to absorb impact and ability to expand intracranial volume, but also result in increased susceptibility to hemorrhage.

BIOMECHANICAL CONSIDERATIONS

- Direct impact, acceleration/deceleration, or rotational shear forces.
- More flexible skull bends inward on impact; surrounding areas bend outward and may fracture distant to point of impact.

- Isolated skull fractures are usually associated with normal mental status unless there is also significant underlying brain injury with mass effect.

PATHOPHYSIOLOGY

- Enhanced vulnerability of cells due to ionic shifts, abrupt depolarization, impaired axonal function and many other impairments of cellular function.
- Confusion or headache usually associated with state of enhanced vulnerability.
- Second impact before brain is fully recovered may result in potentially fatal loss of cerebrovascular autoregulation.
- After a single brain impact, state of increased vulnerability to further injury may persist for 3–5 days, but usually resolves within 1 week.
- “Postconcussion syndrome” may occur: persistent alterations in cognition, behavior, and personality changes, emotional swings.
- Multiple, seemingly trivial, head injuries may cause chronic, cognitive impairments, particularly in athletes.

ASSESSMENT

- The ABCs—Airway Breathing Circulation—are essential.
- Assess for hypotension and acidosis, which may cause secondary injury.
- Bleeding from the scalp may be extensive, unrecognized, and result in hypotension.
- In infants, a high BP and low HR may resemble normal VS for an adult or older child but is abnormal.
- Crying may simulate hyperventilation and temporarily mitigate intracranial hypertension.

HISTORY

- Obtain time, nature of incident, height of fall or force of impact.
- Did the patient have LOC, altered mental status, and what was the duration of symptoms?
- Did the patient have vomiting or seizures?
- Consider pre-existing problems, eg, bleeding diatheses and medications.
- Be alert for inconsistencies in history, especially if it is not congruent with child’s developmental stage.
- Ask carefully about previous head injuries, especially in the high-performing athlete, who may try to hide symptoms because of pressures to perform.

PHYSICAL EXAMINATION

- Complete primary and secondary surveys and GCS.
- Head:
 - Linear hematomas, especially temporal or occipital areas
 - Crepitus, depressed fractures
 - Fullness of fontanelles
 - Check for hemotympanum
 - Blood or CSF from ears or nose
 - Raccoon, battle signs
- Neck:
 - Palpate for any step offs, swelling, crepitus
 - Observe neck movement
- Neurologic exam:
 - Cranial nerves
 - Posturing
 - Asymmetry of movement
 - Baseline mental status

ED MANAGEMENT

RESUSCITATION

- Ensure the ABCs, especially adequate (but not excessive) ventilation.
- Avoid hypotension, control any bleeding promptly.

GUIDELINES FOR IMAGING

- Benefit of imaging versus risks of radiation and possible need for sedation.
- Imaging followed by discharge more cost effective than observation without imaging. No difference in outcome.
- Skull radiography not usually used in the United States.

THE “HYPERACUTE” CT SCAN

- Small group of patients who present initially with significant AMS with or without LOC but who clear up almost completely.
- Initial CT, usually within 1–2 hours of injury, looks normal.
- Child deteriorates several hours later, often fatally.
- Repeat CT scan shows either diffuse axonal swelling or a large subdural hematoma.
- Follow-up CTs now part of trauma service routine for these higher risk patients
- Be sure to clarify patient’s initial, postinjury condition, and mental status.

SPECIAL SITUATIONS

CONCUSSION IN THE ATHLETE

- Even mild brain injury can impair memory and concentration, affecting academic performance especially in children involved in competitive sports.
- Cumulative concussions may have an additive effect on behavioral, neuropsychological, and cognitive function, but studies often flawed.
- Stronger studies seem to indicate that MTBI may be associated with alterations in neuropsychological performance, but recovery usually occurs within 12 months.
- More severe cases of MTBI, especially during preschool years, may cause long-term problems in psychosocial function.

SECOND IMPACT SYNDROME (SIS)

- Second, concussive head injury while the brain is trying to recover from first impact can be fatal.
- May not even involve direct impact but just deceleration to the body.
- Second impact may cause severe loss of cerebrovascular autoregulation, resulting in massive brain swelling and almost inevitable death.
- Return-to-play guidelines are designed to try to prevent SIS.

POSTCONCUSSION SYNDROME

- Persistent symptom complex developed by a minority of head-injured patients, usually resolving over 3 months.
- Three categories of complaints: cognitive, somatic, and emotional (affective).
- Controversy over organic versus functional basis; both processes may be involved.
- Counseling about expected symptoms and suggested coping mechanisms may reduce the duration of symptoms as well as the perception of stress.

DISPOSITION AND DISCHARGE INSTRUCTIONS

- Factors to consider when planning discharge.
 - Caregivers:
 - Do they understand discharge instructions?
 - How reliable are they?
 - How comfortable are they with this plan?
 - How far away do they live?

- Do they have access to transportation, telephone, and EMS?
- Can they get early, reliable, outpatient follow-up?
- Instructions:
 - Should be easy to understand, not too burdensome.
 - OK to allow child to fall asleep.
 - No participation in demanding physical activities the first 2 days after a head injury (toddlers will do what they want—trying to restrict their activities is usually futile).
 - Avoid putting child into stimulating environment for 2–3 days.
 - School-age children would benefit from 2–3 days off school.
 - No computer games for 2–3 days.
- Provide information about expected symptoms and duration. Suggest strategies for coping.
 - Distractibility, restlessness, poor memory, or lack of concentration may be misinterpreted as sloth or deliberate misbehavior.
 - No visible signs of incapacitation in most cases.
 - Important to support and assist, rather than criticize or punish.

THE CONCUSSED ATHLETE AND RETURN TO PLAY (RTP)

- In the US, 300,000 sport-related brain injuries/year in high school sports.
 - Mostly MTBIs, concussions.
 - Football has highest reported numbers, but TBIs occur in other sports.
 - Athletes who have sustained a concussion are often evaluated in ED
 - Some will try to get clearance to play from a different physician.
 - Recognition, response, rest, and rehabilitation are the necessary steps to minimize morbidity.

RECOGNITION

- Recognizing concussion—note that LOC is one sign, but not a requirement for diagnosing concussion. It is the most obvious presentation.

RESPONSE

- Any athlete suspected of having a concussion should be removed immediately from the game or practice.

- The athlete must not return to play (RT) while still symptomatic as it greatly increases risk of more severe symptoms and a longer postconcussive course.
- Impaired reaction time or coordination also poses a greater risk of another injury.
- It is critical to let the young athlete's brain recover sufficiently before risking another concussion.
- The emergency physician should ensure that she/he has explained the risks to both the parents and the athlete.
- Close follow-up is essential to ensure complete resolution prior to RTP.

REST

- Physical and cognitive stressors both aggravate postconcussive symptoms.
 - Concentration, memorization, and learning.
 - Lack of outward signs of concussion makes it difficult for others (teachers, employers, classmates) to realize that there is a problem.
 - Resistance (weight) training increases intracranial pressure and can exacerbate postconcussive symptoms.
 - Total rest is important, even though it can lead to depression and isolation from the team.
 - Emphasize the need for support from family, teachers, coaches, team, and peer group.

REHABILITATION/RETURN TO PLAY

- The emergency physician should be familiar with a concussion grading system when counseling player and parents.
- Use guidelines to determine when it is safe for the athlete to return to play.
- Use the highest-grade concussion to decide which recommendation to follow.
- If there is any doubt, err on the side of caution.

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QUESTIONS

- A 20-month-old child has fallen in the playground while at daycare. There are no eyewitnesses available to say exactly what happened. The mother was called to pick up the child for a medical evaluation, but the worker in charge of the child has gone for the day. The child slept for the entire ride to the hospital is noted to have some vomitus on the clothing. VS are within the normal range for age and the child now appears to be acting appropriately for a toddler. The appropriate management for this child would include

 - A full physical and neurologic exam, a noncontrast CT scan of the brain and observation for several hours.
 - A full physical and neurologic exam, then discharge home with head injury instructions.
 - A full physical and neurologic exam, admission for fluids, and antiemetics.
 - A primary trauma survey followed by rapid sequence intubation.
 - A full physical and neurologic exam, a report to child protective services, and discharge home with reassurance and antiemetics.
- Which of the following are considered to be intermediate-risk criteria for children and adolescents with MTBI?

 - Mild von Willibrand disease
 - Previous neurologic disorder
 - Suspicion of nonaccidental trauma
 - No LOC
 - Brief, generalized, post-traumatic seizure
- A high-school senior comes to the emergency department asking for clearance to play in the final football game of the season the following day. He had been observed overnight 2 days ago following a head injury during the previous game. He insists that he feels fine and that he has to play in order to qualify for a letter and his college scholarship application depends on this.

What is the most appropriate next step?

 - Ask how close he is to qualifying for the letter
 - Ask how many previous concussions he has sustained
 - Ask what his coach thinks
 - Do a CT scan of the head
 - Ask him to do serial 7s.
- A 7-month-old girl is brought in for evaluation after falling down the stairs. She has a “dent” in her mid-forehead. She had cried immediately and moved all extremities as soon as the parents reached her. Her exam is otherwise nonfocal. She recognizes her parents and clings to them, moves all extremities equally and appropriately to avoid medical personnel, but does not interact with any other people in the room. The appropriate next step would be

 - Order a head CT without contrast and also cervical spine films.
 - Order a quick brain MRI.
 - Send the patient home with the parents and good head injury instructions.
 - Observe the patient overnight without imaging.
 - Call neurosurgery immediately to decompress her depressed skull fracture.
- You are the assistant coach for your son’s eighth grade soccer team. The team’s best striker has just hit his head on the goalpost. He was not knocked out, but was quite dazed for a few minutes and vomited once. It is the deciding game for the playoffs and there are 25 minutes of playing time left. He insists that he is now fine and ready to play. The coach, player, and parents are all agitating to have the striker return to play now. This is his first concussion.

 - You decide that this is a first-time grade I concussion and that he can return to play when he has been asymptomatic for 15–20 minutes.
 - You decide that he is play acting because he has been watching too much World Cup soccer and let him return to play now.
 - You tell the player’s parents that, because of the vomiting, he has to go to the ED now to get a CT of his head.
 - You allow him to return to play but insist that he wear a helmet.
 - You send him home to rest.

6. In the scenario described in Question 6, the RTP guidelines are used to
- Ensure that players have enough rest during a game.
 - Reduce the risk of long-term cognitive dysfunction.
 - Protect yourself from litigation.
 - Reduce the risk of second-impact syndrome (SIS).
 - Make players and parents aware of the risks of multiple concussions.
7. Which of the following is true about postconcussion syndrome?
- Most children's symptoms resolve by the end of 3 months.
 - Family adjustment determines the duration of symptoms.
 - It mostly affects emotional functioning.
 - Teachers and peers are usually very supportive.
 - Brain exercises will speed recovery.
8. The following statement regarding second impact syndrome is true:
- A mild second impact while the patient is still symptomatic from the first concussion should not count toward risk assessment for SIS.
 - The second injury may be as trivial as a deceleration, eg, when the player is stopped by another player's limb.
 - Experts agree on the duration of rest needed to prevent SIS.
 - When a player goes down the second time, it is time to terminate his season and advise him to refrain from collision sports.
 - The second injury will be immediately fatal.
9. The decision to discharge a child with a low-risk MTBI depends on which of the following factors?
- How well the caregivers understand the discharge instructions and reasons for return for re-evaluation.
 - If the caregivers have at least an eighth grade educational level for literacy.
 - If the caregivers have had other children and can easily compare the child's level of development
 - If the caregivers promise, they will not sleep or let the child sleep tonight.
 - How well the child has been fed by the caregivers while in the ED.
- possibility of significant mechanism and possibly a period of alteration of LOC. Answer B is incorrect because the child should at least be observed for several hours. Answer C would be correct if the child were persistently vomiting and the CT (not given as part of the answer) were negative. Answer D is not appropriate for this situation but for the child with severe brain injury. Answer E is incorrect because, if the child is still vomiting, she/he should be scanned and at least observed.
2. E. In the context of an otherwise nonfocal neurologic examination and completely resolved postictal period. A, B, and C are exclusion criteria for MTBI and should make the physician suspect a more severe TBI. D is a low-risk criterion.
3. B. The number of concussions that the player has already sustained determines the amount of symptom-free time that must elapse before it is safe for him or her to play again and risk sustaining another concussion. The athlete and coach are under tremendous pressure to perform and will often lie about symptoms or past history to try to gain clearance. A CT scan will generally be negative and not contribute at all to decision-making. Serial 7s may help to see if he can concentrate, but actual physical stress (eg, push-ups or a short sprint) and watching for and asking about symptoms such as headache or signs such as decreased cognitive ability may be more helpful.
4. A. Although she does have a depressed skull fracture, the skull is pliable enough that it is likely that the force required to produce the injury was less severe, and that there is not a significant, underlying brain injury. C-spine films could be used to rule out a cervical spine injury, which is unlikely in this scenario. A C-spine CT would probably result in too much radiation exposure for the yield, and the risks of sedation must also be factored into the risk-benefit equation. The quick brain MRI is not sensitive enough to detect blood. D could be acceptable, but because of the depressed skull fracture, it is important to image the underlying brain tissue. E is incorrect because the neurosurgeons will want to know the condition of the underlying brain before deciding whether to operate.
5. A. The return to play (RTP) recommendations state that he must be asymptomatic for 20 minutes. It is not safe to assume that he was playacting. Since he has recovered quickly and had no LOC, a CT of the head is not indicated. Soccer players cannot wear helmets, and a mild impact, or even just a deceleration, may worsen his condition. If he remains asymptomatic, E is not necessary.

ANSWERS

1. A. The child meets intermediate risk criteria (see Table 15-1) because of concerning or unknown mechanism of injury, unwitnessed trauma with the

TABLE 15-1 Classification of Head Injury Risk and Recommendations for Imaging Studies

RISK	RECOMMENDATION
<p>A. High risk:</p> <ul style="list-style-type: none"> Depressed mental status Focal neurologic findings Depressed or basilar skull fractures Irritability Bulging fontanelle Seizure Vomiting >5 times or >6 h LOC >1 min 	Head CT scans should be obtained in these children.
<p>B. Intermediate risk:</p> <p>B-1. Clinical indicators of possible brain injury:</p> <ul style="list-style-type: none"> Transient LOC (<1 min) 3 or more episodes of vomiting Period of irritability or lethargy, now resolved Alteration of LOC or not at baseline behavior according to caregivers Skull fracture >24 h ago <p>B-2. Concerning or unknown mechanism of injury or suspicion of skull fracture on physical examination:</p> <ul style="list-style-type: none"> Higher force mechanism Falls onto hard surfaces Unwitnessed trauma with the possibility of significant mechanism (e.g., finding child at bottom of stairs or wall) History inconsistent with the physical findings or the child's developmental status 	<p>If two or more of these risk factors are present, or if the duration and severity of the LOC or alteration of behavior is significant, CT scan should be more strongly considered.</p> <p>Consider CT scan or further observation (4–6 h), depending on the clinical situation.</p>
<p>C. Low risk:</p> <ul style="list-style-type: none"> Low-energy mechanism (e.g., fall <3 ft) Asymptomatic for at least 2 h after injury 	This group may be observed at home.

6. D. There have been no studies that show that following the RTP guidelines preserve cognitive function, although that would seem to be a natural consequence of protecting the brain from repeated concussions.

7. A. B is partially correct, but premorbid vulnerability and postinjury brain function can also affect the duration of recovery. C is incorrect because the syndrome also includes cognitive and somatic complaints. D is incorrect because there is usually no obvious outward sign of abnormal brain function as compared with a cast on a fractured limb. Children are expected to do well in school and work hard. Listlessness, poor concentration, and poor performance tend to elicit criticism rather than sympathy. E is incorrect because adequate brain rest immediately postinjury is important.

8. B. A is incorrect because even a mild impact can cause fatal brain swelling. Experts do not all agree about the existence of SIS. D is incorrect unless this constitutes his third concussion of which at least one is a grade 3. E is incorrect because the player sometimes will be able to finish the play before collapsing.

9. A. B is incorrect because most instructions are written at a fifth grade literacy level. C is incorrect: the presence of many other children may make close

observation more difficult. D: it is unnecessary to stay awake all night or keep the child awake. E. It is reassuring if the child tolerates food, but it is not the only criterion for safe discharge.

16 APPROACH TO THE PATIENT WITH RASH

Daniel McManus
Gregory Garra

HISTORY AND PHYSICAL EXAMINATION

- History is very important in the evaluation and treatment of rashes. One should ask: Initial location of rash and how long has it been present? How did the rash spread? What prompted the patient to seek care? Are there associated symptoms, that is, fever? Is there itching, burning, or stinging associated with the rash? Are there any alleviating or aggravating factors? Have there been any exposures to potential allergens, occupational exposures? Any sick contacts?

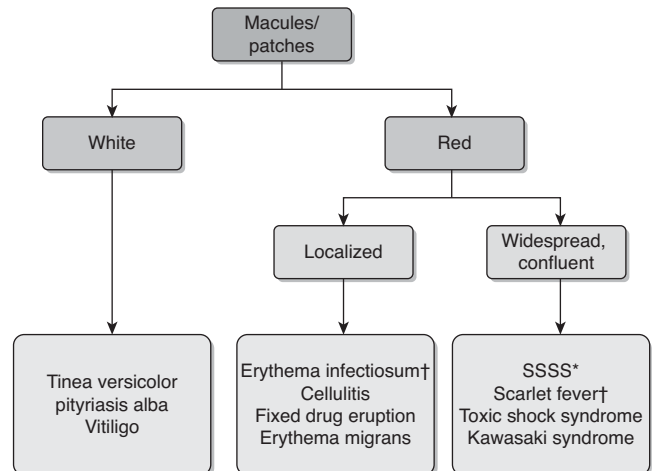
- Physical examination should include a specific exam of the skin as well as a general examination to look for other systemic signs. When examining the skin, you should identify the following: What is the primary lesion? Is a secondary lesion present? Is the lesion palpable? What is the distribution of the lesions? Is an enanthem present? General exam, vital signs.

MORPHOLOGY OF LESIONS

- Primary Lesions include the following:
 - Macule: flat nonpalpable lesion <1 cm in diameter.
 - Patch: flat nonpalpable lesion >1 cm in diameter.
 - Papule: circumscribed, solid raised palpable lesion <1 cm in diameter.
 - Plaque: circumscribed, solid raised palpable lesion >1 cm in diameter.
 - Vesicle: fluid filled raised lesion <1 cm in diameter.
 - Bulla: fluid filled lesion >1 cm in diameter.
 - Pustule: vesicle filled with purulent exudates.
 - Nodule: solid lesion; <1 cm in diameter, at any depth.
 - Tumor: solid lesion; >1 cm in diameter, at any depth.
 - Petichiae: <1 cm pinpoint spots of blood or pigment under skin, round, red.
 - Purpura: >1 cm spots of blood or pigment under skin.
 - Wheal: Evanescent transient papules or plaques with pale center and pink margins.
- Secondary Lesions include
 - Scale: compacted, desquamated layers of stratum corneum.
 - Crust: dried exudate (serum, pus, or blood) on the skin.
 - Excoriation: abrasions generally caused by scratching.
 - Fissure: linear crack or cleavage on skin, with sharply defined margins.
 - Ulcer: depressed lesion with epidermal or dermal tissue loss.
 - Scar: repair and replacement of injured dermis with connective tissue.
 - Lichenification: area of increased epidermal thickness with accentuation of skin.

DIAGNOSTIC FEATURES OF LESION

- By identifying the distribution and configuration as well as whether it is discrete or confluent, one can narrow the differential diagnoses.



† Can appear as vesiculobullous disease in later stage

* Staph Scalded Skin Syndrome may also present with papules and plaques

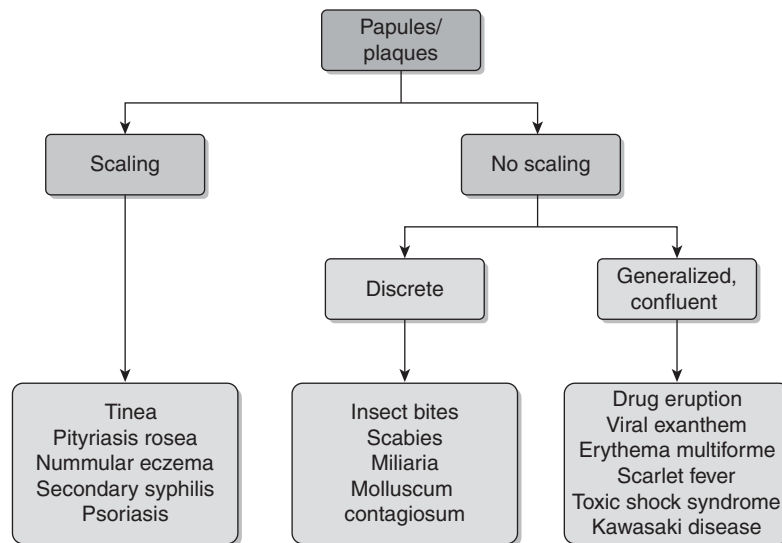
Tinea versicolor	Pink, tan or white, slightly scaling- KOH (+) is scraped
Pityriasis alba	poorly demarcated white, fine scale
Vitiligo	well demarcated, complete depigmentation
Erythema infectiosum	red cheeks followed by net like erythema on trunk
Cellulitis	cardinal signs of inflammation
Fixed drug eruption	violaceous, occurring within hours of offending drug ingestion
Erythema migrans	erythematous border with central clearing, lesion at least 5 cm

FIG. 16-1. Algorithmic approach to a rash with macule/patch primary lesion.

- Distribution: many skin diseases have a preferential area of involvement. Widespread (symmetrical or asymmetrical) or localized (single, clustered, acral, sun exposed)
- Configuration: the general shape or pattern in which the lesions are arranged. Occasionally diagnostic: linear, acriform, circular, or grouped
- Algorithmic Approach: see Figs. 16-1–16-5

TRUE PEDIATRIC DERMATOLOGIC EMERGENCIES

- Stevens–Johnson syndrome: drug reaction, patients toxic appearing, rapid progression of disease, macules progressing to bullae, (+) Nikolsky sign, with less than 10% of body surface area involvement. Mortality 5%.
- Toxic epidermal necrolysis: drug reaction, patients toxic appearing, rapid progression of disease, macules that progress to bullae, (+) Nikolsky sign, with more than 30% of body surface area involvement. Mortality 40%.
- Staphylococcal scalded skin syndrome: exfoliative dermatitis caused by exotoxins released by



Tinea	annular or serpiginous, elevated scaling border, central clearing
Pityriasis rosea	trailing collerette of delicate scale, mainly on trunk, herald patch
Nummular eczema	coin-shaped, no central clearing
Secondary syphilis	reddish-brown or yellow, widely distributed, "great imitator"
Psoriasis	sharply demarcated, silvery scales
Insect bites	grouped, asymmetric, central punctum
Scabies	pruritic, inflammatory papules, white serpiginous burrow
Miliaria	multiple, discrete, fine inflammatory papules
Molluscum contagiosum	umbilicated, flesh colored, dome-shaped, often grouped
Drug eruption	intense erythema, confluent in large areas, pruritic
Viral exanthem	prodrome of fever & constitutional symptoms
Erythema multiforme	variety of lesions, targets, symmetrically distributed
Scarlet fever	sandpaper texture, mucous membrane findings
Toxic Shock syndrome	multisystem involvement
Kawasaki disease	diagnostic criteria

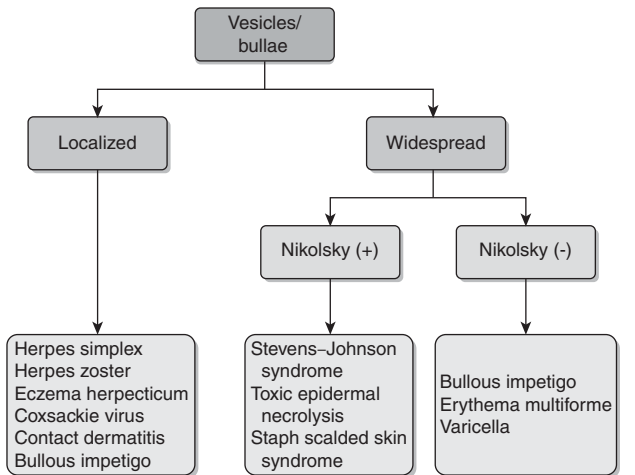
FIG. 16-2. Algorithmic approach to a rash with papule/plaque primary lesion.

staphylococcal bacteria. Generalized erythema progress to bullae with a positive Nikolsky sign.

- Toxic shock syndrome: scarletiform diffuse macular rash, which may have a sandpaper-like character. Due to exotoxins released by staph and strep. Symptoms include: fever >38.9 , rash, shock (SBP <5 th percentile or orthostasis), and multisystem involvement.
- Kawasaki disease: Acute, self-limited vasculitis, most common in males under 5 years of age. The follow-

ing features must be present to make the diagnosis: Unexplained fever $\times 5$ days plus 4 of the following:

- edema or desquamation of the extremities
- bulbar conjunctivitis
- polymorphous rash
- cervical lymphadenopathy
- mucous membrane changes (ie strawberry tongue)
- Meningococcemia: rapidly progressive, classic presentation is fever, photophobia, stiff neck, vomiting



*SSSS is Staphylococcal Scalded Skin Syndrome

Herpes simplex	grouped vesicles, erythematous base
Herpes zoster	grouped vesicle in a dermatomal distribution
Eczema herpeticum	vesicular eruption in a patient with atopic dermatitis
Coxsackie virus	hand, foot, and mouth involvement
Bullous impetigo	fragile, clear/cloudy blisters, collerette like remnant of bulla at rim
Contact dermatitis	linear, angular, or geometric configuration
Bullous impetigo	angular, crust with glistening base (when removed)
SJS	mucocutaneous blistering, epidermal detachment, systemic symptoms
TEN	extensive blistering, epidermal detachment >30%, often drug associated
SSSS	fever, large flaccid bullae, shedding of large sheets of skin
Varicella	lesions in all stages, pruritic, successive crops

FIG. 16-3. Algorithmic approach to a rash with vesicle/bulla primary lesion.

and AMS. Sudden onset of fever and petichial or purpuric rash. Initially may be difficult to distinguish from more benign processes.

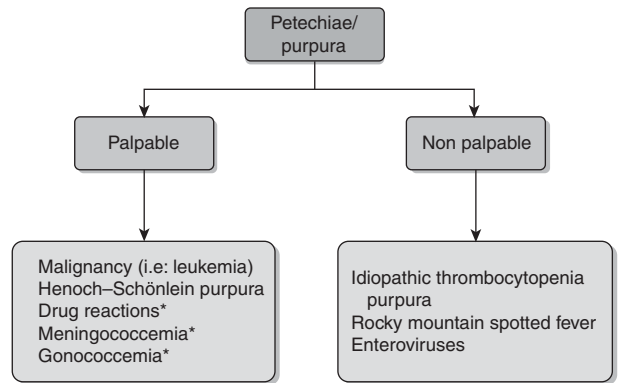
- Anaphylaxis: occurs seconds/minutes from exposure; wheezing, urticaria, angioedema, hypotension, circulatory collapse
- Purpura fulminans: hemorrhagic disease, seen in setting of sepsis and DIC and is rapid in onset.

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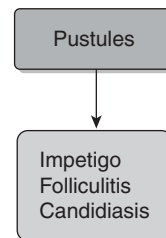
Rosenstein NE, Perkins BA, Stephens DA, et al: Meningococcal disease. *N Engl J Med* 344:1378–1388, 2001.



* may present as palpable or nonpalpable petechiae/purpura

Malignancy	systemic signs and symptoms
HSP	symmetric, acral distribution, abdominal or joint pain
Drug reactions	nontoxic appearance, history of exposure
Meningococemia	influenza like illness with rapid progression
Gonococemia	purpuric pustules, sparse, acral distribution, fever, arthralgia
ITP	pronounced on dependant areas, mucosal bleeding
RMSF	fever, headache, peripherally distributed
Enteroviruses	non toxic appearance, viral prodrome

FIG. 16-4. Algorithmic approach to a rash with petechiae/purpura.



Impetigo	annular, crust with glistening base (when removed)
Folliculitis	hair in center of lesion
Candidiasis	bright, intensely erythematous, poorly defined borders with satellite pustules

FIG. 16-5. Algorithmic approach to a rash with pustule primary lesion.

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QUESTIONS

1. A 6-year-old female presents to the ED complaining of a rash and fever. The mother states that she had been in her usual state of good health until

1 week ago when she developed bronchitis and was given an amoxicillin. There are nonpalpable target like macules and few bullae with a positive Nikolsky sign. You note several lesions on the oral mucosa. What is the next step of management in this patient?

- Reassurance and follow up with PMD
 - Discontinue amoxicillin; tell the patient to avoid beta lactam antibiotics in the future, and d/c home.
 - Initiate fluid resuscitation, discontinue amoxicillin, dermatological consultation, and admit to burn unit
 - Apply a low potency steroid cream to the affected area.
 - Give the patient 2 mg/kg of prednisolone PO.
2. A 5-year-old male presents to the ED with a complaint of rash. You note several distinct lesions with central clearing and a target like appearance. There are no blisters, a negative Nikolsky sign and no mucous membrane involvement. The child is nontoxic appearing and playing happily on the exam room floor. What is the most likely diagnosis?
- Hansen disease
 - Erythema multiforme
 - Meningococemia
 - Stevens–Johnson syndrome
 - Klein–Levin syndrome
3. A 6-month-old otherwise healthy female is brought to the emergency department at 2 am with a complaint of fever. The mother states that she gave the patient Tylenol and Motrin, but that she checked the temperature and it was 101.2°F. The patient is lethargic and tachycardic. You note 2 petichiae on the patient’s abdomen. What is the next appropriate step in management?
- Antipyretics have patient follow up with PMD
 - Blood cultures, LP, antibiotics, fluid resuscitation
 - Surgical consultation for laparotomy
 - CT scan of head, neurological consultation
 - A short course of systemic steroids
4. A 3-year-old male presents with 6 days of a fever. His mother brought him to the emergency room because despite completion of the course of antibiotics her doctor prescribed, the fever has persisted. On examination, you find there to be bilateral conjunctival injection, cervical lymphadenopathy, a strawberry tongue and edema of the hands. What laboratory test should confirm the diagnosis?
- Complete blood count
 - Paired serum and blood IgG

- Western Blot
- All of the above
- None of the above

ANSWERS

- C. This patient has a disorder on the spectrum of Stevens Johnson syndrome or TEN. The patient should be admitted to the hospital burn unit or ICU as her course can be expected to worsen. Steroids have no role in the treatment of SJS or TEN, however cyclosporine and IViG have been used, and however, this therapy generally would not be started in the emergency department.
- B. The patient has target like lesions with no mucosal involvement, does not have full thickness lesions, and is nontoxic appearing, the most likely diagnosis is therefore erythema multiforme minor. This is on the same spectrum of illness as SJS and TEN, but is much less severe and usually self-limited.
- B. This patient likely has early meningococemia, and is likely to quickly deteriorate. Cultures and LP should be done before antibiotic administration, but antibiotic administration should not be delayed.
- E. This patient has Kawasaki disease, for which there is no reliable laboratory test; diagnosis is based on clinical features.

17 NECK MASSES

Raemma Paredes Luck
Robert W. Schafermeyer

NECK MASS

- The emergency physician often must evaluate an infant or child with a neck mass.
- Most of these neck masses are benign and result from reactive lymph nodes caused by viral infections.
- The challenge is to distinguish between the pathologic lesions that need expeditious management versus those neck conditions that are benign but still cause parental anxiety.
- Important elements of the history and physical examination will be highlighted and a discussion of the differential diagnoses and management options will be presented (see Fig. 17-1).

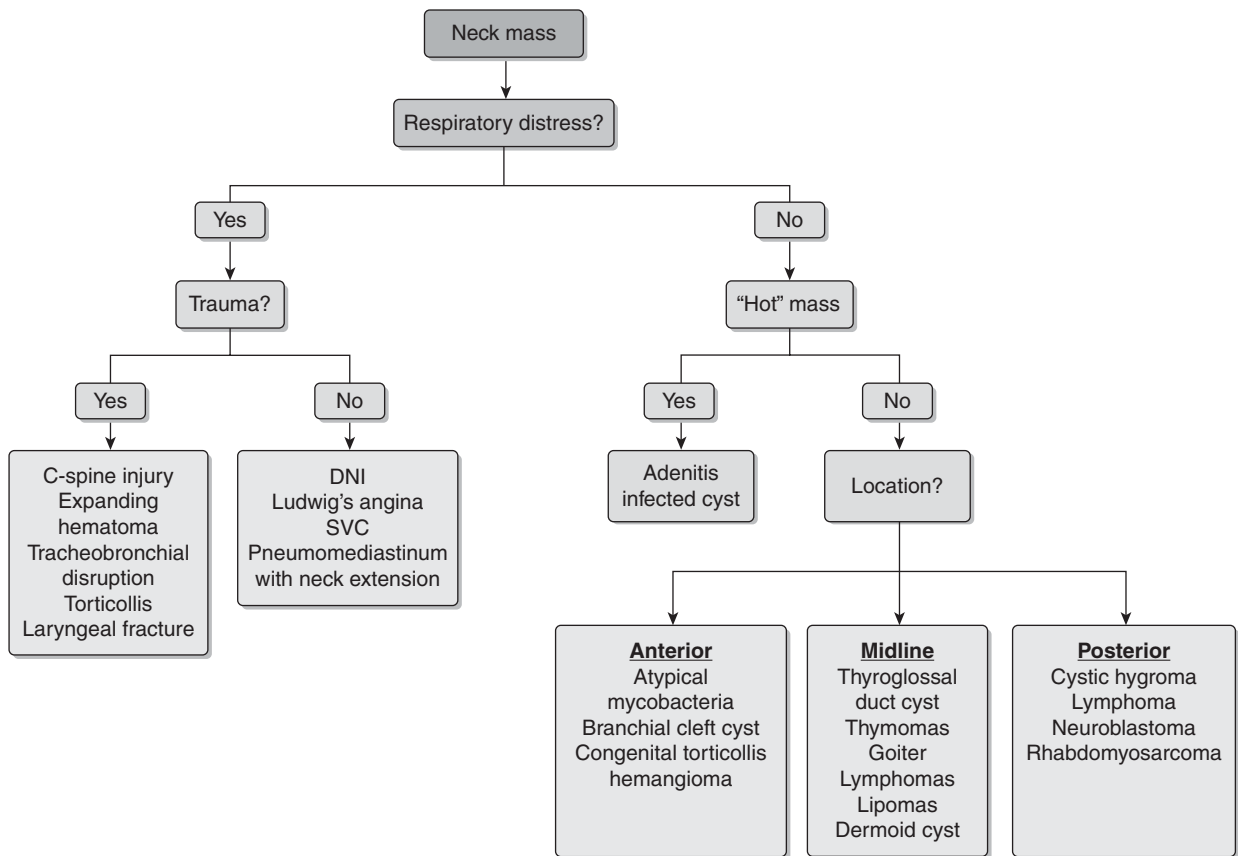


FIG. 17-1. Algorithm on the management of a child with a neck mass.

ANATOMY

- There are several anatomic classifications used to describe the location of neck lesions. Simple method is to divide the neck into two compartments or triangles with the sternocleidomastoid muscle as the common boundary (Fig. 17-1).
- The anterior compartment is defined by the anterior border of the sternocleidomastoid muscle, the lower border of the mandible, the sternum inferiorly, and a line extending from the submandibular symphysis to the sternal notch.
- Vital structures in this compartment include the larynx, trachea, esophagus, the thyroid and parathyroid glands, the carotid sheath, and the suprahyoid and infrahyoid muscles. Lymph node chains in this area include the jugulodigastric chain that lies anterior to the sternocleidomastoid muscle.
- The posterior compartment is defined inferiorly by the clavicle, laterally by the trapezius, and medially by the sternocleidomastoid muscle. Structures in this area include the subclavian vessels, cervical roots of the brachial plexus, spinal accessory nerve, and several lymph node chains.

- Knowledge of the anatomy of the neck and the specific regions drained by the lymph node group will help the clinician in locating the primary infection.
- The posterior part of the tongue, tonsils, sinuses, nasopharynx, larynx, and pharyngeal regions drain into the superficial and deep anterior cervical lymph nodes.
- The anterior scalp, ear canal, pinna, and the conjunctiva drain into the preauricular lymph nodes.
- The temporal and parietal scalp regions drain into the postauricular nodes and the posterior scalp region drain into the occipital nodes.
- The retropharyngeal, prevertebral, and parapharyngeal spaces are potential spaces and an infection in one of these spaces can spread to the other spaces through the fascial planes and cause neck swelling.
- The short neck of a child relative to the large head makes it less susceptible to direct trauma. However, because a large number of vital structures are located in such a small area, even minor neck injuries can become potentially life threatening.

ASSESSMENT OF A PATIENT WITH A NECK MASS AND IN RESPIRATORY DISTRESS

- An infant or a child presenting with a neck mass and respiratory distress should be managed according to the pediatric advanced life support guidelines. Those with a history of a major traumatic injury should be managed according to the advanced trauma life support guidelines.
- Do not force a pediatric patient in respiratory distress to lie down, as it may precipitate cardiopulmonary arrest from a possible retropharyngeal or mediastinal mass compressing the trachea.
- Maintaining the airway is a top priority in a child with a neck mass and respiratory distress.
- Depending on the level of obstruction, administration of oxygen, a nasopharyngeal or oral airway, mask ventilation, or a laryngeal mask airway may be used to support a compromised airway.
- Endotracheal intubation may be necessary either using direct laryngoscopy or fiber-optic methods. A transtracheal jet ventilation may be lifesaving, although can be technically challenging in the young child. An urgent tracheotomy in an unsecured pediatric airway carries considerable risk and should be employed only when other methods fail.
- Pursue the cause of the patient's neck mass once the patient is stabilized with a brief history and a more detailed physical examination. Do a complete examination of the head region, note the consistency, size, and location of the neck mass, any deviation of the trachea, and presence of ecchymosis or subcutaneous emphysema.
- The progression of the neck mass and symptoms such as neck pain, dysphagia, stridor, drooling, hemoptysis, or dysphonia should be noted. Also note any puncture wound, however minor, since this can be misleading and may result in a delay in diagnosis of significant injuries.
- A history of trauma to the neck in a child, who is in distress, should make the clinician consider an expanding hematoma, tracheobronchial disruption, laryngeal injuries, or cervical spine injuries. These conditions are discussed more in depth in Chapter 30.
- Suspect a deep neck infection in a child presenting with dysphagia, stiff neck, trismus, stridor, or muffled speech especially in the absence of trauma.
- Deep neck space infections involving the retropharyngeal and parapharyngeal spaces are potentially life-threatening conditions that continue to be seen (Fig. 17-2).
- These infections originate from a tonsillitis, pharyngitis, or sinusitis that had spread through the regional

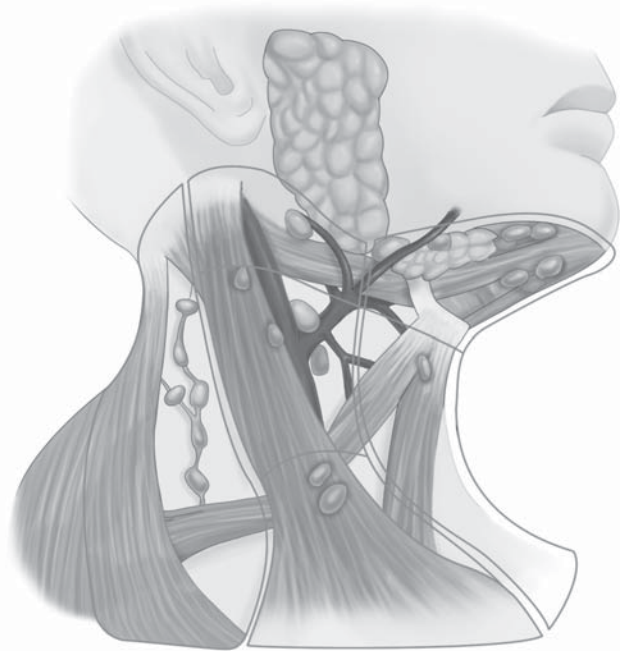


FIG. 17-2. Triangles of the neck and location of lymph nodes.

lymph nodes of the neck. In many cases the etiology cannot be found. The usual organisms cultured are staphylococci, streptococci as well as anaerobic organisms.

- Complications of deep neck infections include spontaneous rupture into the pharynx, extension to the lateral side of the neck, or dissection into the mediastinum and prevertebral space leading to aspiration, airway obstruction, compression of major blood vessels, and death. A suspicion of a possible deep neck infection calls for early consultation with our surgical colleagues.
- Other causes of respiratory distress in an infant with a neck mass are cystic hygromas and hemangiomas (Table 17-1). Cystic hygromas can extend from the tongue to the mediastinum and in rare cases, rapidly enlarge, causing extrinsic compression of the adjacent vital structures. Such rapid enlargement can occur secondary to trauma, infection, or hemorrhage into the cyst.⁷

ASSESSMENT OF A PATIENT WITH A NECK MASS IN NO DISTRESS

- The first step in the evaluation of a neck mass is a thorough history and physical examination. Details help narrow the differential diagnoses.
- The age of the child is important because normal lymph node sizes vary with age.

TABLE 17-1 Life-Threatening Causes of Neck Masses

TRAUMATIC	INFLAMMATORY	CONGENITAL	NEOPLASTIC
Subcutaneous emphysema from tracheal/pulmonary injury	Deep neck infections	Large cystic hygroma	Hodgkin's lymphoma with mediastinal extension
Expanding hematoma	Ludwig's angina Infected cysts		

- In the newborn period, lymph nodes are not palpable, as the infant has not been exposed to many antigens. Neck masses noted in this age group will be mostly congenital in origin.
- Over time, the size of the lymph nodes increases such that palpable or “shotty” nodes up to 1 cm in size in the cervical, axillary, and inguinal areas are considered normal and frequently seen without signs of infection.
- The duration of the neck mass is important. A painless mass noted since birth or shortly after birth suggests a congenital lesion. However, some congenital lesions may not be apparent until years later when there is an obstruction or infection causing it to mimic an inflammatory condition.
- A history of neck pain, hoarseness, stridor, or dysphagia is worrisome and may be due to a mass compressing on a nerve or a vital organ and therefore needs a more emergent workup.
- Presence of systemic symptoms such as fever, night sweats, fatigue, and weight loss is significant and suggests a systemic disease such as tuberculosis, sarcoidosis, or malignancy.
- A history of travel to another country or endemic area, exposure to animals, or ingestion of unpasteurized milk may also provide some clues to the cause of the neck mass.
- Note any limitation in the range of motion of the neck as well as tilting toward one direction.
- The size and consistency of the mass, its mobility, and location are important as congenital lesions consistently appear at certain locations. For example, most branchial cleft sinuses or cysts are located in the lateral neck at the anterior border of the sternocleidomastoid muscle.
- Thyroglossal duct cysts are usually located in the midline anywhere between the base of the tongue and the thyroid. These lesions move with tongue protrusion.
- A soft, nontender, compressible mass in the posterior triangle of the neck is highly suggestive of a cystic hygroma while hemangiomas can be located anywhere in the neck.
- Supraclavicular lymphadenopathy in any age group is a serious concern and should be promptly investigated for an underlying malignancy. Other characteristics of a neck mass that should increase suspicion

of a malignancy include the presence of irregular margins, hard consistency, size of more than 3 cm, adherence to surrounding areas and association with other systemic symptoms.

- Examination of the rest of the body is important to detect clues that will help in narrowing the differential diagnoses. For example, generalized lymphadenopathy is caused by a systemic illness such as human immunodeficiency virus (HIV) infection or tuberculosis.
- Splenomegaly in a child with exudative pharyngitis and enlarged cervical lymphadenopathy suggests infectious mononucleosis.

DIFFERENTIAL DIAGNOSES

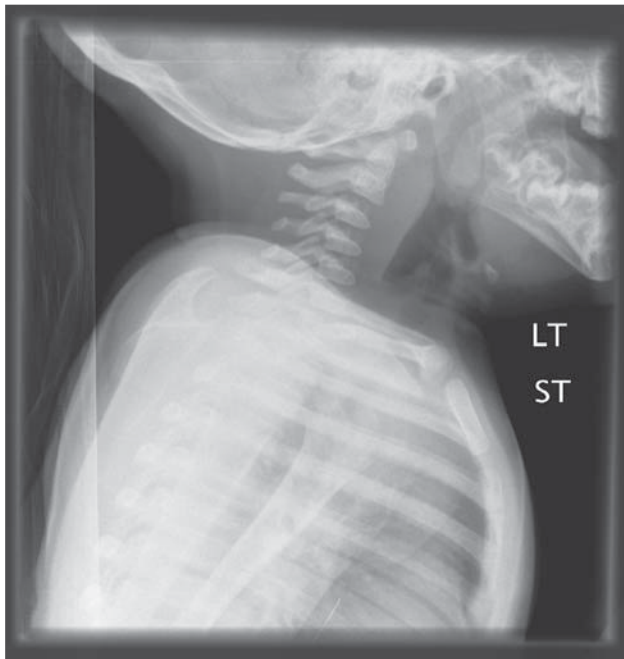
- Adults with an asymptomatic neck mass should be considered to have a malignancy until proven otherwise.
- In contrast, majority of neck masses in children are benign in nature and can be classified into four general categories: inflammatory, congenital, neoplastic, and traumatic conditions (Table 17-2).

INFLAMMATORY CONDITIONS

- The vast majority of pediatric neck masses fall under this category. Viral upper respiratory tract infection is the most common cause of bilateral cervical lymph node enlargement. It is not unusual for viral infections to cause nodal enlargement in two or more noncontiguous sites.
- Most cases of cervical lymphadenopathy are reactive in nature, self-limited, and require no treatment.
- Bacterial infections of the head and neck often cause unilateral lymph node enlargement. Cervical adenitis results when this enlargement is accompanied by local tenderness, redness, and warmth. This is usually accompanied by systemic symptoms such as fever and irritability.
- Because 80% of all cases of acute adenitis are caused by *Streptococcus* and *Staphylococcus*, the antimicrobial therapy of choice is a first-generation cephalosporin, oxacillin, or clindamycin. Anaerobic

TABLE 17-2 Differential Diagnoses of Neck Masses by Location and Etiology

	ANTERIOR TRIANGLE	MIDLINE	POSTERIOR TRIANGLE
Inflammatory	Adenitis from various causes Reactive adenopathy Parotitis Atypical mycobacteria	Adenitis Thyroiditis Ludwig's angina	Adenitis Sialadenitis
Congenital	Branchial cleft cyst Laryngocoele Congenital torticollis	Thyroglossal duct cyst Dermoid cyst	Cystic hygroma
Neoplastic	Hemangioma Neurogenic tumors Salivary gland tumors	Thymomas Lymphoma Lipoma Goiter	Lymphoma Metastatic lesions Neuroblastoma Rhabdomyosarcomas
Traumatic	Hematoma Acquired torticollis	Laryngeal fracture	Hematoma Acquired torticollis

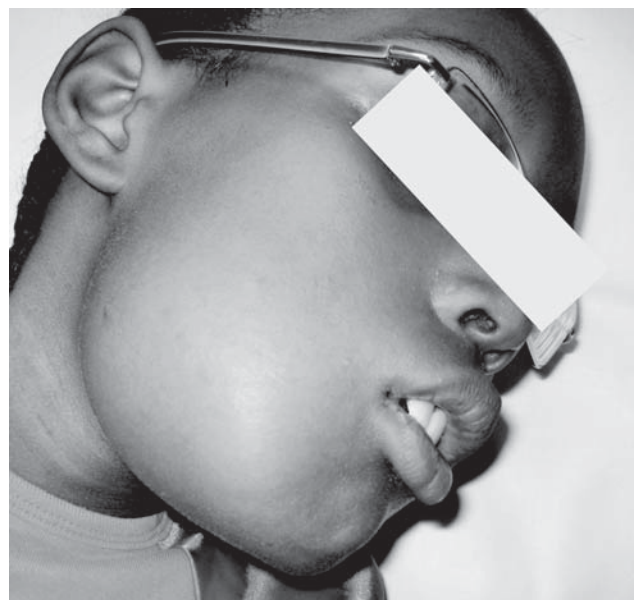
**FIG. 17-3.** Lateral neck of a child with a retropharyngeal abscess showing widening of the retropharyngeal space.

organisms should be considered when the cause of the adenitis is from an odontogenic infection.

- Failure to respond to oral antibiotics, abscess formation, concomitant cellulitis, or systemic toxicity necessitates hospitalization for intravenous antibiotics (Figs. 17-3 and 17-4).
- Computed tomography (CT) of the neck with contrast or ultrasonography is helpful in distinguishing a lesion that has progressed into an abscess.
- A patient with a neck mass that is minimally tender, nonfluctuant, slowly enlarging over a few days or weeks with no obvious source or systemic symptoms will have a wide list of differential diagnoses and

consideration should be given to atypical mycobacterium as well as mycobacterium tuberculosis infection, infectious mononucleosis, cat scratch disease, HIV infection, sarcoidosis, actinomycosis, and toxoplasmosis.

- If a patient has not been treated, a course of an appropriate oral antibiotic is indicated with close follow-up to assess response to treatment.
- Ancillary testing may be helpful. This may include a complete blood count and a chest radiograph to detect pulmonary infiltrates or mediastinal adenopathy.
- Persistence of the mass despite antibiotics, suspicion of malignancy, or other esoteric etiology necessitates a referral to an infectious disease specialist or to an otolaryngologist for a biopsy.

**FIG. 17-4.** Right submandibular neck abscess.

- Atypical mycobacteria, also known as nontubercular mycobacteria and, in particular, mycobacterium avium complex, present as a chronic cervical adenitis and occurs in children between 1 and 5 years of age. The bacteria gain entry from a breakdown in the mucous membranes of the oropharynx and tonsils and then invade regional lymph nodes.
- The usual presentation is that of an enlarged lymph node in the submandibular region that has a rubbery consistency, minimal tenderness, and a dull reddish color. Occasionally, a draining sinus is present.
- The treatment of choice is complete surgical resection or curettage of the node as most atypical mycobacteria respond poorly to antibiotics.
- *Bartonella henselae*, the organism responsible for cat scratch disease, is a common cause of regional lymphadenopathy and usually involves the axillary area but occasionally can affect the cervical, epitrochlear, or inguinal nodes.
- Recent contact with a cat or kitten can be obtained in the majority of patients. A papule at the site of the inoculation is frequently noted, followed in 1 to 2 weeks by the development of tender, indurated, erythematous skin overlying the enlarged node in the lymphatic chain that drains the site of infection. Systemic symptoms such as fever and malaise are seen in about a third of the patients.
- In patients with systemic involvement or painful adenitis, antibiotics such as trimethoprim-sulfamethoxazole, rifampin, azithromycin, ciprofloxacin, or parenteral gentamicin may be effective in ameliorating symptoms.
- Other inflammatory conditions to consider are infections of the salivary glands or sialadenitis. This can present as a tender and swollen mass in the area of the submandibular or parotid glands. Occasionally, the condition can be bilateral as in parotitis caused by the mumps virus.
- Majority can be treated with conservative measures such as hydration, pain relief, application of moist heat, and sialogogues. If you suspect bacterial superinfection, broad-spectrum antibiotics effective against staphylococcus, streptococcus, and anaerobic flora should be initiated.

TRAUMATIC CONDITIONS

- Minor trauma to the neck and a variety of other conditions can cause spasm of the cervical muscles, primarily the sternocleidomastoid.
- Underlying etiologies of torticollis include upper respiratory infection, cervical adenitis, retropharyngeal abscess, atlantoaxial rotatory subluxation and

rarely, dystonic reactions or intracranial and spinal cord tumors. Major traumatic injuries to the neck are covered in Chapter 30.

- Congenital torticollis is suspected when an infant, usually at 2 to 8 weeks of age, presents with an ipsilateral neck mass with the head tilted toward it and the chin in the opposite direction. The cause of congenital torticollis is still not clear, but may be related to bleeding into the sternocleidomastoid muscle from a difficult delivery. The onset of marked facial hypoplasia or asymmetry is an indication for surgical transection of the middle third of the affected sternocleidomastoid muscle.

CONGENITAL MASSES

- Hemangiomas are the most common congenital lesions of the head and neck and are red or purplish in color, flat or raised, and blanch with pressure. They grow rapidly in the first few months of life, slowly regress afterwards, and may disappear with time.
- Close observation is the rule unless the lesions cause airway obstruction, high-output cardiac failure, thrombocytopenia or hemorrhage, and coagulopathy (Kasabach–Merritt syndrome).
- Thyroglossal duct cysts arise from the vestiges of the thyroglossal duct that runs in the middle of the neck from the base of the tongue to the thyroid gland (Fig. 17-5). These cysts enlarge after bouts of upper respiratory infections.
- One clue aside from its midline location is that protrusion of the tongue will retract the lesion. Majority of these cysts manifests between the age of 2 and 10 years, although a third do not become apparent until after the second decade of life.
- Infection is common because of the persistent communication with the base of the tongue and the oral flora. Once the infection has been treated, surgical excision to remove the cyst and its entire tract can follow.
- When primordial lymphatic ducts fail to establish drainage into the venous system, multiloculated cystic masses or cystic hygromas are formed. They are found in areas where lymphatic ducts drain into large veins such as the neck, axilla, and mediastinum.
- The left side of the neck is frequently involved because it is where the thoracic duct enters the subclavian vein. Majority of these are identified at birth, although some may not be diagnosed until the second decade of life or if they become infected. There is a strong association between congenital cystic hygromas of the neck and Turner syndrome.



FIG. 17-5. Patient with thyroglossal duct cyst.

- Anomalous development of the branchial arches and cleft produces brachial cysts, sinuses, fistulae, or skin tags in the neck. Most branchial cleft anomalies originate from the second cleft. These painless cysts or sinuses are usually seen at the anterior border of the middle to lower third of the sternocleidomastoid muscle. They can become secondarily infected.
- Brachial cysts are more commonly diagnosed after the first decade of life while fistulas are usually diagnosed shortly after birth.

NEOPLASTIC MASSES

- Malignant neoplasms of the head and neck account for approximately 5% of all malignancies in childhood. Ninety percent of neck cancers in children are mesenchymal in origin, in contrast to adults where the squamous cell line is involved.
- The most common malignancies of the neck are lymphomas (Hodgkin and non-Hodgkin lymphomas) and soft tissue sarcomas, primarily rhabdomyosarcoma.

- The neck is second to the orbit as a common site of rhabdomyosarcoma. The tumor presents as a rapidly enlarging, painless neck mass that is hard and immobile.
- Other tumors include neuroblastoma and lymph node metastasis from malignancies of the skin and thyroid. These masses tend to be hard and fixed to the underlying structures.

ANCILLARY TESTING

- The vast majority of neck masses are the result of inflammatory or infectious causes that are detected by history and physical examination. Laboratory testing is often not necessary.
- A neck mass that is rapidly enlarging and does not respond to the standard antibiotic regimen or has been present for a few weeks needs further laboratory and radiographic evaluation to narrow the cause. A complete blood count with differential, and either erythrocyte sedimentation rate or C-reactive protein may be obtained.
- Chest radiographs can detect pulmonary infiltrates or mediastinal involvement in patients suspected of having tuberculosis, sarcoidosis, or primary lung tumors. CT or magnetic resonance imaging (MRI) can help differentiate vascular, solid, or cystic lesions. They can delineate the location of a mass and its relation to other structures in the neck.
- Occasionally, the CT scan or MRI can identify a source of infection or the primary tumor. For deep pediatric neck infections, the accuracy of CT scans in differentiating between an abscess and a cellulitis has recently been questioned by several investigators, with one study showing an overall accuracy of only 63%. Ultrasonography has also been used in differentiating solid from cystic lesions.
- Children with chronic or subacute lymphadenopathy should have a purified protein derivative (PPD) tuberculin test done to exclude mycobacterial infections. However, in atypical mycobacterial infections, majority of the chest radiographs are normal and the PPD are usually negative or intermediate.

DISPOSITION

- When the history and physical examination point to a benign condition, the emergency physician should provide parental reassurance.
- Cervical lymphadenopathy in response to specific infections is the most common cause of neck masses. Treatment of the underlying cause is sufficient in most cases.

- A patient with an acute “hot” neck node deserves a trial of antibiotics with close follow-up. Indications for hospitalization include failure of oral antibiotics with progression of symptoms, suspicion of a deep neck infection, systemic toxicity, or respiratory distress.
- Indications for a surgical referral include persistence of the neck mass after 4 weeks; an increase in size during that time; mass size of more than 3 cm; supraclavicular location; and systemic signs and symptoms suggestive of a malignancy.

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QUESTIONS

1. A 1-year-old presents with an anterior neck mass that you determine is located at the lateral neck along the anterior border of the sternocleidomastoid muscle. You determine that this is one of the following:
 - A. Cystic hygroma
 - B. Thyroglossal duct cyst

- C. Brachial cleft cyst
 - D. Lymphoma
 - E. Anterior lymph node
2. A 3-year-old with a URI for 4 days presents with an earache and you note a group of posterior cervical nodes all under 1 cm in size. On exam the TMs are normal. What treatment if any should be given for the cervical nodes?
 - A. Penicillin Vee K
 - B. Zithromycin
 - C. Motrin and follow up
 - D. Rocephin
 - E. Excisional biopsy
 3. A 4-year-old has a sore throat, neck pain and fevers to 103°F (39.5°C). You palpate both a 2 and 1.5 cm anterior node on the right. They are red, hot and tender. Your treatment includes
 - A. Incision and drainage
 - B. Keflex
 - C. Trimethoprim-sulfamethoxazole
 - D. Vancomycin
 - E. Hot compresses
 4. A 5-year-old comes in with a 3-week history of a painless left sided neck mass. You note that it is hard and is 3 cm in diameter. What treatment will you prescribe?
 - A. Keflex for 2 weeks
 - B. Hot compresses and follow up in 2 weeks
 - C. Incision and drainage
 - D. CT Scan of the neck
 - E. Admission for IV antibiotics

ANSWERS

1. C. The size and consistency of the mass, its mobility, and location are important as congenital lesions consistently appear at certain locations. For example, most branchial cleft sinuses or cysts are located in the lateral neck at the anterior border of the sternocleidomastoid muscle. Thyroglossal duct cysts are usually located in the midline anywhere between the base of the tongue and the thyroid. A soft, nontender, compressible mass in the posterior triangle of the neck is highly suggestive of a cystic hygroma while hemangiomas can be located anywhere in the neck. Supraclavicular lymphadenopathy in any age group is a serious concern and should be promptly investigated for an underlying malignancy. Other characteristics of a neck mass that should increase suspicion of a malignancy include the presence of irregular margins, hard consistency, size of more than 3 cm, adherence to

surrounding areas and association with other systemic symptoms.

2. C. The vast majority of pediatric neck masses are due to inflammatory or infectious causes. Viral upper respiratory tract infection is the most common cause of bilateral cervical lymph node enlargement. It is not unusual for viral infections to cause nodal enlargement in two or more non-contiguous sites. Most cases of cervical lymphadenopathy are reactive in nature, self-limited, and require no treatment.
3. B. Bacterial infections of the head and neck often cause unilateral lymph node enlargement. Cervical adenitis results when this enlargement is accompanied by local tenderness, redness, and warmth (Fig. 17-3). This is usually accompanied by systemic symptoms such as fever and irritability. Because 80% of all cases of acute adenitis are caused by *Streptococcus* and *Staphylococcus*, the antimicrobial therapy of choice is a first-generation cephalosporin, oxacillin, or clindamycin. Anaerobic organisms should be considered when the cause of the adenitis is from an odontogenic infection.
4. D. Malignant neoplasms of the head and neck account for approximately 5% of all malignancies in childhood. Ninety percent of neck cancers in children are mesenchymal in origin. The most common malignancies of the neck are lymphomas (Hodgkin and nonHodgkin lymphomas) and soft tissue sarcomas, primarily rhabdomyosarcoma. The neck is second to the orbit as a common site of rhabdomyosarcoma. The tumor presents as a rapidly enlarging, painless neck mass that is hard and immobile. Other tumors include neuroblastoma and lymph node metastasis from malignancies of the skin and thyroid. These masses tend to be hard and fixed to the underlying structures.

18 NEONATAL EMERGENCIES

Jenice Forde-Baker

- In utero, the fetus is protected from metabolic and anatomic anomalies by its relationship with the placenta and maternal circulation. Once delivery occurs, the child must be self-sustaining; and if a congenital problem exists, maternal compensatory effects are no longer available. These congenital abnormalities may not present themselves until hours to weeks after birth.

- The neonatal patient presents a diagnostic challenge for the emergency physician and, thus, neonatal management is generally protocol driven and should include rapid glucose determination, pulse oximetry, complete blood count, basic metabolic blood tests, urine analysis, urine culture, blood cultures and chest x-ray. Additional diagnostics studies may be ordered as the clinical scenario dictates (Fig. 18-1).

CARDIORESPIRATORY PRESENTATIONS

- In utero congenital anomalies are masked by shunts and venous/arterial blood mixing.
- At birth the foramen ovale and ductus arteriosus begin closure and are complete by 7–10 days.
- Because of a neonate's limited number of activities, presenting symptoms are rather vague and can occur days after discharge from the hospital.

CYANOTIC CARDIAC LESIONS

- Most are due to obstruction of pulmonary blood flow or separation of pulmonary and systemic circulations.
- Clinical presentations include dusky appearance, lethargy, and poor feeding.
- Hyperoxygenation test compares ABG on room air with an ABG on 100% oxygen for ten minutes. If supplemental oxygen does not improve hypoxia, it reveals a fixed cardiac lesion.

PULMONARY LESIONS

- Pulmonary lesions are the result of shunting of systemic circulation to pulmonary circulation, causing left to right shunt.
- Excessive pulmonary circulation leads to congestive heart failure.
- Clinical presentations include tachypnea, tachycardia, rhonchi, and hepatomegaly.
- Supplemental oxygen can improve CHF symptoms.
- Viral and bacterial infections can also cause respiratory failure.

VENTRICULAR DYSFUNCTION

- Obstructive/stenotic lesions limit systemic outflow (ie, coarctation of the aorta, aortic stenosis).
- A hypoplastic left ventricle leads to inadequate systemic flow.

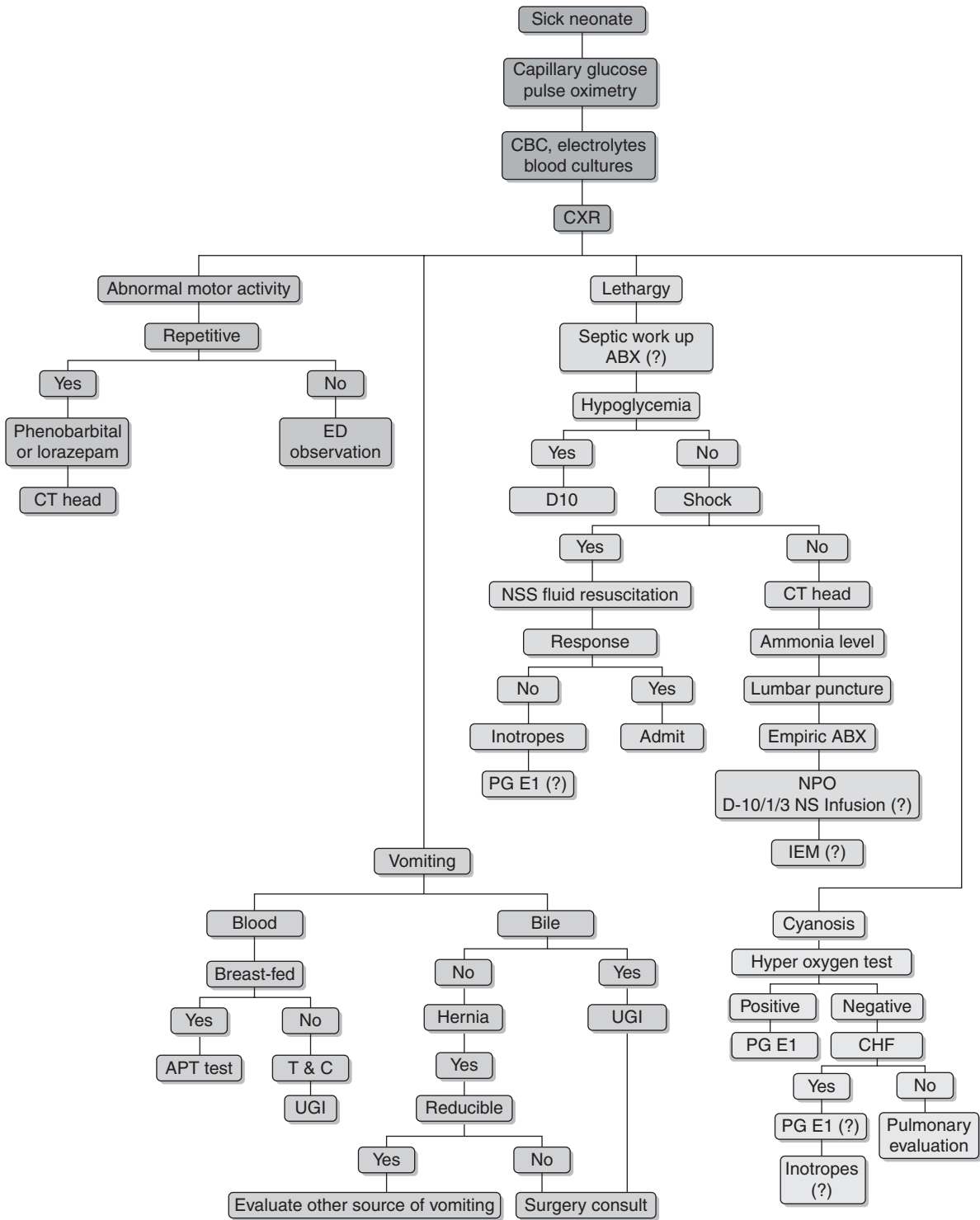


FIG. 18-1. A summary of the approach to the sick neonate. PGE₁, prostaglandin E₁; UGI, upper GI with oral contrast; ABX, antibiotics; IEM, inborn error of metabolism.

TABLE 18-1 Cardiac Lesions Presenting in First Month of Life

PRESENTATION TYPE	CARDIAC LESION
Cyanotic lesions	Tetralogy of Fallot (TOF)
	Transposition of great arteries (TGA)
	Tricuspid atresia (TA)
	Total anomalous pulmonary venous return (TAPVR)
	Truncus arteriosus
Increased pulmonary blood flow	Pulmonary stenosis (PS)
	Atrial septal defect (ASD)
	Ventricular septal defect (VSD)
	Patent ductus arteriosus (PDA)
Left ventricular outflow obstruction or ventricular artesia	Endocardial cushion defect (AV Canal)
	Aortic stenosis (AS)
	Coarctation of aorta
	Hypoplastic left heart syndrome (HLHS)

- Neonates usually present in shock, pallor or ashen color, slow or absent capillary refill and low pulse oximetry. (Table 18-1)

EMERGENCY TREATMENT

- Initial treatment is primarily aimed at restoring fetal circulation through reestablishment of a patent ductus arteriosus.
- Opening an anatomic/physiologic shunt between the pulmonary and systemic circulations permits admixing of oxygenated and deoxygenated blood.
- Restoration of the ductus arteriosus is accomplished through a continuous infusion of prostaglandin E₁ (PGE₁) (0.05–0.1 µg/kg/min).¹ The effect of the prostaglandin may be immediate but frequently takes at least 15 minutes to be seen.
- Prostaglandin should be given if any neonate fails a hyperoxia test or demonstrates severe acute CHF.
- Inotropic support with milrinone, dopamine or dobutamine may be needed if prostaglandin infusion fails.
- Antibiotics and supplemental oxygen support is needed for pulmonary infections.
- Endotracheal intubation should be used if respiratory deterioration or failure ensues.

ENDOCRINE AND METABOLIC DISORDERS

CONGENITAL ADRENAL HYPERPLASIA (CAH)

- Congenital adrenal hyperplasia results from a single enzyme defect, most commonly 21-hydroxylase, in the adrenal steroid genesis pathway.

- These patients have profound salt wasting that leads to severe hyponatremia and hyperkalemia.
- The most common emergency presentation involves dehydration, hypoglycemia, hyponatremia, and hyperkalemia.
- Initial stabilization of these neonates will require vascular access and routine laboratory studies.
- Rapid rehydration at 20 mL/kg of normal saline will generally be sufficient to treat the dramatic hyperkalemia.
- Hypoglycemia is treated with a 10% dextrose slow bolus or infusion.
- Treat hyponatremia with 3% NaCl only if neurological symptoms.
- Hydrocortisone at 25 mg/m² is needed due to is a deficiency of this hormone in CAH.

INBORN ERRORS OF METABOLISM (IEM)

- IEMs are associated with well-defined clinical syndromes and are generally identified early in infancy. Some, however may not present until the patient undergoes a stressful condition such as an infection, dehydration or starvation.
- In contrast, those conditions associated with a single hormone or enzyme defect may go undetected until a toxic metabolite accumulates or an electrolyte or endocrine catastrophe occurs (Table 18-2).
- Presentation will vary given the different areas the toxic metabolites affect, and may be similar to neonatal sepsis.
- Along with the septic evaluation, venous blood gases, liver function panels and ammonia levels should be checked.

TABLE 18-2 Inborn Errors of Metabolism

TYPE	CHARACTERISTIC FINDINGS
Acute metabolic encephalopathy	Lactic acidosis
	Organic acidoses
	Hypoglycemia
	Hyperammonemia
	Amino acid disorders
Jaundice—severe liver dysfunction	Galatosemia
	Tyrosinemia type I
	Fructose intolerance
Cardiac disease	Long-chain fatty acid oxidation disorders
Encephalopathy with seizures	Nonketotic hyperglycinemia
Phenotypic distinctive disorders	Lysosomal disorders
	Glycosylation disorders
	Peroxisomal disorders
Severe hypotonia	Nonketotic hyperglycinemia
	Congenital lactic acidosis
	Congenital disorders of glycosylation
	Lysosomal storage disorders
Nonimmune hydrops fetalis	

- Treatment is aimed at rehydration with electrolyte solutions containing 10% glucose to promote conversion from a catabolic to an anabolic state.

GASTROINTESTINAL

- Vomiting is the most common presenting complaint of neonates with GI abnormalities.
- Pyloric stenosis may present in the neonate and is nonbilious projectile vomiting. It classically presents with a hypochloremic, hypokalemic, and metabolic alkalosis in the face of pronounced dehydration.
- Bilious/bloody emesis is always an ominous finding and warrants early surgical consultation.
- Causes include intestinal malrotation, volvulus, duodenal atresia, peritoneal bands, annular pancreas, and Hirschsprung disease.
- The APT test may be used to distinguish maternal blood from neonatal blood in emesis.
- Evaluation with complete blood cell count, type and screen, electrolyte panel, liver panel and upper GI series can help in diagnosis.
- Bloody diarrhea is also a dangerous sign as it indicates possible bowel ischemia (Fig. 18-2).

SEIZURES

- Perinatal asphyxia is the most common cause of neonatal seizures.
- Cerebral ischemia accounts for 20% of all cases and intracranial hemorrhage, usually subarachnoid, account for another 10%.



FIG. 18-2. Bloody diarrhea in a 5-day-old infant secondary to malrotation and mid gut volvulus.

- Other causes may be infectious, metabolic and or inborn errors of metabolism.
- Initial management includes a CT scan of the head and lumbar puncture.
- Empiric antibiotics should be given without any radiographic delay.
- Initial seizure management includes phenobarbital (20 mg/kg), or lorazepam 0.05 mg/kg) and refractory seizures may require phenytoin (20 mg/kg).
- Hypoglycemia is managed with 2–4 mL/kg boluses of 10% dextrose.
- Hypocalcemia is managed with 4 mg/kg of 5% calcium gluconate.
- Hypomagnesemia is treated with 50% magnesium sulfate at 0.2 mL/kg IM.
- Hyponatremia in a seizing neonate is treated with 1 mL/kg aliquots of 3% saline until the seizure abates.

SURGICAL EMERGENCIES

- Hernias are the most common source of masses occurring in 5% to 30% of low-birth-weight individuals. These present as a lump in the inguinal area of the neonate. Irreducible hernias require a surgical consult, while reducible hernias can have outpatient management.
- Testicular torsion is common in the neonatal period and acutely presents as a red tender swollen hemiscrotum. Ultrasonography confirms the diagnosis and emergent surgical intervention is essential for salvage of the testicle.
- 70% of testicular torsions occur in utero and present as a firm, nontender mass and emergent surgery is not indicated.

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QUESTIONS

- An 8-day-old infant, with history of a normal vaginal delivery, presents to the ED. The parents state that the child has been dusky. Vital signs show a neonate with 90% O₂ saturation, HR 200. What test can immediately help distinguish if there is a fixed cardiac lesion?
 - The Apt test
 - The hyperoxygenation test
 - The A-a gradient test
 - The Winters' formula
 - The anion gap test
- A neonate presents with cyanosis, labored breathing and tachycardia. Initial physical examination reveals, JVD, absent capillary refill and moderate lethargy. Supplemental oxygen has not improved the child status. What should be done next?
 - Dopamine
 - 20 mL/kg of normal saline
 - Prostaglandin infusion
 - Blood transfusion
 - Surfactant
- Congenital adrenal hyperplasia, CAH, crisis presents with severe dehydration, hyponatremia and what other laboratory findings?
 - Hypoglycemia
 - Hyperkalemia
 - Hyperglycemia
 - Hypokalemia
 - Both A and B
- A 2-week-old presents to the emergency department and the parents state that their child has been vomiting a lot. While in the ED during a feeding trial, bilious emesis is noted. Which disposition is safest for the neonate?
 - Call the pediatricians office for a visit the next morning
 - Check blood work and if normal discharge
 - Have an immediate surgical evaluation
 - Make a surgical appointment within 2–3 days for the child
 - Perform an UGI series
- A 9-day infant is brought in by a concerned mother. After the last breast-feeding, the mother noticed blood in some of the milk the baby burped. What test can be done to differentiate mother's blood from infant blood?
 - Apt test
 - Hematocrit
 - Blood type and Rh
 - HemmoCult
 - GastroCult
- A neonate is born prematurely at 34 weeks. During the newborn examination the child begins to seize. What is the most common cause of this seizure?
 - SIDS
 - Hypoxemia
 - Shaken baby syndrome
 - Hypoglycemia
 - Moro reflex
- A seizing neonate presents to the emergency department. Vital signs are a temperature of 99.4, 94% O₂ saturation, 175 beats per minute for heart rate (HR). What are the next steps in management?
 - Head CT
 - Supplemental oxygen
 - Lumbar puncture
 - Phenobarbital 20 mg/kg
 - All of the above
- A 4-week-old infant presents to the ED with inconsolable crying for 2 hours. Vital signs are a temperature of 98.9T, 99% O₂ saturation, HR 180 beats per minute. Physical exam is normal except for a swollen tender scrotal sac. What should be done next?
 - Treat for cellulites of the scrotum
 - Obtain a urine analysis
 - Testicular ultrasound and urological consultation
 - Refer patient for urologic treatment of a hydromel
 - Order CT scan of the abdomen
- A neonate presents with increased sleepiness and decreased po intake according to the mother. Vital signs are temperature of 100.3, 94% O₂ saturation, and HR 200 beats per minute. What should be done in management of this infant?
 - CBC, BMP, UA
 - Blood, urine, and CSF cultures
 - Empiric antibiotics
 - NS IV @ 20 cc/kg
 - All of the above
- A 4-week-old infant presents to the ED with mom with a complaint of constant crying. The child has

not been feeding well and has been vomiting milk violently after meals. Laboratory studies reveal a hypochloremia, hypokalemia and profound metabolic acidosis. What is the likely diagnosis?

- Duodenal atresia
- Imperforated anus
- Pyloric Stenosis
- GERD
- Hirschsprung disease

ANSWERS

- B. As the hyperoxygenation test compares pulse oximetry at room air and 100% supplemental oxygen after 10 minutes. A, the Apt test is a test for maternal blood in neonatal emesis. C, D, and E are calculated formulas for oxygen perfusion, and acidosis.
- C. Any neonate in shock and respiratory distress should be given prostaglandins immediately to help maintain ductus arteriosus flow. All the other choices are indicated for shock after prostaglandins are given.
- E. Both hypoglycemia and hyperkalemia are present in a CAH crisis.
- C. Bilious emesis is an ominous sign and surgery evaluation is the immediate final disposition even if transfer to another hospital is warranted. It most likely represents a malrotation with volvulus or other critical obstructive lesion and any delay in consultation will result in an adverse outcome. Laboratory evaluation is necessary but normal labs should not delay any surgical consultation.
- A. The apt test can determine fetal from maternal blood. B is part of the cell bold count laboratories and C will not answer the question as to whose blood is in the milk. D and E are agents used to detect blood in stool and gastric contents, respectively.
- B. Perinatal asphyxia in is the most common cause of neonatal seizures. SIDS stands for sudden infant death syndrome and C may cause seizures by causing brain damage but is not the most common cause. D is a metabolic cause and E is a normal neonate neurological reflex.
- E. 20% of seizures are caused by intracranial hemorrhage. All the other choices are appropriate for workup and treatment of seizures.
- C. Testicular torsion must be ruled out and urology consultation should be done promptly. A is incorrect because it is a tender swollen scrotum, is not erythematous and torsion must always be excluded from the differential diagnoses. B can be ordered but is not definitive given the physical examination.

D may be done after torsion is ruled out, and E is not needed as this is a urological issue.

- E. This neonate is presenting with fever and as such should have a complete septic workup, which includes answer choices A through D.
- C. A presents with bilious emesis and intestinal obstruction and is a surgical emergency. B is present within the first days of life and characterized by no meconium passage. D is common in the infant population and can be treated with diet modifications and/or H₂ blockers. E is caused by lack of enteric neurons in rectum and/or colon and characterized by abdominal distention and chronic constipation. This may lead to enterocolitis

19 THE TRANSPLANT PATIENT IN THE EMERGENCY DEPARTMENT

Susan M. Scott

Neil Evans

As advances in transplant medicine progress, more and more transplant patients will present to the emergency department (ED) and the pediatric emergency medicine provider must remain aware of the complexities involved in the care of the transplant patient. The transplant service or nearest transplant center should be involved in the patient's care early on during the ED visit. They can serve as an invaluable resource and aid in management and disposition.

COMMON COMPLAINTS PRESENTING TO THE EMERGENCY DEPARTMENT

- The majority of patients will likely require hospitalization sometime during the first 6 months after transplant.
- Common reasons for presentation are fever, rejection, gastrointestinal symptoms, and neurological complaints.
- Fever should be investigated and treated aggressively as most transplant patients are in an immunocompromised state, which may result in rapid deterioration if infection is not detected and treated quickly.
 - Cultures of blood, urine, and when indicated cerebrospinal fluid should be obtained and broad-spectrum antibiotic coverage initiated.
 - Meningitis is a concerning cause of headache in immunocompromised individuals with or without associated neck pain or stiffness.

- Many transplant patients have central venous catheters and these should be considered a source of infection.
- Fever may also be an indicator of graft rejection, which should always be a concern and in the differential of the transplant patient in the ED.
- A wide range of GI symptoms may be encountered including nausea, vomiting, diarrhea, abdominal pain, and bleeding.
- The most common neurological symptom encountered is headache.
 - Common causes of headache encountered in the transplant patient include migraine and tension.
 - Malignant hypertension can also be a concerning cause.
 - Pseudotumor cerebri often is associated with corticosteroid use.

COMMON COMPLICATIONS

- Complications related to medication adverse effects can also lead to an ED visit.
- One of the most common is immunosuppression from maintenance medications with an increased risk of infection.
- The commonly used calcineurin inhibitors like tacrolimus can place a patient at higher risk for nephrotoxicity.
- Corticosteroids, another commonly used transplant maintenance medication, can cause other complications such as hypertension.

IMMUNOSUPPRESSIVE MEDICATIONS

- Corticosteroids are used to induce an immunosuppressed state and to treat acute episodes of rejection.
- Some common adverse effects of corticosteroids include hypertension, headache, psychoses, and hyperglycemia.
- Long-term use can also cause other adverse effects including Cushing's syndrome, growth suppression, decreased bone mineral density, adrenal suppression, glaucoma, and cataracts

AZATHIOPRINE

- Once used as a primary immunosuppressant along with corticosteroids, azathioprine now is used mainly as an adjunctive therapy.
- Common adverse effects of azathioprine include myelo-suppression, hepatotoxicity, pancreatitis, and hypersensitivity reactions such as drug fever.

- Long-term use can lead to increased risk of infection and neoplasm, particularly lymphoma and skin cancers.

CYCLOSPORINE

- Cyclosporine is used either alone or in combination with other medications as prophylaxis for transplant rejection.
 - Common adverse effects include hypertension, seizures, nephrotoxicity, hepatotoxicity, and post-transplant lymphoproliferative disease (PTLD).
 - Monitoring drug levels is important when administering other medications as several drugs alter the metabolism of cyclosporine via the cytochrome P450 isoenzyme.

TACROLIMUS (FK506)

- Tacrolimus is a potent immunosuppressant in the class of calcineurin inhibitors useful in both the maintenance of immuno-suppression and the treatment of acute or chronic rejection.
- Common adverse effects include hypertension, hyperglycemia, encephalopathy, headache, electrolyte imbalance, renal insufficiency, and rash.
- Caution should be taken with patients on both cyclosporine and tacrolimus as the agents are synergistic and can increase toxicities of both agents.

MYCOPHENOLATE

- Used as an adjunct to other immunosuppressants or for acute rejection.
- Common adverse effects include hypertension, rash, pancreatitis, leukopenia, PTLT, and pulmonary fibrosis.

SIROLIMUS

- Can be used as a single agent or in combination with calcineurin inhibitor therapy for maintenance of immuno-suppression or as a rescue agent for acute and chronic rejection.
- Common adverse effects include hypertension, edema, syncope, hypercholesterolemia, acne, nephrotoxicity, and abdominal pain.

MUROMONAB-CD3 (OKT3)

- Used in the treatment of acute rejection episodes resistant to conventional treatment.

- The administration of OKT3, especially the first few doses, can lead to common adverse events attributable to a cytokine release syndrome including hyperpyrexia, shock, pulmonary edema, cardiovascular collapse, and cerebral edema.
- Other associate side effects include aseptic meningitis, increased risk of sepsis, seizures, and PTLD.

INFECTION PROPHYLAXIS

- Most transplant centers use a combination of prophylactic antibiotics, antifungals, and antiviral medications in the postoperative period.
- During the first year after transplant while immunosuppressant levels are high, vaccines may be deferred until lower levels of immune suppression can be reached in order to increase rates of seroconversion after immunization.

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QUESTIONS

1. A 12-year-old male one month after a liver transplant presents with fever, headache and vomiting. Vital signs include HR 140, RR 32, BP 110/85 and

temperature of 38.5°C. He appears ill, complaining of frontal headache and eye pain. Exam is unremarkable with a central line site on the left chest wall, which is clean and dry. The appropriate next steps include

- A. Lumbar puncture
 - B. Blood culture
 - C. Blood culture from central line
 - D. Broad spectrum antibiotics
 - E. All of the above
2. A transplant patient presents to the ED with headache and neck pain. The patient is currently on corticosteroids and tacrolimus. You prepare to do a lumbar puncture. In addition to routine CSF studies what other information should be obtained?
 - A. CT head
 - B. Opening pressure
 - C. Fungal cultures
 - D. Spinal fluid for immunoglobulins
 - E. C spine films
 3. A 7-year-old with a history of a kidney transplant is brought to the ED for watery diarrhea. Two weeks ago she was started on tacrolimus for acute rejection. Her mother reports compliance with her transplant medications. This patient is at an increased risk for which of the following?
 - A. Pulmonary fibrosis
 - B. Hypotension
 - C. Nephrotoxicity
 - D. Hypoglycemia
 - E. Myoglobinuria

ANSWERS

1. E. Transplant patients are immunosuppressed and at high risk for infection. Patients with recent transplantations may have central line sepsis, meningitis, and cultures of blood and spinal fluid should be obtained quickly and broad-spectrum antibiotics started immediately after obtaining cultures.
2. B. The use of chronic corticosteroids places the patient at increased risk of infection as well as the development of pseudotumor cerebri, which can be recognized by elevated opening CSF pressure obtained during performance of a lumbar puncture.
3. C. Nephrotoxicity can occur as a result of tacrolimus therapy. Other adverse effects include hypertension, hyperglycemia, encephalopathy, headache, electrolyte imbalance, cardiomyopathy, seizures, hepatotoxicity, and pancreatitis.

Section 2

SEDATION, ANALGESIA, AND IMAGING

20 PROCEDURAL SEDATION AND ANALGESIA

Amy L. Baxter

INTRODUCTION

Over the last two decades, acknowledgment of the presence and importance of pediatric pain and wider availability of sedative agents have transformed the management of ill and injured patients. Procedural sedation and analgesia (PSA) are now integral to pediatric emergency care. Sedation and analgesia are commonly used for tedious procedures such as MRI, procedures where precision is critical such as ocular procedures or plastic repairs, or for procedures that are painful including fracture reduction, abscess drainage, or burn debridement. The Joint Commission and the American Academy of Pediatrics recognize that sedation is a continuum; safety and monitoring guidelines focus on the ability to rescue a patient from a deeper level of sedation than intended.

LEVELS OF SEDATION

- Anxiolysis or “minimal sedation”
 - Impaired coordination and cognitive function.
 - Patients respond appropriately to verbal stimuli.
- Moderate sedation
 - Retain purposeful response to verbal or light stimuli.
 - Profound relaxation.
- Deep sedation
 - Purposeful response to repeated painful stimulation.
 - Possibility of depressed ventilatory function.
- General anesthesia
 - No response to painful stimuli.
 - Lack of tone can compromise airway reflexes and cardiorespiratory function.

PATIENT ASSESSMENT

- Determine depth of sedation by patient complaint, temperament, treating clinician, and, when appropriate, family preference.
- Consider minimal sedation for patients with allergies, previous high risk history, drug-specific contraindications.
- Moderate or deep procedural sedation:
 - Focused history pertinent chief complaint
 - Use modified SAMPLE approach; see Table 20-1.
 - Assess for obstructive airway concerns:
 - Snoring
 - Recurrent or current stridor
 - Obstructive sleep apnea
 - Morbid obesity
 - Gastroesophageal reflux
 - Recent URI
 - Swallowing problems.
- NPO guidelines:
 - Balance procedure urgency, sedative type, and overall ED acuity.
 - Typical nonurgent requirements: 2 hours clear liquids and 4 hours solids.
- Vital signs: baseline blood pressure, oxygen saturation, and temperature.
- Focused physical exam:
 - Oropharynx, posterior pharynx, perfusion, and respiratory
 - Assess ease of emergent intubation, eg, with Mallampati score, neck mobility
- Assign an American Society of Anesthesiologists (ASA) score.
 - ASA 1 or 2: generally healthy children, good candidates for PSA in the ED.
 - ASA 3: case by case basis.
 - ASA 4 or 5: generally better treated in a formal operating room.

TABLE 20-1 Pediatric Sample History for Sedation

- Signs/Symptoms: Respiratory infections or obstruction? Snoring? Sleep apnea? Stridor? Heart Disease? Gastroesophageal reflux? Swallowing problems?
- Allergies: Include egg, soy, and latex
- Medications: Particularly concurrent opioids, other analgesics
- Past medical and sedation history: Seizures? Family history of, or prior sedation problems?
- Last meal, liquid
- Events leading to need for sedation: Head injury? Previous failed sedation? Bad experiences with needles or health care?

- Consider general anesthesia consult for
 - Any patient who could be dangerous or difficult to intubate.
 - Craniofacial abnormalities, past reconstruction of the trachea, or atlantoaxial instability.
 - A low systemic risk score should not override a concerning airway evaluation.
- Immediately prior to sedation, Joint Commission requirements state that patients must be reassessed to ensure that their physical condition has not deteriorated since the initial examination.
- The AAP guidelines call for a “time out,” documenting the patient’s name, procedure, and reason for procedure.

EQUIPMENT

- Prior to moderate or deep sedation:
 - Suction, oxygen, and monitoring equipment should be turned on; see Table 20-2.
 - Airway crash cart can be readily available but not opened.
 - A bag-valve-mask setup may be left unopened in the room for moderate sedation, but should be open and ready to use for anticipated deep sedation.
- Use of supplemental oxygen is controversial
 - May mask hypoventilation
 - ED sedations with supplemental O₂ have fewer hypoxia events

TABLE 20-2 Equipment for Deep Sedation (Soap Me)

- Suction: Appropriate size suction catheters connected and tested
- Oxygen: Appropriate size mask, bag, and sufficient oxygen flow to inflate anesthesia bag
- Airway: Nasopharyngeal, oropharyngeal airways, LMAs, laryngoscope, blades, endotracheal tubes
- Pharmacy: Advanced life support medications and reversal agents
- Monitors: Cardiac, respiratory, oxygen saturation, and ETCO₂ when appropriate
- Equipment: Case appropriate special equipment (C-arm, defibrillator)

- End-tidal capnometer (ETCO₂) not required by AAP guidelines, but should be used:
 - For children dependent on normal arterial CO₂ tensions, eg, moya moya.
 - When the patient is physically removed from the sedationist (as for an MRI).
 - When respiratory effort is difficult to assess even at the bedside, as in the obese patient with a gluteal abscess who will be sedated prone.

PATIENT MONITORING

- Determined by the intended depth of sedation and procedure.
- Minimal sedation/anoxiolysis generally does not require any additional monitoring.
 - Parenteral doses of opioids require continuous pulse oximetry.
 - Adjunct parenteral benzodiazepines raise sedation level to moderate, and require a minimum of continuous pulse oximetry.
- Moderate or deep procedural sedation.
 - Continuous cardiac and pulse oximetry
 - Intermittent blood pressure monitoring
 - Intermittent respiratory rate monitoring
- Electronic monitoring should be in place prior to sedation initiation. For agitated patients, obtain baseline vitals to ensure the electronic equipment works, and then remove the offending monitor to calm the child prior to induction. Replace when the child is asleep.

PERSONNEL

- Deep sedation: requires an additional person who only monitors the patient.
- Moderate sedation: monitoring practitioner may intermittently assist with the procedure.
- Sedationist: licensed independent practitioner
 - Skills to recognize the signs and symptoms of respiratory depression.
 - Able to provide effective bag-valve-mask ventilation should apnea occur.
- Continue monitoring
 - Until pharmacologic peak of medications is past.
 - Until child’s respiratory and mental status approach baseline.

DISCHARGE CRITERIA

- An appropriate presedation mental and physical baseline.
- Ability to tolerate sips of fluids (if not NPO).
- Ability to maintain head control if they are still in a child-seat.

AFTER-CARE INSTRUCTIONS

- Should reflect the fact that the child has received an agent that may alter mental status and must therefore receive close supervision.
- Caution caregivers to restrict play activities such as bike riding or climbing for 24 hours.
- Properly staffed and prepared EDs have demonstrated low complication rates for all PSA patients, with no published aspirations or deaths. Implementation of current guidelines has further improved hospital sedation safety.

ROUTES OF ADMINISTRATION IV, IM, SQ, PO, TM

Intravenous (IV) administration

- Offers the greatest flexibility to titrate medications
- IV access limiting factor in infants and agitated children
- IV administration is the preferred route when deep sedation is planned
- Ketamine as a single agent does not require IV access

Intramuscular (IM) and subcutaneous (SQ) injections

- Reliable delivery
- Reserve for drugs with well-established dose–response relationships such as ketamine
- Do not use when repeated doses are anticipated

Oral (PO) administration

- Use for medications with predictable actions (eg, benzodiazepines)
- Reasonable when placement of an IV is not possible
- Pharmacokinetics of repeat doses increase risk due to variable absorption
- The most comfortable of all routes of administration.

Transmucosal (TM) administration (buccal, intranasal, rectal)

- More rapid onset than PO administration
- Titration of medications less precise
- Best-researched are rectal methohexital, intranasal midazolam, and intranasal fentanyl.

Inhaled sedatives

- Inhalation of nitrous oxide and methoxyfluorane have been described in children in the ED setting
- Relatively well-tolerated route of delivery
- Painless administration

- Require specialized equipment and patient cooperation.

SEDATIVE AND ANALGESIC AGENTS

Medications most commonly used for PSA are listed in Table 20-3. Physicians intent on using these agents clinically must be familiar with their indications, all of their actions, relative contraindications, and potential alternatives. Physicians caring for children should have a working knowledge of multiple agents and be adept at choosing alternate drugs when their drug of choice is inappropriate in a given circumstance.

MINIMAL SEDATION/ANXIOLYSIS

BENZODIAZEPINES

- Most commonly used sedative hypnotic agents for anxiolysis
- Act on γ -aminobutyric acid (GABA) receptors
- Reversible with the competitive antagonist flumazenil if needed
- Midazolam
 - Short-acting, duration 15 minutes (intranasal) to 45 minutes (oral)
 - Oral doses of 0.5 to 1 mg/kg result in calmness within 15 to 30 minutes
 - Intranasal delivery at 0.3 mg/kg to 0.5 mg/kg effective in 5 to 15 minutes.
 - pH 3.3, burns during nasal administration
 - Rectal doses of 0.45 mg/kg to 1 mg/kg 62% to 93% effective.
 - Paradoxical reactions and/or subsequent irritability (small study data).
 - IV: 1.4% paradoxical agitation
 - Oral: 6% paradoxical agitation
 - Intranasal: 12% paradoxical agitation
 - Rectal: 27% agitation

MIDAZOLAM/OPIOID COMBINATIONS

- Midazolam and Fentanyl resulted in more vomiting during laceration repair.
- Enteral hydrocodone and midazolam have not been adequately studied; peak effect of hydrocodone is 1.3 hours, should be given 30–45 minutes prior to midazolam for maximum effect.

NITROUS OXIDE

- Inhaled sedative analgesic
- Administered as an oxygen–nitrous oxide mixture.

TABLE 20-3 Common Pediatric Procedural Sedation and Analgesic Agents

MEDICATION	ROUTE	DOSE (mg/kg)	*TYPICAL MAXIMUM	ONSET IN MINUTES	DURATION	COMMENTS
Sedative Analgesics						
Morphine	IV, IM, SQ	0.1–0.15	8	5	2–4 h	Treat itching with diphenhydramine
Fentanyl	IV, IN	0.001–0.002 (1–2 µg/kg/dose)	0.075 (75 µg/dose)	1–2	20 min	Rigidity with rapid administration
	TM	0.010–0.015 (10–15 µg/kg/dose)		30		Increased vomiting when combined with midazolam, not recommended
Hydromorphone	IV	0.01–0.02	2	30	4–6 h	
Hydrocodone	PO	0.2	10	30	4–6 h	
Codeine	PO	1–1.5	60	30	4–6 h	Up to 15% of children would not metabolize to effective metabolite
Sedatives						
Midazolam	IV	0.05–0.1	2	2	30 min–2 h	
	IN	0.3–0.5	12	5–15	1–3 h	
	PR, PO	0.5–1 mg/kg		30	60–90	
Pentobarbital	IV	2.0–6.0	200	2–5	2–4 h	
Thiopental	IV	3.0–5.0	500	0.5	20 min	Intubation doses
	PR	15–40	1200	10–30	60–90	17% defecation
Methohexital	IV	1–3	100	1	20 min	
	PR	18.0–25.0		6–20	1–2 h	
Chloral Hydrate	PO	50–75	1000	60–90	10–24 h	Postdischarge deaths, not an ED drug
Propofol	IV	1–2	75	0.5	20 min	Egg/soy allergy
	Infusion	100 µg/kg/min			5 min	
Etomidate	IV	0.3	20	0.5	10 min	Do not use if septic
Other Agents						
Ketamine	IV	1.0–1.5	60	2	45–90 min	
	IM	4–5	100	5	1–2 h	
	PO	10	250	35	3 h	
Nitrous Oxide	Inhalation	30–70%		1–2	1–2 min	
Diphenhydramine	PO	1.0–1.5	50	15	2–4 h	
Dexmedetomidine	IV	1–2 µg/kg over 10 min		5–6	45–85	Hypotension and bradycardia
	Infusion	1.5 µg/kg/h				
	Intranasal	1 µg/kg		45	90	
	Buccal	4 µg/kg		15	30	
Olanzapine (Zydis)	PO	Repeat of child's normal dose				Existing psychiatric history
Risperidone (M-Tabs)	PO	6 and up, 1 mg		15		Existing psychiatric history
Ziprasidone	IM	10 mg		15		Existing psychiatric history
Reversal Agents						
Naloxone	IV, IM	0.1	2 mg	2 min	20 min	
Flumazenil	IV	0.01	0.2 mg	2 min	30 min	

*Typical maximum dose = effective in most patients. Since all patients respond differently, some patients may require more, in which case precautions in monitoring the patient should be taken.

- Onset of action of 2 to 3 minutes and a similar duration of action.
- No metabolism, direct excretion through exhalation.
- Dose-dependent sedation and analgesia produced.
 - Minimal clinical efficacy is a 30% nitrous oxide to 70% oxygen mixture.
 - 50% to 50% mixture most common.
 - Concentrations of 51% to 70% nitrous are considered moderate sedation.
 - Nitrous with adjunct opioids can result in deep sedation.
- Contraindicated with trapped air (eg, pneumocephalus or pneumothorax)
- May cause ear pain in patients with otitis media.
- Vomiting in 10% to 20% of patients
 - Improved by slow increase in percentage of N₂O.
 - One study found that eating small amounts eliminated vomiting.
- Avoid when there is concern for increased intracranial pressure (ICP), but studies suggest ketamine is safe for intracranial trauma without impending herniation due to preservation of cerebral perfusion pressure disproportionate to increased ICP.
- History of schizophrenia in first degree relative is a “relative” contraindication
- The ketamine clinical practice guideline considers the age <3 months be an absolute contraindication with risks outweighing benefits.

MODERATE SEDATION

- As the distinction between minimal and moderate sedation is clinically nuanced, institutional guidelines often arbitrarily define “moderate” based on types and doses of medications.
- Combinations of IV opioid and benzodiazepines are likely to result in moderate sedation.
- Adverse events may be more common than with “deep” sedatives due to synergistic respiratory depression.
- Sedation/analgesia with combinations of fentanyl/midazolam has more complications and respiratory depression than ketamine/midazolam.

KETAMINE

The dissociative analgesic ketamine has become the most commonly used sedative for painful emergency procedures.

- Produces a trance-like cataleptic state, disrupting the cortical and limbic systems’ communication.
- Onset in 2 minutes.
- Immediately after initial bolus, minimal response to repeated painful stimulation.
- Mild sympathomimetic effects
 - Spontaneous respiratory drive is maintained
 - Decreased bronchospasm
 - Raised systemic blood pressure
 - Tachycardia.
- For induction, 4 to 5 mg/kg IM or 1–1.5 mg/kg IV.
- Contraindications:
 - Avoid in children at risk for increased intraocular pressure.

DEEP SEDATION

- Most sedative-hypnotic agents resulting in deep sedation work through central GABA receptors.
- Delivery of subtherapeutic doses causes disinhibition and may result in agitated, uncontrolled behavior with an increased risk of laryngospasm.
- Treatment of this state is through administration of more of the sedative hypnotic agent to take the child through the plane of disinhibition to one of somnolence.

BARBITURATES

- Variability in effects related to their lipid solubility and rate of central nervous system penetration.
- No pain with administration; may be hyperalgesic
- Drugs used for ED sedation include pentobarbital and methohexital.
- Methohexital—ultrashort acting
 - Can be used as a continuous bolus

- 1–2 mg/kg IV over 30 seconds, profound sedation within 30 seconds.
- Apnea up to 10% with induction, minimal respiratory effects during maintenance infusion.
- Rectal administration of 25 to 30 mg/kg, onset of sedation within 5 minutes, transient airway events in 4% to 10%.

PENTOBARBITAL—SHORT-ACTING

- Administered either IV or IM.
- Up to 6 mg/kg in two to three rapid push divided doses, stacking doses every 30 seconds if the preceding one does not result in sleep.
- Success rate of 97% for diagnostic studies.
- More complications and less effective than etomidate for CTs.
- “Pentobarbital rage” can occur in up to 7% of patients.
- Contraindicated in patients with known temporal lobe seizures.

PROPOFOL

- Ultrashort-acting sedative hypnotic.
- Highly lipid soluble agent.
- Clinical effects within 30 seconds with a duration of 6 to 8 minutes.
- 2 mg/kg IV bolus, with supplemental boluses of 1 to 2 mg/kg maintain the sedation.
- For prolonged sedation, a continuous infusion of 100 to 150 $\mu\text{g}/\text{kg}/\text{min}$.
- ED-only studies have started with 1 mg/kg when adjunctive opioids have been used.
- Propofol burns with injection, which can be mitigated by a 1 mg/kg lidocaine mini-bier block applied with tourniquet for 1 minute prior to delivery.
- Contraindications include severe allergies to egg, soy, or propofol.
- Airway events secondary to apnea or profound relaxation of tone; anticipate need for repositioning of airway at induction; jaw thrusts needed 3% to 4% of the time.
- Brief bag-valve-mask in 4/52 methohexital patients and 2/51 propofol.
- Hypotension with good perfusion is common; supplemental IV fluids do not change outcome.
- Supplemental O_2 decreases the incidence of the most common side effect, mild hypoxia; ETCO_2 may help distinguish between central or obstructive apnea.
- Serious adverse events are rare when deep sedation guidelines are followed and ED practitioners trained in airway management administer the drug.

- Propofol blunts sympathetic cardiac response; vagal stimulation can result in bradycardia; adjunct atropine should be readily available.

ETOMIDATE

- Imidazole sedative hypnotic.
- 0.3 mg/kg in a fast IV push brings on sedation within 30 seconds.
- Good cardiorespiratory adverse event profile; only one reported case of pediatric apnea when used as a single agent.
- No analgesia.
- Side effects
 - Burning with injection
 - Brief myoclonus up to 22% of the time, less common in pediatric patients.
 - Myoclonus can be reduced by pushing more rapidly, or by giving small 0.015 mg/kg doses of midazolam prior to administration.
 - Adrenal suppression can occur transiently with even a single dose; avoid in potentially septic or critically ill patients.

DEXMEDETOMIDINE

- α_2 -adrenoceptor agonist
- Sedative, analgesic, and anxiolytic actions.
- Minimal respiratory depression
- Decrease hypotension and bradycardia by loading over 10 minutes
- Recovery over 20–40 minutes
- 2 $\mu\text{g}/\text{kg}$ IV bolus over 10 minutes is effective, and reduces adverse airway events
- Contraindicated for patients on digoxin; may cause complete heart block.
- Combinations of dexmedetomidine and ketamine are less satisfactory than ketamine/propofol.
- Buccal and intranasal routes of 4 $\mu\text{g}/\text{kg}$ may be promising future applications for agitated children without an IV.

SELECTION OF PSA AGENTS

- PSA selection is dictated by the intended effect on a child.
- If cooperation for a painless diagnostic procedure is desired, then a sedative hypnotic agent will be the drug of choice.
- Use of a narcotic analgesic to produce somnolence may require such a large dose that respiratory depression becomes a risk.

- Use of a pure sedative with no analgesic properties to provide cooperation for a painful procedure is equally inappropriate.
- Combinations of different agents may be used to take advantage of the desired properties of each.
- The most frequent combinations pair short-acting sedatives with short-acting narcotics such as fentanyl/midazolam or fentanyl/propofol.
- Caution should be maintained with such combinations because not only will the desired effects be enhanced but adverse effects such as respiratory depression may also be increased.
- Doses lower than those of either agent alone should be used initially when combining potent agents.
- Combining a local anesthetic with a sedative can also produce effective patient control.
- Regional anesthesia will permit pain control, and a sedative can be used to produce anxiolysis and cooperation for the procedure.
 - Procedures such as lumbar punctures and laceration repairs are examples of where this technique is applied.

Potential agents for different desired clinical outcomes are listed in Table 20–3.

CHILDREN WITH SPECIAL HEALTH CARE NEEDS

- Patients who already carry a psychiatric diagnosis (autism and schizophrenia)
 - Orally dissolving tablets of atypical antipsychotics
 - Repeat a dose of regular daily medication.
 - Intramuscular midazolam more effective and safer than IM haloperidol
 - Ziprasidone: 20 mg IM results in sedation in 15 minutes in adults.
 - Dexmedetomidine: Onset of sedation in a mean time of 27 minutes using 5 µg/kg of buccal dexmedetomidine.
- Children with special health care needs (CSHCN) are usually still appropriate candidates for PSA.
- The approach to PSA in this population is the same as for any other child, except that selection of the PSA agent must take into account not only the child's acute problem but also preexisting conditions.
- Coordinate sedation with a child's medication schedule: a child on neuropsychiatric or seizure medications who becomes somnolent following routine dosing of the medication may undergo a painless diagnostic study a short time after receipt of their last medication dose.
- Children with cardiovascular problems should be managed with agents such as fentanyl, which have little blood pressure or heart rate effects.
- Children with respiratory pathology or anatomic upper airway difficulties may best be served with regional anesthesia or a drug with minimal ventilatory effects such as ketamine.
- Children with hepatic or renal failure may be more sensitive to barbiturate and other drugs, requiring prolonged postprocedure observation.
- Emergency physicians should also not hesitate to involve anesthesiology colleagues to help with special needs patients, particularly those who are ASA class 3 or above.

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QUESTIONS

1. A 5-year-old child needs moderate procedural sedation for a supracondylar fracture reduction that is anticipated to be painful. When considering medication selection which of the following do not contain analgesic properties?
 - A. Propofol and etomidate
 - B. Pentobarbital and nitrous oxide
 - C. Dexmedetomidine and methohexital
 - D. Propofol and dexmedetomidine
 - E. Nitrous oxide and midazolam
2. A 7-year-old girl presents to the ED in need of a complex laceration repair of her face. You decide to administer the dissociative analgesic ketamine as your agent of choice for this procedure. Which adjunct agents improve the safety profile of ketamine for this patient?
 - A. Anticholinergics
 - B. Benzodiazepines
 - C. Propofol
 - D. Ondansetron
 - E. None of the above
3. It is important for clinicians to understand the guidelines and indications for procedural sedation administered in the ED. Which of the following is true regarding monitoring and equipment for intended deep sedation?
 - A. The AAP guidelines require end-tidal carbon dioxide monitoring
 - B. The AAP guidelines require a designated personnel to monitor who does not assist with the procedure
 - C. All monitoring equipment must be attached to the patient before initiating deep sedation
 - D. An open crash cart is required
 - E. None of the above
4. Which of the following is associated with increased risk of airway events during procedural sedation?
 - A. Single agent deep sedatives compared to combination medications for moderate.
 - B. Sedation.
 - C. Oral procedures.
 - D. Patient's history of snoring.
 - E. Supplemental oxygen.
 - F. Use of end tidal CO₂ monitoring.
5. A 5-year-old girl presents with a large abscess to her leg and needs an incision and drainage procedure. Procedural sedation is selected in order to perform the procedure on this very anxious and frightened patient. In this patient which of the following is the appropriate use of adjunctive local anesthesia?
 - A. It is only indicated for fracture reductions.
 - B. It is not needed when procedural sedation is used.

- C. It is important and indicated when using procedural sedation.
- D. It is only necessary when using sedatives without analgesic components.
- E. It increases overall risk by prolonging the duration of the procedure.

ANSWERS

1. A. When selecting medications for moderate procedural sedation it is important to understand the purpose of the sedation and which agents provide adequate analgesia and which are solely sedative hypnotics and anxiolytics. Use of a pure sedative with no analgesic properties to provide cooperation for a painful procedure is inappropriate. Neither propofol nor etomidate provides analgesia. While nitrous oxide have some analgesic properties, propofol and to a lesser degree methohexital are barbiturates and may cause increased sensitivity to pain. Dexmedetomidine, like clonidine, provides moderate analgesia as well as the sedation.
2. E. Large meta-analyses demonstrate increased risk of airway events with anticholinergics, benzodiazepines, propofol, and younger age. While the incidence of vomiting is lower with ondansetron, there is no evidence that it improves the safety or decreases adverse respiratory events. Most emesis occurs at home after sedation. Of note, studies evaluating propofol and ketamine mixes find a decreased incidence of vomiting, ranging from 0 to 3%.
3. B. The AAP guidelines stress the ability to rescue the patient from a deeper level of sedation than intended. During deep sedation, there must be one licensed independent practitioner whose only responsibility is to monitor the patient. However, when the monitoring equipment itself agitates the patient and can contribute to increased risk with induction, it is permissible to check that the monitors are functioning and reattach them to the patient after sedation is initiated. A crash cart must be readily available, but need not be opened.
4. D. The history of obstructive sleep apnea or severe snoring has been associated with an increased risk of airway events. Use of supplemental oxygen decreases hypoxic events, and although end-tidal CO₂ monitoring identifies more periods of apnea, its effect on safety for patients who are being sedated with the practitioner at the bedside remains to be established. The combination of fentanyl and midazolam has been associated with more adverse

events than ketamine as a single agent or ketamine with midazolam. Combining medications that have synergistic sedative effects results in more adverse events than combining medications with counterbalancing side effects (eg, propofol and ketamine).

5. C. Procedural sedation is most risky when patients are disinhibited as they descend toward somnolence. By definition, only during general anesthesia will make patients not respond to painful stimuli. Therefore, during any procedural sedation the administration of pain will result in patients becoming more stimulated and less sedate. As this stage of disinhibition is the most risky, it is important to reduce pain from procedures as much as possible using local or regional analgesics. While some sedatives have analgesic properties (ketamine, nitrous, dexmedetomidine) their effects alone are either brief or insufficient for most severely painful procedures (abscess drainage, joint aspiration, laceration repair).

21 PAIN MANAGEMENT

Amy L. Baxter

INTRODUCTION

- Pain is the most common reason a patient presents for health care.
- The Joint Commission (JCAHO) directs pain be monitored with other vital signs.
- Pain is often undertreated for children in emergency department (ED) settings.

UNDERTREATING PEDIATRIC EMERGENCY PAIN

- In 1987, Anand et al. first demonstrated morbidity from untreated pain in neonates.
- Pain in infants impacts neuronal development, pain perception, and emotionality.
- Untreated pain is remembered even by preverbal children, affecting later compliance, willingness to donate blood, and decreased immunization. For example, untreated pain from LPs increases pain response with subsequent procedures.

PAIN ASSESSMENT

- Wong–Baker FACES scale has become routine in the US (Fig. 21-1).
- A more parametric scale is the faces pain scale—revised, used throughout the world (Fig. 21-2).
- Visual analog scales are validated in ED populations and improve treatment.
- Pain change after the first morphine dose predicts sickle cell patient admission.

PROCEDURAL PAIN MANAGEMENT

VENIPUNCTURE

- Needle sticks are the most common and greatest source of procedural pain in the world.
- Topical anesthetics raise the action potential threshold blocking the fast pain impulse.
- Eutectic mixture of local anesthetics (EMLA) is prilocaine 2.5% and lidocaine 2.5%, and reduces IV catheter insertion after a minimum of 45 minutes. EMLA can be left on up to 4 hours penetrating up to 5 mm. Numbness lasts one hour after

WHICH FACE SHOWS HOW MUCH HURT YOU HAVE NOW?

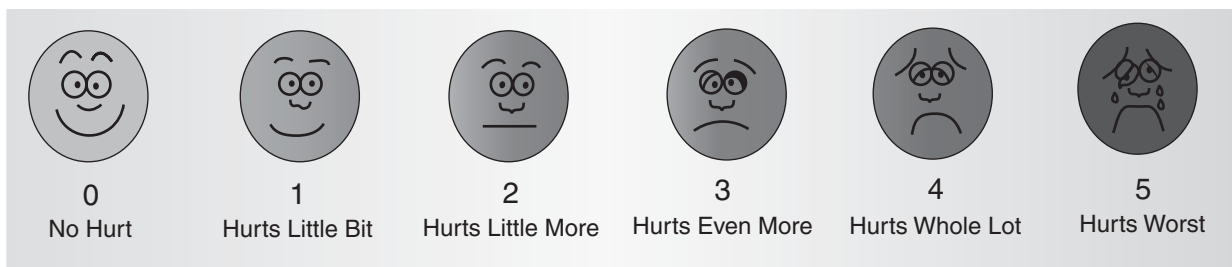


FIG. 21-1. Wong Baker FACES pain scale. From Hockenberry MJ, Wilson D. *Wong's Essentials of Pediatric Nursing*. 8th ed. St. Louis; 2009. Used with permission. Copyright Mosby.

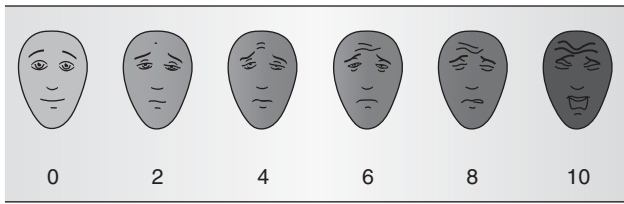


FIG. 21-2. Faces Pain Scale—Revised. For correct use and translations, visit www.painsourcebook.ca. (With permission from Hicks CL, von Baeyer CL, Spafford PA, van Korlaar I, Goodenough B. The Faces Pain Scale—Revised: toward a common metric in pediatric pain measurement. *Pain*. 2001;93(2): 173–183.)

removal. Vasoconstriction resolves in 1–2 hours. Methemoglobinemia is rare, more likely in preterm infants. A purpuric rash can occur in 1% to 2%, particularly in atopic patients.

- LMX-4 (previously called ELA-Max) places 4% lidocaine into liposomes. Effective in 30 minutes, it works as well as EMLA for venipuncture pain.
- LMX-4 improves cannulation success on the first attempt (74% vs 55%, $p = 0.03$) and significantly lowers time of insertion and pain scores.
- LMX-4 does not cause methemoglobinemia.
- Tetracaine and lidocaine mixed 7%/7% in a self-contained patch Synera (Endo Pharmaceuticals) for children 3 years and older contains a heating element that causes local vasodilation. The mixture was tested as the S-Caine patch without the heating element, and showed good pain control and IV start success in children.

LIDOCAINE DEVICES AND TECHNIQUES

- Iontophoresis uses a low-voltage electrical current to drive lidocaine through skin. Time of application is at a minimum 10 minutes.
- Injecting buffered lidocaine using a 30 G needle prior to venipuncture is fast, inexpensive and effective.

- J-tip (National Medical Products, Irvine, CA), puts lidocaine under the skin via a jet of compressed carbon dioxide, with better pain relief than EMLA. 81% found J-tip lidocaine administration painless.
- If the prepared area is not subsequently used, the cost recurs in a newly chosen site.
- Cold spray (Pain Ease, ethyl chloride, Gebauer) applied directly to the penetration site has been shown effective in children older than 8. The advantages are efficacy in seconds and the option to use at another site instantly if needed.
- Figure 21-3 provides an algorithm for balancing optimal pain control with time available.

LACERATIONS—TOPICAL

- Lidocaine, epinephrine, and tetracaine (LET or LAT) are the standard of care for pediatric wound repair.
- Place a shred of LET-soaked cotton or LET mixed with methyl-cellulose directly in the wound before irrigation and repair. Avoid placing gauze or absorptive surfaces near the LET.
- Numbness (seen as blanching) has onset at 20 minutes; anesthesia lasts 20 minutes after removal. LET alone gives sufficient pain control for 90% of pediatric facial lacerations, decreasing length of stay by 30 minutes compared to lidocaine injection.
- LET has 50% efficacy in extremities; two small studies did not find ischemia. Avoid LET when it will blanch vermilion border landmarks. EMLA has been used for laceration repair but is less effective.

LACERATIONS—INFILTRATION

- Perform prior to cleaning.
- Decrease pain of lidocaine infiltration by
 - Warm to room temperature.
 - Inject slowly.
 - Inject with the smallest gauge needle possible
 - Inject from within rather than around the wound.

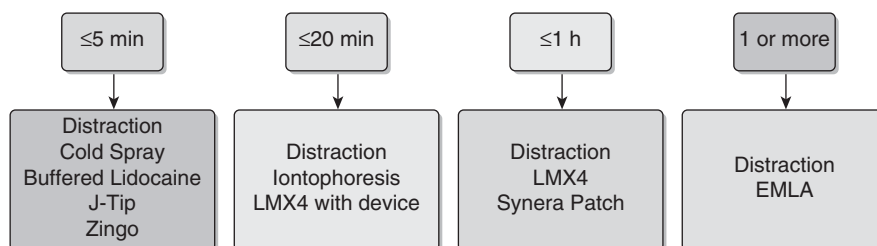


FIG. 21-3. Options for venipuncture pain (in order of increasing cost).

- Buffer acidity with one part sodium bicarbonate (NaHCO_3) to nine parts lidocaine.
- Vibrate skin proximal to the site of infiltration.
- Consider regional nerve blockade for foot lacerations, facial lacerations, and palmar lacerations
- Local injection equivalent pain to digital block for lacerations
- Local infiltration may be less painful for smaller lacerations.

ABSCCESS

- Often requires procedural sedation
- Adequate pain control decreases amount of sedative needed
 - Apply topical anesthetic over intact skin with adequate time
 - Infiltrate lidocaine in a circular field block prior to incision
- Pain with packing removal
 - Consider penrose rather than gauze.
 - Apply LET/LMX mixture prior to removal (moistens packing, numbs incision).

LUMBAR PUNCTURE (LP)

INFANTS UP TO 2 MONTHS

- In 2003 only 30% of physicians reported performing LPs with no analgesia.
- Topical anesthetics improve success rates.
- Place LMX-4 prior to collection of blood and urine for emergent LPs.
- EMLA appropriate for urgent or elective LPs.
- Oral 24% sucrose on a pacifier provides pain relief during the procedure up to 2 months.

OLDER INFANTS

- Topical anesthetic does not penetrate to dura, additional lidocaine needed.
- Add buffered injected lidocaine or J-Tip to reduce pain.
- Local anesthetics with or without an anxiolytic may be adequate.

CHILDREN

- For children anticipating multiple LPs (eg, leukemia, pseudotumor cerebri) deep sedation improves subsequent fear and posttraumatic reactions.

- Inadequate pain control causes increased pain response with subsequent LPs.
- EMLA decreases the pain of LPs in older children.
- Infiltrate 1 mL per 10 kg of body weight up to 5 mL buffered lidocaine.
- Fentanyl 1 mcg/kg IV or 1.5 mcg/kg IN further improves comfort.

PENILE PROCEDURES – PARAPHIMOSIS REDUCTION

- Meatal LMX-4 for 30 minutes was equivalent to EMLA for 45 minutes.
- Consider a dorsal penile nerve block if reduction requires multiple attempts.
- Oral, intranasal, or IV opioids prior to the procedure will help.
- Consider nitrous oxide if available.

URETHRAL CATHETERIZATION

- 2% lidocaine jelly one to two minutes prior to catheterization effective for adults.
- Pediatric research equivocal.

FRACTURES AND BURNS

- Splint fractures and provide initial oral or intranasal opioid prior to radiographs.
 - Acetaminophen and hydrocodone or oxycodone: onset 30 minutes, duration 4 hours.
 - Intranasal fentanyl onset 3 minutes, duration 30 minutes.
- For severe fractures or burns, initial parenteral dose of 0.1 mg/kg of morphine is often inadequate; consider 0.15 mg/kg.
- Opioid monotherapy for debriding or reduction can result in inadequate pain control or respiratory depression after procedure completion.
- Consider regional pain control for fracture reduction or burn debridement
 - Bier block: an IV is placed on the affected side, and plain lidocaine and saline are administered after inflation of a double-blood pressure cuff to 250 mm Hg.
 - A short-acting anxiolytic may be considered during the procedure.
 - A hematoma block and concurrent nitrous oxide performed as well as ketamine.
 - Femoral or axial nerve blocks require specific training to apply effectively.

TABLE 21-1 Common Pediatric Analgesic Agents

MEDICATION	ROUTE	DOSE (mg/kg)	TYPICAL MAXIMUM ^a	DURATION	COMMENTS
Sedative analgesics					
Meperidine	IV, IM	1.0–2.0	100 mg/kg	3–4 h	Chronic accumulation risk
Morphine	IV, IM, SQ	0.1–0.15	10 mg/kg	2–4 h	
Fentanyl	IV	0.001–0.002	0.05 mg/kg	20 min	Rigidity, apnea, lower dose <6 mo
	TM	0.005–0.010			Rigidity, apnea
Remifentanyl	IV	0.001	0.05 mg/kg	4–6 min	
Hydromorphone	IV	0.01–0.02	2 mg/kg		
Hydrocodone	PO	0.2	10 mg/kg	4–6 h	
Codeine	PO	1–1.5	60 mg/kg	4–6 h	
Reversal agents					
Naloxone	IV, IM	0.1	2 mg	20 min	
Flumazenil	IV	0.01	0.2 mg	30 min	

^aTypical maximum dose represents dose that is effective in most patients. Since all patients respond differently, it is possible that for some patients a dose in excess of this dose may be required. If such a higher dose is utilized, precautions in monitoring the patient should be taken.

SICKLE CELL PAIN

- Patients with sickle cell anemia will average 0.8 painful episodes annually.
- 60% require hospital based care, as many as 42% of children inpatient.
- Crisis pain is worse than postoperative or labor pain.
- Crisis frequency dulls physiologic response, so severe pain may not cause typical physiologic changes such as increased heart rate or blood pressure.
- Anti-inflammatory (ibuprofen, ketorolac) plus opioid mainstay of treatment.
- Patients or families often know most effective opioid and starting dose from previous experience; involve them in care.
- Lack of any pain relief after the first opioid dose most predictive for admission.

ANALGESIC AGENTS—SEE TABLE 21-1

PURE ANALGESIA

- Acetaminophen
 - 15 mg/kg dosing for antipyrexia and minor extremity pain
 - Rectal administration may require 40 mg/kg as a loading dose
 - Subsequent rectal doses of 20 mg/kg
- Ibuprofen
 - Not yet available in parenteral formulations
 - Effective at doses of 10 mg/kg in 20–30 minutes
- Ketorolac effective in prostaglandin-mediated conditions, such as biliary or renal colic

ANALGESIA/ANESTHESIA

- Opioid analgesics create dose-dependent pain relief with mild sedative effects.

- Nausea and transient itching or urticaria from histamine release possible.
- Reverse cardiovascular/respiratory depression with naloxone or nalmepphine.

Morphine

- Administer with diphenhydramine or hydroxyzine if history of histamine-release itching
- Neonates may need less per kilogram than patients older than 6 months.
- Children 6 months to 11 years metabolize rapidly and may need frequent redosing.

Meperidine

- Metabolized to an active metabolite, normeperidine, with longer serum half-life.
- Metabolites can result in seizures after multiple doses.

Hydromorphone

- More potent semisynthetic narcotic analgesic.
- May be more effective for renal colic pain, abdominal pain, or sickle cell anemia patients.

Fentanyl citrate

- Synthetic short-acting narcotic approximately 100 times more potent than morphine.
- Rapid onset and 20-minute duration of action
- Fewer cardiovascular effects
- Chest wall rigidity when administered too rapidly, particularly in infants.
 - Reversible with naloxone
 - Skeletal muscle paralysis and intubation may be required.

- Unique in that fentanyl may be administered through transmucosal routes. For intranasal use the standard IV formulation.
- Reduce dose for neonates and <6-month-old infants.

Codeine, hydrocodone, and oxycodone

- Less potent, often administered orally in combination with a pure analgesic
- Codeine poorly metabolized by up to 15% of Asian patients and 7% to 10% of Caucasian patients have decreased metabolism into the active morphine analog.
- Hydrocodone and Oxycodone may be better metabolized into the active morphine.

BEHAVIORAL PEDIATRIC PAIN MANAGEMENT

PREPARATION

- Present concrete details about procedural steps and physical sensations, not rationale.
- Content should be clear, concise, and age-appropriate.
- Include coping skills, such as relaxation techniques (eg, deep breathing, progressive muscle relaxation) or distraction (eg, imagery, watching a movie, finding, and counting tasks).
- Prepare parents and allow them to ask question,
- Parental anxiety is strongly predictive of children's medical distress.
- Parental distraction, praise and encouragement are beneficial; provide reassurance, avoid criticism or minimizing ("Oh, that didn't hurt") or apologizing may increase child distress.

PROCEDURAL INTERVENTIONS

- Distraction is equally effective across gender and ethnic groups.
- Distraction may be most effective for children younger than 7 years of age.
- Distraction stimuli include movies, interactive toys, virtual reality, music, bubbles, and short stories.
- Performing other tasks (eg, cough, finger tapping) works well for older patients.
- Involve multiple sensory modalities (eg, vision, hearing, touch)
- Include parents as coaches.
- Distraction should be done prior to, during, and following the procedure.
- For young infants sucrose, nonnutritive sucking, and skin-to-skin contact are effective.

- Performing procedures in a "position of comfort" diminishes procedural pain. Placing children in parent's arms on a gurney rather than a papoose improves results.

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QUESTIONS

1. Which opioid can be administered through mucous membranes in pediatric patients?
 - A. Hydrocodone
 - B. Hydromorphone
 - C. Oxycodone
 - D. Fentanyl
 - E. Meperidine

2. A 14-year-old male presents with abdominal pain from renal colic and is in moderate distress from the pain. Which of the following opioid may be most effective for this patient?
 - A. Hydrocodone
 - B. Hydromorphone
 - C. Meperidine
 - D. Morphine
 - E. Fentanyl
3. Which of the following is a helpful coaching technique to decrease distress?
 - A. Reassuring
 - B. Praising
 - C. Apologizing
 - D. Minimizing
 - E. Criticizing
4. A 10-year-old female has a 3 cm simple laceration of the left leg. You plan to use lidocaine for local analgesia. The pain of the anesthetic injection can be minimized by which of the following
 - A. Cooling the lidocaine before injection
 - B. Injecting the medication rapidly
 - C. Injecting from within rather than around the wound
 - D. Using an 18 gauge needle or larger
 - E. Buffering with one part lidocaine to nine parts potassium hydroxide
5. A 15-year-old female with a history of sickle cell disease presents with a complaint of bilateral leg pain and back pain. She states the pain is typical of her pain crises. Which is true in regards to the management of this patient.
 - A. Vital signs change when pain is severe and indicate degree of pain.
 - B. Failure to improve with first dose of opioid is predictive of need for admission.
 - C. Decreasing crisis frequency is associated with increasing mortality.
 - D. Sickle cell pain is not as severe as postoperative pain.
 - E. Patients will have approximately 2–4 severe pain crises per year.

ANSWERS

1. D. Fentanyl is unique in that it is the only narcotic that can be administered through transmucosal routes. Current studies using the intranasal route use the standard IV formulation. Reduced doses should be used in neonates and very young infants since they are more sensitive to the respiratory depressant effects.
2. B. Because of the partial prostaglandin mediated inflammation and pain, hydromorphone may be

more effective for appendicitis or renal colic pain relief than the other options. Hydrocodone is oral and a poor choice when a patient is NPO, Meperidine accumulates when used long term and metabolites may result in seizures

Fentanyl is extremely short acting and not a good long-term solution, though initiating relief with transmucosal fentanyl while IV access is established is a reasonable option.

3. B. Praising, distraction and relaxation techniques are useful behavioral interventions that can be used during painful procedures. Excessive reassurance, criticism, or apologizing may exacerbate distress.
4. C. Pain during infiltration of lidocaine can be reduced by injecting from within the wound rather than through intact skin. Other methods of reducing pain include warming the drug to room temperature, injecting slowly, using the smallest gauge needle possible, and buffering the lidocaine (nine parts lidocaine to one part sodium bicarbonate).
5. B. Acute vital sign changes are uncommon by the time a patient presents to the emergency department. Nonresponse to a first parenteral opioid is a good predictor of need for admission. Patients will have approximately one severe crisis per year, and increased number of crises is predictive of mortality. The pain of a crisis is usually severe and considered worse than postoperative or labor pain.

22 IMAGING

Wendy C. Matsuno

INTRODUCTION

Technology has provided us with a variety of imaging studies to help us reach a diagnosis. This chapter will review the most commonly available imaging modalities and discuss the various considerations for optimal visualization and patient safety.

ULTRASOUND

- Ultrasound technology was first commercially available in the late 1960s as a rigid contact B-mode machine that was able to take still pictures only.
- In the late 1970s, the first real-time ultrasonography machine was introduced, and by the mid-1980s

ultrasound machines started to be used in the emergency department (ED).

PHYSICS AND PATHOPHYSIOLOGY

- Ultrasound employs sound waves, a form of nonionizing radiation, to compressed and decompressed molecules in tissue.
- The transducer, also known as the probe, is held by the examiner and emits sound waves. A linear probe emits sound waves at a higher frequency, which makes it ideal for viewing superficial structures. The curved probe emits sound waves at a lower frequency, which allows deeper penetration and is useful for deep structures (eg., organs).
- Sound waves are reflected back to the transducer by very dense, high impedance objects (eg, gallstone). These objects appear white on the image screen and are referred to as echogenic. The surrounding fluid filled areas, which do not reflect the sound wave, appear black (anechoic).
- Refraction can make visualization difficult, so the transducer should be held perpendicular to the area of interest. Sound waves can also be scattered in various directions by irregular, small material (eg, gas), thus to prevent attenuation and scatter an acoustic window is often used, to view distant structures through an area of good penetration (eg, full bladder).
- Doppler works by registering the calculated change in frequency of sound waves that return to the transducer from a moving object (eg, red blood cells). Doppler by itself does not provide structural information, thus it is most helpful when used concurrently with real-time ultrasonography.

SAFETY CONSIDERATIONS

- Thermal injury and cavitation are hypothetical risks that have been raised regarding ultrasonography.
- The thermal mechanism postulates heat is produced by the absorption of ultrasound by the tissues and may cause tissue damage. Thermal damage is unlikely with diagnostic ultrasonography, as the intensity and time of exposure needed to produce such a temperature change is significantly greater than what is currently used for imaging.
- The cavitation mechanism can be divided into transient and stable cavitation. Transient cavitation occurs at high intensities where gas-filled bubbles enlarge, then suddenly collapse resulting in a localized temperature change, thermal decomposition of water, and release of free radicals. In stable cavitation, which occurs at

low intensities, the gas-filled bubbles vibrate and may cause shearing stress to surrounding tissue. The low intensity level and short duration of exposure make the occurrence of cavitation unlikely.

BENEFITS AND LIMITATIONS

- Ultrasound does not use ionizing radiation and is considered safe at current diagnostic settings.
- Ultrasound is a noninvasive, painless procedure that does not require the patient to be completely still; thus, sedation is not needed.
- Small, portable machines make ultrasound easily accessible at the bedside.
- The skill of the person operating the ultrasound machine can limit the quality of the image and the ease by which structures are identified.
- The larger body habitus of some pediatric patients can make visualization of internal structures difficult.

DIAGNOSTIC AND PROCEDURAL CONSIDERATIONS

- Pyloric stenosis can be confirmed by the finding of a thickened pylorus muscle of ≥ 0.3 cm or by an elongation of the pylorus to ≥ 1.4 cm. The sensitivity of ultrasound for pyloric stenosis is reported to be 97% to 100% and with a specificity of 99% to 100%.
- Testicular torsion can be detected on ultrasound with a sensitivity of 78% with a specificity of 97%. In cases of high clinical suspicion, a negative study should not rule out torsion.
- Ectopic pregnancy can be detected by transvaginal ultrasound with a sensitivity 90% to 100% and a specificity of 88% to 95%.
- Ovarian torsion can be diagnosed on transabdominal and transvaginal ultrasound with a sensitivity and specificity of 88% and 87%, respectively. Color Doppler flow can be seen in 57% to 62% of torsed ovaries; thus, the presence of flow does not rule out ovarian torsion.
- Appendicitis is determined on transabdominal ultrasound by the presence of a noncompressible, tubular structure that measures longer than 6 mm in diameter. The sensitivity and specificity of ultrasound to diagnose appendicitis in children is 80% to 92% and 86% to 98%, respectively.
- Focused abdominal sonography for trauma (FAST) has been used in adult trauma patients to evaluate for intra-abdominal injury based on the concept that free fluid (eg, blood) collects in the dependent portions of the supine trauma patient. The evaluated areas are the

pericardial sac, Morrison's pouch, splenorenal recess, and the pelvis. A modified FAST also looks at the paracolic gutters bilaterally. The sensitivity and specificity for FAST to detect hemoperitoneum is 80% and 96%, respectively.

DIAGNOSTIC RADIOGRAPHS (X-RAYS)

- The discovery of x-rays was made by Roentgen in 1895.

PHYSICS AND PATHOPHYSIOLOGY

- An x-ray is a collection of electromagnetic energy called a photon, which ionizes atoms that they encounter, hence labeling x-rays as a form of ionizing radiation.
- Approximately 1% of x-rays navigate all the way through the patient to the film. The remainder of the x-rays are either absorbed or scattered.
- Absorption of an x-ray results in a white appearance on the film because the x-ray does not penetrate through the given object onto the film.
- Scattered x-rays appear as a gray color, which decreases the quality of the image. An antiscatter grid is used, to absorb the scattered x-rays.

SAFETY CONSIDERATIONS

- X-rays can cause damage when they are absorbed or scattered by an atom causing electrons to shoot off and ionize surrounding atoms.
- Lead shields are often used to protect body parts that are not being imaged. The high atomic number of lead prevents x-rays from penetrating through it.

BENEFITS AND LIMITATIONS

- The benefit of using diagnostic radiography is that one is able to get a quick view of a large area with a relatively low amount of radiation.
- One limitation of x-ray is that it is a two-dimensional image of a three-dimensional subject, thus multiple views may be needed.
- Radiolucent objects cannot be seen on film (eg, plastic).

DIAGNOSTIC AND PROCEDURAL CONSIDERATIONS

- Diagnostic x-ray is the standard for diagnosing fractures and is generally used to evaluate for chest and abdominal pathology.

- Fluoroscopy is similar to diagnostic x-rays except that the fluorescent screen is viewed directly with the help of an image intensifier. Fluoroscopic procedures allow real-time images, but the drawback is that the image quality is poorer and the patient is exposed to more radiation. Common fluoroscopic procedures include fracture reductions and contrast studies.

COMPUTED TOMOGRAPHY

- Computed tomography (CT) was developed in the late 1960s by the British engineer Geoffrey Hounsfield.

PHYSICS AND PATHOPHYSIOLOGY

- CT has a narrow opening where the x-ray source emits a thin, fan-shaped beam that is received by x-ray detectors. The x-ray source and detectors rotate simultaneously around the patient and the information is processed by a computer to create a three-dimensional image.
- X-rays are a form of ionizing radiation that can release electrons from atoms and molecules. In the case of water molecules, hydroxy radicals are released that can damage deoxyribonucleic acid (DNA) causing strands to break and bases to be harmed. DNA can also be directly ionized by x-rays.
- The damage to DNA can be fixed by the cell in most cases; however, double-strand breaks are less easily mended and incorrect repairs that can lead to point mutations, translocations, gene fusions, and ultimately cancers.

SAFETY CONSIDERATIONS

- The absorbed dose is the radiation dose delivered to an organ and is measured in grays (Gy), where 1 Gy is equal to 1 joule (J) of absorbed radiation energy per kilogram.
- The organ dose represents the deposition of the radiation within the organ.
- The effective dose takes into account the amount of radiation each organ receives as well as the radiosensitivity of the specific organs. The effective dose is measured in mSv.
- Children are more vulnerable to ionizing radiation from CT scans than adults because the tissues and organs of children are more radiosensitive and the latent period between the time of exposure and the development of a potential cancer is longer.

- Radiologists and CT technicians should adjust parameters to use the radiation dose that is as low as reasonably achievable (ALARA) to get an adequate scan so radiation exposure is decreased.

BENEFITS AND LIMITATIONS

- CT scans are readily available and can be performed rapidly, so sedation is rarely needed.
- The main limitation of CT is that it provides a significant radiation exposure, especially in the pediatric age group.
- Another limitation is that the patient needs to be transported to the radiology department, which may limit its use in unstable patients.

DIAGNOSTIC AND PROCEDURAL CONSIDERATIONS

- When the diagnosis of appendicitis cannot be determined clinically or on ultrasound, an abdominal CT scan can be obtained with a sensitivity and specificity of 87% to 100% and 83% to 97%, respectively.
- In pediatric trauma patients, CT scans can provide a great deal of information quickly.

MAGNETIC RESONANCE IMAGING

- Developed in the 1980s. Since then, it has become available in most institutions.
- Provides better evaluation of soft-tissue and organs.
- Generally not considered to be a routine emergency department study, but there are conditions that require emergent MRI.

PHYSICS AND PATHOPHYSIOLOGY

- MRI uses radiofrequency energy, which is a type of nonionizing radiation.
- This energy is absorbed by tissue and is expressed as the specific absorption rate (SAR).
- Radiofrequency energy can cause an increase in body temperature, which in certain patients with medical challenges (eg, fever, obesity, hypertension, etc.) can theoretically cause tissue damage.
- MRI systems also employ a static magnetic field usually in the range of 0.2 to 3 Teslas (T).
- The FDA states that clinical MRI machines that use a static magnetic field up to 8 T can be used without significant risk to the patient.

SAFETY CONSIDERATIONS

- The biggest safety hazard is the risk that ferromagnetic objects (eg, metallic clips or coils, internal defibrillators/pacemakers, ferromagnetic foreign bodies or foil containing medicine patches) may be brought into the MRI magnetic field.
- The isocenter of the magnet exerts a constant force, which can subject a ferromagnetic object to a rotational force.
- The spatial area outside the magnet exerts a gradient field where the greatest force is closer to the magnet and decreases further out from the magnet. In this area, ferromagnetic objects are subjected to a translational force in the direction of the magnet.
- Patients with tattoos may experience minor adverse effects (eg, irritation). The FDA states that the risk of not having an MRI done when it is indicated is likely greater than the risk of the potential complications.
- MRI can subject the patient to high levels of acoustic noise so ear protective devices are used. To be consistent with Occupational Safety and Health Administration guidelines for industrial workers, the noise level should be kept below 100 dBs when protective gear is in place.

BENEFITS AND LIMITATIONS

- The benefit of MRI is that it provides a more detailed image for most organs and soft tissue than a CT scan and does not use ionizing radiation.
- The limitation of MRI is that it cannot be used in patients with ferromagnetic objects.
- MRI cannot be performed at the bedside and requires a significantly longer time to complete.
- MRI also requires radiology interpretation.
- In children, MRI scans often require the use of sedation, which adds other potential safety risks.

DIAGNOSTIC AND PROCEDURAL CONSIDERATIONS

- MRI in the emergency department setting is usually reserved for emergent conditions, such as cord compression and strokes.

CONCLUSION

- Diagnostic imaging is vital to the practice of pediatric emergency medicine.
- Practitioners should be aware of the different imaging modalities and be aware of the radiation risk of x-ray and CT studies.

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QUESTIONS

1. A 7-week-old male presents with projectile nonbilious vomiting after every feed. The parents report poor weight gain, irritability, and decreased urine output. Which imaging study would be best to diagnose the likely diagnosis?
 - A. Ultrasound
 - B. X-rays
 - C. CT scan
 - D. Fluoroscopy
 - E. MRI
2. A 5-year-old male was involved in a high-speed motor vehicle collision and has a lap belt contusion. He is not ventilating well. You decide to proceed with rapid sequence intubation. You are concerned about associated intra-abdominal injury. What is the best study to evaluate for abdominal injury?
 - A. Upright chest X-ray
 - B. FAST scan
 - C. Lateral decubitus film
 - D. CT scan with IV contrast
 - E. Fluoroscopy
3. A 12-year-old male presents with the acute onset of left testicular pain for the last 5 hours. The testicle is high riding, red and swollen. There is no cremasteric reflex. What is the next best course of action?
 - A. X-ray
 - B. Urinalysis
 - C. Ultrasound with Doppler flow studies
 - D. CT scan
 - E. Urology consult
4. A 17-year-old female presents with abdominal pain. On physical examination she is noted to have lower abdominal pain and adnexal fullness. She does not remember when she last had her menses. What would be the next best course of action?
 - A. X-ray
 - B. Ultrasound
 - C. CT scan
 - D. Enema
 - E. Fluid trial
5. A 2-year-old male presents to the emergency department with a history of swallowing a coin. He appears nontoxic and is in no respiratory distress on examination. What is the next best step in management?
 - A. X-ray
 - B. Ultrasound
 - C. CT scan
 - D. Fluoroscopy
 - E. GI consult
6. A neonate presents with bilious emesis and dehydration. After resuscitation and stabilization what would be the diagnostic imaging study of choice?
 - A. Abdominal x-ray
 - B. Ultrasound
 - C. CT scan
 - D. Fluoroscopy
 - E. Surgical consult
7. When discussing CT scans with the parents of your patient, they ask about the risks of radiation to their child. You tell them that radiation in children
 - A. Is less likely to cause damage in a child because they are still growing
 - B. Is the same in adults as most hospitals use the ALARA principle
 - C. Is negligible and carries no risk
 - D. Is variable depending on the child
 - E. Is more likely to cause potential cancer in a child because tissues and organs of children are more radiosensitive and the latent period between the time of exposure and the development of a potential cancer is longer

ANSWERS

1. A. Ultrasound—Pyloric stenosis can be confirmed by the finding of a thickened pylorus muscle of ≥ 0.3 cm or by an elongation of the pylorus to ≥ 1.4 cm. The sensitivity of ultrasound for pyloric stenosis is reported to be 97% to 100% and with a specificity of 99% to 100%. X-ray can be part of the evaluation process, but would not be diagnostic. CT scan and fluoroscopy may be able to diagnose a pyloric stenosis, but has considerable radiation to the patient and would not be the best study. MRI scan would require sedation and would pose unnecessary risk to the patient when an ultrasound could be done instead.
2. B. FAST scan—Focused abdominal sonography for trauma (FAST) that evaluates the pericardial sac, Morrison's pouch, splenorenal recess, pelvis, and paracolic gutters can detect free fluid in the abdomen indicating abdominal injury. It can be done easily at the bedside while other interventions are being done. While not diagnostic of a particular injury, the finding of free fluid would indicate an abdominal injury. X-ray and fluoroscopy can be done at the bedside, but would not be able to evaluate for abdominal injury. CT scan and MRI need to be done in the Imaging department.
3. E. Urology consult—With the clinical presentation of this patient there is a high concern for testicular torsion and with the onset of symptoms closely approaching 6 hours it would be best to consult a urologist. An ultrasound may be able to diagnose the condition, but may delay definitive treatment. Also ultrasound has a sensitivity of 78%, thus cases could be missed. An x-ray would not contribute to a diagnosis. A urinalysis may point to other infectious causes, but would not be the next best course of action. A CT scan would not be appropriate in this case.
4. B. Ultrasound—The ultrasound would be able to check for an intrauterine or ectopic pregnancy. Depending on the gestational age an abdominal or transvaginal ultrasound may be utilized. An x-ray may be able to pick-up an obstructive process or mass, but with the history an ultrasound would be a better course of action along with β -hCG testing. A CT scan would not be the next best course of action, but may be indicated depending on the ultrasound results. An enema and fluid trial would not be the next best course of action.
5. A. X-ray—An x-ray to check the location of the radio-opaque foreign body would be the next best step in management. Should the coin be lodged in the trachea or in the esophagus, a GI /surgical consult would then be considered. Fluoroscopy and CT scan would be able to pick up the object, but would expose the patient to more radiation and would not be warranted. An ultrasound would not be able to easily detect the location of the coin.
6. D. Fluoroscopy—Fluoroscopy in the form of an upper gastrointestinal series (UGI) would be the diagnostic study of choice to diagnose malrotation with volvulus. While an abdominal x-ray and surgical consult are also indicated, they would not be the diagnostic imaging study of choice. An ultrasound may be able to pick up abnormalities, but would likely lead to a obtaining a more diagnostic UGI. A CT scan may be able to pick up the abnormality, but would likely delay diagnosis and would not be the diagnostic imaging study of choice.
7. E. Is more likely to cause potential cancer in a child because tissues and organs of children are more radiosensitive and the latent period between the time of exposure and the development of a potential cancer is longer. The radiation risk in a child is increased as compared to an adult due to the developing tissue and the increased time in which they have to develop possible cancers. The ALARA principle should decrease the amount of radiation that a child receives since the concept is to use the least amount of radiation necessary to get the study done adequately.

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Section 3

RESUSCITATION

23 AIRWAY MANAGEMENT

Loren G. Yamamoto

PEDIATRIC AIRWAY ANATOMY

- Airway is smaller (incremental narrowing is more critical)
- Large tongue
- Cricoid ring is the narrowest portion of airway (endotracheal tube (ETT) can pass through cords, but might not pass cricoid)
- Epiglottis is softer
- Airways are less rigid (worse in tracheomalacia and laryngomalacia)

RECOGNIZING A DIFFICULT AIRWAY PATIENT

- Conditions with known airway difficulties (eg, Down syndrome and Pierre Robin syndrome)
- Malformations that affect the tongue, mandible, and neck
- Tumors, masses, swelling, and edema (eg, burns, chemical inhalations, and allergic reactions) in the mouth and neck area.
- Laryngomalacia and tracheomalacia place patients at higher risk for airway obstruction.
- Head trauma, neck trauma, and multiple trauma can injure the airway and require cervical spine immobilization making visualization of the larynx potentially more difficult.
- Noisy breathing, harsh/frequent cough could indicate infections such as croup, bacterial tracheitis, retropharyngeal abscess, and occasionally epiglottitis.

- Patients with an altered sensorium and patients who are pharmacologically sedated are at higher risk for airway compromise as the oral structures relax and fall posteriorly over the airway when the patient is in a supine position.

CLINICAL ASSESSMENT OF AIR EXCHANGE AND THE DEGREE OF AIRWAY OBSTRUCTION (USE ALL THREE IF POSSIBLE)

- Visual: chest movement and exaggerated respiratory effort
- Auscultation: assess quantity and resistance of air exchange
- Technology: end-tidal CO₂ monitoring. Pulse oximetry measures oxygenation, which will decline with diminishing air exchange.

AIRWAY MANAGEMENT

- The ability to rescue a patient from an airway emergency in order to maintain oxygenation is a vital skill that emergency physicians must possess.
- Most airway repositioning maneuvers move the posterior portion of the tongue anteriorly so that it does not block the airway.

IN TRAUMA PATIENTS, CERVICAL SPINE IMMOBILIZATION MUST OFTEN BE MAINTAINED

- Jaw thrust
- Chin lift

IF CERVICAL SPINE MOVEMENT IS NOT A CONCERN, THEN OTHER MANEUVERS THAT CAN BE ATTEMPTED WITH VARYING DEGREES OF SUCCESS, SUCH AS:

- Sniffing position: putting a thick towel beneath the occiput to bring the face and jaw more anterior.
- Shoulder roll: placing a towel roll under the patient's scapulae and upper thoracic spine while permitting the head to tilt backward. This does the opposite of raising the occiput, but the backward tilt of the head can often raise the posterior portion of the tongue. An excessive head tilt can stretch and compress the airway.
- Placing the patient on his/her side with the face slightly downward permits gravity to move the tongue forward.
- Placing the patient prone. While this permits gravity to move the tongue forward and allow secretions to drain out of the mouth, it does not permit easy access to the airway for other manipulations such as laryngoscopy. However, bag-mask ventilation (BMV) can be done in this position. This position might be especially optimal for a patient with epiglottitis in respiratory failure: an exaggeration of the "tripodding" position preference (erect, leaning forward), to keep the epiglottis off the airway.

PHARMACOLOGIC TREATMENT

- Aerosolized epinephrine can improve air exchange in croup and other conditions resulting from upper airway edema. Note that the standard dose of 0.5 mL of 2.25% racemic epinephrine is equal to 5.5 mg (5.5 mL of 1:1000) epinephrine.
- Aerosolized and systemic corticosteroids can also reduce airway swelling caused by inflammation.
- Anticholinergics (ipratropium) and albuterol can bronchodilate the medium and smaller airways.

OXYGEN ADMINISTRATION FOR SPONTANEOUSLY BREATHING PATIENTS

- Nasal cannula: enriches oxygen concentration in the oral-nasal region
- Blow-by: for fussy children who won't wear a device
- Oxygen mask: can deliver a moderately high oxygen concentration.
- Nonrebreather oxygen mask: can deliver a very high concentration of oxygen with the nonrebreather bag properly inflated at maximum oxygen flow rates.

- Rusch bag and mask (anesthesia-type bag): closed system that can deliver very high oxygen concentration and positive pressure (ventilation or continuous positive airway pressure (CPAP)) at high flow rates.
- Do not use a self-inflating bag and mask since this does NOT deliver any oxygen for spontaneously breathing patients.

RESCUE BREATHING AND BMV ARE A VITAL SKILL THAT MUST BE PRACTICED FOR PROFICIENCY

- Airway repositioning is generally required simultaneously.
- The two-rescuer technique is easier and usually more effective.
- The single-rescuer technique is more difficult to perform optimally.
- Over-inflation with BMV can result in gastric distention, which can restrict lung expansion and place the patient at risk for gastric regurgitation.
- Applying cricoid pressure (the Sellick maneuver) can reduce gastric inflation to some degree.
- Excessive ventilation volumes and pressures can also increase intrathoracic pressure impeding venous return.

VENTILATION BAG TYPES

- Self-inflating bag is stiffer (Fig. 23-1). Does not require a gas source.
- Rusch bag (also called anesthesia bag) is more floppy and it normally collapses (Fig. 23-2). It is a closed system. No outside air is entrained if a proper mask seal is maintained, so 100% oxygen can be easily delivered if 100% oxygen enters the bag.
- The term "ambu" bag can refer to both types of bags.

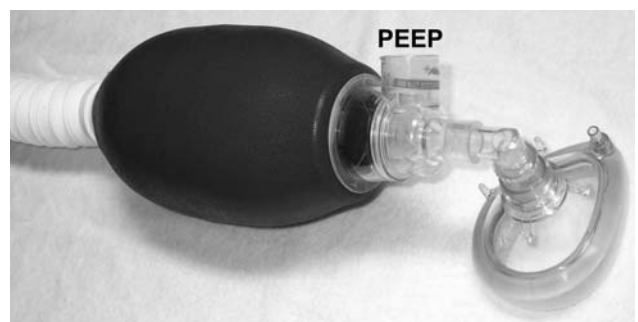


FIG. 23-1. Self-inflating bag. The tail is on the left. High flow oxygen entering the tail increases the FiO_2 entering the bag. This particular model has an opening for an optional PEEP valve.

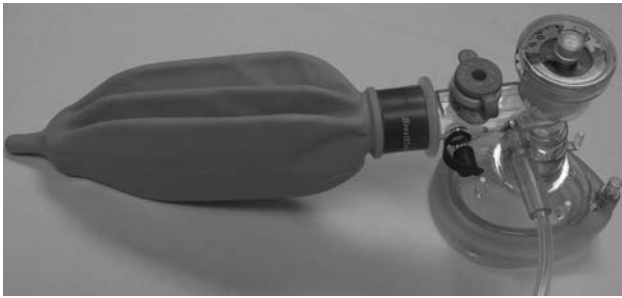


FIG. 23-2. Rusch bag and mask. Note the built-in manometer.

HOW DO THE BAG TYPES DIFFER IN USE AND FUNCTION?

- Self-inflating bag is stiffer. It does not require a gas tank or oxygen outlet. It only delivers oxygen if the bag is squeezed so it should not be used for blow-by oxygen for spontaneously breathing patients. Room air enters its intake so maximum FiO_2 is below 100%. When oxygen flow rate is maximal and the tail of the bag is extended, FiO_2 can approach 100%.
- Large self-inflating bag can deliver larger tidal volumes at a greater inspiratory force than a Rusch bag. This can be useful for poorly compliant lungs or transtracheal ventilation through a narrow catheter.
- Rusch bag is a closed system so it can deliver 100% FiO_2 . It can be used for blow-by oxygen in a spontaneously breathing patient and it can also deliver CPAP through a mask or ETT.
- Another useful application of the Rusch bag and mask is that it can be used to visibly monitor ventilation in a spontaneously breathing patient. If the gas flow is adjusted optimally with a proper mask seal, the bag will collapse and expand as an indicator of air exchange.
- Both bag types can deliver positive end-expiratory pressure (PEEP) but the self-inflating bag needs a PEEP attachment for this functionality.

ADVANCED AIRWAYS

- The laryngeal mask airway is an alternative to endotracheal intubation. As with any airway device, training and practice is required for proficient use.
- Tracheal intubation is best accomplished via the rapid sequence intubation (RSI) method with direct visualization using a laryngoscope. Other techniques that have been described are blind nasal tracheal intubation, blind oral intubation via palpation, and intubation via fiberoptic or video laryngoscopy visualization and guidance.

RSI IS DESCRIBED IN A SERIES OF THE FOLLOWING STEPS (A SEQUENCE)

Step 1. Patient assessment, preparation, and resuscitation

- Assess the patient and perform immediate resuscitation measures such as BMV, if necessary.
- Practicing equipment preparation and posting Table 23-1 facilitates this readiness.

Step 2. Premedications

- Depends on the clinical circumstances and practitioner preference.
- An abbreviated list of these includes atropine and lidocaine.

Step 3. Hyperoxygenation

- Administer maximal oxygen to spontaneously breathing patients.
- For apneic or hypoxic patients, use BMV (with cricoid pressure) to maximally oxygenate the patient prior to intubation.

Step 4. Cricoid pressure (Sellick maneuver)

- This occludes the esophagus and reduces the risk of passive regurgitation.
- Also, this positions the larynx more posteriorly to facilitate visualization.
- Use with caution in infants; the airway can easily be compressed and distorted.

Step 5. Paralyzing agent

- Selecting succinylcholine versus rocuronium is controversial.
 - Succinylcholine has a shorter duration, but a higher risk of malignant hyperthermia, and hyperkalemia.
 - Rocuronium has a longer duration of paralysis but a lower risk of adverse reactions.
- Principles known as priming and defasciculation prolong the time to intubation.

Step 6. Sedation agent

- Table 23-1 describes a basic method of selecting a sedation agent.
- Selection criteria may be separated into patients with head trauma (or increased ICP), hypotension, and respiratory failure due to asthma.
- Thiopental has cerebroprotective properties, but it is a myocardial depressant and can lower blood pressure.
- Ketamine increases blood pressure and bronchodilation, but it also increases ICP.
- Etomidate is a more intermediate agent and is purported to be a universal RSI sedative because of less adverse effects.
- Benzodiazepines and propofol have been used but have significant disadvantages.

TABLE 23-1 RSI Drugs, Doses (Mg/Kg), Sizes, Distances

AGE	2 Mo	6 Mo	1 y	3 y	5 y	7 y	9 y	11 y	12 y	14 y	16 y	ADULT
Average weight (kg)	5	8	10	15	19	23	29	36	44	50	58	65
Preoxygenation												
Adjunctive agents (optional):												
Atropine (0.01–0.02 mg/kg): Use in all children or with ketamine.	0.1	0.15	0.2	0.3	0.3	0.4	0.5	0.5	0.5	0.5	0.5	0.5
Lidocaine (1.5 mg/kg): Lowers ICP	8	12	15	22	28	35	44	54	66	75	90	100
Sellick maneuver												
Sedative												
Hypotension												
Etomidate (0.3 mg/kg):	1.5	2.4	3.0	4.5	6	7	9	11	13	15	17	20
Head trauma without hypotension												
Etomidate (see above) or												
Thiopental (3–5 mg/kg):	15–25	24–40	30–50	45–75	57–95	70–115	90–145	110–180	130–220	150–250	170–290	195–325
Status asthmaticus:												
Ketamine (1–2 mg/kg):	5–7	8–16	10–20	15–30	19–38	23–46	29–58	36–72	44–88	50–100	58–100	65–100
Paralyzing agent:												
Succinylcholine (1.0–1.5 mg/kg):	8	12	15	25	30	40	50	55	60	65	70	80
Rocuronium (0.6–1.0 mg/kg):	4	6	9	12	15	20	25	30	40	45	50	60
Intubate (tube size):	3.5	3.5	4.0	4.5	5.0	5.5	6.0	6.5	7.0	7.0 female, 8.0 male		
Tube depth at lip (cm):	11	12	13	14	15	16	18	19	20	22	22	22
Laryngoscope blade size:	1	1	1	2	2	2	2	2	3	3	3	3

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FIG. 23-3. ETCO₂ colorimetric detector. This particular model is small, made for neonates. Color changes from purple to yellow if CO₂ is detected. (With permission from Gausche-Hill M, Fuchs S, Yamamoto L, eds. *Instructor's Toolkit to Accompany APLS the Pediatric Emergency Medicine Resource*. 4th rev ed. Sudbury, MA: Jones and Bartlett Publishers; 2008.)

- Using no sedative is a serious consideration in hypotensive or septic shock patients since any sedative could result in cardiovascular collapse.
- Give the sedative before the paralyzing agent or vice versa.
- Giving the paralyzing agent first reduces the time to intubation. Since the paralyzing agent takes 60–90 seconds to achieve sufficient paralysis, the sedation agent can be given during this waiting time.
- Giving the sedation agent first (as listed in Table 23-1) permits the patient to avoid the sensation of becoming paralyzed.
- Regardless of which is given first, the paralyzing agent and the sedation agent should be given in rapid sequence.

Step 7. Intubation

- ETT size selection is critical in children.
- The common formula cited is $4 + (\text{age} \div 4)$.
- Using a length-based resuscitation system or posting Table 23-1 in the resuscitation room is more reliable.

Step 8. Confirmation of tracheal intubation (use more than one method and one method must be ETCO₂)

- Colorimetric ETCO₂ (Fig. 23-3) confirms that the trachea is intubated in most instances. Avoid the problem of using an adult-size unit in an infant. Prolonged use will disable these due to water-vapor saturation.
- An ETCO₂ monitor quantifies the ETCO₂ (which can be correlated to the patient's venous or arterial

PCO₂), displays the actual waveform of ETCO₂ production, and can be used continuously and indefinitely. A typical square waveform reliably confirms tracheal intubation, while a nonsquare waveform raises concerns that the trachea is not intubated (Fig. 23-4).

- ETCO₂ is not produced in the absence of pulmonary perfusion. Inadequate CPR will result in no ETCO₂.

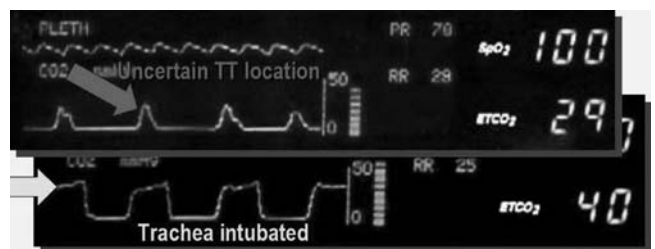


FIG. 23-4. ETCO₂ monitor wave forms. This tracing shows a pulse oximeter waveform on the top. The upper pair of tracings shows an irregular peaked ETCO₂ waveform (top arrow), which does not confirm tracheal intubation. The lower tracing (bottom arrow) shows a regular square-wave ETCO₂ waveform that confirms tracheal intubation. (With permission from Gausche-Hill M, Fuchs S, Yamamoto L, eds. *Instructor's Toolkit to Accompany APLS the Pediatric Emergency Medicine Resource*. 4th rev ed. Sudbury, MA: Jones and Bartlett Publishers; 2008.)

- Auscultation can confirm equal chest aeration, and the absence of gastric breath sounds.
- Visualize chest rise and fall with ventilation and condensation visible in the ETT.
- Improvements in oxygenation and resuscitation parameters are consistent with tracheal intubation.
- Non-ETCO₂ methods do not definitively confirm tracheal intubation; however collectively, the presence of all of these highly support successful tracheal intubation.
- The esophageal detector bulb method (Fig. 23-5). Squeeze the bulb, and then apply it to the ETT. Rapid inflation suggests that the tube is in the trachea or the pharynx. Slow inflation suggests that the tube is in the collapsible esophagus.
- If tracheal intubation is in doubt, direct visual confirmation with the laryngoscope should be attempted.
- Optimize the ETT depth based on the clinician's visual intubation depth (during laryngoscopy) and the ETT-depth guidelines in Table 23-1.

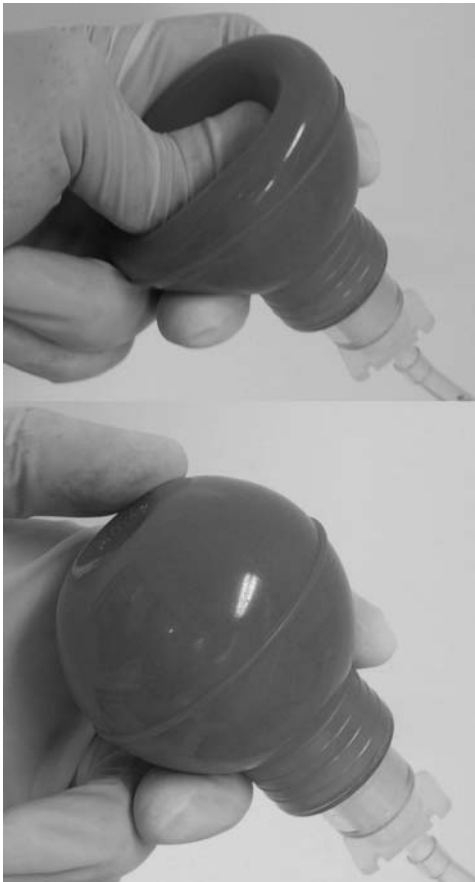


FIG. 23-5. Esophageal detector device (turkey baster). This bulb attaches to the ETT. By squeezing the bulb, it should rapidly inflate if the trachea is intubated. Slow inflation indicates that the esophagus is intubated.

- Secure the ETT (tape or commercial ETT securing products).
- Obtain a chest x-ray to confirm the placement of the tip of the ETT.

THE CAN'T VENTILATE, CAN'T INTUBATE SCENARIO

Airway access through the anterior neck.

- Cricothyrotomy (also known as cricothyroidotomy and transtracheal ventilation). The cricothyroid membrane is punctured with a large IV catheter or an airway quick catheter directed inferiorly. Attach the catheter hub to a 3.0 ETT connector. Use substantial pressure to ventilate with maximal oxygen. A jet ventilation setup can deliver greater tidal volumes through an IV catheter (Fig. 23-6).
- Commercially prepared quick airway kits are better than a large-bore IV catheter. This requires practice.
- An emergency surgical airway such as a tracheostomy or surgical cricothyrotomy is complication prone, but in the “can't intubate, can't ventilate” scenario, this might be the only option to oxygenate the patient.
- Avoid the complication of explosive combustion by avoiding electrocautery in the presence of high oxygen flow.

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QUESTIONS

1. A 10-month-old infant presents with respiratory distress and hypoxia (room air oxygen saturation 89%). A self-inflating bag with a mask is placed in front of the patient's mouth and nose. The tubing

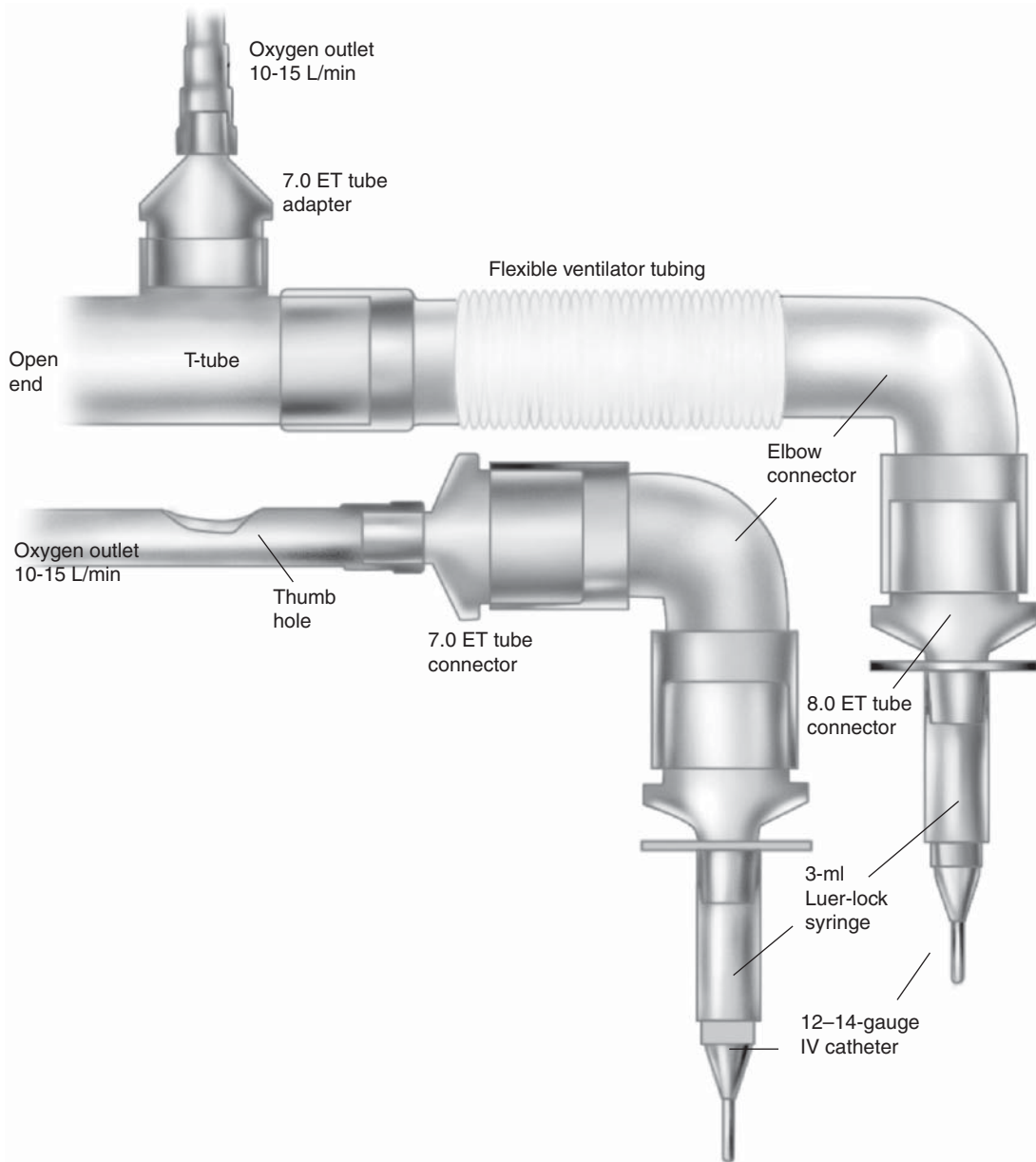


FIG. 23-6. This jet ventilation setup can be used for transtracheal ventilation via an IV catheter. By occluding the open end or the thumbhole, high-pressure oxygen is forced through the transtracheal catheter. Releasing these results in passive exhalation. This diagram shows connection to the IV catheter using a 3 mL syringe and an 8.0 ETT connector. Instead, a 3.0 ETT connector can connect directly into the hub of the IV catheters. However, the connector has no Luer lock onto the catheter hub, and under high inflation pressures, this could pop off. (With permission from Yamamoto LG. Emergency airway management—rapid sequence intubation. In: Fleisher GR, Ludwig S, Henretig FM, eds. *Textbook of Pediatric Emergency Medicine*. 5th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2006:89.)

- is attached to an oxygen outlet at 15 L/minute. The patient's oxygen saturation remains at 89% despite this. Which of the following best explains the most likely explanation?
- The patient has methemoglobinemia
 - The oxygen flow meter is attached to a room air line.
 - No oxygen passes through the mask of a self-inflating bag unless the bag is compressed.
 - The pulse oximeter is inaccurate in hypoxic states.
 - The patient is in respiratory failure and needs to be intubated.
- A 3-year-old child with known congestive heart failure and recurrent pulmonary edema presents to the ED with respiratory distress and hypoxia (room oxygen saturation 85%). A Rusch bag and mask are used to administer oxygen and the oxygen saturation increases to 97%. The Rusch bag has which advantage over the self-inflating bag?
 - The Rusch bag and mask can deliver CPAP (continuous positive airway pressure)
 - The Rusch bag and mask can deliver FiO_2 in excess of 50%
 - The Rusch bag and mask can deliver high inflation pressures.
 - The Rusch bag can be attached to an endotracheal tube.
 - The Rusch bag can be attached an ETCO_2 monitor.
 - A 2-year-old head trauma patient is being transferred to a children's hospital after initial stabilization. The patient is intubated, sedated with thiopental, and paralyzed with rocuronium. He is being hand ventilated using a Rusch bag. His oxygen saturation is 100%. Three minutes prior to arrival at the destination hospital, the oxygen tank of the ambulance is unexpectedly depleted and the Rusch bag deflates. A backup oxygen tank is also empty. What is the best management option?
 - Mouth-to-mouth rescue breathing.
 - See if the ambulance has edrophonium or sugammadex to reverse the rocuronium.
 - Close the tail of the Rusch bag
 - Use a self-inflating bag
 - Extubate the patient.
 - An obtunded 5-year-old child arrives in the ED after being struck by a car. His airway is patent, but he is hypoventilating so BMV is initiated. His heart rate is 130 and his BP is 65/40. IV fluids and O negative PRBCs are infusing. He is intubated using rapid sequence intubation while maintaining C-spine immobilization. Bag ventilation through the colorimetric CO_2 detector and ETT shows no change in the color on the detector. The most likely reason for this is
 - Wrong-size (infant or adult) detector device used.
 - The oxygen flow tube is mistakenly connected to a room air source.
 - Poor pulmonary perfusion.
 - The ETT is in the esophagus.
 - The patient's PCO_2 is too high.
 - A 2-year-old child is undergoing an MRI scan under propofol sedation. From the MRI control and monitoring room, it is very difficult to see the patient's face and chest rise and fall making a visual assessment of ventilation unreliable. It is also impossible to assess ventilation via auscultation because the MRI scan is very noisy. How should ventilation be continuously assessed during the MRI scan under sedation?
 - End-tidal CO_2 monitoring
 - EKG monitoring
 - Visual monitoring of the patient's color
 - EEG monitoring
 - Noninvasive blood pressure monitoring
 - A 7-year-old boy is brought in to the emergency department following rescue from an apartment fire. The child sustained multiple facial burns. Shortly after arrival to the emergency department, patient becomes apneic. You proceed with rapid sequence intubation. Which of the following suggests that the trachea is intubated?
 - Oxygen saturation declines
 - The ETT is taped at the correct depth
 - The esophageal detector bulb (Fig. 23-5) rapidly inflates
 - ETCO_2 monitor shows a spiking waveform (not square wave)
 - Vomitus is suctioned from the ETT.
 - You visit an emergency medicine center in a country that has a very low immunization rate. A 3-year-old child arrives with noisy breathing, shortness of breath, and fever. His temperature is 40°C (104°F). He is diaphoretic and weak in appearance. He is sitting up and leaning forward (tripoding) with audible stridor. His oxygen saturation is 91% on room air. Oxygen is applied. His oxygen saturation declines and his stridor is audibly declining indicating that his ventilation is declining. You attempt to intubate him, but visualization is extremely difficult and his oxygen saturation declines to 40%. The best way to bag mask ventilate this patient is to
 - Use the two person technique in the supine position
 - Use the two person technique in the prone position

- C. Use the one person technique in the lateral position
- D. Use CPAP
- E. Place a rapid surgical airway using electrocautery

ANSWERS

1. C. It is critical to remember that self-inflating bags provide no air (oxygen) flow unless the bag is compressed. It is logical that the OSAT will be unchanged in this scenario since no O₂ is being delivered.
2. A. The Rausch bag can deliver CPAP but the self-inflating bag cannot. Both devices can deliver FiO₂ > 50%, deliver high inflation pressures, can be used with an endotracheal tube, and can be attached to an ETCO₂ monitor.
3. D. If available, a self-inflating would be the best option and does not need an external source of gas flow to deliver room air. Mouth-to-mouth rescue breathing could be used as a last resort if there were no self-inflating bag available. Extubating the patient or reversing paralysis is unnecessary and inappropriate. Closing the tail of the Rausch bag would not help deliver oxygen if there is no flow.
4. D. If the patient has a heart rate and blood pressure, and the CO₂ detector shows no color change, it must be assumed that this is an esophageal intubation. Even with poor pulmonary perfusion, there should be some color change. PCO₂ is never too high to demonstrate a color change if the child is being ventilated. In a 5-year-old either an infant or adult detector should produce a color change.
5. A. The most reliable monitor of ventilation in a sedated patient is end-tidal CO₂ monitoring. Pulse oximetry should also be monitored, but was not an option. All other options in this patient would only be useful after the patient sustained a significant hypoxic event.
6. C. A rapidly inflating esophageal bulb detector demonstrates that the ETT is in a rigid structure (trachea). A declining oxygen saturation, a spiking wave form on ETCO₂ monitor, and vomitus suctioned from the ETT suggest that there is an esophageal intubation.
7. B. In a patient with supraglottitis, it has been demonstrated that in an emergency a patient can be ventilated using the two-person technique until a surgical airway can be established. Using the prone position may help to relieve the obstruction.

24 RESPIRATORY FAILURE

Lynette L. Young

Respiratory failure is the most common cause of cardiac arrest in pediatric patients. It is important to recognize respiratory distress early so that actions can be taken to avoid respiratory failure whenever possible. If respiratory failure does occur, prompt intervention will give the patient the best chance for survival with the least risk of neurologic sequelae. Young children have less physiologic reserve and can deteriorate very rapidly. In a critical situation, the emergency physician has the task of not only making quick resuscitation management decisions but must also consider age-related anatomic differences, appropriate equipment (Table 24-1), and drug dosage differences when caring for infants and children.

ANATOMY AND PHYSIOLOGY

- Young infants may be obligate nose breathers and obstruction of the nasal passages can cause distress.
- Children have more flexible chest walls and less developed chest muscles. Their diaphragms are more prone to fatigue.

TABLE 24-1 Endotracheal Tube Size and Length and Size of Laryngoscope Blades by Age^{a,b}

AGE	SIZE (mm)	TYPE
Endotracheal Tubes		
Newborn	3.0	Uncuffed
Newborn–6 mo	3.5	Uncuffed or cuffed (not in newborn)
6–18 mo	3.5–4.0	Uncuffed or cuffed
18 mo–3 y	4.0–4.5	Uncuffed or cuffed
3–5 y	4.5	Uncuffed or cuffed
5–6 y	5.0	Uncuffed or cuffed
6–8 y	5.5–6.0	Uncuffed or cuffed
8–10 y	6.0	Cuffed
10–12 y	6.0–6.5	Cuffed
12–14 y	6.5–7.0	Cuffed
Laryngoscope Blades		
<2.5 kg	0	Straight
0–3 mo	1.0	Straight
3 mo–3 y	1.5	Straight
3 yr–12 y	2.0	Straight or curved
Adolescent	3.0	Straight or curved

^aUncuffed tracheal tube size (mm) = (age in years/4) + 4 = internal diameter of endotracheal tube or patient's fifth digit.

^bCuffed tube size (mm) = (age in years)/4 + 3 = internal diameter of cuffed endotracheal tube. Depth can be calculated by taking the internal diameter and multiplying by 3.

- Anatomical differences along with the limited alveolar space in young children make them vulnerable to respiratory failure from many causes.
- Tachypnea is a universal finding in infants and young children in respiratory distress. Due to limited lung reserve, an increased respiratory rate is required to augment minute ventilation and facilitate elimination carbon dioxide. The increased work of breathing can lead to muscle fatigue and respiratory failure.
- Infant and children have the potential to desaturate more quickly with intubation. Children have about twice the oxygen consumption rate and proportionally smaller functional residual capacity.

PERTINENT HISTORY

- Infants may present with feeding problem.
- The presenting complaint for an older child may be wheezing or decreased activity.
- Patients with significant past medical history of prematurity with bronchopulmonary dysplasia, asthma, or cystic fibrosis may have increased risk for respiratory compromise.

PHYSICAL EXAMINATION

- Evaluate mental status early as it is an important indicator of respiratory status. Restlessness, anxiety, and irritability may indicate significant respiratory insufficiency. Extreme agitation, lethargy, and somnolence are ominous signs of potential respiratory arrest.
- Increased respiratory rate is present in all patients with respiratory distress; however, normal respiratory rate is age dependent and is influenced by factors such as body temperature and mental state.
- Retractions result from the use of accessory muscles of respiration and imply significant respiratory distress. Nasal flaring may be seen.
- Grunting is most often seen in infants and indicates severe respiratory distress from primary lung disease or systemic illness (sepsis).

LABORATORY STUDIES

- Pulse oximetry is a rapid and useful way to assess a patient's oxygen status. It does not give any information about PCO_2 or acid-base status and can be unreliable in low perfusion states.
- Measurement of arterial blood gas provides a relatively comprehensive picture of respiratory and acid-base status. Strictly speaking, respiratory failure occurs when PaO_2 is less than 60 mm Hg despite

supplemental oxygen of 60%, or $PaCO_2$ is greater than 60 mm Hg.

- Arterial blood gas measurement must be viewed in the context of a patient's baseline respiratory status; patient with underlying lung disease may have chronic hypoxia and CO_2 retention.

ASSISTED VENTILATION

INDICATIONS

- Progressive muscle fatigue (common in infants and young children)
- Apnea
- Inability to maintain a patent airway (altered mental status)
- Refractory shock (to decrease the work of breathing)
- Controlled ventilation (as in increased intracranial pressure)
- Progressive hypoxia or hypercarbia despite maximum therapy.

BAG-VALVE-MASK VENTILATION

- Effective bag-valve-mask (BVM) ventilation of infants and children is a vital skill (Chapter 23).
- BVM ventilation is recommended for infants and children in the prehospital setting especially if the transport time is short.
- Minimizing the time of bagging is important to reduce the possibility of gastric distention and aspiration during intubation.

ADVANCED AIRWAY MANAGEMENT

- Endotracheal intubation and rapid sequence intubation are discussed in Chapter 23.
- A well-positioned and proper-sized endotracheal tube will have an air leak when ventilation is applied at 15–20 cm water level.
- Cuffed endotracheal tubes may be used in the hospital for infants beyond the newborn period.
 - A cuffed endotracheal tube may be helpful in situations of poor lung compliance, high airway resistance, and large glottic air leak.
- Clinically correct endotracheal tube placement is confirmed by observing adequate chest wall expansion and auscultating bilateral breath sounds.
- The laryngeal mask airway with a deflatable mask-like projection at the distal end is an alternative for a patient with a difficult airway.

- Mechanical ventilation is sometimes necessary for an ED patient awaiting admission or transport.
 - The usual tidal volume in an infant or child is 8–10 mL/kg.
 - In the ED setting, PEEP is usually set at 3–5 cm H₂O.
 - Pressure ventilators are used mainly in neonates and young infants.

NONINVASIVE MECHANICAL VENTILATION

- Noninvasive mechanical ventilation (NIPPV) such as continuous positive airway pressure (CPAP), bimodal positive airway pressure (BiPAP), and humidified high-flow nasal cannula (Vapotherm) can provide respiratory support without the risks associated with intubation.
- Benefits may include improved oxygenation and ventilation with decreased muscle fatigue.
- With CPAP there is continuous pressure delivered through the entire respiratory cycle. It can be delivered by face mask in children and binasal prongs in pretermatures, neonates, and infants.
- BiPAP cycles between a higher inspiratory positive airway pressure and lower expiratory positive airway pressure. Successful use of BiPAP requires a cooperative patient and a good-fitting mask.
- NIPPV can be beneficial in acute management of children with asthma, cystic fibrosis, and neuromuscular disease presenting with respiratory distress.
 - Do not use NIPPV in patients who are obtunded, vomiting, hypotensive, and have cardiac dysrhythmias.
 - Any patient receiving NIPPV should be observed closely for deterioration and the ED physician should be prepared for tracheal intubation if necessary.

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QUESTIONS

1. A 3-week-old male was diagnosed with RSV bronchiolitis 2 days ago by his pediatrician. His mom brings him to the ED because of difficulty in breathing. Which of the following is universally present in infants with significant respiratory distress?
 - A. Coughing
 - B. Wheezing
 - C. Difficulty bottle feeding
 - D. Tachypnea
 - E. Stridor
2. A 13-month-old female has history of cough and runny nose for 5 days. She now has fever. On examination she is tired appearing and has decreased breath sounds in her right lung base. A difference in children compared to adults that can lead to children being more vulnerable to respiratory distress is
 - A. The chest walls of children are more rigid.
 - B. The chest muscles of children are more developed.
 - C. The diaphragm of a child is more prone to fatigue.
 - D. The young infant's nose has relatively larger passages.
 - E. The alveolar area for gas exchange is relatively larger.
3. An 18-month-old female is being transported to the ED by paramedics. She had a prolonged seizure and

- is apneic. The transport time to the ED is 5 minutes. The recommendation for ventilation is
- BVM ventilation
 - Endotracheal intubation
 - Laryngeal mask airway
 - Continuous positive airway support
 - Combitube
- A 5-year-old male was riding his bike and was hit by a car. He was intubated with a 5.0 mm cuffed endotracheal tube and transported to the ED. You are confident that the correct size tracheal tube was chosen and that it is well positioned when an audible air leak is heard with ventilation at a pressure of
 - no leak audible
 - 0–5 cm H₂O
 - 15–20 cm H₂O
 - 25–30 cm H₂O
 - >30 cm H₂O
 - A child with moderate persistent asthma is brought to the emergency department. He is in moderate respiratory distress but does not appear to need endotracheal intubation at this time. You decide to use noninvasive mechanical ventilation. In which of the following scenarios would noninvasive mechanical ventilation be the most beneficial for this patient?
 - The patient becomes less responsive.
 - The patient becomes hypotensive
 - The patient becomes hypoxic.
 - The patient becomes more agitated.
 - The patient becomes bradycardic.
 - There is a 9-year-old male in the ED with status asthmaticus. He is in severe respiratory distress despite maximal conventional medical therapy. The ED physician could next consider:
 - High flow, warmed, humidified nasal canula oxygen
 - Nonrebreather mask oxygen
 - IV fluids
 - Chest tube
 - BiPAP
 - A. In the out-of-hospital setting, it has been shown that endotracheal intubation by paramedics compared to bag-mask-ventilation did not improve survival or neurologic outcomes in the pediatric patient. In 2005, AHA recommended that if there is a short prehospital transport time that young children and infants be BVM ventilated.
 - C. A well-positioned and proper-sized endotracheal tube will have an air leak when ventilation is applied at 15–20 cm water level.
 - C. Noninvasive mechanical ventilation benefits may include improved oxygenation and ventilation with decreased muscle fatigue, thus avoiding intubation.
 - E. BiPAP has been used to treat status asthmaticus in pediatric patients in the ED who were refractory to conventional medical therapy with success. This patient should already be receiving oxygen as he is in severe respiratory distress.

25 SHOCK

Jonathan Marr

HYPOVOLEMIC SHOCK

- Most common cause of shock worldwide
- Diarrhea, acute hemorrhage following trauma, burns, and DKA account for the majority of causes of hypovolemic shock.
- Volume depletion and loss of oxygen-carrying capacity both contribute to physiologic dysfunction and ultimately death.

DISTRIBUTIVE SHOCK

- Types include septic, anaphylactic, and neurogenic shock.
- Inappropriate distribution of blood volume causes inadequate perfusion due to poor vascular tone and integrity.
- In sepsis, the loss of the adaptive immune system by apoptosis creates an immunosuppressive state rather than previously thought pro-inflammatory state.
- Anaphylaxis is an immune-mediated hypersensitivity reaction involving IgE and histamine.
- Common causes of anaphylaxis are foods, medications, envenomations, and iatrogenic (blood products, contrast, latex, and vaccines).
- Neurogenic shock results in loss of sympathetic tone from trauma.

ANSWERS

- D. Infants and young patients with significant respiratory distress will be tachypneic. Coughing, wheezing, difficulty bottle feeding and stridor may be present with respiratory distress but tachypnea is the finding that is virtually always present.
- C. The diaphragms of children are more prone to fatigue. The nasal passages of infants and young children may become obstructed, their chest wall is more flexible, their chest muscles are less developed, and their alveolar space is limited.

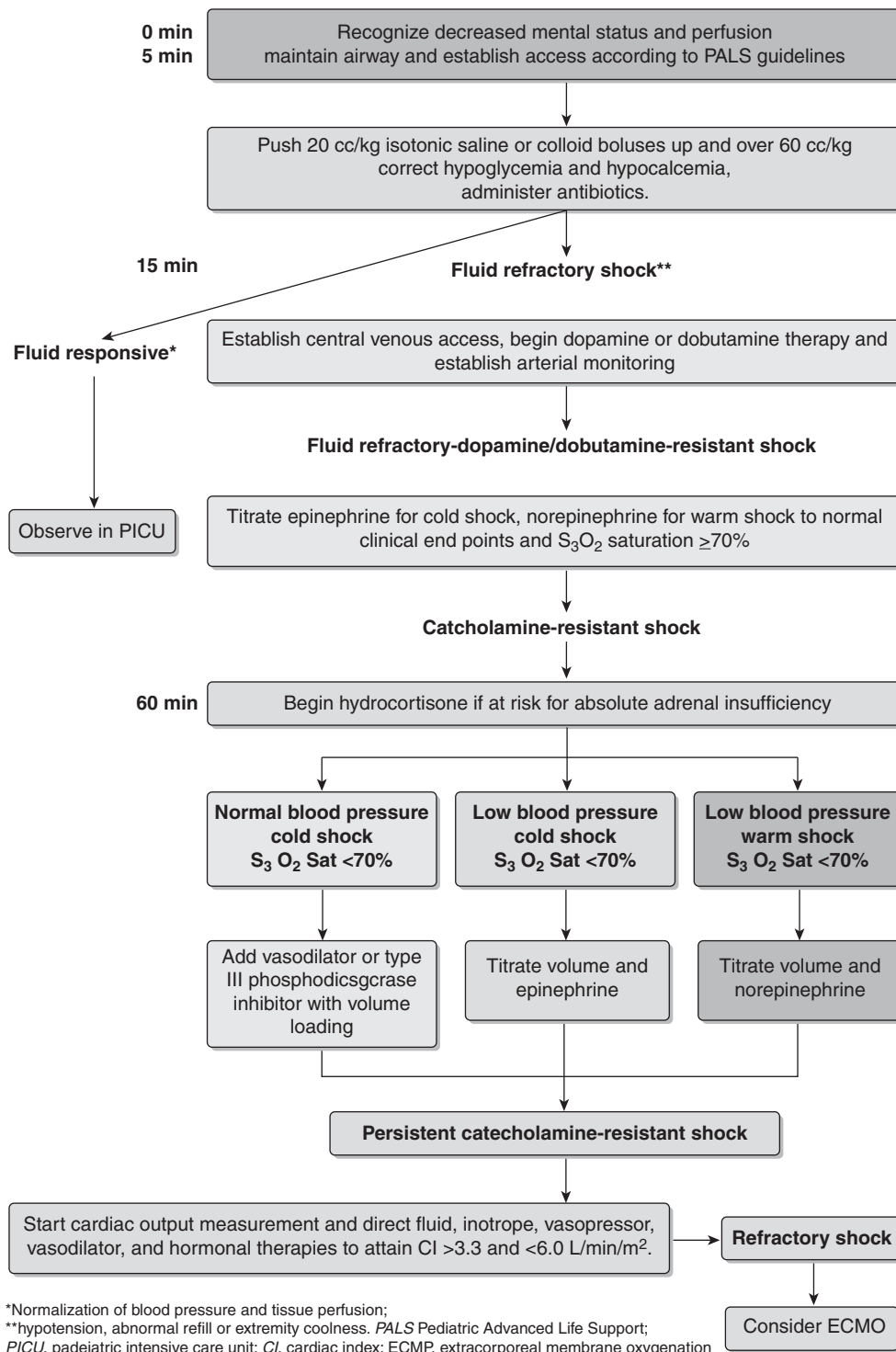


FIG. 25-1. Approach to management of pediatric septic shock. (With permission from Dellinger RP, Levy MM, Carlet JM, et al. Surviving sepsis campaign: international guidelines for management of severe sepsis and septic shock: 2008. *Crit Care Med.* 2008;36:296.)

CARDIOGENIC SHOCK

- Structural or functional (contractility and rhythm) abnormalities in the cardiac cycle can cause cardiogenic shock.
- Low cardiac output and high systemic vascular resistance is the hallmark.
- The neurohormonal response to cardiogenic shock compounds problems to poor cardiac output.

OBSTRUCTIVE SHOCK

- Cardiac output is compromised by physical obstruction from blood from the heart.
- Causes include tamponade, pneumothorax, pulmonary embolism, and congenital heart lesions (i.e., coarctation, aortic arch anomalies, and hypoplastic left heart syndrome).

RECOGNITION

- Beware of tachycardia in child without a source.
- Hypotension in a child heralds impending cardiac arrest.
- Be on the look out for a child with effortless tachypnea as it may reflect compensation for metabolic acidosis.
- Shock is a clinical diagnosis, but labs studies (ie., blood gas, hemoglobin, and bedside glucose) may offer supporting information that can be helpful in guiding treatment.

TREATMENT/INTERVENTIONS

- Early and aggressive intervention (Fig. 25-1) with goal-directed therapy (Table 25-1).

TABLE 25-1 Goal-Directed Management of Pediatric Shock

1. Recognize shock at time of triage
 - a. Hypotension alone with bounding pulses in warm shock
 - b. Diminished peripheral perfusion alone (diminished peripheral compared with central pulses and capillary refill >2 s) in compensated cold shock
 - c. Combination of hypotension with diminished peripheral perfusion in decompensated cold shock
2. Transfer patient immediately to shock/trauma room and amass resuscitation team
3. Begin nasal oxygen and establish intravenous access using 90 s for peripheral attempts
4. If unsuccessful after 2 peripheral attempts, consider intraosseous access
5. Palpate for hepatomegaly; auscultate for rales
- 6a. If liver is up and if no rales are present, push 20 mL/kg boluses of isotonic saline or 5% albumin up to 60 mL/kg in 15 min until improved perfusion or liver comes down or patient develops rales. Give 20 mL/kg pRBCs if unresponsive hemorrhagic shock [18]
- 6b. If liver is down, beware of cardiogenic shock, and give only 10 mL/kg bolus of isotonic crystalloid. Begin PGE1 to maintain ductus arteriosus in all neonates
7. If capillary refill >2 s and/or hypotension persists during fluid resuscitation, begin IO/peripheral epinephrine at 0.05 µg/kg/min
8. If at risk for adrenal insufficiency (e.g., previous steroid exposure, Waterhouse Friderichsen, or pituitary anomaly) give hydrocortisone as bolus (50 mg/kg) and then as infusion titrating between 2 and 50 mg/kg/d
9. If continued shock, use atropine (0.2 mg/kg) plus ketamine (2 mg/kg) for sedation for central line placement. If mechanical ventilation is required, use atropine plus ketamine plus neuromuscular blocker (in skilled hands) for induction for intubation
10. Direct therapy to goals
 - a. Capillary refill <3 s
 - b. Normal blood pressure for age
 - c. Improving shock index (HR/SBP)

pRBC, packed red blood cells; PGE1, prostaglandin E1; IO, intraosseous. Reprinted from Carcillo JA, Han K, Lin J, et al. Goal-directed management of pediatric shock in the emergency department. *Clin Ped Emerg Med.* 2007;8:165, Copyright 2007, with permission from Elsevier.

TABLE 25-2 Vasoactive Agents

DRUG	DOSE	MECHANISM	CLINICAL EFFECT
Dopamine	2–20 µg/kg/min	β ₁ stimulation α ₁ stimulation at >10 µg/kg/min	↑ HR and contractility ↑ SVR
Epinephrine	0.05–1 µg/kg/min	β ₁ stimulation at 0.05 µg/kg/min α ₁ stimulation when >0.3 µg/kg/min	↑ HR and contractility ↑ SVR
Norepinephrine	0.05–1 µg/kg/min	α ₁ stimulation predominates β ₁ stimulation	↑ SVR ↑ HR and contractility
Dobutamine	2–20 µg/kg/min	β ₁ stimulation α ₂ stimulation	↑ HR and contractility ↓ SVR
Milrinone	50–75 µg/kg load 0.5–1 µg/kg/min	↑ cAMP by inhibiting phosphodiesterase	↑ contractility Vasodilation of pulmonary and arterial vessels

HR, heart rate; SVR, systemic vascular resistance; cAMP, cyclic AMP.

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- Restore oxygen delivery by airway support, correction of anemia, and improve blood volume.
- Improve perfusion with fluid resuscitation, unless a congestive heart failure is the likely etiology.
- Reduce oxygen demand (reduce work of breathing, sedation, paralysis, and pain).

PHARMACOLOGIC AGENTS

- Following volume resuscitation, vasoactive agents stimulate receptors in tissue which augment contractility, heart rate, and increase vascular resistance.
- Phosphodiesterase agents are unique as they have both inotropic and vasodilatory properties.
- For complete list of agents with doses and clinical effects, please see Table 25-2.

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QUESTIONS

1. A 6-month-old male is brought to the emergency department with 3 days of vomiting and diarrhea. His other two siblings at home have similar symptoms but are getting better. His last wet diaper was over 12-hours ago and family is concerned since he is very sleepy and not his usual self. On examining, his HR is found to be 170, while sleeping with effortless tachypnea and no fever is noted. Skin is cool and perfusion is 3-4 seconds and the child is listless. The most appropriate step would be
 - A. Obtain stool cultures, if child tolerates liquids, discharge home with supportive care
 - B. Administer PO zofran and then start oral hydration in the ED
 - C. Place IV and start fluids at maintenance
 - D. Place IV and push isotonic fluids until perfusion normalizes
 - E. Begin epinephrine drip at 0.1 µg/kg/min
2. A 4-year-old female presents to the emergency department for coughing and itching 10 minutes after eating great-grandmother's freshly baked peanut butter cookies. She is vomiting in triage and on examining the child is found to have mild swelling of her lips and appears frightened with urticarial lesions on her face and arms. She is diffusely wheezing in both lung fields, and oxygen saturation is 89%. Blood pressure is 110/78. The next most appropriate intervention would be
 - A. Oral diphenhydramine
 - B. Corticosteroids
 - C. Ranitidine
 - D. Epinephrine (1:1,000) intramuscular
 - E. Epinephrine (1:1,000) intravenous
3. An 8-year-old male involved in an ATV crash is brought by EMS to your emergency department with a suspected spinal cord injury. His heart rate is 48, BP is 82/46, and has both arms paralyzed and legs with poor rectal tone. Which of the following is most likely to be found in patients with neurogenic shock?
 - A. Fever
 - B. Euthermia
 - C. Hypothermia
 - D. Poikilothermia
 - E. Peripheral vasoconstriction
4. The most appropriate vasoactive agent for neurogenic shock is
 - A. Epinephrine
 - B. Dopamine
 - C. Dobutamine
 - D. Milrinone
 - E. Diphenhydramine
5. A 4-year-old male with leukemia and central line was recently discharged home to spend the holiday season with the family. He returns to the emergency department since he has been having fever for the last 24 hours. In triage, he is pale and lethargic with increased work of breathing, and perfusion of 6 seconds. His blood pressure is 60/palp, heart rate is 190, and respiratory rate is 48, and oxygen saturation is not recorded. He is started on rapid isotonic fluid resuscitation. Which of the following therapeutic end points would indicate that

- resuscitation is successful and fluids can be slowed down?
- Heart rate of 185
 - Perfusion of 4 seconds with weak but palpable pulses
 - Normal mentation
 - Urine output 0.5 cc/kg/hr
 - BP=74/40
6. A 3-day-old male born at home to a mother who received no prenatal care is brought to the emergency department with poor feeding, sweating and sleepiness since this morning with episodes of pallor. The child is pale, blue, and dusky with oxygen saturation in triage of 78% despite blow by oxygen applied by the nurse. The femoral pulses are weak and perfusion is poor and the infant has increased work of breathing with a rate of 68/min with significant retractions. A loud murmur is noted and an enlarged liver is present on abdominal exam. The child likely has
- Hypovolemic shock from dehydration
 - Septic shock from Group B streptococcus
 - Cardiogenic shock from complex congenital heart disease
 - Pneumothorax
 - Cardiogenic shock from acquired heart disease
7. Based on the above scenario which of the following is recommended?
- Normal saline bolus 40 cc/kg
 - Prostaglandin E2 drip
 - Oxygen delivery at 100%
 - Cardioversion
 - Dobutamine drip
8. A 14-year-old male playing basketball has acute onset of dyspnea and shortness of breath. He tries to continue to play and collapses on the court. Upon arrival to the emergency department, he is fading in and out of consciousness. HR 55, BP 70/30, and has oxygen saturations of 84% on room air. His breath sounds are very diminished on the right and you notice tracheal deviation to the left. After oxygen is applied, the next most appropriate step is
- IV access and IV fluids
 - Intubation
 - Needle thoracocentesis
 - Portable chest x-ray
 - Begin CPR
9. A 6-year-old boy with 4 days of fever, URI symptoms, and generalized malaise comes to the emergency department for extreme fatigue. Vitals: T 99.3, HR 140, RR 33, BP 75/44, and oxygen saturations of 90%. Significant findings on exam include:
- a tired male who is arousable with slight pallor with a gallop rhythm and enlarged liver with a full abdomen and weak pulses. CXR shows cardiomegaly and upon questioning parents, he has no known cardiac problems. Your next step would be to
- Place on oxygen
 - Intubate
 - Give 40 cc/kg bolus
 - Start dopamine drip
 - Obtain EKG
10. An echocardiogram is done on the patient in question #9 and the cardiologist reads it as a dilated cardiomyopathy. The cardiologist would most likely recommend the following treatments:
- Dopamine
 - Milrinone
 - Epinephrine
 - Norepinephrine
 - Corticosteroids

ANSWERS

- D. This child is in compensated shock and based on the pediatric assessment triangle (appearance, color, and work of breathing), he requires emergent intervention. Heart rate of 170 while sleeping with poor perfusion indicates that compensatory tachycardia is not sufficient to maintain perfusion. Without acute and aggressive intervention, the child will progress to decompensated shock with hypotension and likely cardiac arrest. Oral zofran with oral hydration is not likely to be successful since the child is listless. Running IVF at maintenance will not correct the deficit already present. Placing two large-bore IVs and pushing NS is the most appropriate intervention. Following HR, perfusion, color, and mental status can help guide fluid resuscitation. Sometimes, 60 cc/kg of NS is necessary to correct physiologic imbalances. A bedside glucose would be helpful since a 6-month-old child can easily become hypoglycemic with persistent vomiting and poor oral intake in just a day.
- D. This child is having an anaphylactic reaction to peanuts. She has involvement of three systems: skin, pulmonary, and GI. While oxygen and albuterol/racemic epinephrine nebulizer should be started, epinephrine 0.01 mL/kg of 1:1000 IM to the thigh is first-line therapy for acute anaphylaxis and would be the most appropriate step. Autoinjectors (EpiPen) come in 0.3 mg (for >30 kg) and 0.15 mg (for 10–30 kg). Early use

of epinephrine is thought to prevent the biphasic response of allergic reactions. Oral benadryl, ranitidine, and steroids will not work fast enough. According to the 2005 expert consensus practice of Allergy and Immunology specialty guidelines, corticosteroids have no role in the acute management of anaphylaxis. (Joint Task Force on Practice Parameters for the American Academy of Allergy, Asthma and Immunology; the American College of Allergy, Asthma and Immunology; and the Joint Council of Allergy, Asthma and Immunology: The diagnosis and management of anaphylaxis: an updated practice parameter. *J Allergy Clin Immunol* 115:3:S483–S523, 2005. (**Practice Guideline**).

3. C. Loss of sympathetic tone from a spinal cord injury results in bradycardia and hypotension. There is also concomitant dilation of vessels leading to increased heat loss and subsequent development of hypothermia if not monitored appropriately. Temperature regulation involves the hypothalamus and fever would not be expected with a spinal cord injury. Poikilothermia is the inability to regulate temperature and is typically described for reptiles.
4. A. Epinephrine stimulates both alpha and beta receptors, increasing peripheral vascular resistance and heart rate, respectively. Dopamine is an indirect-acting vasopressor since norepinephrine must be released endogenously and subsequently metabolized to an active form. Dobutamine and milrinone will decrease peripheral vascular resistance and are helpful for those in cardiogenic shock. Diphenhydramine is not a vasoactive agent.
5. C. This child is in septic shock. Recommendations are rapid fluid resuscitation via two large-bore IVs. If this cannot be achieved in the first minute, intraosseous placement is recommended. Large volumes of fluids may be necessary during distributive shock—60–100 cc/kg have been reported. Therapeutic end points include normal mentation, normalization of heart rate, perfusion <2 seconds with normal pulses, and urine output of 1 cc/kg/h. Suggested reading includes: Dellinger RP, Levy MM, Carlet JM, et al: Surviving sepsis campaign: International guidelines for management of severe sepsis and septic shock: 2008. *Crit Care Med* 36:296, 2008.
6. C. Complex congenital heart disease is often detected with prenatal ultrasound during pregnancy. This child has heart failure based on the enlarged liver and murmur on exam. Unlike adult heart failure, pediatric heart failure will manifest as subtle history findings such as poor feeding, poor weight gain, diaphoresis with feedings, or difficulty breathing with feedings. Cyanosis with congenital heart disease indicates the presence of the following: tetralogy of Fallot, total anomalous pulmonary venous return, transposition of the great vessels, truncus arteriosus, tricuspid atresia, aortic arch anomalies, and hypoplastic left heart syndrome. The delayed presentation of cardiogenic shock is likely due to the fact that the patent ductus arteriosus is beginning to close, compromising systemic perfusion and increasing pulmonary flow creating pulmonary edema, V/Q mismatch, and increasing work of the heart. Acquired heart disease would be very unlikely in a 3-day old.
7. B. This infant has a ductal-dependent congenital heart lesion causing heart failure. This means that the closure of the patent ductus arteriosus (PDA) is compromising flow to the systemic circulation and shunting the blood to the lungs. Giving more fluid will compound the problem and worsen the heart failure. 100% oxygen will cause the pulmonary bed to vasodilate creating increased blood flow and worsening the pulmonary edema. In this unique instance, oxygen is not your friend. Furthermore, oxygen facilitates closure of the patent ductus arteriosus and can worsen perfusion systemically. Cardioversion has no role as this is not a rhythm problem. Prostaglandin E2 (Prostin) is a drip to keep the PDA open and maintain systemic perfusion while balancing flow to the lungs, thus improving V/Q mismatch. A side effect is apnea so be prepared to intubate. Intubation with sedation and paralysis would likely help this infant since the increased work of breathing increases oxygen consumption and compounds the heart failure.
8. C. This adolescent has a tension pneumothorax and requires immediate needle thoracentesis. There is an external obstruction to blood flow from the heart and until this is removed, cardiac output will be compromised. Fluids and chest x-rays have a secondary role following thoracentesis. Intubation could make the tension pneumothorax worse by giving positive airway pressure.
9. A. This child is in cardiogenic shock. Oxygen via nonrebreather mask will increase oxygen delivery to tissues. Fluid management is important since overzealous fluid administration will worsen contractility since the heart will be working on the wrong side of the Frank–Starling curve. Diuresis with lasix would probably be helpful. Intubation would not be recommended at this time, although may be necessary after maximizing medical management. Dopamine will cause tachycardia and increase afterload which increases myocardial

oxygen demand (bad for cardiogenic shock) and increases the work on the heart. An EKG is indicated, but oxygen delivery is the first priority.

10. B. Milrinone is a phosphodiesterase inhibitor and is effective in heart failure since it increases contractility and decreases peripheral vascular resistance. Dopamine and epinephrine cause tachycardia and increase peripheral vascular resistance. Norepinephrine increases afterload by increasing peripheral vascular resistance and thus makes the heart work harder.

26 CARDIOPULMONARY RESUSCITATION

Alson Inaba

INTRODUCTION

- Unrecognized respiratory failure and/or shock are the most common etiologies of cardiopulmonary arrest in children.
- Early recognition and stabilization of respiratory distress and shock are essential in preventing cardiopulmonary arrest.
- Primary cardiac dysrhythmias are not a common etiology of cardiopulmonary arrest in children.

THE IMPORTANCE OF INCORPORATING HIGH-QUALITY BASIC LIFE SUPPORT INTO ADVANCED LIFE SUPPORT

- High-quality CPR is essential to survival from cardiopulmonary arrest. Refer to Table 26-1 for the five essential components of high-quality CPR.
- The code leader must ensure the quality of chest compressions by checking for and documenting palpable pulses during compressions.
- The correct compression to ventilation ratio for infants and children in the ED who do not yet have an advanced airway in place is 15 compressions to 2 ventilations.
- Avoid overzealous ventilations during CPR as this may cause a decrease in venous return to the heart, which will then result in decreased cardiac output during chest compressions.
- The key points of the 2010 PALS guidelines are summarized in Table 26-2.

TABLE 26-1 Dr. Al's "5 & 2" Rule for High-Quality CPR

Five critical components when performing chest compressions:

1. Correct hand position—the heel of one hand placed over the sternum at the nipple line in children and one finger breath below the nipple line in infants. When two-rescuer CPR is performed in infants, use the two-thumb-encircling hands technique to compress the sternum with the thumbs and squeeze the infant's chest with the encircling fingers.
2. Push hard—compress the chest at least 1/3 of the AP depth of the chest (about 2 inches in children and 1.5 inches in infants).
3. Push fast—compress at a rate of at least 100 compressions per minute. *Note:* If you perform chest compressions at the same beat as the popular Bee Gee's song Stayin Alive, you will achieve a rate of 100 compressions/minute.
4. Allow complete recoil of the chest in order to allow the heart to refill with blood before the next compression.
5. Minimize interruption of chest compressions; perform uninterrupted CPR in 2-min intervals before reassessing the patient.

AIRWAY AND VENTILATION

- Although uncuffed ETTs are used more often in pediatrics, cuffed tubes may be used in the in-hospital setting under certain circumstances (ie, poor lung compliance, large glottic leak, and high airway resistance). Cuff pressure must be monitored and kept < 20 cm H₂O.
- The correct ETT size can be determined by using the formula: (Age in years)/4 + 4.
- Length-based resuscitation tapes (ie, Broselow tape) provide the correct ETT size based on the child's length. The correct size of other equipment used for

TABLE 26-2 Key Changes and Major Points of Emphasis in the 2010 PALS Guidelines

1. Ventilation to compression ratios for infant and child CPR:
 - Lone rescuer: 30:2.
 - Two-rescuer: 15:2.
 - CPR with an advanced airway in place: 8–10 breaths/min, and compressions at 100/min.
 - Apnea with a pulse: 12–20 breaths/min.
2. Palpable pulses during compressions assesses quality of CPR.
3. Avoid overzealous ventilations during CPR.
4. AEDs can now be safely and effectively used in infants and children of all ages. Pediatric attenuator devices are recommended only for infants and children <25 kg. If an attenuator is not available, use the AED in the standard adult fashion. Pediatric attenuator devices should not be used in children >25 kg.
5. Cuffed ETTs may be used under certain circumstances; however, the cuff pressure must be kept <20 cm H₂O.
6. The vascular route (IV or IO) for medication administration is highly preferred over endotracheal administration.
7. High-dose epinephrine is no longer routinely recommended and may be harmful.
8. VF and PVT (pulseless ventricular tachycardia) are now treated with single defibrillations followed immediately by 2-min cycles of high-quality CPR. Third and subsequent defibrillations should be at least 4 joules/kg but should not exceed 10 joules/kg (or the maximum adult dose for the specific defibrillator).
9. Minimize interruptions of chest compression.

respiratory complications (ie, suction catheters and chest tubes) is also provided on these length-based resuscitation tapes.

- Other useful equipment calculations based on the size of the ETT include the following:
 - Suction catheter for any given ETT size = $ETT \times 2$
 - Lip-to-tip distance to tape the ETT at the lip = $ETT \times 3$
 - Chest tube size = $ETT \times 4$
- Confirm proper ETT size selection, proper depth of insertion, and proper ventilation techniques by observing symmetric rise and fall of the chest.
- ETT tube placement should also be confirmed via CO_2 detection devices.
- During cardiopulmonary arrest, the lungs are poorly perfused even with high-quality CPR, and therefore the CO_2 detection device may not detect sufficient amounts of CO_2 despite the ETT being correctly positioned in the trachea. Under this circumstance document symmetric rise and fall of the chest, good bilateral breath sounds, and lack of distension of the stomach during ventilations.
- Check the manufacturer's specifications for the CO_2 detection device since the standard size capnometer that is used in adults and older children may not be able to detect CO_2 in small infants. There are smaller capnometers specifically designed to detect exhaled CO_2 for neonates and infants < 15 kg.
- Continuous capnography or capnometry monitoring may be beneficial in assessing the quality of chest compressions during CPR. An abrupt and sustained rise in end-tidal CO_2 may be observed just prior to clinical identification of the return of spontaneous circulation, so use of continuous end-tidal CO_2 monitoring may spare the rescuer from interrupting chest compressions for a pulse check.

VASCULAR ACCESS PRIORITIES FOR MEDICATION ADMINISTRATION

- Intravenous and/or intraosseous access are the preferred methods for medication administration during any resuscitation. The absorption of medications given via the ETT route is unpredictable and should only be used if vascular access cannot be established.
- IO lines can be inserted into infants and children.
- The preferred site for an IO needle is the proximal tibia (approximately 2–3 cm inferior to the tibial tuberosity over the flat medial portion of the tibia).
- An alternative site for IO placement in older children is the distal tibia (2–3 cm proximal to the medial malleolus).

- It may be possible to aspirate blood when an IO line is placed, which can be sent for various laboratory studies (ie, blood gases, electrolytes, and hematocrit) that may be useful in guiding further management during the resuscitation.
- Infusion of medications and fluids through an IO needle should flow easily and the site should be constantly monitored for soft tissue swelling which would indicate that the IO access has infiltrated.
- An IO line is only a temporary means of vascular access so other attempts at intravenous access should be secured.
- Any medication that can be administered through an IV can be administered through an IO line.

ESTIMATION OF A CHILD'S WEIGHT FOR MEDICATION ADMINISTRATION

- Length-based tapes (ie, Broselow tape) are recommended by the PALS guidelines to estimate the child's weight for medication calculations during the resuscitation. These length-based tapes also provide the resuscitation equipment sizes for ETTs, IV catheters, suction catheters, and chest tubes.
- Length-based tapes do not take the child's body habitus into consideration so clinical judgment will be necessary for extremely thin and extremely obese children.
- If you do not have a length-based tape, the child's weight can be estimated based on the child's age. Refer to Table 26-3.

TABLE 26-3 Estimating A Child's Weight Based on the Child's Age

Full-term neonate = 3–3.5 kg.
Doubles birth weight by 4–6 mo.
Triples birth weight by 1 year of age (1-year-old child = ~10 kg).
The target ages according to this formula are the odd numbered years.
Start at 10 kg for the 1-year-old and simply increase the weight in increments of 5 kg for each subsequent odd numbered target year until age 11 years. After 11 years of age, increase the weight in 10-kg increments in order to compensate for the rapid growth spurt during the adolescent period.
Age = weight:
1 year old = 10 kg
3 years old = 15 kg
5 years old = 20 kg
7 years old = 25 kg
9 years old = 30 kg
11 years old = 35 kg
13 years old = 45 kg
15 years old = 55 kg
17 years old = 65 kg

TABLE 26-4 Clinical Conditions Associated with a High Risk for Developing Dysrhythmias

Congenital heart defects (uncorrected defects and postoperative complications)
Congenital complete heart blocks (i.e., maternal systemic lupus erythematosus)
Myocarditis
Rheumatic heart disease
Kawasaki disease with coronary artery involvement
Prolonged QT syndrome (familial or drug-induced)
Aberrant AV conduction pathways
Comotio cordis
Severe hypoxia
Profound hypothermia
Electrolyte abnormalities (potassium, calcium magnesium disturbances)

A SYSTEMATIC APPROACH TO ARREST AND PREARREST DYSRHYTHMIAS

Although dysrhythmias are not as common in pediatric patients as compared to adults, there are a select group of infants and children who may have a higher risk for developing dysrhythmias. Refer to Table 26-4.

- Any resuscitation scenario can be approached by asking the following two questions (Refer to Fig. 26-1):
 - Is there a pulse? If the child has no pulse, there can be only one of four possible rhythms.
 - If a pulse is present, then what is the child’s heart rate and is the child hemodynamically stable or unstable?
- There are only two general categories of treatment for the four pulseless rhythms:
 - Shockable rhythms: ventricular fibrillation and pulseless ventricular tachycardia
 - Nonshockable rhythms: asystole and PEA
- In addition to treating the rhythm, consider looking for reversible causes of dysrhythmia. The six H’s and six T’s that are taught in PALS include the following:
 - H’s: hypovolemia, hypoxemia, hypothermia, hypoglycemia, hyper/hypokalemia, and hydrogen ion excess (metabolic acidosis).
 - T’s: tamponade, tension pneumothorax, toxins, trauma, cardiac thromboembolus (myocardial infarction), and pulmonary thromboembolus (PE).
- The mnemonic “P-A-T-H. is also useful (Table 26-5).”

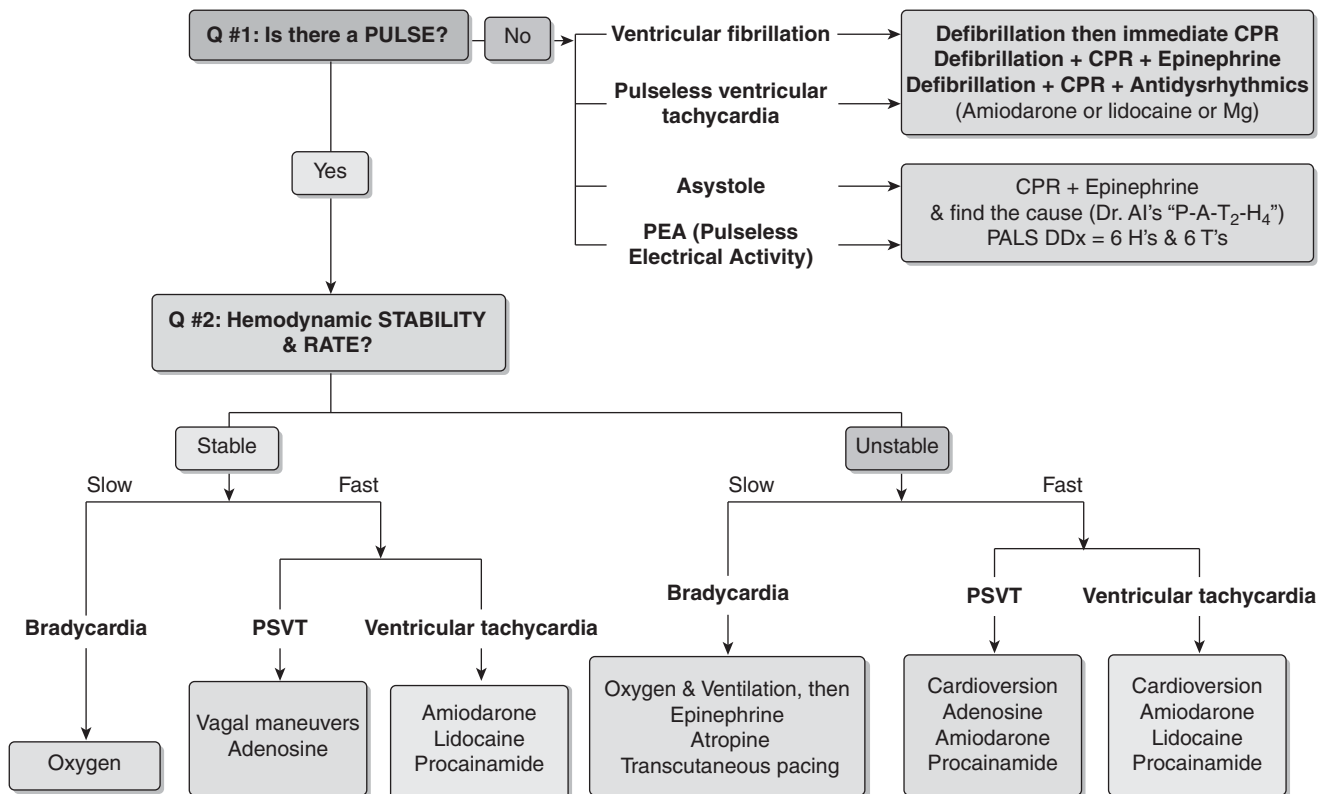


FIG. 26-1. Dr. Al’s simplified and systematic approach to pediatric dysrhythmias.

TABLE 26-5 Reversible Causes of Cardiopulmonary Arrest in Children**“P-A-T2-H4”:**

When confronted with a child in PEA, use the asystole treatment algorithm and “head down the right PATH in the algorithm”:

P = Pneumo-thorax

A = Acidosis

T = Tamponade

T = Toxins

H = Hypovolemia

H = Hypoxemia

H = Hyper/Hypokalemia

H = Hypothermia

Note: The most common cause of PEA in children is hypovolemia. Myocardial and pulmonary thromboses are rare in children.

PAROXYSMAL SUPRAVENTRICULAR TACHYCARDIA (PSVT)

- In young children, PSVT has nonspecific symptoms such as fussiness, lethargy, tachypnea, pallor, and/or difficulty feeding, and can be difficult to recognize. Failure to correct PSVT can lead to congestive heart failure and cardiogenic shock.
- In older children, PSVT has symptoms of palpitations, difficulty breathing, and/or chest discomfort.
- Treatment of pediatric PSVT depends upon the child’s hemodynamic status.
- During any conversion attempt (ie, vagal maneuvers, adenosine, or cardioversion) run a continuous rhythm strip to monitor and document the response to each conversion attempt and also to capture the initial resulting rhythm after the conversion. Capturing a delta wave immediately after the conversion would suggest an underlying accessory conduction pathway such as WPW.
- Avoid verapamil in infants and young children due to the high incidence of profound hypotension and cardiovascular collapse with verapamil in this age group.

SYMPTOMATIC BRADYCARDIA

- Bradycardia is the most common prearrest rhythm in pediatrics and it is usually associated with severe hypoxia, hypotension, and/or metabolic acidosis.
- Hypoxemia and hypoventilation are the most common etiologies of bradycardia in children. Therefore, the first step in the treatment of symptomatic bradycardia in children is to ensure adequate oxygenation and ventilation.
- Other etiologies of bradycardia may include toxic ingestions, medications, increased intracranial pressure, hypothermia, and heart blocks.
- Children who remain bradycardic despite adequate oxygenation and ventilation will require chest compressions and medications.

- Begin chest compressions in a child with signs of poor systemic perfusion when the heart rate is <60 beats/min.
- Epinephrine is preferred over atropine as the first-line agent in the treatment of pediatric bradycardia.

VENTRICULAR TACHYCARDIA (WITH A PULSE)

- The majority of children with ventricular tachycardia have some underlying condition(s) that predisposes them (Table 26-4).
- Other etiologies in children include toxic ingestions and various electrolyte abnormalities (ie, hyperkalemia, hypocalcemia, and hypomagnesemia).
- Avoid amiodarone and procainamide as treatments if congenital or acquired prolonged QT syndrome is suspected as the underlying etiology of the ventricular tachycardia.

VENTRICULAR FIBRILLATION AND PULSELESS VENTRICULAR TACHYCARDIA (PVT)

- Consider ventricular fibrillation or pulseless ventricular tachycardia in cases of commotio cordis or in a child who suddenly arrests.
- In a recent study of pediatric in-hospital cardiac arrests, a shockable rhythm was present during some point during the resuscitation in up to 25% of the cases.
- There has been no change in the recommended energy levels for the first and second defibrillations. However the energy levels for the third and subsequent defibrillations may be increased from a minimum of 4 joules/kg up to a maximum of 10 joules/kg (but not to exceed the adult maximum dose).
- After each defibrillation, immediately resume high-quality CPR for 2 minutes before reassessing the rhythm and pulse.

ASYSTOLE AND PEA

- Asystole is the most common pulseless arrest rhythm in pediatrics.
- Survival rate from PEA is slightly better than asystole provided that the resuscitation team rapidly identifies and treats the underlying reversible etiologies of PEA. Refer to Table 26-5.
- The most common etiology of PEA in children is hypovolemia. Always consider a rapid fluid bolus in addition to CPR and epinephrine when treating PEA.
- (Refer to Tables 26-6 and 26-7 for the key summary points of the treatment of dysrhythmias)

TABLE 26-6 Summary of Pediatric Dysrhythmia Management**Asystole and PEA:**

- CPR (reassess after 2-min intervals).
- Epinephrine (q 3–5 min).
- Treat the underlying cause of PEA (Table 26–5).

VF and PVT:

- Defibrillation followed immediately by 2 min of CPR.
- Defibrillation followed immediately by 2 min of CPR + epinephrine (q 3–5 min).
- Defibrillation followed immediately by 2 min of CPR + (amiodarone or lidocaine or magnesium).

VT (with a pulse):

- Unstable: Immediate cardioversion.
- Stable: Amiodarone or lidocaine or procainamide (*Note:* Avoid concurrent use of amiodarone and procainamide).

PSVT:

- Unstable: If IV access is immediately available administer adenosine while preparing for cardioversion if adenosine fails to convert the PSVT. If IV access is not immediately available and/or if the patient is hemodynamically unstable, perform immediate cardioversion.
- Stable: Various vagal maneuvers (Valsalva maneuver, ice water slurry in a bag applied to the face, blowing on an occluded straw, and/or blowing on the distal end of a syringe in an attempt to blow out the plunger).
- Adenosine if vagal maneuvers fail to convert the PSVT.

Bradycardia:

- Unstable: Ensure adequate ventilation and oxygenation.
 - CPR (reassess after 2-min intervals).
 - Epinephrine.
 - Atropine if suspect an increase in vagal tone or cholinergic poisoning.
 - Cardiac pacing.
- Stable: No emergent treatment is required.

TABLE 26-7 Resuscitation Medications—Defibrillation and Cardioversion Doses**Epinephrine:**

- IV or IO dose (Standard Dose = SD) = 0.01 mg/kg (which equals 0.1 mL/kg of the 1:10,000 epinephrine solution)
- PEDS = SD stands for Pediatric Epinephrine Dosing Story = Slide the Decimals
- Starting with the patient's weight in kg slide the decimal point one spot over to the left to determine the correct volume (milliliters) of the 1:10,000 epinephrine solution to draw up. Then after that volume (milliliters) of epinephrine is administered, slide the decimal point one more spot over to the left to document the correct amount (milligram dose) of the 1:10,000 epinephrine that was given.
- ETT dose (High Dose = HD) = 0.1 mg/kg (which equals 0.1 mL/kg of the 1:1,000 epinephrine solution)

Adenosine:

- First dose = 0.1 mg/kg IV/IO (maximum = 6 mg/dose).
- Second and subsequent doses = 0.2 mg/kg IV/IO (maximum = 12 mg/dose).

Note: Because of the extremely short half-life of adenosine, all doses must be rapidly administered followed by 10–20 mL of NS flush to rapidly get the medication into the central circulation. A proximal vein is preferred over a distal IV site.

Amiodarone (maximum = 300 mg/dose):

- VF and PVT = 5 mg/kg rapid IV/IO bolus.
- Stable VT = 5 mg/kg IV/IO slowly over 20–60 min to avoid the hypotensive effects of amiodarone.
- PSVT unresponsive to adenosine = 5 mg/kg IV/IO slowly over 20–60 min to avoid the hypotensive effects of amiodarone.

Note: Avoid the concurrent use of other medications that can also prolong the QT interval (i.e., procainamide).

Lidocaine (does not cause QT prolongation like amiodarone and procainamide):

- 1 mg/kg IV/IO.

Atropine:

- 0.02 mg/kg IV/IO (minimum of 0.1 mg/dose to avoid paradoxical bradycardia). Maximum single dose of 0.5 mg for a child and 1 mg for an adolescent. May repeat to a total maximum dose of 1 mg in a child and 2 mg in an adolescent.

Dextrose:

- 0.5 g/kg IV/IO (Table 26–8)

Sodium bicarbonate:

- 1 mEq/kg IV/IO (for severe metabolic acidosis, hyperkalemia or cyclic antidepressant toxicity).

Magnesium sulfate (maximum = 2 g/dose):

- 25–50 mg/kg (rapid IV/IO push for PVT because of torsades or slowly over 10–20 min for VT with a pulse because of torsades).

Calcium chloride (10% solution = 100 mg/mL):

- 20 mg/kg IV/IO (equals 0.2 mL/kg of the 10% solution) over 30–60 min.

Note: When calcium and bicarbonate are being infused in an emergent situation flush the IV line with NS after each medication to avoid the formation of an insoluble precipitate in the IV line.

TABLE 26-7 (Continued)

Procainamide:	15 mg/kg IV/IO over 30–60 min for stable VT or for PSVT unresponsive to adenosine.
Defibrillation:	First dose; 2 joules/kg
	Second dose; 4 joules/kg
	Third and subsequent doses; At least 4 joules/kg but not to exceed 10 joules/kg or the adult maximum
Cardioversion:	Start at 0.5–1 J/kg then may increase up to 2 J/kg.

TABLE 26-8 Dr. Al's "Hawaii Five-O" Rule for the Treatment of Hypoglycemia

A bolus of 0.5 g/kg of dextrose will raise the patient's serum glucose by approximately 60–100 mg/dL. If the child remains symptomatic after the first dose of 0.5 g/kg, the same dose may be repeated. Although D10W is the most commonly used dextrose solution in pediatrics, a simple method to quickly draw up 0.5 g/kg of dextrose using any of the four available dextrose solutions is as follows:

(The numeric concentration of the dextrose solution) × (The number of mL/kg of that particular dextrose solution) = the number 50 always.

(Dextrose solution concentration) × (mL/kg of that solution) = 50:

5% × 10 mL/kg

10% × 5 mL/kg

25% × 2 mL/kg

50% × 1 mL/kg

Note: D₅₀W solutions should first be diluted to a less concentrated solution before administration to infants and younger children.

SPECIAL ETIOLOGIES OF CARDIOPULMONARY ARREST AND POSTRESUSCITATION CONSIDERATIONS

- Monitor for and treat hyperthermia, seizures, and various electrolyte abnormalities after resuscitation.
- Avoid routine use of hyperventilation unless the patient is exhibiting signs of impending cerebral herniation.
- Consider using pressors in the post-resuscitation phase since myocardial depression is common in children who survive cardiopulmonary arrest.
- Monitor for and treat hypoglycemia. Refer to Table 26-8 for an easy and quick method to calculate how to administer 0.5 g/kg of IV dextrose.

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QUESTIONS

1. EMS is bringing in a 2-year-old child in full cardiopulmonary arrest. The child has an intraosseous line in the right proximal tibia. The correct dose of epinephrine to administer via the intraosseous line is
 - A. 0.1 mg/kg of the 1:10,000 solution
 - B. 0.01 cc/kg of the 1:10,000 solution
 - C. 0.1 mg/kg of the 1:1,000 solution
 - D. 0.01 mg/kg of the 1:10,000 solution
 - E. 0.1 mL/kg of the 1:1,000 solution

2. A 4-year-old child in full cardiopulmonary arrest has been intubated with an appropriate sized ETT and chest compressions are continued. What is the correct ventilation rate for this intubated child who is still requiring chest compressions?
 - A. 30 compressions to 2 ventilations
 - B. 15 compressions to 2 ventilations
 - C. One ventilation every 2–3 seconds
 - D. 30–40 ventilations per minute
 - E. 8–10 ventilations per minute
3. An average size 6-year-old boy presents to the ED with pulseless ventricular tachycardia after ingesting grandmother's medications. The Broselow length-based tape is not available to determine this child's weight for the resuscitation medication dosing. What is the estimated weight of an average size 6 year old?
 - A. 13 kg
 - B. 18 kg
 - C. 22kg
 - D. 28 kg
 - E. 32 kg
4. A 6-month-old infant is brought to the ED by EMS with a 2 day history progressive lethargy. The initial rhythm on the CR monitor reveals a narrow complex bradycardic rhythm at a rate of about 60 beat per minute. What is the most common etiology of bradycardia in infants and children?
 - A. Head trauma with increased intracranial pressure
 - B. Toxic ingestions
 - C. Congenital heart block
 - D. Hypovolemia due to vomiting and/or diarrhea
 - E. Hypoxia and hypoventilation
5. A 4-month-old unresponsive infant is rushed to the ED by EMS. The infant is cyanotic and has no pulse. However the CR monitor reveals a narrow complex rhythm at a rate of 80–100 beats per minute. What is the most common cause of pulseless electrical activity (PEA) in children?
 - A. Hypoxia
 - B. Toxic ingestions
 - C. Tension pneumothorax
 - D. Cardiac tamponade
 - E. Hypovolemia
6. A 7-year-old boy with myocarditis who was transferred to your ED suddenly becomes unresponsive. The CR monitor reveals ventricular fibrillation. What would be the appropriate initial defibrillation dose for this average size 7 year old?
 - A. 25 J
 - B. 30 J
 - C. 50 J
 - D. 75 J
 - E. 100 J
7. A 4-year-old girl presents to the ED with a 1 hour history of sudden onset chest pain while watching TV. She has no previous cardiac problems and was perfectly fine prior to the onset of her chest pain. The CR monitor reveals a very regular narrow complex rhythm at a rate of approximately 230 beats per minute. She is alert with a BP=95/60 and brisk capillary refill in all extremities. Based on her current hemodynamic status, the initial treatment for her tachycardia would be
 - A. To have her blow into an occluded straw
 - B. A 20 cc/kg fluid bolus of normal saline
 - C. To administer adenosine 0.1 mg/kg via a rapid IV bolus
 - D. To perform immediate cardioversion
 - E. To administer amiodarone 5 mg/kg via rapid IV bolus
8. A 4-year-old child presents to the ED in septic shock. You are preparing to intubate the child but there is no Broselow tape to guide you in selecting the appropriate sized ETT for this child. According to the ETT estimation formula that is taught in PALS, the appropriate sized ETT for this 4-year-old child would be
 - A. 3.0
 - B. 4.0
 - C. 5.0
 - D. 6.0
 - E. 7.0
9. After you have intubated and stabilized the child in the above scenario, the patient suddenly becomes cyanotic and bradycardic while being transferred to the PICU. You suspect that the ETT may have accidentally slid down into the right mainstem. While you are assessing the breath sounds, your colleague is checking to see at what depth the ETT is taped at the lip. What would be an appropriate lip-to-tip distance in this child?
 - A. 10 cm
 - B. 12 cm
 - C. 15 cm
 - D. 18 cm
 - E. 21 cm
10. You have just resuscitated a 2--year-old child with PEA due to profound hypovolemia. A bedside glucose check reveals a blood glucose level of 30 mg/dL. The appropriate amount of dextrose to administer in this child would be
 - A. 60 mL of D10W
 - B. 30 mL of D5W

- C. 80 mL of D25W
- D. 20 mL of D50W
- E. 150 mL of D10W

ANSWERS

1. D. The correct epinephrine solution to administer via the intraosseous line is the 1:10,000 solution. The correct dose of epinephrine is 0.01 mg/kg. Therefore, the correct answer is D. The concentration of the 1:10,000 epinephrine solution is 0.1 mg/mL and the entire 10 mL ampule contains a total of 1 mg of epinephrine. The quick method to accurately calculate the correct volume (mL) and dose (mg) of 1:10,000 epinephrine can be remembered by my “**PEDS=SD**” method. Refer to the epinephrine section in Table 26-7.
2. E. When a patient has an advanced airway in place, the compressions and ventilations are performed asynchronously. The chest compressions are performed at a rate of 100 compressions per minute and ventilations through the advanced airway are performed as a rate of 8–10 ventilations per minute (which is one ventilation about every 6 seconds). Choices C and D are incorrect because this provides too much ventilation in a patient requiring chest compressions. Choices A and B are incorrect because these are the compression to ventilation ratios to use in patients in cardiopulmonary arrest who do not yet have an advanced airway in place. Refer to Table 26-2.
3. C. The average weight of a 5-year old is 20 kg and the average weight of a 7-year old is 25 kg. Therefore, the estimated weight of an average size 6-year old would be somewhere between 20 and 25 kg. Refer to my easy methods of estimating a child’s weight based on the child’s age (Table 26-3).
4. E. Hypoxia and hypoventilation are the most common causes of bradycardia in pediatrics. Therefore, the initial approach to any pediatric patient with symptomatic bradycardia is to ensure that the patient is adequately oxygenated and ventilated. Patients with persistent symptomatic bradycardia despite adequate oxygenation and ventilation will require chest compressions (if the heart rate is <60) and epinephrine. Although head trauma, increased intracranial pressure, toxic ingestions, and heart block can all cause bradycardia, hypoxia and hypoventilation are the most common causes of pediatric bradycardia. Choice D is incorrect because tachycardia is the initial cardiovascular compensation for hypovolemia.
5. E. Profound hypovolemia is the most common etiology of PEA in pediatrics. Therefore, in addition to high-quality CPR and epinephrine, one should always consider a rapid fluid bolus in any child with PEA. The key to successfully resuscitating a child from PEA is to rapidly identify and treat the underlying reversible causes of PEA. Refer to Table 26-5.
6. C. The initial defibrillation dose for pediatric patients is 2 J/kg. Based on the average weight of a 7-year old (refer to Table 26-3) which is approximately 25 kg, the correct initial defibrillation dose would be 50 J. The second defibrillation dose should be at 4 joules/kg, which would be equal to 100 joules in a 25 kg 7 year old. However based on the 2010 AHA PALS guidelines, third and subsequent defibrillation doses should be at least 4 joules/kg but should not exceed 10 joules/kg (or the maximum adult dose for that particular defibrillator device). In other words although 10 joules/kg for a 25 kg child would be 250 joules, if the defibrillator that is being used has a maximum dose of 200 joules for an adult patient, then the maximum joules that should be used for this 7 year old patient would be 200 joules.
7. A. This is a case of hemodynamically stable paroxysmal supraventricular tachycardia (PSVT) or SVT. The treatment approach to PSVT is based on the child’s hemodynamic status. Children who are very hemodynamically unstable with signs of poor perfusion should undergo immediate cardioversion if IV access is not readily available. If an IV access is available, you may try a dose of adenosine while you are preparing for cardioversion. Patients with PSVT who are hemodynamically stable may first undergo conversion attempts with various vagal maneuvers while adenosine is being prepared. Vagal maneuvers that can be attempted are dependent on the child’s ability to cooperate. Applying a bag of crushed ice and water to the face may be tried in infants and children. Older children who are able to cooperate may try other vagal maneuvers such as blowing into an occluded straw or blowing on the distal tip of a syringe in an attempt to blow out the plunger. Choice C would be the next best choice if the child failed to convert with various vagal maneuvers and remained hemodynamically stable. Choice D is incorrect because this child is very stable from a hemodynamic standpoint and does not need immediate cardioversion. Choice A is incorrect because a fluid bolus will do nothing to convert PSVT. Choice E is incorrect because although amiodarone can be used to convert PSVT that has failed conversion with adenosine, a rapid amiodarone bolus will cause hypotension and convert a hemodynamically stable

patient into an unstable patient with hypotension. Refer to Table 26-6.

8. C. The formula for estimating the appropriate ETT size in pediatrics is: $(\text{Age in years})/4 + 4$.
9. C. The formula for estimating an appropriate lip-to-tip distance for any ETT is: $(\text{ETT size}) \times 3$. In this scenario, the 4-year-old child was intubated with a 5.0 ETT. Therefore, an appropriate lip-to-tip distance would be approximately 15 cm at the lip.
10. A The correct amount of dextrose to administer for symptomatic hypoglycemia is 0.5–1 g/kg. The easy method to calculate 0.5 g/kg is my “Hawaii Five-O” method. Refer to Table 26-8. Based on the method to estimate a child’s weight based on the child’s age (Table 26-3), the average weight of a 2-year-old child would be approximately 12–13 kg. Therefore, based on the “Hawaii Five-O” rule, the appropriate amount of dextrose would be 10 mL/kg of D5W which in this case would be equal to 120–130 mL of D5W, or 5 mL/kg of D10W which in this case would be equal to 60–65 mL of D10W.

The more concentrated forms of dextrose D25W and D50W are usually used in older children.

27 NEONATAL RESUSCITATION

Paul J. Eakin

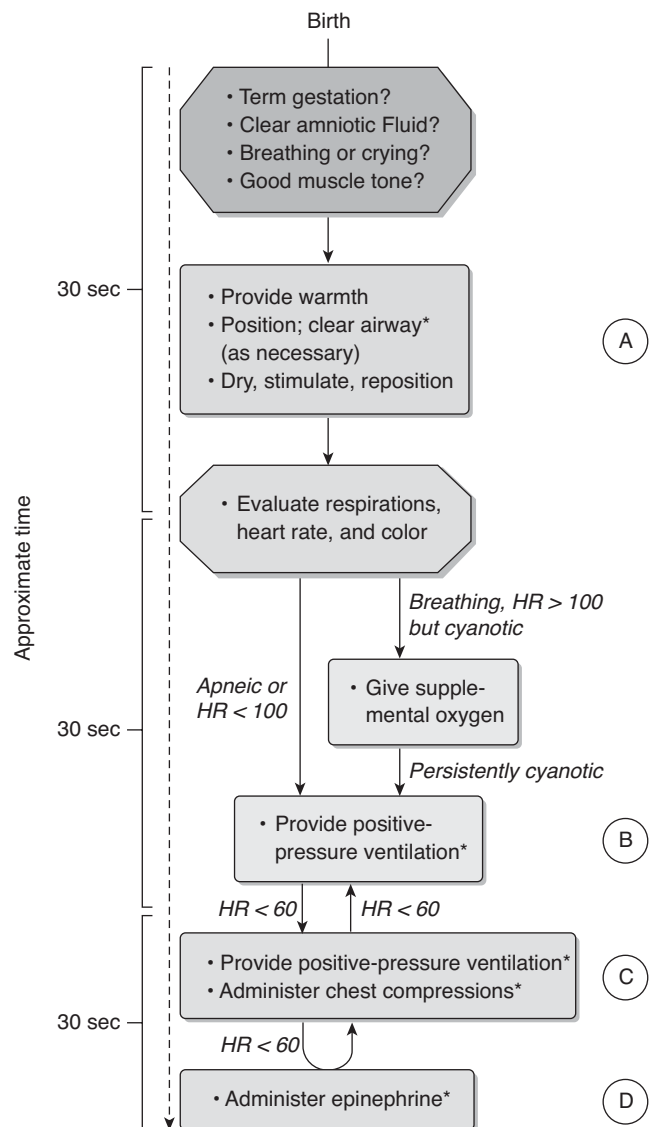
INTRODUCTION

- The vast majority of infants will transition from intrauterine life with little or no intervention.
- After birth and clamping of the umbilical cord, the infant is no longer connected to the placenta and the lungs become the only source of oxygen.
- Systemic blood flow increases as the pulmonary artery pressure decreases and the lungs are filled with air, the amniotic fluid is absorbed, and perfusion of the lungs is established.
- The ductus arteriosus begins closing stimulated by rising blood oxygen levels and more blood is shunted into the lungs initiating the mature circulation.
- Births that occur outside of the delivery room are often complicated by lack of prenatal care, trauma, or prematurity.
- Key maternal history to elicit includes the following:
 - Gestation of the pregnancy
 - Maternal diabetes, hypertension, or drug abuse (especially narcotics)
 - Prolonged rupture of membranes (>18 hours)

- Meconium stained or foul-smelling amniotic fluid
- Severe vaginal bleeding
- Prolapsed umbilical cord

ASSESSMENT OF NEWLY BORN INFANT

- Current American Heart Association guidelines describe several questions which help predict which infants will not require resuscitation (Fig. 27-1):
 - Was the baby born after a full-term gestation?



* Endotracheal intubation may be considered at several steps.

FIG. 27-1. Algorithm for neonatal resuscitation. (Used with the permission of the American Academy of Pediatrics. *Textbook of Neonatal Resuscitation*. 5th ed. American Academy of Pediatrics and American Heart Association; 2006.)

TABLE 27-1 The Apgar Score

PARAMETER	SCORE 0	SCORE 1	SCORE 2
Color	Blue, pale	Body pink, extremities blue	Totally pink
Muscle tone	None, limp	Slight flexion	Active, good flexion
Heart rate (beats/min)	0	<100	>100
Respiration	Absent	Slow, irregular	Strong, regular
Reflex irritability (response to nasal catheter)	None	Some grimace	Good grimace, crying

To calculate Apgar score, add numbers for all parameters together.

- Is the amniotic fluid clear of meconium and without evidence of infection?
- Is the baby breathing and crying?
- Does the baby have good muscle tone?
- If the answer to all of the questions is yes, the infant will likely not require significant resuscitation.
- If the answer to any of these questions is no, then resuscitation may be required and should take place in an orderly fashion (Fig. 27-1).
- The Apgar score is a method of objectively measuring the newborn's condition and response to resuscitation. The Apgar score is assigned at 1 minute and again at 5 minutes, based on respirations, heart rate (best evaluated by palpating the umbilicus), central color, muscle tone, and reflex irritability (Table 27-1).
 - Neonates with a 1-minute Apgar score > 7 require minimal resuscitation other than drying and stimulation.
 - Scores between 4 and 6 indicate that mild to moderate asphyxia has occurred and more vigorous resuscitation may be needed, including supplemental oxygen and vigorous stimulation.
 - One-minute Apgar scores <3 indicated moderate to severe asphyxia and aggressive resuscitation should be initiated immediately.

RESUSCITATION

- Prior to resuscitation, appropriate equipment should be assembled (Table 27-2).
- The infant should be dried with warm towels and placed supine under a radiant warmer to avoid hypothermia.
- The head should be put into a “sniffing” position and oral and nasal suctioning should be performed (Fig. 27-2).
- Vigorous newborns usually only require drying and suctioning of the mouth for stimulation.
- If the newborn does not respond to this tactile stimulation with good respiratory effort, positive pressure ventilation (PPV) should be initiated.
- Free-flow oxygen should be provided to all infants with central cyanosis (evidenced by cyanosis of the mouth and tongue).

TABLE 27-2 Supplies for Neonatal Resuscitation

RESUSCITATION TRAY (STERILE)	RESUSCITATION EQUIPMENT
Bulb syringe	Radiant warmer
DeLee suction trap	Wall suction with manometer
Endotracheal tubes (2.0, 2.5, 3.0, 3.5, and 4.0 mm)	Oxygen source with flow meter
Suction catheters (6, 8, 10, and 12F catheter)	Resuscitation bag (250–500 mL) with manometer
Endotracheal tube stylet	Laryngoscope
Umbilical catheter (3.5, 5F catheter)	Laryngoscope blades (Miller 0 and 1)
Syringes (5, 10, and 20 mL)	Charts with proper drug doses and equipment sizes for various sized neonates.
Three-way stopcock	Warmed linens
Feeding tubes (5, 8F catheter)	
Towels	
Umbilical cord clamps	
Scissors	

- Oxygen is administered at 5 L/min using a face mask or flow inflating bag mask.
- Acrocyanosis, which is cyanosis of the hands and feet, represents peripheral vasoconstriction and is not an indication for oxygen.
- PPV with 100% oxygen is indicated if the infant remains apneic or gasping, the heart rate is <100 beats/min after 30 seconds of initial resuscitation, or has central cyanosis despite supplemental oxygen.
- Ventilation is performed using either cushioned mask with a flow inflating bag or a T-piece connector.

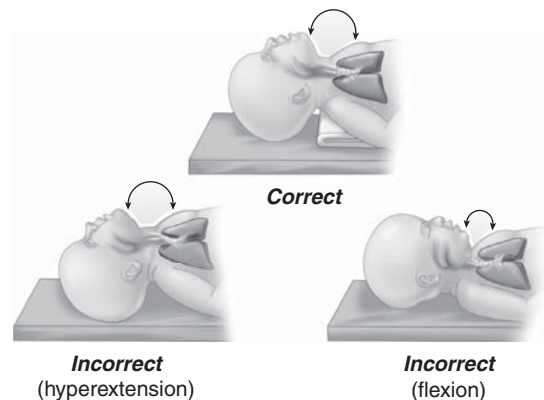
**FIG. 27-2.** Sniffing position.

TABLE 27-3 Endotracheal Tube Sizing

TUBE SIZE (mm) (Internal Diameter)	WEIGHT (g)	GESTATIONAL AGE (wk)
2.5	Below 1000	Below 28
3.0	1000–2000	28–34
3.5	2000–3000	34–38
3.5–4.0	Above 3000	Above 38

Used with the permission of the American Academy of Pediatrics. *Textbook of Neonatal Resuscitation*. 5th ed. American Academy of Pediatrics and American Heart Association; 2006.

- The mask must make a tight seal over the nose and mouth and the pop-off valve may need to be bypassed because of high peak inspiratory pressures needed for the initial breaths (up to 40 cm H₂O).
- It is important to use the lowest pressure that will adequately give chest rise to avoid iatrogenic pneumothorax.
- Bag-mask ventilation at 40–60 breaths/min is continued for 30 seconds and the infant is then reassessed.
 - Assisted ventilation can be discontinued once the heart rate is >100 beats/min; the infant is breathing spontaneously and improvements in color and tone are seen. If the heart rate remains <100 beats/min, assisted ventilation is continued.
 - If the heart rate remains <60 beats/min, despite assisted ventilation, chest compressions are initiated and the infant should be intubated.
- Indications for endotracheal intubation during neonatal resuscitation include the following:
 - Poor response to or the inability to provide adequate PPV
 - Need for endotracheal suctioning or chest compressions
 - Extreme prematurity
 - Diaphragmatic hernia
- Technique for endotracheal intubation:
 - The size of the endotracheal tube depends on the weight or gestation of the newly born infant (Table 27-3).
 - A laryngoscope with a straight blade (Miller) is utilized, using a 0 or 1 blade.
 - Successful endotracheal intubation is evidenced by bilateral chest rise and improvement in heart rate, color, and muscle tone.
 - The use of an end-tidal carbon dioxide monitor can assist with confirmation of proper tube placement, even in very low-birth-weight infants.
 - Care must be made not to advance the ET tube too far, which will result in mainstem bronchus intubation.
 - The result will be diminished breath sounds over half of the chest. The tube should be slowly withdrawn, until equal bilateral breath sounds are auscultated.

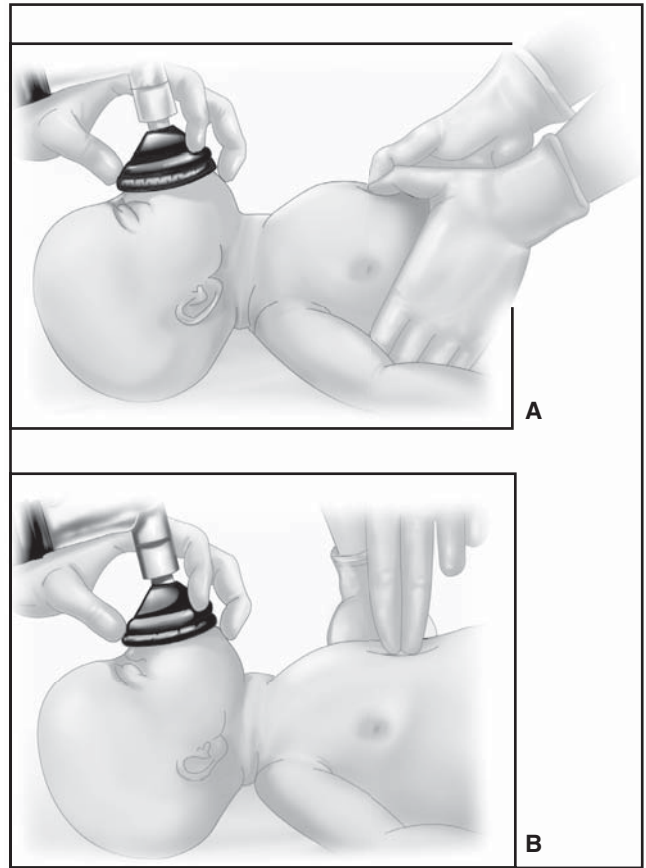


FIG. 27-3. Chest compression techniques. (A) Thumb-circling hand technique and (B) two-finger technique.

- A rough formula for depth of intubation is as follows: depth of tube insertion at gums (in cm) = 6 + infant's weight (in kg)
- Laryngeal mask airways (LMAs) have been shown to be effective for ventilation of term or near-term newborns.
 - They may be considered when bag-mask ventilation or intubation has been unsuccessful.
 - LMAs should only be used by providers experienced in their use.
- Chest compressions are indicated for asystole or when the heart rate remains <60 beats/min, despite 30 seconds of effective positive pressure ventilation.
 - There are two different techniques for administering chest compressions (Fig. 27-3), with the two thumb-circling hand technique being more effective
 - Compressions should be given over the lower third of the sternum to a depth of roughly one-third the diameter of the chest cavity (Fig. 27-4)
 - There should be a 3:1 compression to ventilation ratio with ventilation and compression coordinated to avoid simultaneous delivery.

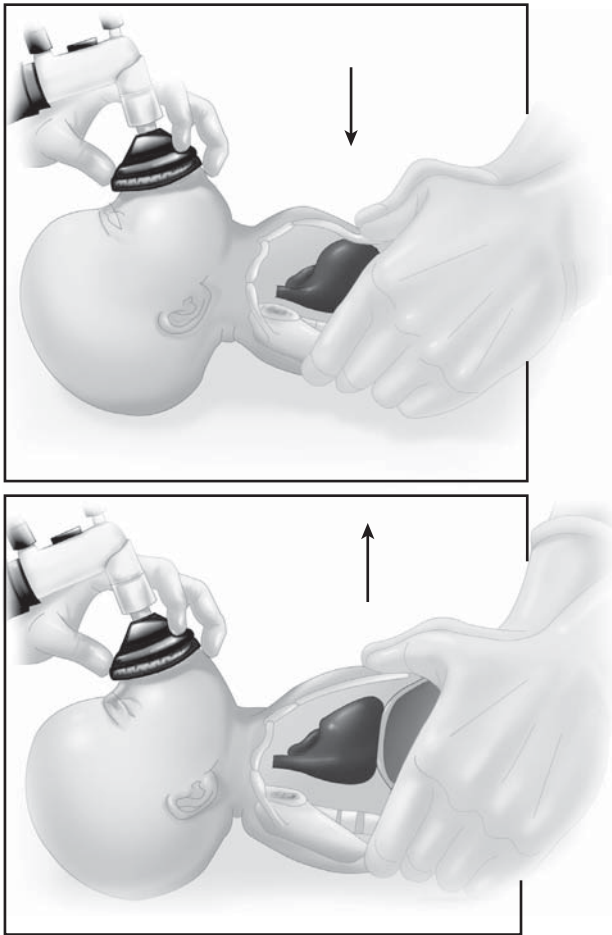


FIG. 27-4. Correct depth of chest compressions.

- There should be 120 events/min (90 compressions and 30 ventilations) resulting in an event roughly twice a second
- Every 30 seconds, respiratory effort, color, pulse, and muscle tone should be reassessed.
- Chest compressions and ventilation can be discontinued when the heart rate is >60 beats/min.

VASCULAR ACCESS

- The umbilical vein is the preferred route for intravenous access in the newly born.
 - The umbilical vein can be identified as a thin-walled structure usually in the 11- or 12-o'clock position in the umbilical cord as opposed to the paired, more muscular umbilical arteries.
 - After cleaning the umbilical cord, the cord is cut aseptically 1–2 cm above the skin line, perpendicular to the cord.
 - A 3.5 or 5 F umbilical catheter can be advanced 2–4 cm or until blood can be easily aspirated from

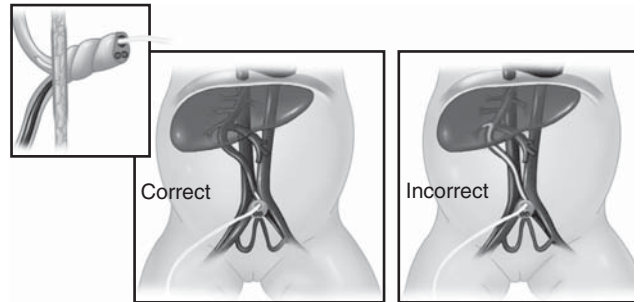


FIG. 27-5. Correct placement of umbilical venous catheter.

the catheter (Fig. 27-5). Deeper advancement of the catheter should be avoided.

- Once the catheter is adequately in place, it should be secured with sutures.

PHARMACOLOGIC AGENTS

- Epinephrine is the most important medication used in infant resuscitation.
 - It is indicated in asystole and in bradycardia with a heart rate less than 60 after 30 seconds of effective ventilation with 100% oxygen and chest compressions.
 - The dose is 0.1–0.3 mL/kg (0.01–0.03 mg/kg) of 1:10,000 solution.
 - Higher dosing is not recommended and may have deleterious effects.
 - The preferred route is IV, although the endotracheal route may be utilized.
 - Epinephrine may be repeated every 3–5 minutes, reassessing the patient between doses.
- Hypoglycemia is often seen in premature infants and infants born to diabetic mothers.
 - Signs of hypoglycemia include jitteriness, hypo or hypertonia, and seizures.
 - Glucose should be administered to maintain normoglycemia (>40 – 50 mg/dL).
 - 2 to 4 mL/kg of D₁₀W given intravenously.
 - Until the infant is stabilized and feedings are established, glucose should be infused at a rate of 6–8 mg/kg/min.
- Naloxone is a direct narcotic antagonist used to reverse respiratory depression when maternal narcotics were administered within 4 hours of delivery.
 - The dose of naloxone is 0.1 mg/kg given either intravenously or intramuscularly.
 - Intratracheal administration of naloxone is not supported by current recommendations.
 - Naloxone should not be given if there is maternal chronic narcotic abuse because severe acute withdrawal syndrome and seizures can occur.

- Sodium bicarbonate may be helpful during prolonged resuscitation for the treatment of hyperkalemia or metabolic acidosis after adequate ventilation has been established.
 - The dose is 1–2 mEq/kg (4.2% solution) given slowly IV over 2 minutes.

VOLUME EXPANSION

- Conditions associated with hypovolemic shock include:
 - Placental abruption
 - Placenta previa
 - Avulsed umbilical cord
 - Twin-to-twin transfusion
 - Premature cord clamping
- Babies in shock appear pale, have weak pulses, delayed capillary refill, and are often persistently bradycardiac despite well-administered resuscitation.
- Volume expansion is given in aliquots of 10 mL/kg of normal saline or lactated Ringer's solution, followed by cross-matched packed red blood cells if there is a poor response to the crystalloid solution.
- Uncross-matched O-negative blood should be administered if a full type and cross-match cannot be obtained in a timely fashion.

SPECIAL SITUATIONS

MECONIUM

- The likelihood of meconium-stained fluid is much higher in infants who are postmature or have in utero fetal distress.
- Meconium aspiration in the newborn can cause
 - severe pneumonitis
 - airway obstruction
 - respiratory failure
 - persistence of fetal circulation
 - profound hypoxia.
- Routine obstetrical suctioning of the oropharynx and nasopharynx after delivery of the head for infants born through meconium stained fluid is no longer advised.
- Further management is based on the activity of the infant, not the consistency of the meconium.
- Vigorous newly born infants do not require endotracheal suctioning of meconium.
- This should be performed only on depressed infants (absent or depressed respirations, heart rate <100 beats/min, and poor muscle tone).
- The technique is to visualize the trachea using a laryngoscope, intubate the trachea, and then suction

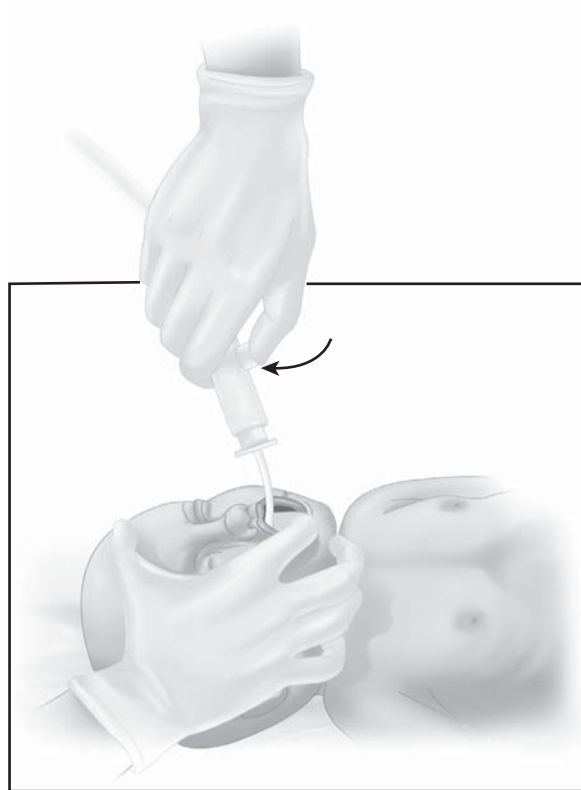


FIG. 27-6. Using meconium aspiration device.

the meconium by attaching the ET tube to wall suction (Fig. 27-6). Multiple attempts and fresh ET tubes may be necessary if the meconium is especially thick or adherent.

- Positive pressure ventilation should be instituted if the infant becomes severely depressed or bradycardic.

PREMATURITY

- Prematurity is defined as birth at less than 37 weeks of gestation; however, most complications occur in more premature infants:
 - Pulmonary immaturity and lack of surfactant
 - Intraventricular hemorrhage
 - Perinatal hypoxia and asphyxia
- For extremely premature infants, the decision to resuscitate can be very complex medically and ethically.
- In general, resuscitation should be pursued when an infant is born with spontaneous heart rate and respirations.
- Withdrawal of care may be considered if there are no signs of life (no heart beat or respiratory effort) after 10 minutes of continuous and adequate resuscitation.

DIAPHRAGMATIC HERNIA

- A diaphragmatic hernia allows abdominal contents to enter the left chest cavity, thereby displacing the lungs.
- The infant will present with respiratory distress, cyanosis, and often has a scaphoid abdomen.
- Bag-mask ventilation should be avoided because it will lead to gastric distension and further respiratory compromise.
- A gastric tube should be placed to decompress the stomach and oxygen should be administered.
- Prompt intubation and positive pressure ventilation is indicated.

OMPHALOCELE/GASTROSCHISIS

- Congenital defects of the umbilical ring result in herniation of abdominal contents into the amniotic fluid.
- Omphalocele describes herniation within the umbilical cord where intestinal contents may be covered by a thin layer of peritoneum. Cardiovascular malformations are frequently seen with this disorder.
- Gastroschisis describes a herniation adjacent to the umbilical cord (usually to the right) and the externalized abdominal contents will be seen.
- Both of these conditions are highly prone to evaporative fluid and heat loss, which may result in hypovolemic shock.
- To prevent this, saline-soaked gauze and a sterile plastic bag should be placed over the protruded abdominal contents. Vascular access should be obtained and fluid resuscitation initiated.
- A gastric tube should be placed and the patient should be transferred to a tertiary center with a neonatal intensive care unit and a pediatric surgeon

PNEUMOTHORAX

- Pneumothorax is reported to occur in 1–2% of term newborns and is often asymptomatic, although premature infants and infants with meconium aspiration syndrome are at higher risk.
- During neonatal resuscitation and PPV, pneumothorax can rapidly lead to tension pneumothorax.
- Infants with this condition will be tachypneic, retracting, grunting, and tachycardic.
- As the tension pneumothorax progresses, the infant may become bradycardic and shocky.
- Transthoracic illumination may be helpful in localizing the affected lung because the breath sounds may be difficult to ascertain.

- Needle decompression is the treatment for tension pneumothorax and can be performed using a 20-gauge needle antiseptically advanced either in the fourth intercostal space in the anterior axillary line or the second intercostal space in the midclavicular line of the affected lung.

INFANT OF A DIABETIC MOTHER

- Infants born to a mother with glucose intolerance are exposed in utero to increased glucose levels and respond with elevated insulin production.
- The infant is prone to
 - Severe hypoglycemia
 - Polycythemia
 - Respiratory distress
 - Intrapartum asphyxia
 - Hyperviscosity syndrome
 - Renal vein thrombosis
 - Birth defects
 - Large for gestational age.
- These infants need to be monitored very closely for hypoglycemia and respiratory distress.

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QUESTIONS

- Paramedics bring a 16-year-old pregnant female who is in active labor into the emergency department. Which of the following historical elements are associated with infants requiring significant resuscitation?
 - 38-week gestation
 - History of maternal yeast infection
 - Gravidity
 - Rupture of membranes 1 hour prior to arrival
 - Maternal history of chronic methadone therapy
- A full-term infant is delivered in your emergency department and the amniotic fluid is meconium stained. You evaluate the infant under the radiant warmer and note gasping respirations, pale color, and heart rate of 85. The best initial step is
 - Immediate endotracheal intubation and suctioning of meconium
 - Vigorous drying and stimulation
 - Initiate chest compressions
 - Intubation and ventilation
 - Bag-mask ventilation
- A term newborn is emergently delivered in the ED. After drying and stimulation, you note cyanosis of the face and mouth, respiratory distress, and a scaphoid abdomen. Breath sounds are diminished over the left chest. Which of the following is contraindicated in the care of this child?
 - Placement of umbilical vein catheter
 - Bag-mask ventilation
 - Immediate intubation
 - Consultation with neonatology
 - Checking bedside glucose
- You are involved with resuscitation of a 32-week infant that was born in the ambulance on the way to the hospital. The infant did not respond to bag-mask ventilation and intubation was performed. Despite adequate chest compressions and ventilation for 30 seconds, the heart rate continues to be less than 60 beats/min. Which of the following is the most appropriate in the management of this infant?
 - Administer sodium bicarbonate at a dose of 1 meq/kg intravenously
 - Administer naloxone at a dose of 0.1 mg/kg intravenously
 - Administer epinephrine at a dose of 0.1 mL/kg of the 1:10000 solution
 - Administer epinephrine at a dose of 0.1 mg/kg of the 1:10000 solution
 - Administer dextrose in a dose of 2 mL/kg of a 10% solution.
- A code blue is called overhead during your shift in the ED. You arrive to the newborn nursery to evaluate an infant born 8 hours earlier with sudden onset of respiratory distress. The infant is tachycardic, tachypneic, grunting, cyanotic, and breath sounds are diminished over the right lung fields. The tracheal appears to be deviated to the left. Which of the following should be done immediately?
 - Call for portable chest X-ray
 - Obtain bedside glucose
 - Endotracheal intubation
 - Needle decompression of the right chest
 - Obtain intravenous access
- A frantic couple brings their 4-hour-old newborn to the ED after he had a 2-minute-long-generalized seizure. The pregnancy and midwife attended home delivery were uncomplicated. On examining, you note a tired appearing infant with decreased muscle tone. A bedside glucose is 30 mg/dL and a peripheral IV has been placed in the right saphenous vein. What medication should be administered to this infant?
 - Administer lorazepam 0.1 mg/kg IV
 - Administer naloxone at a dose of 0.1 mg/kg IV
 - Administer dextrose in a dose of 2 mL/kg of a 25% solution
 - Administer dextrose in a dose of 2 mL/kg of a 10% solution
 - Administer dextrose in a dose of 10 mL/kg of a 10% solution
- You are assisting with resuscitation of a 32-week infant born to a mother with prolonged rupture of membranes > 24 hours. After 1 minute of resuscitation, the infant continues to be limp and pale, with poor muscle tone. Heart rate is 60 and respiratory effort is poor and irregular. When you suction his nose, only a slight grimace is noted. What is the 1-minute Apgar score for this infant?
 - 3
 - 5
 - 7
 - 8
 - 10
- A term infant is born after a pregnancy complicated by placental abruption and vaginal bleeding. On examination, the infant is pale, tachycardic, and has poor muscle tone. You decide to administer volume expansion. What is the preferred route for providing vascular access?
 - Intraosseous line
 - Peripheral IV

- C. Umbilical venous catheter
D. Femoral line
E. Internal Jugular line
9. A 32-week preterm infant is delivered emergently at home and transported to your ED via ambulance. Resuscitation efforts had been initiated and you decide that intubation will be necessary. What is the correct uncuffed endotracheal tube size for this 1600-g infant?
A. 2 mm
B. 2.5 mm
C. 3 mm
D. 3.5 mm
E. 4 mm
10. You are called to the delivery suite following delivery of a term infant. The infant had been dried and stimulated and is under the radiant warmer. He was noted to have cyanosis to lips and face so blow by oxygen was initiated. As you arrive, continued cyanosis and a heart rate of 80 are noted. What is the next best step in resuscitation of this infant?
A. Reposition airway and dry vigorously
B. Administer epinephrine
C. Initiate chest compressions
D. Endotracheal intubation
E. Bag-mask ventilation
- vascular access. Immediate intubation would assist this infant's respiratory status. D and E are steps that may be taken in the care of this infant and are not contraindicated.
4. C. Several medications may be used during neonatal resuscitation. Epinephrine is indicated for bradycardia despite adequate chest compressions and ventilation. The correct dose is 0.1 mL/kg (0.01 mg/kg) of the 1:10000 solution. Sodium bicarbonate is used during prolonged resuscitation to treat acidosis or hyperkalemia. Naloxone is used to treat narcotic-induced respiratory depression. Dextrose is used to treat hypoglycemia.
5. D. This infant has a tension pneumothorax and is rapidly progressing to respiratory failure. Needle decompression is the fastest way to relieve the respiratory and cardiovascular compromise. Chest X-ray may assist in confirming the diagnosis, but will delay the definitive treatment. Endotracheal intubation and IV access may be useful options in this scenario, but will only delay definitive treatment. This scenario does not seem to be consistent with hypoglycemia, so bedside glucose determination has limited value.
6. D. The likely etiology of this seizure is hypoglycemia, which was confirmed by the bedside glucose. The correct dosage of dextrose for hypoglycemia is 2 mL/kg for a 10% solution. 25% dextrose is too hyperosmolar for a peripheral vein. Anticonvulsants are not indicated in this clinical situation. Naloxone could be considered, but there was no history of maternal narcotic exposure.

ANSWERS

1. E. Chronic maternal narcotic exposure places the infant at risk for respiratory depression. 38-week gestation is considered full term. Prolonged rupture of membranes may increase risk of intra-uterine infection, but is defined as more than 18 hours. Gravidity and maternal yeast infection are not considered risk factors.
2. A. For depressed infants born through amniotic fluid, the best initial step is to perform endotracheal suctioning. Immediate stimulation or ventilation may result in meconium aspiration syndrome and worsening respiratory depression. For vigorous infants with HR >100 and showing good respiratory effort, drying and stimulation should be initiated because endotracheal suctioning is not indicated. Chest compressions are indicated if heart rate remains below 60 beats/min despite 30 seconds of adequate ventilation.
3. B. This patient has a congenital diaphragmatic hernia and bag-mask ventilation will lead to distension of abdominal contents and worsening respiratory distress. Umbilical vein catheter will allow for
7. A. The Apgar score is way of objectively measuring the newborn's condition and response to resuscitation. The Apgar score is assigned at 1 minute and again at 5 minutes, based on respirations, heart rate (best evaluated by palpating the umbilicus), central color, muscle tone, and reflex irritability (Table 27-1). This patient is given one point for heart rate of 60, one point for slow, irregular respiratory effort, and one point for grimacing when nose was suctioned. A score less than 3 indicates that moderate to severe asphyxia has taken place and aggressive resuscitation efforts should be initiated.
8. C. Umbilical venous catheter is the preferred vascular access site in infants, especially those with hypovolemic shock. In newborn infants, catheterization of the umbilical vein can usually be obtained rapidly. IO line can be placed in newborns, but will usually not be as easily achieved as UV line. The site for placement is the medial aspect of the proximal tibia and due to a small intraosseous space,

access may be attempted with 20 or 22 gauge spinal needle. A peripheral IV line can be very useful, but in shocky infants, placement may be quite difficult. Central vascular access will not be as rapid as UV catheterization.

9. C. The size of endotracheal tube varies according to gestation and weight (Table 27-2). In general, a 3.5 mm tube should be utilized for a normal sized, term infant.
10. E. Continued cyanosis and heart rate < 100 , despite that supplemental oxygen is an indication for positive pressure ventilation with bag mask. Airway positioning and stimulation have already been performed and it is unlikely that this will help. Endotracheal intubation may be considered depending on response to bag-mask ventilation. Epinephrine is utilized for asystole or bradycardia despite ventilation and chest compressions.

Section 4

TRAUMA

28 EVALUATION AND MANAGEMENT OF THE MULTIPLE-TRAUMA PATIENT

Michael Gerardi

NATURE OF INJURIES AND UNIQUE PEDIATRIC ASPECTS

- Blunt trauma is predominant mechanism in children accounting for 80–90% of trauma Table 28-1.
- Motor vehicle crashes (MVCs) account for as 50% of all childhood trauma deaths, and mortality climbs steeply at 13 years and peaks at 18 years.
- Falls from heights and against fixed objects account for 25–30% of deaths, drowning 10–15%, and burns 5–10%.
- Boys are injured twice as frequently as girls.
- Infants (birth to 12 months), toddlers (1–3 years), and preschoolers (3–5 years) are at greatest risk from falls. They have a proportionately larger head and a higher center of gravity and therefore tend to sustain a higher proportion of isolated closed-head injuries. Infants and toddlers are also at risk of child abuse.
- School-age children (6–12 years of age) are most commonly victims of unintentional trauma, especially motor vehicle-related trauma, as pedestrians, bicyclists, or unrestrained passengers.
- Adolescents (13–19 years of age) engage in many risk-taking behaviors and are at risk for homicide and suicide.
- Children have physiologic and psychologic responses to trauma that are different from those seen in adults.
 - Kinetic energy from injury is distributed over a smaller area and impacts a greater proportion of the total body volume.
 - Musculoskeletal compliance is greater in children and they have less protective muscle and subcutaneous tissue.
 - The increased flexibility and resilience of the pediatric skeleton and surrounding tissues permits external forces to be transmitted to the deeper internal structures.
 - Always consider the possibility of internal injury, even in the absence of external signs of trauma.
- Head injuries are more common in children and account for a large percentage of morbidity and mortality.
- Children's brains have a higher ratio of white to gray matter and thus more resilience in withstanding blunt trauma but more susceptibility to axonal shearing forces and cerebral edema.
- Children's shorter necks and relatively heavier heads make them especially vulnerable to traumatic forces and sudden movements.
- A younger child's short, fat neck makes it difficult to evaluate neck veins and tracheal position.
- The pediatric airway has the most dramatic and critical differences between children and adults (Table 28-2).
 - A child's larynx is located in a more cephalad and anterior position.
 - The epiglottis is tilted almost 45° in a child and is more floppy, making manipulation and visualization for intubation more difficult.
 - The cricoid cartilage is the narrowest portion of the airway and this plus abundant loose columnar epithelium, limits the size of the endotracheal tube, and is the reason that uncuffed tubes are frequently used in children younger than 8 years of age. However, there are certain injuries where a cuffed tube will be preferred.
- The pediatric thorax is more pliable allowing transmission of a greater amount of blunt force to the underlying tissues.

TABLE 28-1 Leading Causes of Trauma in Children by Age Group

<1 y	1–4 y	5–9 y	10–14 y
Suffocation	Motor vehicle traffic	Motor vehicle traffic	Motor vehicle traffic
Motor vehicle traffic	Drowning and submersion	Drowning and submersion	Drowning and submersion
Drowning and submersion	Fire and burn	Fire and burn	Fire and burn
Fire and burn	Suffocation	Suffocation	Suffocation

- The diaphragmatic muscle is much more distensible in a child. A child's mediastinum is also very mobile. Therefore, the mediastinum and abdominal organs are subject to sudden, wide excursions that can be dramatic, such as occurs in tension pneumothorax.
- The diaphragm inserts at a nearly horizontal angle from birth until approximately 12 years of age, in contrast to the oblique insertion in the adult. This, in effect, causes abdominal organs to be more exposed and less protected by ribs and muscle. Therefore, apparent insignificant forces can cause serious internal injury.
- Young children are primarily diaphragmatic or "belly" breathers, making them dependent on diaphragmatic excursion for ventilation.
- Long bones in children differ from the adult because of the presence of growth plates or epiphyses and increased compliance.
 - The epiphyseal–metaphyseal junctions are relatively weak and ligaments are stronger than the growth plate. This weakness predisposes a child to disturbances of the growth plate.
 - Increased compliance of bone results in significant absorption of energy without radiographic signs of fracture even though there is bony damage. The physical examination is often more sensitive than radiographs for growth plate and long bone fractures.

TABLE 28-2 Comparison of Infant and Adult Airways

	INFANT	ADULT
Head	Large, prominent occiput, assumes sniffing position when supine	Flat occiput
Tongue	Relatively larger	Relatively smaller
Larynx	Cephalad position, opposite C2–3	Opposite C4–6
Epiglottis	"Ω" or "U" shaped, soft	Flat, flexible
Vocal cords	Short, concave	Horizontal
Smallest diameter	Cricoid ring, below cords	Vocal cords
Cartilage	Soft	Firm
Lower airways	Smaller, less developed	Larger, more cartilage

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PEDIATRIC TRAUMA SYSTEMS

- Differences in mechanisms and injury patterns together with immature anatomic features and the developing physiology result in unique responses to major trauma, driving the need for specialized pediatric resources optimally at a hospital specializing both in pediatric and trauma care.
- When a pediatric trauma center is not available, this role should be fulfilled by the adult trauma center with the largest volume of pediatric patients.

PREHOSPITAL CARE ISSUES

- Considerations in the field care of the traumatized child include endotracheal intubation, IV access, immobilization, and rapid transport.
- Trauma deaths can be reduced by 25% when a system provides personnel trained to perform airway management and develops guidelines for ground versus aeromedical transport.
 - Rural systems may require more aggressive initial treatment in the field because of transport times that are three to four times greater than those in urban areas.
- Vascular access is a difficult procedure under the best of circumstances and is often a reason for delay in transport of a critically ill child. It is reasonable for traumatized children to be transported immediately without vascular access if a short transport time is expected. Vascular access can be attempted en route to avoid prolonged scene time.
- Intraosseous (IO) infusion (Fig. 28-1) should be used as a quick access for crystalloid infusion if attempts at intravenous cannulation are unsuccessful after 90 seconds.
 - IO lines can be placed successfully in the field 80% of the time.

INITIAL ASSESSMENT AND MANAGEMENT GUIDELINES

- Identify and treat life-threatening injuries immediately; then, identify injuries requiring operative intervention (Fig. 28-2).

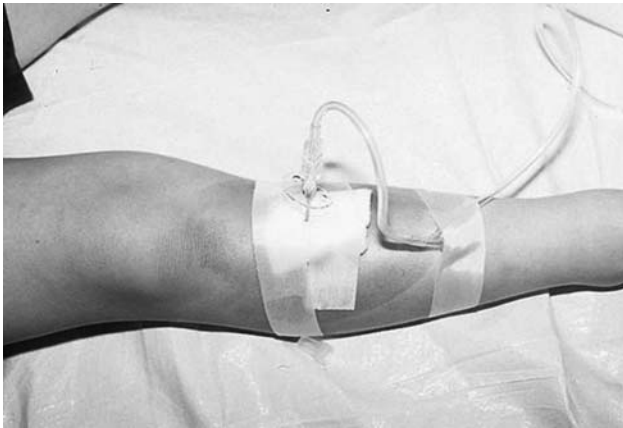


FIG. 28-1. IO lines can be used to ensure adequate fluid administration.

- Examine for non-life-threatening injuries and initiate specific therapy (Table 28-3).
- Use recognized criteria for transferring a patient to a trauma center or activating an in-house trauma team (Tables 28-4 and 28-5).
- Equipment tables or a Broselow tape should be readily available in the resuscitation room as an aid in determining tube and catheter sizes.
- Continually reassess children with serious injuries, repeating vital signs every 5 minutes during the primary survey and every 15 minutes while awaiting transfer or operative intervention.

PRIMARY SURVEY

- The primary survey and resuscitation are carried on simultaneously, including evaluation of the airway, stabilization of the cervical spine (C-spine), adequacy of breathing, and ventilatory effort.



FIG. 28-2. A team approach is required for management of multisystem trauma.

TABLE 28-3 Initial Approach to the Pediatric Trauma Patient

1. Before arrival
Prepare all equipment
Mobilize trauma team and call for assistance (respiratory therapy, nurses, radiology technician)
Have O-negative blood on standby
2. First 5 min
Assess respiration, oxygenation; ventilate if necessary
Intubate or attain surgical airway if indicated
Maintain C-spine immobilization
Check pulse oximetry reading
Cardiac and blood pressure monitoring
Consider end-tidal CO monitoring if available
Perform needle or tube thoracostomy if tension pneumothorax suspected
Treat obvious wounds: Apply pressure to external hemorrhage; dressing to sucking chest wound
3. Second 5 min
Reassess airway, ventilation, oxygenation, temperature
Evaluate level of consciousness/neurologic status
Assess perfusion
Volume resuscitation: 20 mL/kg with crystalloid and repeat as necessary
Consider uncross-matched or O-negative blood
Send laboratory specimens: Type and cross, CBC, amylase, liver transaminases, BUN, creatinine, glucose, electrolytes, ABG, urinalysis
Needle pericardiocentesis, thoracotomy, and aortic clamping if indicated
Nasogastric tube, urinary catheter
4. Next 10 min/secondary survey
Reassess airway, ventilation, oxygenation, perfusion, neurologic status, and disability
Assess head, neck, chest, abdomen, pelvis, neurologic examination, extremities
Tube thoracostomy if indicated
Reduce vascular-compromising dislocations
Administer drugs: Tetanus toxoid, antibiotics, analgesics, sedatives
Lateral neck, chest, and pelvis radiographs
ECG
Start to make arrangements for transfer, admission, and movement to operating room or ICU
5. Next 10 min
Reassess airway, ventilation, oxygenation, perfusion, neurologic status, and disability
Document resuscitation; talk to family
Splint fractures; dress wounds
Ultrasound, CT scan, IVP, DPL, as indicated
Consider more invasive monitoring devices central venous line, arterial line

- Evaluate the circulatory status and control hemorrhage then perform a neurologic screening examination.
- Finally, perform a thorough physical with the patient fully undressed yet kept warm.
- Vital signs vary by age and a table or chart with normal values should be readily available (Table 28-6).

AIRWAY

- Secure the airway while concomitantly stabilizing the neck. Use the jaw thrust maneuver to open the airway and clear the oropharynx.

TABLE 28-4 Criteria for Trauma Activation

Trauma team to ED

Physiologic

- Cardiopulmonary arrest
- Respiratory arrest
- Hypotension
- Neurologic failure (Glasgow Coma Scale score <8)
- Trauma score <12

Anatomic

- Penetrating (gunshot or stab) wound to head, chest, or abdomen
- Facial/tracheal injury with potential airway compromise
- Burn >30% body surface area (BSA); Inhalation airway burn
- Major electrical injury

Trauma alert

Mechanism

- Ejected from motor vehicle
- Extraction time of >20 min
- Fatality of another passenger in MVC
- Intrusion of vehicle >20 inch by collision
- Vehicle traveling >20 mph in pedestrian accident or passenger unrestrained in MVC (>35 mph restrained)
- Fall >20 ft
- Run over by vehicle
- Lightning

Anatomic

- Significant injuries both above and below the diaphragm
- Two or more proximal long bone fractures
- Burn of 15%–30% BSA (second/third degree)
- Traumatic amputation of limb proximal to wrist or ankle
- Crush injury of torso
- Spinal injury with paralysis

- Bony C-spine injuries are less common in children but they are at high risk for cervical cord injuries, which should be assumed until a normal examination and an adequate C-spine series is obtained.
- Indications for endotracheal intubation include inability to ventilate the child by bag-valve-mask methods, need for prolonged airway control, prevention of aspiration in coma, or the need for controlled mild hyperventilation in patients with serious head injuries and children with a GCS score of 8 or lower.
 - It should also be considered for flail chest with pulmonary contusion and in patients with shock that is unresponsive to fluid volume.

TABLE 28-5 Reasons for Transfer of Pediatric Trauma Patients for Tertiary Care

I. Mechanism of trauma

A. Falls

1. Falls >10 ft involving patients <14 y old
2. Falls from second floor or higher

B. Motor vehicle–crash passenger

1. Evidence of high-impact velocity motor vehicle accident
 - a. Shattered windshield
 - b. Evidence of intrusion into the passenger compartment
 - c. Bent steering wheel
2. Rollover incident with unrestrained victim
3. Ejection of the patient from the vehicle
4. Death of an occupant within the same passenger compartment
5. Extraction time >20 min

C. Auto vs. pedestrian incident at >20 mph and victim <15 y old

D. Major burns

E. Blast injuries

II. Physiology

A. Total trauma score of ≤12

B. Pediatric trauma score ≤8

C. Unstable vital signs (age appropriate)

D. Compromise of airway, breathing, or circulation, or need for protracted ventilation

E. Severely compromised neurologic status (Glasgow Coma Scale of ≤8)

III. Injuries

A. Penetrating injuries involving the head, neck, chest, and abdomen or groin

B. Two or more proximal long bone fractures

C. Traumatic amputation proximal to either the wrist or the ankle

D. Evidence of neurologic deficit because of spinal cord injury

E. Flail chest, major chest wall injury, or pulmonary contusion

F. Penetrating head injury, open-head injury, or CSF leak

G. Suspicion of vascular or cardiac injury

H. Severe maxillofacial injuries

I. Depressed skull fracture

- Orotracheal intubation is the most reliable means of securing an airway.
 - Use an uncuffed tube in children <8 years of age, although some may use cuffed tubes for certain types of injury. Appropriate tube size is approximated by the diameter of the nostril or the diameter of the child’s fifth finger (Table 28-7).
 - Emergency intubation should always be accomplished via the oral approach. Nasotracheal intuba-

TABLE 28-6 Pediatric Vital Signs

AGE	WEIGHT, ^a kg	RESPIRATORY RATE	HEART RATE	SYSTOLIC BP ^b
Preterm	2	55–65	120–180	40–60
Term newborn	3	40–60	90–170	52–92
1 mo	4	30–50	110–180	60–104
6 mo–1 y	8–10	25–35	120–140	65–125
2–4 y	12–16	20–30	100–110	80–95
5–8 y	18–26	4–20	90–100	85–100
8–12 y	26–50	12–20	60–110	90–115
>12 y	>40	12–16	60–100	100–130

^aWeight estimate: $8 + [2 \times \text{age (in y)}] = \text{weight (in kg)}$.

^bBlood pressure minimum $70 + [2 \times \text{age (in y)}] = \text{systolic blood pressure}$; $\frac{2}{3} \times \text{systolic pressure} = \text{diastolic pressure}$.

TABLE 28-7 Equipment Sizes for Pediatric Trauma

AGE	MASK SIZE	ORAL AIRWAY	NASAL AIRWAY	LARYNGOSCOPE BLADE	ENDOTRACHEAL TUBE (mm)	FOLEY CATHETER	OROGASTRIC TUBE (F)	SUCTION CATHETER	CHEST TUBE	VASCULAR CATHETER	IO NEEDLE (G)
Newborn	Infant	0		0	3–3.5	5–8	5 or 8 feeding	8	12–18	20–22	
6 mo	Infant/child	1	12	1	3.5	8	8	8	14–20	20–22	17
1 y	Child (s)	1–2	12	1	4.0	8	10	8	14–24	20–22	17
3 y	Child (s)	2	16	2	4.5	10	10	10	16–28	18–22	15
5 y	Child (m)	3	16	2	5.0	10	10–12	10	20–32	18–20	15
6 y	Child (m)	3	16	2–3	5.5	10–12	12	10	20–32	18–20	
8 y	Sm med	4	20	2–3	6.0	12	14	12	24–32	16–20	
12 y	Med lg	4–5	24–28	3	6.5	12–16	14	12	28–36	16–20	
16 y	Med lg	5	28–30	3–4	7.0–8.0	14–18	16–18	12–14	28–40	14–18	

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tion should not be performed in a child younger than age 9.

- Intubation may be necessary to maintain an adequate airway, yet may be difficult because of poor airway visualization, seizures, agitation, or combativeness. Prolonged intubation procedures can lead to an increase in ICP, pain, bradycardia, regurgitation, and hypoxemia.
 - Rapid sequence induction (RSI) can greatly facilitate intubation and reduce adverse effects significantly (see Chapter 23).
- Emergency physicians must be able to secure an airway when unable to perform orotracheal or nasotracheal intubation. There are several options:
 - *Cricothyrotomy* has a role for patients in whom there is extensive central facial or upper airway injury or when there have been unsuccessful attempts at orotracheal intubation. However, it is difficult and hazardous in children and is not recommended in children younger than the age of 10. Complication rates are as high as 10–40%.
 - *Tracheostomy* is time-consuming and hazardous in the ED, in addition to requiring surgical skill.
 - *Needle cricothyrotomy with transtracheal jet ventilation (TTJV)* currently is the preferred surgical method of choice to secure an emergency airway in children. A percutaneous technique and TTJV has several advantages over a surgical cricothyrotomy in the ED.
 - Complications include subcutaneous emphysema, bleeding, and catheter dislodgment.
 - TTJV provides a lifesaving and temporary airway, which should be adequate for 45 minutes to 2 hours, until endotracheal intubation can be achieved.
 - The O₂ source for TTJV *must be a high-pressure* source directly from the wall and not from a regulator valve. A low psi must be used initially in children and the provider should look for adequate chest excursion. The psi can be adjusted upward until adequate chest rise is observed and is the best indicator of adequate tidal volume. The inspiration:expiration ratio is 1:3 or 1:4 (Table 28-8).

TABLE 28-8 Parameters for Transtracheal Jet Ventilation

	INITIAL PSI	ESTIMATED TIDAL VOLUME
Adult	30–50	700–1000 mL
8 y to young teens	10–25	340–625
5–8 y	5–10	240–340
<5 y	5	100

BREATHING

- Acceptable ventilation only occurs if there is adequate spontaneous air exchange with normal O₂ saturation and CO₂ levels.
- Pulse oximetry is mandatory and end-tidal CO₂ monitoring should be used to confirm and monitor endotracheal tube placement.
- Hypoxemia may manifest as any combination of and degree of agitation, altered mental status, cyanosis, poor end-organ function, poor capillary refill, and desaturation on pulse oximetry.
- Children with respiratory failure must have positive pressure ventilation (PPV) started immediately.
 - Signs that indicate that a child has inadequate ventilation include tachypnea, nasal flaring, grunting, retractions, stridor, and wheezing.
 - Reasons for compromised ventilatory function include depressed sensorium, airway occlusion, restriction of lung expansion, and direct pulmonary injury.
 - Restriction of lung expansion by gastric distention is more likely to occur in young children because of the limitation of diaphragmatic excursion. This problem is addressed by early placement of an oro- or nasogastric tube.
- Ventilation with a bag-valve-mask device is initiated to treat inadequate ventilation. Cricoid pressure should be applied when ventilating a patient with a bag and mask to prevent gastric insufflation.
- At this phase of the resuscitation, immediate attention and treatment of tension or hemopneumothorax is required, if present or suspected.
 - Children are especially sensitive to mediastinal shift in tension pneumothorax and needle decompression should be performed immediately if any of the following are present:
 - Decreased breath sounds, refractory hypotension, hypoxia, or radiographically confirmed hemopneumothoraces.
 - Massive hemothorax may present as absent breath sounds, dullness to percussion on the affected side of the chest, and hypotension.
 - Operative thoracotomy should be considered when the initial drainage is greater than 15 mL/kg or the chest tube output exceeds 4 mL/kg/h.

CIRCULATION

- The initial circulatory assessment and treatment includes identifying and controlling both external and internal hemorrhage and assessing perfusion.

Vascular access for fluid infusion and phlebotomy are additional goals.

- The child's volume status and perfusion are estimated by assessment of pulse, skin color, and capillary refill time and obtaining a blood pressure.
 - A palpable peripheral pulse correlates with a blood pressure above 80 mm Hg, and a palpable central pulse indicates a pressure above 50–60 mm Hg.
 - A euvolemic, euthermic patient's capillary refilling time, assessed after blanching, will be 2–3 seconds.
 - Control external hemorrhage by direct pressure. Application of extremity tourniquets or hemostats to bleeding vessels should be avoided unless there is exsanguinating hemorrhage.
 - Trendelenburg position may be of benefit in low perfusion states to maintain central circulation.
- Absent pulses or cardiac arrest in a child with traumatic injuries portends a poor outcome.
- In children with penetrating chest or abdominal trauma, a resuscitative thoracotomy can be lifesaving if vital signs were lost recently.
- During resuscitation of traumatic arrest, standard advanced cardiac life-support algorithms should be followed and there should be early administration of blood products.
- Fluid boluses should be administered early and may temporize until pericardiocentesis and resuscitative thoracotomy is undertaken.

- Obtain vascular access in a rapid and safe manner, with a capability to infuse the greatest possible volume of fluid.
 - Consider an IO line in a severely injured child if vascular access is difficult. Any fluids, medications, or blood products can be given through this line.
 - If central venous access is desired, the femoral vein is the easiest site because of identifiable landmarks and relative ease of the procedure compared to other sites in children.
 - Ultrasound guided placement of central lines has improved safety and ease of inserting them.
 - Administer fluid boluses in aliquots of 20 mL/kg and repeat as necessary until perfusion improves.
 - Since traumatic shock is caused by blood loss, packed red blood cells should be used without hesitation when it becomes apparent that there is significant blood loss and crystalloids are inadequate for volume replacement and if the initial hemoglobin level is 8 or less.

DISABILITY

- *Disability* is assessed by performing a rapid neurologic examination to determine level of consciousness and pupil size and reaction to light.
- The Glasgow Coma Scale is a more quantitative measure of level of consciousness (Table 28-9).

TABLE 28-9 Pediatric Glasgow Coma Scale

SCORE	0–1 (y)	>1 (y)	0–2 (y)	2–5 (y)	>5 (y)
Eye opening					
4	Spontaneously	Spontaneously			
3	To shout	To verbal command			
2	To pain	To pain			
1	No response	No response			
Best motor response					
6		Obeys command			
5	Localizes pain	Localizes pain			
4	Flexion withdrawal	Flexion withdrawal			
3	Decorticate	Decorticate			
2	Decerebrate	Decerebrate			
1	No response	No response			
Best verbal response					
5			Appropriate cry, smiles, coos	Appropriate words and phrases	Oriented, converses
4			Cries	Inappropriate words	Disoriented, converses
3			Inappropriate cry	Cries/screams	Inappropriate words
2			Grunts	Grunts	Incomprehensible sound
1			No response	No response	No response

Note: A score is given in each category. The individual scores are then added to give a total of 3–15. A score of <8 indicates severe neurologic injury.

TABLE 28-10 AVPU Method for Assessing Level of Consciousness

A	Alert
V	Vocal stimuli: Responds
P	Painful stimuli: Responds
U	Unresponsive

- In the midst of the trauma resuscitation, the AVPU system (Table 28-10) is helpful in following mental status changes.

EXPOSURE

- Completely undress the patient in order to perform a thorough assessment.
- Children have a larger body surface area to weight ratio, so preventing hypothermia is a constant concern.

RESUSCITATION

- This phase occurs simultaneously with the primary survey.
- Ensure adequate oxygenation and ventilation of all trauma victims.
- Vascular access is the next priority via percutaneous or cutdown cannulation of the upper or lower extremity veins.
 - Establish two large-bore intravenous lines, with size guided by the size of available veins.
- Send blood for type and cross-match, complete blood count, serum electrolytes, liver transaminases, and amylase.
 - Liver transaminase elevation in the acute trauma setting serves as a marker of liver injury that might not be clinically apparent.
- Send a blood gas for any patient who may have significant volume loss, respiratory compromise, or

concomitant toxic exposure (eg, carbon monoxide poisoning in a burn patient).

- Perform an assessment for shock and determine whether or not adequate organ perfusion exists. Shock after trauma is almost always due to hypovolemia.
 - *Hypovolemic shock* occurs most commonly after major trauma and is caused by blood loss (Table 28-11).
 - The blood volume of the child makes up 8–9% of the total body weight.
 - Determination of volume depletion and shock is difficult in children, and multiple parameters must be used. Hematocrit can be normal in the face of acute blood loss—and blood pressure alone is an insensitive indicator of shock, especially when determining treatment priorities.
 - Pulse and respiratory rate and mental status are more sensitive in identifying early stages of shock.
 - *Cardiogenic shock* after a major childhood injury is rare but could occur because of cardiac tamponade or direct cardiac contusion. It should be suspected if there are dilated neck veins in a patient with decelerating injury, penetrating chest trauma, or sternal contusion.
 - *Neurogenic shock* presents with hypotension without tachycardia or vasoconstriction and is usually due to spinal cord injury.
- Normal saline (NS) or Ringer's lactate (LR) is the fluid of choice for initial resuscitation of the pediatric trauma victim.
 - Fluid replacement can be divided into two phases: (1) initial therapy and (2) total replacement (Tables 28-12 and 28-13). The initial resuscitative fluid should be isotonic crystalloid solution. Give an initial infusion of 20 mL/kg as rapidly as possible.
 - After a rapid 20 mL/kg bolus over 10 minutes, the child should be reassessed.
 - Repeat fluid boluses up to four times if necessary. If the child continues to be unstable, 10–20 mL/kg

TABLE 28-11 Therapeutic Classification of Hemorrhagic Shock in the Pediatric Patient

	BLOOD LOSS PERCENT OF BLOOD VOLUME ^a			
	UP TO 15	15–30	30–40	≥40
Pulse rate	Normal	Mild tachycardia	Moderate tachycardia	Severe tachycardia
Blood pressure	Normal or increased	Decreased	Decreased	Decreased
Capillary refill	Normal	Positive	Positive	Positive
Respiratory rate	Normal	Mild tachypnea	Moderate tachypnea	Severe tachypnea
Urinary output	1–2 mL/kg/h	0.5–1.0 mL/kg/h	0.25–0.5 mL/kg/h	Negligible
Mental status	Slightly anxious	Mildly anxious	Anxious and confused	Confused and lethargic
Fluid replacement (3:1 rule)	Crystalloid	Crystalloid	Crystalloid + blood	Crystalloid + blood

^aAssume blood volume to be 8%–9% of body weight (80–90 mL/kg).

TABLE 28-12 Classification for Fluid Resuscitation in Shock

	CLASS I	CLASS II	CLASS III	CLASS IV
Blood loss%/blood volume	Up to 15%	15%–30%	30%–40%	40% or more
Pulse rate	Normal	Mild tachycardia	Moderate tachycardia	Severe tachycardia
Blood pressure	Normal/increased	Normal/decreased	Decreased	Decreased
Capillary blanch test	Normal	Positive	Positive	Positive
Respiratory rate	Normal	Mild tachypnea	Moderate tachypnea	Severe tachypnea
Urine output	1–2 mL/kg/h	0.5–1 mL/kg/h	0.25–0.5 mL/kg/h	Negligible
Mental status	Slightly anxious	Mildly anxious	Anxious/confused	Confused/lethargic
Fluid replacement	Crystalloid	Crystalloid	Crystalloid + blood	Crystalloid + blood

packed red blood cells or whole blood need to be infused urgently.

- The 3:1 rule is commonly used in replacing lost blood with crystalloid as follows: 300 mL of crystalloid for each 100 mL of blood loss.
- If the initial hemoglobin value is <7, blood should be given immediately since this level of hemoglobin overwhelms compensatory mechanisms and increases cellular hypoxia.
- If a child is not responding, suspect continued bleeding and look for other causes of refractory shock, such as tension pneumothorax or hypoxemia.

TABLE 28-13 Guidelines for Fluid Resuscitation in Shock

Mild shock (15%–25% of blood volume loss)

Initial volume:

20 mL/kg LR or NS

If no improvement, repeat 20 mL/kg LR or NS

Total volume:

If improved, run LR or NS at 5 mL/kg/h for several hours

If child remains stable, adjust intravenous rate downward toward maintenance levels

Maintenance after volume is restored:

10 kg: 100 mL/kg/24 h

10–20 kg: 1000 mL 50 mL/kg/24 h

20 kg: 1500 mL 20 mL/kg/24 h

Moderate shock (25%–40% of blood volume loss)

Initial volume:

20 mL/kg/LR or NS; repeat immediately if not improved

If no improvement, alternative therapy includes:

20–40 mL/kg LR, or NS again, or 10–20 mL/kg packed red blood cells, or operative intervention

10–20 mL/kg packed red blood cells if initial Hgb < 7.0

Total volume:

If improved, run LR or NS at 5 mL/kg/h for several hours

If a child remains stable, adjust intravenous rate toward maintenance levels

May need transfusion depending on clinical response and hematocrit

Severe shock (>40% of blood volume loss)

Initial volume:

Push LR or NS until colloid available

Push packed red blood cells or whole blood

Surgery

Total volume:

Replace loss with type-specific blood

- Insert a Foley catheter and use urinary output as a straightforward, readily available monitor as directed: 1 mL/kg/h for children >1 year of age and 2 mL/kg/h for children <1 year of age. Urinary output may help assess perfusion and intravascular status.
- While restoring or immediately after attaining adequate perfusion, place a urinary and gastric catheter.
 - If there is blood at the urethral meatus, in the scrotum or abnormal placement of prostate on rectal examination, urinary catheterization must be done after a retrograde urethrogram (RUG) proves that the urethra is intact.
- Nasogastric tube insertion should be avoided or performed with the utmost care to avoid passage into the brain via a cribiform plate fracture when a patient has blood coming from the ears, nose, or mouth.
- Measure and monitor the patient's body temperature. Hypothermia must be avoided and/or corrected. Use radiant warmers, warmed IV fluids, cover exposed body parts, and raise the room temperature.
- Obtain radiographs at this time, but limit them to C-spine, chest, and pelvic films until the patient is initially resuscitated.
- It is important to obtain surgical consultation in any significantly injured child as early in the evaluation as possible (Table 28-5).

SECONDARY SURVEY AND DEFINITIVE CARE

- Once life-threatening conditions identified in the primary survey are stabilized, perform a timely, directed evaluation of each body area, proceeding from head to toe. Continuously reassess vital signs and any abnormal conditions identified in the primary survey every 15 minutes at a minimum.
- The components of the secondary survey include a history, a complete head to toe examination, laboratory studies, radiographic studies, and problem

identification. Use an AMPLE history to determine the mechanism of injury, time, status at scene, changes in status, and complaints that the child may have. This includes allergies, medications, past medical and surgical history, last meal time, and events preceding the injury.

- Complete laboratory and radiologic studies that were not done during the initial resuscitation.
- A decision regarding disposition can usually be made at this point during most resuscitations.

HEAD EXAMINATION

- Reevaluate pupil size and reactivity. Perform a conjunctival and fundal examination for hemorrhage or penetrating injury. Assess visual acuity by determining if the patient can read, see faces, recognize movement, and distinguish light versus dark.
- Palpate the skull and mandible looking for fractures or dislocations.
 - Although relatively uncommon, infants may become hypotensive from blood loss into either the subgaleal or epidural space.
 - Unlike adults, vomiting and altered mental status, such as amnesia, commonly occur in head injured children and do not necessarily imply increased ICH.
 - However, persistent vomiting, progressive headache, palpable skull defect, or an inability to observe a patient's mental status (eg, they are going to the operating room) are some of the indications for an immediate head CT scan.

CERVICAL SPINE

- Injuries of the C-spine are not common in children but the presence of one or more of several injuries increases the risk.
 - These include injuries above the clavicles, injuries from falling >1 floor, motor vehicle–pedestrian crash at >30 mph, unrestrained or poorly restrained occupant of a MVC, sports injuries, etc.
 - Children tend to have injuries of the upper C-spine and cord.
 - In very low-risk injuries, the C-spine can usually be cleared in the ED with a normal C-spine films and a normal clinical examination.
 - C-spine films must show all seven C-spine vertebrae and that the patient should be awake, cooperative, and free of other distracting painful injuries before ruling out a cervical injury. Children with high-risk mechanisms of injury

should have three views: anteroposterior (AP), an odontoid, and lateral views.

- The child, performing the movements voluntarily, should actively flex, extend, and rotate the neck with no symptoms or signs of spasm, guarding, pain, or tenderness.
- The incomplete development of the bony spine, the relatively large size of the head, and the weakness of the soft tissue of the neck predispose to spinal cord injury without radiographic abnormality (SCIWORA).
- Patients with altered sensorium cannot be cleared despite negative films and the collar should remain in place while further testing and imaging studies are completed.
- Special considerations are required in four situations:
 1. The child who requires immediate intubation because of airway compromise should not have airway management delayed waiting for C-spine film(s). The safety of oral intubation with in-line C-spine immobilization has been demonstrated in multiple studies.
 2. If such a child who is intubated is at high risk for C-spine injury, then a CT scan of the upper cervical vertebrae should be done when a head CT scan is performed.
 3. If an injured patient arrives with a helmet in place and does not require immediate airway intervention, then lateral C-spine can be done prior to removing the helmet. There should be careful attention to maintaining C-spine immobilization while removing the helmet.
 4. Penetrating injuries to the neck requiring operative intervention should have entry and exit sites noted with opaque markers on AP and lateral films of the C-spine.

CHEST

- Expose and visually inspect the chest for wounds requiring immediate attention.
 - Sucking chest wounds require a sterile occlusive dressing.
 - A flail chest component could be splinted but the patient may need intubation to do so.
 - Roll the patient, keeping in-line spine immobilization, and look for posterior wounds. Auscultate the chest and evaluate for pneumothorax, hemothorax, or cardiac tamponade.
 - A hemodynamically unstable child should undergo immediate needle decompression thoracentesis if there is reason to suspect blunt or penetrating

injury to the thorax. After thoracentesis, tube thoracostomy(ies) should be done.

- Impaled objects protruding from the chest should be left in place until the child undergoes surgery.
- If the chest radiograph reveals a widened mediastinum or apical cap, or other signs suggesting aortic injury or there is a history of significant deceleration injury, CT angiography of the chest is indicated. Aortography may be needed in select circumstances.
 - Air lucencies on chest radiography appearing to be of intestinal origin should be considered evidence of a diaphragmatic injury. Any penetrating injury to abdomen or lower chest carries a risk of diaphragmatic injury.

ABDOMEN

- During the secondary survey, determining the exact etiology of an abdominal injury is secondary to determining whether or not an injury is present.
- Retroperitoneal injuries are difficult to identify, unless there is a high index of suspicion.
- Signs suggesting abdominal injury include abdominal wall contusion, distention, abdominal or shoulder pain, and signs of peritoneal irritation and shock. Penetrating wounds to the abdomen usually need immediate operative intervention Figure 28-3.
- The role of the focused assessment sonography in trauma (FAST) is unclear. While it may identify intra-abdominal blood that is not enough information to dictate management other than to mandate immediate laparotomy in such a patient who is also hypotensive and not responding adequately to crystalloid and packed red blood cell expansion.



FIG. 28-3. Children with bruises from seatbelts should be checked for deceleration injuries.

- DPL, although rarely done, may have a role in the hypotensive, injured child because it is valuable in deciding whether or not a patient needs immediate laparotomy.
 - In children, after emptying the bladder with a Foley catheter, use a midline approach above or below the umbilicus. Instill 10 mL/kg of LR if the initial aspirate is not grossly bloody.
 - A positive DPL >100,000 RBCs may be due to a laceration of the liver or spleen but this would not be an indication for surgery. Greater than 80% of these patients will stop bleeding under observation without an operative intervention.

PELVIS

- Palpate the bony prominences of the pelvis for tenderness or instability.
- Examine the perineum for laceration, hematoma, or active bleeding.
- Blood loss from pelvic fractures can be critically significant and difficult to control, leading to a fatal hemorrhage. If there is major pelvic disruption, then the patient will need to be transferred to a trauma center. Consider binding pelvis if an open-book-type fracture.

PERINEUM/RECTUM

- The perineum should be examined for contusions, hematomas, lacerations, and urethral bleeding. If there is no blood and prostate is normal, then place a Foley catheter.
- Perform a rectal examination prior to placing a urinary catheter. Determine sphincter muscle tone, rectal integrity, prostatic position, presence of a pelvic fracture, and the presence of blood in the stool.
- For the female patient, a vaginal examination should also be considered in the secondary survey.

EXTREMITY EXAMINATION

- Look at all extremities looking for deformity, contusions, abrasions, intact sensation, penetrating injuries, pulses, and perfusion.
- The presence of a pulse does not exclude a proximal vascular injury or a compartment syndrome.
- Palpate long bones circumferentially assessing for tenderness, crepitation, or abnormal movement.
- Straighten severe angulations of the extremities if possible and apply splints and traction.
- Open fractures and wounds should be covered with sterile dressings. Inspect soft tissue injuries for foreign bodies, irrigate to minimize contamination, and debride devitalized tissues.

- Remember to check for fractures involving the bones of the hands, wrists, and feet since they are commonly missed until the patient regains consciousness.

BACK EXAMINATION

- Examine the back, particularly in cases of penetrating trauma, looking for hematomas, exit or entry wounds, or spine tenderness.
- With the neck immobilized log roll the patient for examination.

SKIN

- Examine for evidence of contusions, burns, penetration sites, petechiae, and signs of abuse.

NEUROLOGIC EXAMINATION

- Obtain an additional Glasgow Coma Score and perform an in-depth evaluation of motor, sensory, and cranial nerves. Check the fundi and look for rhinorrhea.
- Level of consciousness, pupillary examination, and sensorimotor examination, as quantified in the Glasgow Coma Scale, are invaluable in identifying a change in mental status. Presence of paresis or paralysis suggests a major neurologic injury. Conversely, lack of neurologic findings does not eliminate the possibility of a cervical cord injury, especially when the patient has a distracting injury and/or pain.

ADDITIONAL TREATMENT AND TESTS

- Provide tetanus prophylaxis (toxoid and possibly tetanus immune globulin).
- Consider the psychosocial aspects of traumatic injuries.
- Permit parents at the child's bedside as soon as the child's clinical status is stabilized.

BURNS

- Burns are the second most common cause of accidental death among children <4 years of age and may often be a significant part of the problem in a multiply injured patient (See Chapter 138 on BURNS).

IMAGING

- A child with major blunt trauma needs three basic radiographs immediately, the AP chest, AP pelvis, and C-spine films.

- Chest films are more sensitive than clinical examination in young children for detecting hemothorax and pneumothorax. Check for widening of the mediastinum and fractured ribs.
- Pelvic fractures are important clinical indicators, in that 80% of children with multiple fractures of the pelvis have concomitant abdominal or genitourinary injuries.
- During the secondary survey, thoracolumbar and extremity films can be completed as indicated.

CT SCAN OF HEAD

- Indications for CT of the brain include a Glasgow coma scale (GCS) <14, deteriorating neurologic examination, posttraumatic seizures, prolonged lethargy, prolonged vomiting, significant loss of consciousness, amnesia, and confounding medical problems, such as hemophilia.

CT SCAN OF ABDOMEN

- Indications for abdominal CT include (1) a hemodynamically stable victim of blunt trauma with clinical signs of intra-abdominal injury; (2) hematuria >20 RBCs per high-powered field or even minimal hematuria with a history of deceleration injury; (3) worrisome mechanism of trauma in the presence of neurologic compromise (Table 28-14).
- Positive findings on abdominal CT are significantly increased if three of the following are present: Gross hematuria, lap belt injury, assault, or abuse as a mechanism of trauma, positive abdominal findings such as tenderness, trauma score <12, and significant neurologic compromise (GCS <10).
- During the CT, the Foley catheter should be clamped to evaluate the bladder and the nasogastric tube should be pulled into the esophagus to avoid artifact.

TABLE 28-14 Dose of Contrast Media for Radiographic Studies

AGE (y)	DOSE
Intravenous: 60% hypaque	
0 to 9	1 mL/0.45 kg bolus
10 or more	50 mL, followed by infusion of 50 to 100 mL during scan
Oral: 1.5% hypaque (20 mL to 1 L of fluid given po or NG)	
0–2	100 mL
3–5	150–200 mL
6–9	200–250 mL
>9	300–1000 mL
Adult	1000 mL
Oral: Gastrografin, 20 mL/kg via NG tube 20 min prior to scan	

- A limitation of CT is the lack of sensitivity in diagnosing injuries to hollow viscous organs (bladder, intestinal perforations/rupture). If intraperitoneal fluid is found by CT scan with no apparent injury to the spleen or liver, consider injury to the bowel, bladder, or a vascular injury.
 - If injury to bowel or bladder is confirmed, laparotomy is indicated.

ULTRASONOGRAPHY

- Ultrasonography may be an alternative to CT scan in selected cases or when CT scan is not available. Ultrasound can diagnose injuries to the liver, spleen, and kidneys and can document intraperitoneal fluid.

UROLOGIC STUDIES

- If a patient has gross blood at the meatus, or the integrity of the urethra is in doubt because of the possible pelvic fracture, then a retrograde urethrogram should be performed.
 - The study is performed by instilling contrast through a Foley catheter that has been inserted into the distal urethra and partially inflated (0.5–1.0 mL of saline in the balloon). If the urethra is found not to be damaged, the catheter can be advanced to perform a cystogram.
- A 1-shot intravenous pyelogram is a very useful and reasonable test to perform in the ED to evaluate renovascular status if the patient is too unstable for CT scan. This will determine the absence or presence of blood supply to one or both kidneys and it may also show the function of the upper ureters.

INJURY SEVERITY MEASURES

THE TRAUMA SCORES

- The revised trauma score (Table 28-15) was originally developed for rapid assessment, triage, measuring progression of injury, predicting outcome, and assisting in quality assessment. It is useful in the overall management of trauma patient but is less sensitive for severe injury to a single organ system. It is straightforward to calculate for all trauma patients. It allows for standardization of triage protocols and for scientific comparisons between groups of patients and institutions.
- The pediatric trauma score (Table 28-16) was developed to reflect the unique injury pattern in children. It incorporates age into the score.

TABLE 28-15 Revised Trauma Score^a

REVISED TRAUMA SCORE	GLASGOW COMA SCORE	SYSTOLIC BLOOD PRESSURE (mm Hg)	RESPIRATORY RATE (breaths/min)
4	13–15	>99	10–20
3	9–12	76–89	>29
2	6–8	50–75	6–9
1	4–5	1–49	1–5
0	3	0	0

^aA score of 0–4 is given for each variable then added (range, 0–12). A score ≥ 11 indicates potentially important trauma.

TABLE 28-16 Pediatric Trauma Score^a

VARIABLES	+2	+1	–1
Airway	Normal	Maintainable	Unmaintainable
CNS	Awake	Obtunded/LOC	Coma/Decerebrate
Body weight	>20 kg	10–20 kg	<10 kg
Systolic BP	>90 mm Hg	50–90 mm Hg	<50 mm Hg
Open wound	None	Minor	Major
Skeletal injury	None	Closed fracture	Open/multiple fractures
Cutaneous	None	Minor	

^aA score of +2, +1, or –1 is given to each variable, and then added (range –6 to 12). A score ≥ 8 indicates potentially important trauma. LOC = loss of consciousness.

- Score >8: Associated with 100% survival
- Score <8: Should transfer patient to pediatric trauma center
- Score <0: 100% mortality
- The trauma score or pediatric trauma score can be used to triage patients and predict outcome but exercise caution if used to predict functional outcome.

DISPOSITION/TRANSFER

- Facilities that receive trauma victims should have the appropriate personnel and patient care resources committed at all times.
- Children, therefore, can and should receive appropriate care at all hospitals where EMS policies have established that the facility is capable of receiving patients with life-threatening conditions.
- In a hospital without pediatric surgical consultants, early transfers to a pediatric trauma center, adult trauma center, or pediatric intensive care unit should be considered.
- Establish contingency plans for transfers, prearranged agreements between institutions prospectively (Table 28-5).

- Children who appear to have clinical brain death should be considered for continued resuscitation since they may be candidates for organ donation and procurement.
- When dealing with a traumatized child, the physician must communicate openly and clearly with the family.
- Disposition and treatment decisions and progress reports need to be presented frequently, succinctly, and with sensitivity. Parents should be allowed to see the child and/or accompany him or her as soon as is practical.

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QUESTIONS

1. The most common cause of traumatic death in children 1–14 years of age is which of the following?
 - A. Falls
 - B. Drowning
 - C. Motor vehicle crashes
 - D. Burns
 - E. Abuse
2. Which of the following statements regarding pre-hospital care of injured children is correct?
 - A. Prehospital advanced airway management has not been shown to decrease trauma deaths
 - B. Prehospital success rate for endotracheal intubation is < 30%
 - C. It is reasonable for traumatized children to be transported immediately without vascular access
 - D. Intraosseous infusion is too complicated and time-consuming for use in the field
 - E. Urban systems may require more aggressive treatment in the field
3. The primary survey and initial resuscitation phases of management occur simultaneously, usually during the first 5–10 minutes of care and include which of the following?
 - A. Cross-table lateral of the cervical spine
 - B. FAST scan
 - C. Evaluation of pupillary size and reactivity
 - D. Ordering of blood for type and cross-match
 - E. Log-rolling of the patient for thorough examination
4. A 4-year-old child has been involved in a motor vehicle crash and arrives in the ED unresponsive with the only evidence of trauma being blunt head trauma. He is breathing spontaneously at a rate of 20 breaths/min, without evidence of airway obstruction. What is the most appropriate approach to the airway and breathing?
 - A. Ventilate by bag-valve mask as needed
 - B. Administer oxygen and allow for spontaneous respiration
 - C. No intervention is indicated
 - D. Intubate to prevent aspiration but allow spontaneous ventilation
 - E. Intubate and use controlled ventilation to achieve mild hyperventilation
5. Which of the following techniques is an appropriate option for an 8-year-old child in respiratory distress after a traumatic event?
 - A. Retrograde intubation
 - B. Needle cricothyrotomy with transtracheal jet ventilation
 - C. Surgical cricothyrotomy
 - D. Tracheostomy
 - E. Nasotracheal intubation
6. Unique characteristics of the pediatric cervical spine predispose children to which type of injury?
 - A. Spinal cord injury without radiographic abnormality
 - B. Hangman's fracture
 - C. Bilateral facet dislocation

- D. Lower > upper vertebral injuries
E. Burst fracture of the lower cervical vertebrae
7. A 10-year-old child has sustained multiple trauma but has arrived in the ED with normal vital signs. FAST scan shows evidence of intra-abdominal blood. What is the most appropriate management?
A. Immediate laparoscopy
B. Admit for serial examinations
C. CT scan
D. Repeat FAST scan
E. Diagnostic peritoneal lavage
8. Which of the following injuries is most likely to be missed by CT scan?
A. Pancreatic tear
B. Aortic disruption
C. Duodenal rupture
D. Splenic rupture
E. Liver laceration
9. A 16-year-old male victim of a motor vehicle crash is suspected of having a pelvic fracture and is found to have blood at the penile meatus. Which of the following studies is indicated?
A. One-shot IVP
B. Upright KUB
C. CT of the pelvis
D. Voiding cystourethrogram
E. Retrograde urethrogram
10. The pediatric trauma score was developed to reflect the unique injury pattern in children. In what parameter are the two scores significantly different?
A. CNS status
B. Airway and breathing
C. Blood pressure
D. Body weight
E. Parameters are the same but the weighting is different
- versus aeromedical transport. The prehospital success rate for endotracheal intubation varies from 48 to 89%. Intraosseous infusion should be used as a quick access for crystalloid infusion if attempts at intravenous cannulation are unsuccessful after 90 seconds. IO lines can be placed successfully in the field 80% of the time. Rural systems may require more aggressive initial treatment in the field because of transport times that are three to four times greater than in urban areas.
3. D. Ordering of blood for type and cross-match, as well as for other baseline parameters is appropriately done in the primary survey/resuscitation phase of care, when IV access is being established. Radiologic and ultrasound evaluations are part of the secondary survey. Assessing neurologic status in the primary survey deals primarily with responsiveness and does not include pupillary evaluation. Log-rolling for complete examination is also a key part of the secondary survey.
4. E. In the setting of head trauma, endotracheal intubation is used to protect the airway and to allow for controlled ventilation to achieve mild hyperventilation, which will reduce cerebral blood flow and protect against cerebral edema.
5. B. Needle cricothyrotomy with transtracheal jet ventilation is the preferred surgical method of choice to secure an emergency airway in children. Retrograde intubation and surgical cricothyrotomy are not indicated in children. Tracheostomy is time consuming and hazardous in the ED. Nasotracheal intubation should not be performed in a child younger than age 9.
6. A. Unique characteristics of the pediatric C-spine predispose to ligamentous disruption and dislocation injuries without radiologic evidence of bone injury.
7. C. Indications for abdominal CT scanning include a hemodynamically stable victim of blunt trauma with clinical signs of intra-abdominal injury. FAST scan can detect blood in the abdomen but cannot determine the source of that blood. CT scanning will, in most cases, define the source of the bleeding and help to delineate the best course of management, whether it be surgery or observation. Immediate laparoscopy would be indicated if the child were hemodynamically unstable. Admission for serial examinations may be appropriate once the source is defined. Repeat FAST scans may be used but will not by themselves define the best management. Diagnostic peritoneal lavage has a similar limitation in that it can detect blood but cannot define the source.

ANSWERS

1. C. Motor vehicle crashes account for as many as half of childhood trauma deaths and are the most common of causes of childhood death. Drowning, burns, and suffocation are other leading causes.
2. C. It is reasonable for traumatized children to be transported immediately without vascular access if a short transport time is expected. Vascular access can be attempted during transport in order to avoid prolonged scene time. It has been demonstrated that trauma deaths can be reduced by 25% when a system provides personnel trained to perform airway management and develops guidelines for ground

8. C. A limitation of CT scanning is the lack of sensitivity in diagnosing injuries to hollow viscous organs such as the bladder and intestine. CT is quite sensitive for injury to the aorta and to solid organs.
9. E. If a patient has gross blood at the meatus, or the integrity of the urethra is in doubt because of a pelvic fracture, then a retrograde urethrogram should be performed.
10. D. The pediatric trauma score was developed to reflect the unique injury pattern in children and incorporates body weight into the score, which the revised trauma score does not.

29 HEAD TRAUMA

Kimberly S. Quayle

INTRODUCTION

- More than 7000 children die each year as a result of traumatic brain injury, while another 60,000 are hospitalized and an additional 500,000 seek care in emergency departments. Boys are injured twice as commonly as girls.
- Among children who die from trauma, 90% have an associated brain injury.
- Pediatric brain injury leads to major morbidity from physical disability, seizures, and developmental delay.
- The most common cause of head injury in children is falls. Severe injuries are caused by motor vehicle collisions (with the child as occupant or pedestrian), bicycle collisions, and assaults, including child abuse.

PATHOPHYSIOLOGY

- Primary brain injury occurs as a result of direct mechanical damage inflicted during the traumatic event.
- Secondary injuries occur from hypoxia, ischemia, or increased intracranial pressure.
- Anatomic features, specific injuries, and intracranial pressure physiology are important components in the pathophysiology of pediatric brain injury.

ANATOMY

- Figure 29-1 shows the anatomy of structures overlying the brain with the illustration of possible injuries.

- Movement of the brain within the vault along the uneven base of the skull may injure brain tissue. The unyielding, mature skull can contribute to brain injury when brain edema or an expanding hematoma develops. Herniation across compartments can cause compression of vital structures, ischemia from vascular occlusion and infarction.
- In infants, the open sutures and thin calvarium produce a more flexible skull capable of absorbing greater impact. The disproportionately large size and weight of the head compared to the rest of the body of infants and young children contribute to an increased likelihood of head injury.
- Incomplete myelination of the infant brain contributes to greater plasticity and this flexibility permits more severe distortion between skull and dura and cerebral vessels and brain, increasing susceptibility to hemorrhage.

SPECIFIC INJURIES

- Scalp bleeding can lead to hemodynamically significant blood loss from relatively small lacerations, especially in infants and young children.
- A subgaleal hematoma is an extensive soft tissue swelling that occurs several hours or days after the injury and is commonly associated with a skull fracture.
- The presence of a skull fracture is associated with a higher likelihood of intracranial injury; however, the absence of a skull fracture does not exclude the presence of intracranial injury.
- “Growing fractures” may occur after a skull fracture in children younger than 2 years of age when associated with a dural tear. Rapid brain growth postinjury is associated with the development of a leptomeningeal cyst, which is an extrusion of cerebrospinal fluid or brain tissue through the dural defect.
- Basilar skull fractures typically occur at the petrous portion of the temporal bone and are associated with the clinical signs of hemotympanum, cerebrospinal fluid otorrhea, cerebrospinal fluid rhinorrhea, periorbital ecchymosis (raccoon eyes), or postauricular ecchymosis (Battle’s sign).
- Most epidural hematomas occur in combination with a temporal skull fracture and meningeal artery bleeding. Signs and symptoms include headache, vomiting, and altered mental status, which may progress to signs and symptoms of uncal herniation with pupillary changes and hemiparesis.
- Subdural hematomas usually result from tearing of the bridging veins and are often associated with more diffuse brain injury. They may progress more slowly

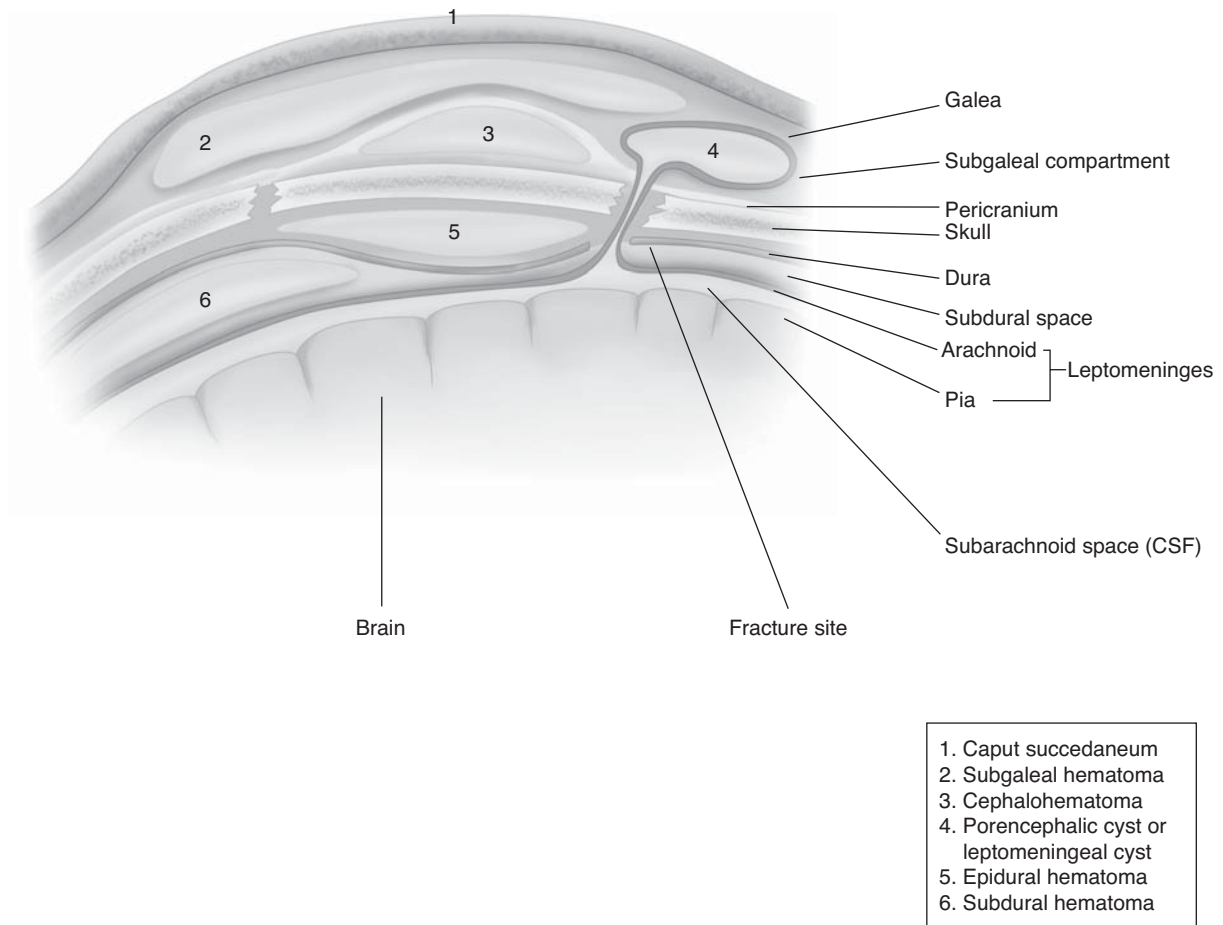


FIG. 29-1. Traumatic head injuries.

- | |
|---|
| 1. Caput succedaneum |
| 2. Subgaleal hematoma |
| 3. Cephalohematoma |
| 4. Porencephalic cyst or
leptomeningeal cyst |
| 5. Epidural hematoma |
| 6. Subdural hematoma |

- than epidural bleeds, with symptoms commonly including irritability, vomiting, and alterations in mental status.
- Parenchymal contusions are caused as the brain moves across the bony irregularities of skull or by shearing injury of the brain. Signs and symptoms may include decreased level of consciousness, focal neurologic findings, and seizures.
- Penetrating injuries result from sharp-object penetration or gunshot wounds. Extensive brain injury is common and severity depends on the path of the object and location and degree of associated hemorrhage.
- A concussion is defined as a “trauma-induced alteration in mental status that may or may not involve a loss of consciousness.” Additional symptoms may include vomiting, headache, dizziness, visual changes, as well as cognitive impairments and abnormal behavior. Most symptoms resolve after 48 hours; however, some symptoms may linger for weeks to months in a “postconcussive” syndrome.

- Diffuse brain swelling occurs more often in children than in adults. The swelling usually results from a shearing or acceleration–deceleration injury. Prolonged coma or death may occur.
- Nonaccidental trauma in infants and young children may result in the constellation of subdural hematoma, subarachnoid hemorrhage, and localized or diffuse brain edema. Retinal hemorrhages, rib fractures, long bone fractures, and external signs of injury may also be present. Common symptoms of nonaccidental traumatic brain injury in infants may include lethargy, vomiting, irritability, seizures, apnea, and alteration in consciousness.

INTRACRANIAL PRESSURE AND CEREBRAL EDEMA

- As intracranial pressure rises to abnormal levels (>15–20 mm Hg), cerebral perfusion becomes impaired and irreversible ischemic damage to the brain ensues.

- An intracranial mass or hematoma will occupy the fixed intracranial space, compress the normal brain tissue, and reduce blood flow.
- Cytotoxic cerebral edema occurs with fluid accumulation within damaged brain and glial cells. Interstitial cerebral edema results from decreased absorption of fluid following brain trauma. Vasogenic cerebral edema occurs as the endothelial cell barrier is compromised and leakage of fluid into the perivascular brain tissue occurs.
- As brain and blood volumes increase, the ventricular spaces become compressed until redistribution is not possible.
- If the cerebrospinal fluid pathways are compressed by edematous tissue, cerebrospinal fluid outflow ceases, and ventricular dilation and hydrocephalus can occur.
- Cerebral blood flow is often increased in head-injured children, possibly due to a loss of normal autoregulatory mechanisms leading to increased risk for brain swelling. Hypoxia and hypotension of the injured patient may also contribute to diffuse brain edema.
- Causes of diffuse brain swelling are likely multifactorial, including hyperemia, excitotoxic neurotransmitters, enhanced inflammatory response, and increased blood–brain permeability.
- Past medical history should include prior history of seizures, neurologic abnormalities, bleeding disorders, and immunization status.
- Child abuse should be suspected for a witnessed report of abuse, a history insufficient to explain the injuries present, a changing or inconsistent history or a developmentally incompatible history.
- Physical evaluation begins with assessment for airway obstruction. Establish an airway by positioning, suctioning, placing an oral airway or intubation. Maintain cervical spine control in children with significant head injury until cervical spine injury is excluded.
- Assess ventilation by observing chest expansion, auscultate breath sounds, and assess for cyanosis or respiratory distress. Hypoventilation is treated with 100% oxygen, bag-valve-mask ventilation, and subsequent intubation of the trachea with rapid sequence technique. End-tidal CO₂ should be monitored in any significant head injury.
- The use of ketamine and succinylcholine is controversial because of concerns for possible increased intracranial pressure associated with their use.
- RSI for intubation is contraindicated in patients with major facial or laryngeal trauma or distorted facial and airway anatomy.
- Evaluate the circulation by checking heart rate, peripheral pulses, and perfusion. Control any life-threatening hemorrhage and maintain blood pressure so that there is adequate cerebral perfusion. Treat hypotension initially with isotonic fluid boluses.
- Hypovolemic shock is rare after an isolated head injury, so other sources for hypovolemia must be identified.
- After the rapid ABC assessment, ascertain the level of consciousness and categorize the patient as alert, responsive to verbal stimulus, responsive to painful stimulus or unresponsive (AVPU). Evaluate the pupillary response.
- Examine the scalp and palpate for depressions. Evaluate the fontanel in the infant. Look for signs of basilar skull fracture. Evaluate extraocular movements, muscle tone, spontaneous movements, and posture. An older child should move the extremities in response to the examiner's request. Palpate the neck for tenderness or deformities.
- Note any stereotyped posturing. Decorticate posturing signifies damage to the cerebral cortex, white matter or basal ganglia. Decerebrate posturing suggests damage to the midbrain.
- In adults and older children, the Glasgow Coma Scale (GCS) evaluates for eye opening, best motor response, and best verbal response (Table 29-1). Use of this scale in infants and young children is limited

HERNIATION SYNDROMES

- Diffusely or focally increased intracranial pressure may produce herniation.
- Cingulate herniation occurs as one cerebral hemisphere is displaced underneath the falx cerebri to the opposite side.
- A transtentorial or uncal herniation occurs when a mass lesion or hematoma forces the ipsilateral uncus of the temporal lobe through the space between the cerebral peduncle and the tentorium. This causes an ipsilateral dilated nonreactive pupil, and a contralateral hemiparesis. As the brainstem becomes depressed, consciousness wanes progressing to apnea and death.
- Herniation of the cerebellar tonsils downward through the foramen magnum causes medullary compression resulting in bradycardia, respiratory arrest, and death.

ASSESSMENT

- Assessment begins with a detailed history of the traumatic event, such as the mechanism of injury and the time and location of injury. Note the signs and symptoms, including loss of consciousness, seizures, vomiting, headache, visual changes, altered mental status, weakness, and amnesia.

TABLE 29-1 Glasgow Coma Scale^a

Eye opening (E)	Spontaneous	4
	To speech/voice	3
	To pain	2
	No response	1
Motor response (M)	Obeys commands	6
	Localizes pain	5
	Withdraws to pain	4
	Abnormal flexion-decorticate	3
	Abnormal extension-decerebrate	2
Verbal response (V)	No response	1
	Oriented	5
	Confused/disoriented	4
	Inappropriate words	3
	Incomprehensible sounds	2
	No response	1

^aE + M + V = coma score (range: 3–15).

due to this age group's underdeveloped verbal skills, so modifications have been made for the preverbal child (Table 29-2).

- The neurologic status of a head-injured child must be reassessed regularly, particularly with regard to level of consciousness and vital signs. The frequency of reassessment should be dictated by the condition of the child.

DIAGNOSTIC STUDIES

- In children with serious injuries, complete blood count, type and cross-match, electrolytes, and coagulation studies should be done. Arterial blood gases, toxicology screens, and ethanol levels are obtained as indicated.

TABLE 29-2 Glasgow Coma Scale^a Modified for the Preverbal Child

Eye opening (E)	Spontaneous	4
	To speech/voice	3
	To pain	2
	No response	1
Motor response (M)	Spontaneous movements	6
	Withdraws to touch	5
	Withdraws to pain	4
	Abnormal flexion	3
	Extensor response	2
Verbal response (V)	None	1
	Coos, babbles	5
	Irritable, cries	4
	Cries to pain	3
	Moans to pain	2
	None	1

^aE + M + V = coma score (range: 3–15).

With permission from Reilly PL, Simpson DA, Sprod R, et al. Assessing the conscious level in infants and young children: a pediatric version of the Glasgow Coma Scale. *Childs Nerv Syst.* 1988;4:30.

- Cervical spine films should be obtained in alert patients with neck pain or neurologic deficits and in all unconscious patients.
- Computed tomography (CT) of the head is the diagnostic method of choice for identification of intracranial pathology in patients with acute head trauma.
- Skull radiographs are not routinely recommended; however, they may be useful in certain clinical situations, such as screening in young infants with scalp hematomas, in cases of suspected nonaccidental trauma or when CT is not readily available.
- Children with severe injuries including those with altered mental status, focal neurologic deficits or penetrating injuries should undergo emergent head CT and prompt neurosurgical consultation. Children with a known skull fracture or signs of a basilar or depressed skull fracture should also undergo head CT.
- Growing concerns regarding radiation exposure to the developing brain have prompted the development of guidelines for the management of children with minor head injuries.
- A recently published guideline discusses the consideration of head CT versus observation with the presence of scalp hematomas, who are asymptomatic, have had a low mechanism of trauma for children younger than two years.
- The guideline discusses the consideration of head CT versus observation in children who had minimal or no loss of consciousness and are alert, have normal mental status and normal neurologic exam in children 2 years and older.
- Decision to obtain the CT or observe may be influenced by physician experience, multiple versus isolated findings, worsening symptoms, or signs after emergency department observation, age <3 months and parental preference.

TREATMENT FOR SEVERE HEAD INJURY

- The goal of management of head injury in children is to prevent secondary injury to the brain; therefore, prevention of hypoxia, ischemia, and increased intracranial pressure is essential.
- Prompt neurosurgical intervention is necessary in the majority of seriously head-injured or multisystem-injured children.
- Endotracheal intubation and controlled ventilation are almost always required for patients with severe head injury. Intubate any child with a GCS of 8 or less.
- Avoid hypoxemia and hypercarbia.
- Modest hyperventilation (PCO₂ 35–40 Torr) is recommended for managing increased intracranial pressure.

More aggressive hyperventilation may be needed to treat acute deterioration with suspected herniation, but other treatment may be more appropriate. Monitor end-tidal CO₂ continuously in these patients.

- Preserve brain perfusion by maintaining normal intracranial pressure and normal mean arterial pressure. Hypotension should be treated with isotonic fluid boluses and inotropic medications as needed to maintain an adequate mean arterial blood pressure and cerebral perfusion pressure.
- Mannitol is often used to maintain optimal intracranial pressure by reducing intravascular volume. Fluid restriction may be used in conjunction; however, cerebral perfusion pressure must be maintained.
- Hypertonic saline (3%) has been studied in children with traumatic brain injury, but is not routinely used in the emergency setting.
- Syndromes of inappropriate antidiuretic hormone secretion or diabetes insipidus may occur; therefore, fluid balance and electrolyte status must be followed closely.
- Most children with serious injuries are treated with fosphenytoin either to treat active seizures or for prophylaxis.
- Seriously brain-injured children must be monitored in an intensive care setting. Cardiopulmonary monitors, noninvasive blood pressure monitors or indwelling arterial catheters, and urinary catheters are commonly used.
- Intracranial pressure monitors may be used to detect acute changes in intracranial pressure, to limit indiscriminate therapies, to control intracranial pressure, and to reduce intracranial pressure directly by cerebrospinal fluid drainage.
- The patient should be positioned with a 30° elevation of the head. Sedation and analgesia should be used as needed to limit wide fluctuations in intracranial pressure.
- Coordination of the care for these children should involve neurosurgical, pediatric, and critical care physicians, most often in a tertiary care pediatric center.

DISPOSITION FOR MINOR HEAD INJURY

- Patients who do not meet criteria for imaging may be observed at home by a reliable caretaker with careful instructions to return the child to medical attention if symptoms develop, worsen, or persist.
- Children who have normal head CT scans and normal mental status and neurologic examinations may also be observed at home, as the development of delayed intracranial injuries in these patients is rare.

- Children with isolated nondepressed skull fractures without intracranial injury may also be observed at home.
- If children have persistent alteration in mental status despite a normal head CT, they should be admitted for observation with serial neurologic examinations.
- Children with protracted vomiting may need intravenous hydration and admission.
- Children with suspected child abuse should also be admitted for observation as well as evaluation of the social situation.
- If a responsible caretaker cannot be identified, hospital admission for observation for the first 24 hours is warranted.
- Young athletes with concussions should delay return to play for at least 1 week after the injury if the child had symptoms of a concussion. With a history of any loss of consciousness, return to play should be 2–4 weeks after the injury.
- Children with symptomatic head injury or isolated nondepressed skull fractures should be seen by their primary physician 24 hours after the emergency department visit.

PROGNOSIS

- Over the past 20 years, morbidity and mortality secondary to pediatric traumatic brain injury has dramatically declined.
- Infants and children have long been thought to have better functional outcomes following a severe brain injury.
- New data suggest that children younger than 4 years of age have the poorest outcomes, possibly related to the high incidence of nonaccidental trauma as cause for severe injury in this age group.
- Early identification of neurobehavioral deficits is an important part of follow-up in children with significant head injury.

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QUESTIONS

1. A 2-month-old infant presents with apnea and alteration in mental status. She has multiple contusions on her scalp, trunk, and extremities without a history of injury. You do not palpate any bony crepitus. What is the most likely finding on her head CT?
 - A. Epidural hematoma
 - B. Depressed skull fracture
 - C. Epidural hematoma with associated uncal herniation.
 - D. Subdural hematoma with diffuse brain swelling
 - E. Basilar skull fracture
2. A 10-month-old male presents with a 3 × 3 cm forehead hematoma after hitting his head on a table while walking 3 hours ago. He did not lose consciousness and he has been acting normally according to his parents. His physical examination is otherwise normal. What is the most appropriate management for this child?
 - A. Observation at home if caretakers are reliable
 - B. Four-hour observation in the emergency department
 - C. Overnight hospital admission for observation
 - D. Evaluation by head CT
 - E. Head CT followed by hospital admission for observation
3. A 7-year-old male presents after he was ejected during a high-speed motor vehicle crash. He moans and opens his eyes briefly to painful stimuli and he exhibits decorticate posturing. What is the most appropriate first step in his management?
 - A. IV Mannitol
 - B. Transfer to a level 3 Pediatric Trauma Center
 - C. Intubation with cervical spine control
 - D. Head CT
 - E. IV 3% saline
4. A 13-year-old male presents after a head injury during a football game. He had a helmet-to-helmet hit with another player, and was unconscious for approximately 2 minutes after the injury. He has amnesia for the event, but has otherwise returned to baseline. His parents ask when he can return to playing football. What is your answer?
 - A. Three days if cleared by his pediatrician
 - B. One week if cleared by his coach
 - C. Two weeks if cleared by his pediatrician
 - D. Eight weeks if he has no other symptoms
 - E. He should not play for the remainder of the season
5. A 10-year-old girl falls from a tree and develops a severe headache, vomiting, and increasing lethargy. A head CT shows a left-sided epidural hematoma and a skull fracture. Where is the most likely location for the skull fracture?
 - A. Occipital skull fracture
 - B. Left-sided temporal skull fracture
 - C. Right-sided frontal fracture
 - D. Left-sided frontal fracture
 - E. Basilar skull fracture
6. A 15-year-old male suffers a severe head injury. He is hypoventilating and is hypotensive. His GCS is 8. You are about to intubate him. What drug should you use for induction?
 - A. Ketamine
 - B. Thiopental
 - C. Etomidate
 - D. Propofol
 - E. None of the above
7. What increases the infant brain's susceptibility to hemorrhage?
 - A. Thinner and more flexible skull
 - B. Disproportionate size of the head
 - C. Open sutures
 - D. Immature blood vessels
 - E. Incomplete myelination of the brain
8. A 6-month-old infant suffers a severe head injury from a motor vehicle crash. She is intubated and ventilated for transport to a pediatric trauma center. What PCO₂ should the transport team target for this patient?
 - A. 35 Torr
 - B. 50 Torr
 - C. 20 Torr
 - D. 25 Torr
 - E. 45 Torr
9. A right-sided uncal herniation will cause which of the following?
 - A. Left nonreactive pupil with left hemiparesis
 - B. Right nonreactive pupil with left hemiparesis

- C. Right nonreactive pupil with right hemiparesis
 D. Left nonreactive pupil with right hemiparesis
 E. Bilateral nonreactive pupils with right hemiparesis
10. A 12-month-old infant sees her pediatrician two months after suffering a widely diastatic parietal skull fracture without underlying brain injury. What complication might her pediatrician find on her examination?
 A. Leptomenigeal cyst
 B. Microcephaly
 C. Macrocephaly
 D. Subgaleal hematoma
 E. Battle's sign

ANSWERS

- D. Subdural hematomas and diffuse brain swelling are common injuries associated with nonaccidental trauma. Epidural hematomas are less common and usually due to more focal blow to the head or from a fall.
- A. An infant with an isolated frontal scalp hematoma can be safely observed at home. If the child had mild symptoms such as vomited twice or was sleepy, observation for 4 hours may be reasonable. CT is usually not needed for frontal hematomas without other symptoms.
- C. For a child with a severe head injury and Glasgow Coma Score less than 8, intubation with cervical spine control is the first step in management. He will likely need the CT scan and transfer to appropriate level of care after initial airway management and stabilization. 3% Saline has not been proven of benefit at this time.
- C. A child with a loss of consciousness should not return to sports play for 2–4 weeks and should be cleared by a physician before starting practice. The child must be symptom free. The guidelines for return to play continue to evolve and vary somewhat between the organizations. If the child has had a severe concussion or a second concussion, they will most likely have to avoid practice and games for the rest of the season.
- B. An epidural hematoma is most commonly associated with a temporal skull fracture. Occipital fracture with epidural hematoma is rarer and is usually of venous origin. The other fractures are only rarely associated with an epidural hematoma.
- C. Etomidate has a lower likelihood of causing hypotension. Thiopental is neuroprotective but can cause hypotension as can propofol. Ketamine remains controversial for use in head trauma patients. With a GCS of 8, the patient is still aware of his environment and should receive an induction agent.
- E. Incomplete myelination of the brain leads to increased plasticity of the brain, which permits more distortion between skull and dura and cerebral vessels and brain. While the other answers are true about an infant, they are not associated with the susceptibility of hemorrhage.
- A. Modest hyperventilation to 35–40 Torr is recommended for managing increased intracranial pressure with severe head injuries. The extremes of hyperventilation and hypoventilation can worsen brain injury.
- B. An uncal herniation will cause an ipsilateral nonreactive pupil and contralateral hemiparesis. Uncal herniation has ipsilateral eye findings and contralateral motor findings.
- A. An infant with a skull fracture may develop a “growing skull fracture” or leptomenigeal cyst when the skull fracture is associated with a dural tear and rapid brain growth postinjury.

30 PEDIATRIC CERVICAL SPINE INJURY

Julie C. Leonard
Jeffrey R. Leonard

HIGH-YIELD FACTS

- Cervical spine injury should be suspected in any child who has suffered traumatic respiratory arrest and rapid sequence orotracheal intubation should be performed with in-line cervical spine stabilization.
- Standard radiographic screening for children consists of the AP, lateral, and open-mouth views. The open-mouth view can be omitted in young, uncooperative children.
- CT scan is considered the diagnostic imaging of choice in children with altered mental status, focal neurological findings, or inadequate or abnormal plain three-view series.
- Because of differences in the anatomy and physiology, children sustain proportionally more upper cervical spine fractures and spinal cord injuries as compared to adults.
- Spine immobilization is indicated when cervical spine injury is suspected, but there are complications

to immobilization and it should be discontinued as soon as possible during trauma evaluation.

EPIDEMIOLOGY

- Cervical spine injury affects less than 1% of children undergoing emergency department trauma evaluation.
- The estimated mortality associated with cervical spine injury in children is 17%; however, this rate may be as high as 60% in children ≤ 8 years.
- This increased risk of mortality is likely associated with proportionately higher rate of upper cervical spine injury in young children.
- Motor vehicle collisions are the most common cause of cervical spine injuries in children.
- Neonates may suffer cervical spine injuries from birth trauma, particularly in the case of breech or forceps deliveries.
- Sports-related injuries, pedestrians hit by motor vehicles, and falls are common mechanisms of cervical spine injury in older children and teenagers, while gunshot wounds and stabbings are causes of cervical spine injuries in the late teenage years.

ANATOMY AND PHYSIOLOGY

- Recognition of the developmental anatomy of the pediatric cervical spine is critical in differentiating fracture from normal age-appropriate anatomy, as they may appear nearly identical radiographically.
- The atlas (C1) has three primary ossification centers: one anterior arch and two neural arches. By age 3, the neural arches are typically fused to form the solid posterior ring of C1. The neurocentral synchondrosis fuses by age 7.
- Four identifiable ossification centers are present in the developing axis (C2). The neural arches of C2 fuse posteriorly by age 3. The body of C2 fuses with the neural arches and the dens between ages 3 and 6. However, the subdental synchondrosis may be seen until ages 10–11.
- Each level of the subaxial cervical spine (C3–C7) follows the same developmental pattern involving three primary ossification centers: a centrum for the body and two neural arches. The neural arches fuse by age 3, whereas the body fuses with the neural arches by ages 3–6. Secondary ossification centers in the transverse and spinous processes are present by puberty and fuse completely by the third decade.
- Relative to adults, the cervical spine in children is hypermobile due to a relatively large head size, immature neck musculature, underdeveloped ligaments, incompletely ossified bone, anterior wedging

of vertebral bodies, absent uncinate processes, and shallow, horizontally oriented facets.

- The fulcrum of motion in the pediatric cervical spine is at C2–C3 rendering the upper cervical spine more prone to injury. By age 14, the fulcrum migrates to C5–C6 making the biomechanics of the cervical spine and the injuries sustained similar to those observed in adults.

EVALUATION AND MANAGEMENT

- All trauma evaluations begin with attention to the ABCs: airway, breathing, and circulation.
- Airway obstruction is common in severely injured trauma patients and the emergency practitioner must be cognizant of the potential for a cervical spine injury, which could be worsened by excessive motion of the spine. The cervical spine should be stabilized while performing the jaw-thrust maneuver.
- Spine-injured patients may become hypopneic due to diminished diaphragmatic activity or intercostal muscle paralysis; when hypoventilation is suspected, humidified oxygen should be provided and ventilation assisted as needed.
- Manual in-line cervical stabilization, rapid sequence induction, and oral endotracheal intubation are the preferred techniques to achieve airway stabilization in children with suspected cervical spine injury (Fig. 30-1). Intubation should not be delayed for radiographic clearance of the cervical spine.
- Hypotension in the traumatically injured child may be secondary to spinal shock. The pulse, which is rapid in hypovolemic shock, is slow in spinal shock. In addition to volume repletion, vasopressors, such as dopamine, may be necessary.
- Patients in spinal shock are more sensitive to temperature variations and require warming or cooling if subjected to extreme environmental temperatures.
- Protect areas of the body that may have lost sensation from hard, protruding objects, as they may cause skin necrosis, especially on long transports.
- Paresthesia, a pins and needles sensation or numbness or burning, may be an indicator of spine injury.
- Lhermitte's sign is a transient shock-like or electrical sensation transmitted down the spine during neck flexion and/or rotation and suggests a nerve root injury.
- Horner's syndrome (ptosis and a miotic pupil) suggests a cervical cord injury.
- Paresis or paralysis of the arms or legs should always suggest spine injury.
- Priapism is only present in approximately 3–5% of spine-injured patients, but indicates that the sympathetic nervous system is involved.

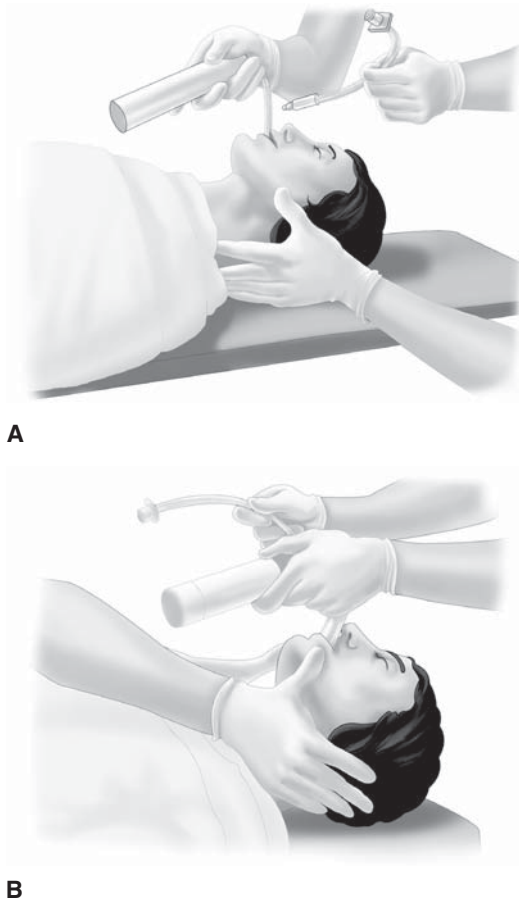


FIG. 30-1. In-line stabilization for endotracheal intubation: (A) above and (B) below.

- Absence of the bulbocavernosus reflex indicates a complete spinal cord injury. To elicit the bulbocavernosus reflex, a finger is inserted into the rectum, and then the glans of the penis or the head of the clitoris is squeezed. A normal response is a reflex contraction of the anal sphincter.
- There are also characteristic cord syndromes (Table 30-1).
- A head tilt may be associated with a rotary subluxation of C1 on C2 or a high cervical injury.
- The prayer position (arms folded across the chest) may signify a fracture in the C4–C6 area.
- Injuries at C5: patients can flex at the elbows, but are unable to extend them.
- Injuries at C6–C7: patients can flex and extend at the elbows.
- Injuries at the T1 level allow finger and wrist flexion.
- If a child may be at risk for cervical spine injury, take steps to maintain neutral cervical spine positioning. In children younger than 8 years, supine positioning

TABLE 30-1 Syndromes Associated with Spinal Cord Injury

Spinal shock <ul style="list-style-type: none"> • Flaccid below level of lesion • Absent reflexes • Decreased sympathetic tone • Autonomic dysfunction (including hypotension) • Sensation may not be preserved; if absent = total cord transection (poor prognosis) 	Brown-S' equard syndrome <ul style="list-style-type: none"> • Hemisection • Ipsilateral loss of <ul style="list-style-type: none"> ◦ Motor function ◦ Proprioception • Contralateral loss of sensation: <ul style="list-style-type: none"> ◦ Pain ◦ Temperature
Central cord syndrome <ul style="list-style-type: none"> • Diminished or absent upper extremity function • Preserved lower extremity function • Associated with extension injuries 	Anterior cord syndrome <ul style="list-style-type: none"> • Complete motor paralysis • Loss of pain and temperature sensation • Preservation of position and vibration sense • Associated with severe flexion injuries

without shoulder padding results in cervical kyphosis due to a relatively large head (Fig. 30-2). In adults, however, supine positioning causes relative cervical lordosis. Thus, as a child grows, padding may be required in either the shoulder or occipital regions to provide neutral positioning.

- Clinical criteria for clearing the cervical spine after blunt trauma have been established in adults. Until further evidence for clinical risk stratification in pediatric cervical spine injury emerges, it is reasonable to rely on the National Emergency X-ray Utilization Study screening criteria (posterior midline cervical tenderness, altered alertness, distracting injury, intoxication, and focal neurological findings) coupled

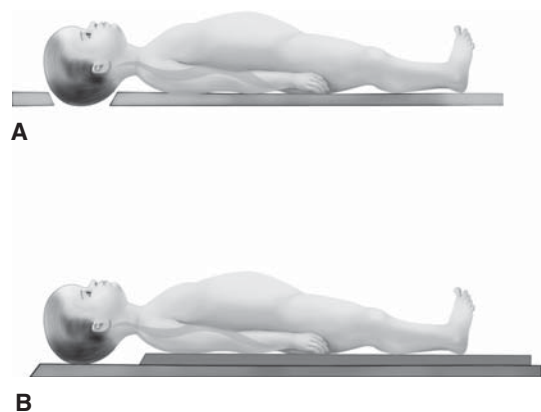


FIG. 30-2. Backboard modifications for children. (A) Young child on a modified backboard that has a cutout to recess the occiput, obtaining a safe supine cervical positioning. (B) Young child on a modified backboard that has a double-mattress pad to raise the chest, obtaining a safe supine cervical positioning.

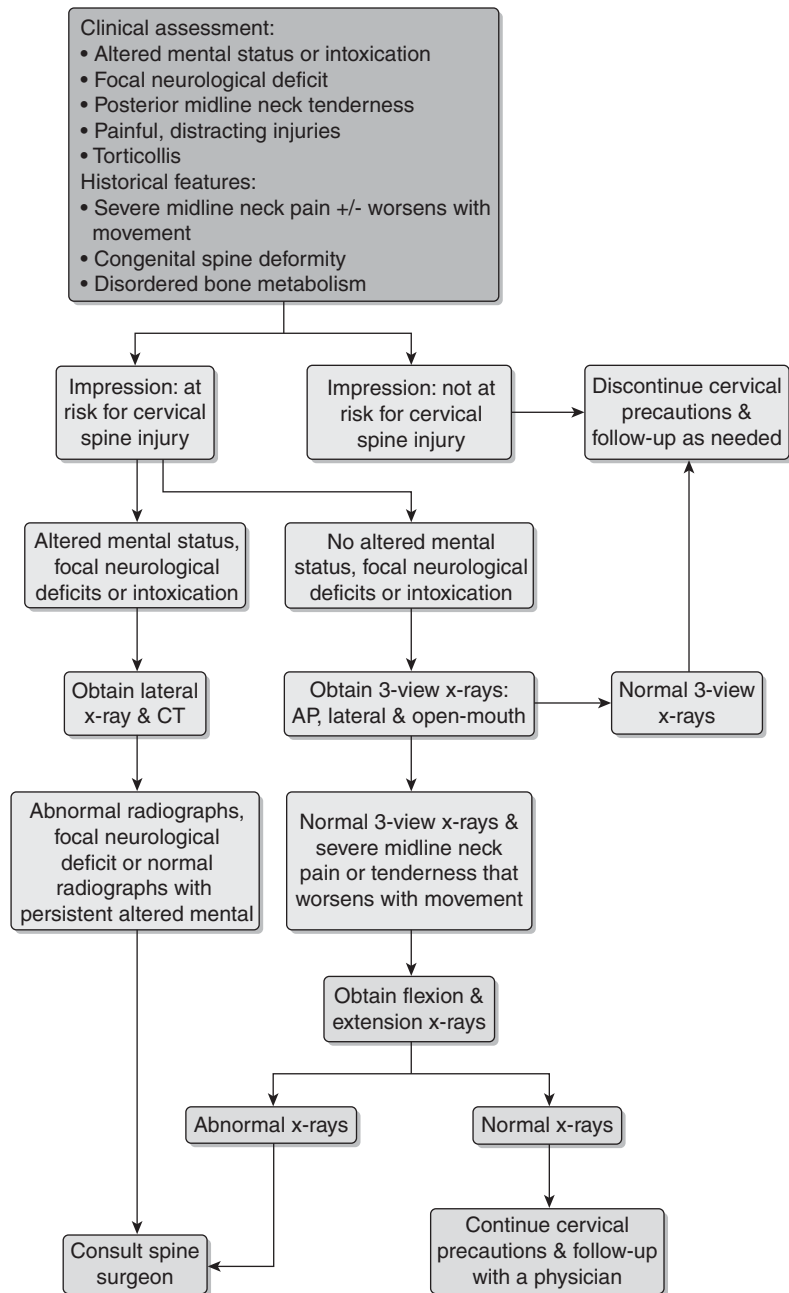


FIG. 30-3. Algorithm for cervical spine clearance in blunt trauma injury.

with the complaint of severe neck pain, torticollis and difficulty or severe neck pain with neck movement, congenital spine deformity or disordered bone metabolism to determine risk in children. Figure 30-3 is a suggested algorithm for cervical spine clearance in children following blunt trauma.

- All trauma victims should be unstrapped and removed from the rigid long board, which is used for extrication and patient transfers in the out-of-hospital setting. This aspect of full spine immobilization is known to

be associated with adverse effects. The cervical collar can be discontinued once it has been determined either clinically or radiographically that the patient is free of cervical spine injury.

- High-dose methylprednisolone (30 mg/kg) followed by 5.4 mg/kg/h for 23 hours is considered an acceptable option in the treatment of spinal cord injuries in children. However, there is controversy and the clinician should consult with their trauma and neurosurgical colleagues.

TABLE 30-2 Criteria for Clearing the Lateral Spine

- All seven vertebral bodies are seen clearly, including the C7-T1 junction
- The posterior cervical line is properly aligned as are the four lordotic curves, which are the anterior longitudinal ligament line, the spinolaminar line, and the lips of the spinous processes
- The predental space is 4 to 5 mm
- All vertebrae are free of fractures and changes in density, which would suggest compression fractures or metastatic lesions
- The intervertebral and interspinous spaces are not more than 11 degrees at a single interspace.
- There is no fanning of spinous processes, which would suggest posterior ligament disruption
- The prevertebral soft tissue distance is less than 7 mm at C2 and less than 5 mm at C3–C4. Note that in children <2 y of age, the space may appear widened if it is not an inspiratory film
- No dislocation at the atlantooccipital region

ANALYSIS OF RADIOGRAPHS

- The standard screening series cross-table lateral view (CTLV), anteroposterior view (AP), and open-mouth view (OM) should be used to screen alert and stable children for c-spine injury
- The steps in evaluating the CTLV are presented in Table 30-2.
- On the AP, symmetry of longitudinal alignment of vertebral bodies, facets, pillars, and spinous processes is assessed and evidence of linear or compression fractures as well as more subtle indicators such as stepoffs, subluxations, and malalignments are sought on the CTLV.
- On the OM, in addition to fractures of the occipital condyles, C1 and C2, the alignment of the atlantooccipital and atlantoaxial joints, alignment of the margins of the lateral arches of C1 with C2, and the position of the odontoid between the lateral arches of C1 are evaluated.
- The Waters view can be substituted for the OM in younger children. This view allows visualization of the odontoid projected through the foramen magnum.
- The swimmer's view to delineate the lower cervical spine, particularly the junction of C7 and T1.
- Flexion and extension views are used to evaluate the stability of the ligamentous structures of the c-spine.
- Supine oblique views allow better visualization of the pedicles and the neural foramina.
- In a child with altered mental status, focal neurological findings or inadequate or abnormal plain three-view series, thin-section CT scan is considered the diagnostic imaging of choice.
- MRI is the modality of choice for assessing the supportive soft tissues of the spine and the spinal cord itself. Additionally, MRI can be used to complete cervical spine injury clearance in the hemodynamically stable child with persistent altered mental status.

INJURY PATTERNS

ATLANTOCCIPITAL DISLOCATION

- Involve the disarticulation of cervical spine at the craniocervical and are severe and frequently fatal injuries.
- The incidence is more than double of that observed in adults (15% vs. 6%). which is thought to be related to anatomic differences.
- Mechanisms of injury include motor vehicle collisions or other high-impact trauma and results from excessive motion of the head relative to the upper cervical spine.

ATLAS FRACTURES

JEFFERSON FRACTURE

- A burst fracture of the atlas resulting from axial loading.
- True Jefferson fractures have four separate fracture lines involving both the anterior and posterior rings of C1 bilaterally.
- “Jefferson fracture” is also loosely applied to two- or three-point fractures of the atlas.
- Neurological injury is uncommon with Jefferson fractures because of the large canal diameter at this level and the propensity for burst fragments to project outward.
- Atlas fractures are stable unless there are severe compressive forces causing a combined C1–C2 fracture complex or disruption of the transverse ligament.
- C1 injuries are initially evaluated with plain cervical spine x-rays. The Atlanto-dens interval, noted on the CTLV, is used to assess the transverse ligament with values in excess of 5 mm indicating disruption. The OM can also be used to assess transverse ligament stability. According to the Rule of Spence, a lateral mass overhang of C1 over C2 greater than 6.9 mm indicates transverse ligament disruption.

ATLANTOAXIAL INJURIES

- The atlantoaxial articulation provides rotational movement of the cervical spine. Because of its structural design, the atlantoaxial articulation is predisposed to a subset of traumatic injury patterns.
 - Translational atlantoaxial subluxation (AAS) and dislocation are rare and highly morbid injuries. They are the result of high-energy mechanisms, such as high-speed motor vehicle–pedestrian collisions. Survivors present with severe head injury, while others present with the complaint of neck

pain or subtle signs of myelopathy or C2 sensory changes. The most common form of AAS is anterior translation of C1 on C2 and rupture of the transverse ligament of the atlas. Severe hyperextension injuries with fracture of the anterior arch of C1 or the odontoid process define posterior translation.

- Atlantoaxial rotatory subluxation (AARS) is common in children who are predisposed due to inherent ligamentous laxity. AARS may occur in the context of severe traumatic injury; however, more frequently it is the result of minor trauma and can occur spontaneously in the setting of pharyngeal, upper respiratory tract or cervical infection (Grisel's syndrome) and iatrogenically during positioning for head and neck surgery. Patients with AARS are usually neurologically intact, but have a painful "cock robin" torticollis characterized by chin rotation to the contralateral side and flexion of the neck. Atlantoaxial rotatory fixation (AARF) results from C1–C2 facet dislocation and trapping of robust synovium in the articular surfaces. Severe neck pain and muscle spasms may also prevent reduction of this position. While plain x-rays may be suggestive of the injury, if the diagnosis of AARS/ AARF is suspected, dynamic CT scan will confirm the diagnosis and determine whether the AARS/AARF results from a true bony lock or from soft tissue blockage of rotation.

AXIS FRACTURES

- In children <7 years of age, the weakest point in the C2 body–dens complex is the cartilaginous subdental epiphysis. While uncommon, significant displacement of the fractured dens can result in neurological deficit. Without displacement of the fracture, it may be difficult to distinguish injury from normal anatomy; however, anterior angulation of the dens is common and may aid in identifying epiphysiolysis.
- True fractures of the odontoid process are seen in older children and adolescents. Type I odontoid fractures, which involve the superior portion of the dens, are rare and caused by avulsion of the alar ligament usually in the setting of unstable craniocervical junction injuries. Type II odontoid fractures involve the base of the neck of the dens and are considered unstable. Type III odontoid fractures extend into the body of C2 such that the fracture fragment incorporates the entire odontoid process and a portion of the body of C2.
- Bilateral pars interarticularis fractures of C2 or the Hangman's fracture result from hyperextension with axial loading. The Hangman's fracture may also result in anterior subluxation of C2 on C3. A critical distinction must be made between pathological C2–C3 subluxation and C2–C3 pseudosubluxation—a common physiologic finding on pediatric C-spine x-rays in which there is <4 mm of subluxation of C2 on C3 and normal spinolaminar alignment.
- Os odontoideum is a term used to describe a bony fragment with smooth cortical margins located cranially to the body of the axis, independent from a small or hypoplastic dens and may appear to look like a healed type I or type II dens fracture. The etiology of may be congenital nonunion of the dens to the C2 body or it may be related to fracture in utero or in early childhood. The clinical significance of os odontoideum is determined by flexion–extension x-rays, which are used to evaluate the stability of the cruciate ligaments.

SUBAXIAL CERVICAL SPINE INJURIES

- Prior to 8 years of age, injuries in this region are uncommon.
- Mechanisms which produce vertical axial loading cause compression or burst fractures of the vertebral body. Compression fractures are generally considered stable and usually heal without surgical intervention. Burst fractures, however, can be unstable and involve retropulsion of bony fragments into the spinal canal.
- Flexion may result in an avulsion fracture, which is identified by a small teardrop of bone observed at the anteroinferior margin of the vertebral body. A true teardrop fracture involves disruption of the facet joints, the anterior and posterior longitudinal ligaments, and the disk, and is considered unstable. MRI is used to evaluate subluxation or ligamentous damage, allowing the clinician to distinguish between a simple avulsion and a true teardrop fracture.
- Hyperflexion with or without distraction may result in facet dislocation. Hyperflexion with rotation can cause unilateral facet dislocation, whereas hyperflexion alone may result in bilateral facet injuries. Terms used to describe facet dislocation of varying degrees include perched, jumped, sprung, or locked.
- Flexion–distraction injuries can cause ligamentous disruption without fracture, potentially resulting in occult cervical instability with normal plain radiographic findings. Flexion–extension views can reveal movement that would have otherwise been missed on neutral lateral views.
- Posterior ligamentous disruption may present with increased interspinous distance on lateral x-ray or sagittal CT scan reconstructions.
- Hyperflexion or direct impact to the spinous process produces avulsion of a subaxial spinous process. The

most common level of spinous process fracture is C7 and is termed as “clay shoveler’s fracture.”

- Lateral mass, transverse process, or uncinat fractures occur with lateral hyperflexion.
- Spinous process, laminar, and pedicle fractures are produced with hyperextension. Milder hyperextension followed by flexion is associated with classic “whiplash” strain of the cervical musculature.
- Separation of the vertebral body from the end plate through the epiphysis is termed a physeal fracture. Diagnosis is critical because certain subtypes of physeal fractures are unstable and require operative stabilization.

SPINAL CORD INJURY WITHOUT RADIOGRAPHIC ABNORMALITY (SCIWORA)

- SCIWORA was originally used to describe traumatic myelopathy in individuals with no radiographic evidence of vertebral injury on plain x-rays, myelogram, or CT scan.
- The true incidence of SCIWORA in children is unknown, but estimates range from 4% to 66% of all children with spinal cord injuries.
- The neurological examination on initial presentation is predictive of long-term outcome in SCIWORA. Children with mild deficits at presentation regain full function; however, the more severely injured children tend to make a limited recovery.
- It is believed that SCIWORA occurs because the inherent hypermobility of the pediatric spine allows for transient deformation of the spinal column without fracture or ligamentous disruption at the expense of the spinal cord.
- With widespread use of MRI in traumatically injured patients, the definition and epidemiology of SCIWORA is continuing to evolve.

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QUESTIONS

1. A 2-year old male is brought to the ED after a fall from his mother’s arms. He is playful, and his exam is normal. Cervical spine radiographs are order and the radiologist reads the x-ray as showing a C2 fracture. The child most likely is displaying which of the following on x-ray?
 - A. Pseudosubluxation
 - B. Type I odontoid fracture
 - C. Ossification center of C2
 - D. Type III odontoid fracture
 - E. Jefferson fracture.
2. An unresponsive toddler comes into the ED after a motor vehicle accident. He is in a cervical collar and needs to have his airway secured. The best course of action would be which of the following treatment options?
 - A. Blind nasotracheal intubation
 - B. Emergency cricothyroidotomy

- C. Oral intubation with in-line traction
 D. Fiberoptic intubation
 E. Laryngeal mask airway placed without removal of the cervical collar
3. A 16-year-old otherwise healthy male comes into the emergency room after being involved in a motor vehicle accident. He has multiple long-bone fractures. The patient is noted to be hypotensive with a normal to low heart rate. He is suffering from which of the following conditions?
 A. Spinal shock
 B. Hypovolemic shock
 C. Septic shock
 D. Cardiogenic shock
 E. Equipment error
4. A pediatric patient presents with a known spinal cord injury. On examination, he can flex at the elbow but cannot extend his arm and has no movement of his hand and wrist. His injury is best localized to which level?
 A. C5
 B. C6
 C. C7
 D. T1
 E. C2
5. A 5 year-old presents after being involved in a high-speed motor vehicle accident. On exam, he is found to have a sensory level, and a C7 motor level. Which of the following is the best describes corticosteroid use in this situation?
 A. An option of the provider though no data exists to support their efficacy.
 B. A clinical standard with high-dose methylprednisolone to be given within 24 hours of injury.
 C. A clinical standard with high-dose methylprednisolone to be given within 8 hours of injury.
 D. Completely contraindicated in children.
 E. A clinical standard in children because it prevents post-traumatic ischemia.
6. A 12-year-old presents after falling from his bike. He is complaining of neck pain and is in a cervical collar. The correct initial imaging study for this patient would be which of the following?
 A. Computed tomography scan
 B. Magnetic resonance imaging
 C. Lateral x-ray
 D. Three-view cervical spine x-rays
 E. Flexion/Extension x-rays
7. A 5-year -old presents with neck pain after being involved in a motor vehicle accident. The x-rays show angulation at C2 with a lucency seen below the dens. The most likely diagnosis would be which of the following injuries?
 A. Type I odontoid fracture
 B. Type III odontoid fracture
 C. Os odontoideum
 D. Synchrondrosis fracture of C2
 E. Normal anatomy of the dens in this age group
8. A 4-year-old is seen in the ED after a fall. X-rays show 2 mm of subluxation of C2 on C3. On exam, the child has no neck pain. The correct next step would be which of the following for this patient?
 A. CT scan of the cervical spine
 B. Place in cervical collar
 C. Consult neurosurgery for further evaluation
 D. Treat with methyprednisolone
 E. Observation
9. A 9-year-old presents with her head turned to the right and tilted to the left. When doing ROM exercises, she cannot bring her head past midline. She has no neck pain. X-rays are read as normal. The correct diagnosis is which of the following?
 A. Atlanto-axial rotatory subluxation (AARS)
 B. Pseudosubluxation
 C. Ligamentous injury
 D. SCIWORA
 E. Torticollis
10. A 5-year-old is examined after falling out of a window. X-rays and MRI show no fractures or evidence of spinal cord injury. The patient has persistent motor weakness. The correct next step in management would be which of the following?
 A. Start methyprednisolone
 B. Observation
 C. Spinal fusion
 D. Halo immobilization
 E. Repeat MRI in 1 week

ANSWERS

1. A. The ossification centers at C2 are not fused at this age. Odontoid fractures at this age are exceedingly rare, and a Jefferson fracture is a fracture through the anterior and posterior arch of C1. This child's normal neurological exam and the absence of neck pain make a fracture less likely.
2. C. Oral intubation with in-line traction with rapid sequence intubation is the preferred choice of providers. Cervical immobilization makes direct laryngoscopy almost three times harder. Blind nasotracheal intubation is unreliable and technically difficult. Emergency cricothyroidotomy is contraindicated in small children because of the small size of the cricothyroid membrane. Fiberoptic intubation is generally not the first intervention

because of the skill required and availability of equipment. Laryngeal mask airway with inline traction is an option following failed oral intubation, but has not been studied in children.

3. A. The correct answer for this teenager is spinal shock. Hypovolemic shock is not correct because the patient would be tachycardic. Septic shock would also have an elevated heart rate along with hypotension. Cardiogenic shock is correlated with tachycardia and a weak, thready pulse.
4. A. C5. A patient with a C4 motor level would not be able to move his shoulder and may have impaired diaphragm function. At C6 and C7, the patient would be able to flex and extend at the elbow. A patient with a T1 level would finger flexion and wrist flexion, and a patient with a C2 level would be a ventilatory assisted quadriplegic.
5. A. Within the National Acute Spinal Cord Injury Trial, the investigators proposed using high-dose methylprednisolone with 8 hours of spinal cord injury. However, within the study, no children 13-years-of age and younger were included, and recent statements by CNS indicate that this is an option of the provider and is not a standard. It is not contraindicated in children.
6. D. Although the use of x-rays in the pediatric trauma patient is understudied, it is reasonable to start with three-view x-rays to initially evaluate a trauma patient. This statement is based on the NEXUS criteria and the subset analysis of pediatric patients showing good sensitivity in detecting injured children. Drawbacks of this study include the absence of young children and the limited number of true injuries. Both CT and MRI, are high cost and in the case of CT deliver a significant radiation dose to the pediatric patient. These studies should be used as secondary imaging study if physical exam and/or x-rays warrant further study. No data exist that show that isolated lateral x-rays or flexion/extension x-rays should serve as an initial screening
7. D. This is a fracture through the synchondrosis of C2. All of the other fractures describe uncommon fractures in the pediatric population. Type I odontoid fractures are at the tip of the dens. Os odontoidem is a congenital abnormality of C2 and not an acute fracture. Jefferson fractures are through the arch of C1, both anterior and posterior arches. A type III odontoid fractures is exceedingly rare in this age group, and extends into the body of C2.
8. E. Pseudosubluxation of C2 on C3 is a common entity seen in the pediatric population. It is defined as subluxation of 2 on 3 though the spinolaminar

line does maintain alignment. This entity should be considered in patients with these x-ray findings in the absence of neck pain. They do not need to be placed in the cervical collar and to be seen by a neurosurgeon. Therapy for spinal cord injury is also not indicated.

9. A. Atlanto-axial rotatory subluxation or AARS is the correct answer. These children have a clinically obvious head tilt in the absence of neck pain, differentiating AARS from torticollis. The x-rays are negative differentiating it from pseudosubluxation, and there are no neurologic deficits associated with AARS differentiating it from SCIWORA.
10. B. This patient has a spinal cord injury without radiographic abnormality. This has previously been defined as neurological deficit in the absence of radiographic abnormality. This however was initially defined in the pre-MRI era. The correct answer for this patient is observation. This occurs in up to 25% of pediatric patients. These patients do not need spinal stabilization, halo immobilization, or methylprednisolone. In addition, there is no need for repeat imaging as well. These deficits are usually transitory and resolve within several hours to several weeks.

31 THORACIC TRAUMA

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Wendy Ann Lucid
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JUST THE FACTS

- Trauma is the most common cause of morbidity and mortality in children of age 1–14 years and accounts for 5–10% of pediatric hospital admissions.
- Thoracic trauma poses unique challenges as even seemingly benign mechanisms of injury have the potential to cause severe injuries and death in infants and young children.
- Despite its low incidence, thoracic injury remains a significant cause of pediatric death secondary to trauma.

MECHANISM OF INJURY

- Blunt trauma accounts for approximately 85% of chest injuries in children.
- Most common mechanisms of thoracic injury include motor vehicle crashes, falls, and nonaccidental trauma

TABLE 31-1 Pediatric Anatomy and Physiology

Incomplete ossification of bony structures	Diminished functional residual capacity
Greater flexibility of thoracic cage	Greater mobility of mediastinal structures
More flexible ligamentous structures	Large cardiac reserve; delayed recognition of shock
Underdeveloped supporting musculature	Higher oxygen consumption per unit body mass
Narrow, short trachea	
More compressible trachea	

(infants and toddlers); pedestrian and sports-related injuries (school-aged children); and again motor vehicle crashes, extreme sports, violence, and suicide (adolescents).

- Most common injuries sustained include pulmonary contusion, pneumothorax, hemothorax, pneumothorax, and rib fractures.
- Thoracic trauma in children is more often associated with multi-system injury and is a marker of injury severity.
- Penetrating trauma is an independent predictor of mortality; urgent and immediate evaluation and treatment by the surgical trauma team are necessary.
- Gunshot wounds to the chest are associated with abdominal injuries in 30–40% of cases.

PATHOPHYSIOLOGY

- There are critical differences in the pediatric anatomy that affect a child's risk of sustaining significant injuries from thoracic trauma (Table 31-1).
- Increased compliance of cartilaginous ribs allows impact forces to dissipate, resulting in significant injury with few or no apparent external signs of trauma.
- Thoracic trauma decreases respiratory efficiency, leading to earlier hypoxia, respiratory fatigue, and failure.
- Mediastinal structures are more mobile and prone to major injury.

CLINICAL FINDINGS

- Physical signs of thoracic injury may be subtle in the child, even with severe injury.
- Children may present with little to no external evidence of thoracic trauma.
- The quality of respirations may vary from rapid, labored, shallow, or seemingly no distress with varying degrees of hypoxia.

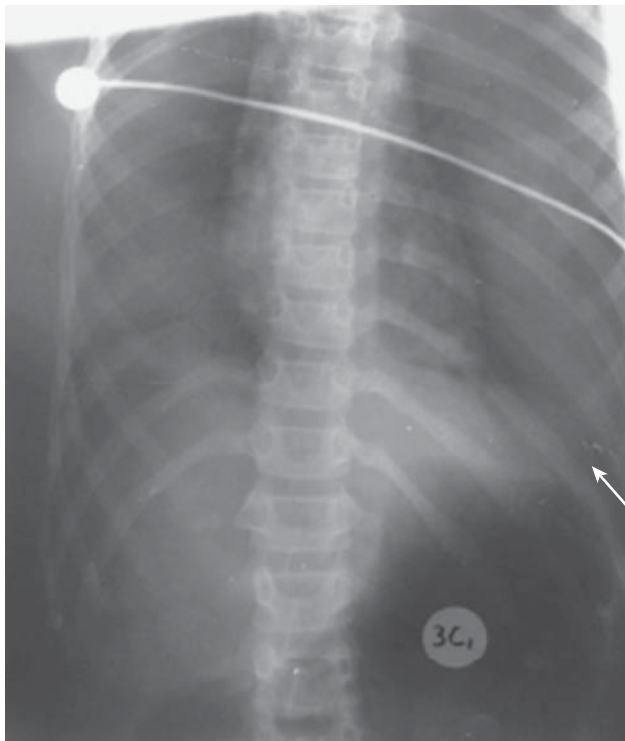
- Pulmonary auscultation may reveal hyperresonance or absent breath sounds (eg, pneumothorax), decreased breath sounds and dullness to percussion (eg, hemothorax), or normal breath sounds (eg, small pulmonary contusions).
- Absence of hypoxia does not exclude the presence of serious or life-threatening thoracic injuries; central cyanosis can be absent in hemorrhagic shock due to a relative decrease in the amount of unsaturated hemoglobin.
- Hypotension is a late sign of shock; children have a remarkable ability to compensate for significant hemorrhage and may remain in a state of compensated hypovolemic shock until up to 40% of their blood volume is lost.
- Children maintain their cardiac output during states of compensated hypovolemic shock by increasing their heart rate. [Cardiac output is dependent upon both heart rate and stroke volume ($CO = HR \times SV$)].
- Several clinical findings are independent predictors of thoracic injury: abnormal chest auscultation findings, hypotension, abnormal external thoracic examination, and elevated age-adjusted respiratory rate (strength of association listed in decreasing order).

MANAGEMENT OF THORACIC INJURY

- Priority for management lies in the recognition of injury and stabilization of the airway, breathing, and circulation.
- Obtain airway patency, while protecting and immobilizing the cervical spine using a jaw thrust maneuver. Avoid using a head-tilt, chin-lift mechanism when positioning the neck.
- Provide adequate oxygenation with 100% FIO₂ and breathing support with bag-valve mask ventilation.
- Consider early rapid sequence intubation if prolonged ventilatory support may be necessary.
- Circulatory support should focus on immediate assessment of key vital signs (heart rate, quality of central and distal pulses, and blood pressure) and state of perfusion (color, capillary refill time in seconds).
- Initiate fluid resuscitation with two large-bore intravenous lines and infusion of isotonic crystalloid solutions, such as normal saline or lactated ringers. If hemorrhage shock is suspected, consider transfusion of donor blood products.
- Useful diagnostic studies include an arterial blood gas, baseline hemoglobin/hematocrit, chest radiograph, and chest computed tomography (CT) if there is a high likelihood for thoracic injury not detected on plain film radiographs.

SPECIFIC INJURIES AND MANAGEMENT PULMONARY CONTUSION, LACERATION, AND HEMATOMA

- Pulmonary injuries are the most common type of thoracic trauma in children.
- Tachypnea is the chief physiologic response to hypoxia and should be identified early.
- Pulmonary contusions or “the bruised lung” interfere with gas exchange, leading to inadequate oxygenation and hypoventilation.
 - Diagnosis is by chest radiograph or chest CT scan.
 - Treatment is to prevent hypoxia and respiratory failure with supplemental oxygen and close monitoring.
 - Mechanical ventilation and higher positive end-expiratory pressures (PEEP) may be necessary.
 - Additional treatments include fluid restriction, early mobilization, pain control, and early treatment of secondary pneumonia.
- Pulmonary lacerations are often associated with penetrating trauma, but may result from rib fractures. Surgical repair is necessary when the laceration is associated with ongoing bleeding or air leakage.
- Pulmonary hematoma is uncommon, generally a self-limited injury, and rarely progresses to lung abscess.



A

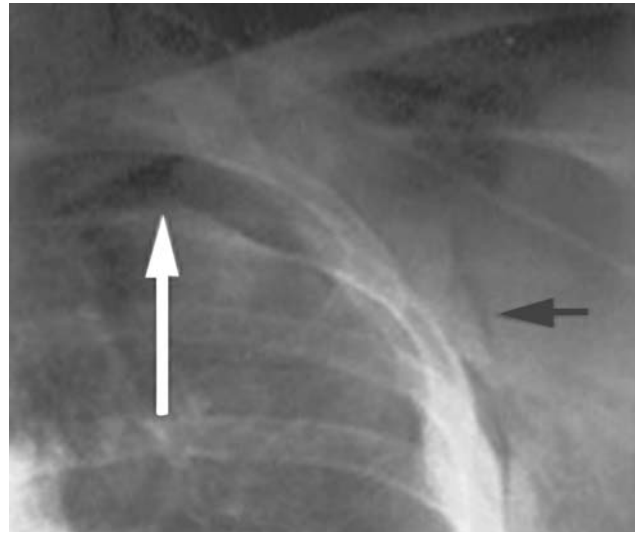
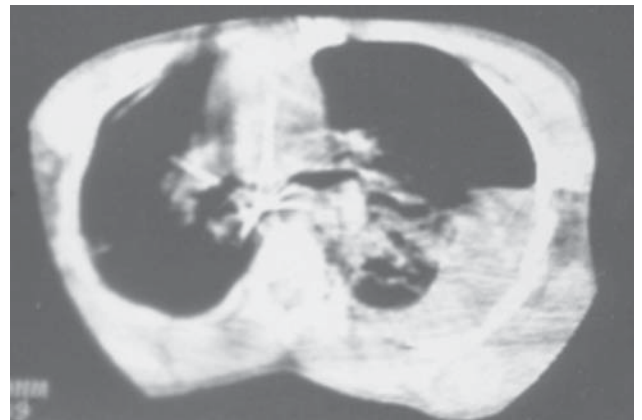


FIG. 31-1. A CXR demonstrating a pneumothorax (white arrow) and overlying subcutaneous emphysema (black arrow), which can be seen tracking up to the neck.

PNEUMOTHORAX

- A pneumothorax is an abnormal air collection in the pleural space and occurs in approximately one-third of cases of pediatric thoracic trauma (Figs. 31-1 and 31-2).



B

FIG. 31-2. (A) A chest x-ray of a 12-year-old boy who was struck by a car. This hastily done, poor-quality film was taken when the child’s mental status began to deteriorate. Note the left costophrenic angle, which is surprisingly deep. This is an example of a deep sulcus sign seen in an anterior pneumothorax. (B) The chest CT scan of the same patient clearly demonstrates the anterior pneumothorax.

- The majority of pneumothoraces have associated injuries (eg, associated pulmonary contusion).
- A small, uncomplicated pneumothorax is often asymptomatic; traumatic pneumothoraces have the potential to expand into a more serious tension pneumothorax.
- A tension pneumothorax puts pressure on mediastinal structures, affects cardiac output, and, if untreated, may rapidly lead to cardiovascular collapse.
- Treatment for small, isolated traumatic pneumothoraces include observation for at least 6 hours with a repeat chest radiograph. If there is no size increase or underlying parenchymal injury and the patient remains clinically stable, the patient may be discharged to return in 24 hours for a repeat evaluation.
- For large pneumothoraces, a large-bore chest tube will allow for the evacuation of air, blood (Fig. 31-6).
- Stable patients with small, insignificant pneumothoraces may need a chest tube if they will have mechanical ventilation or emergency air transport.

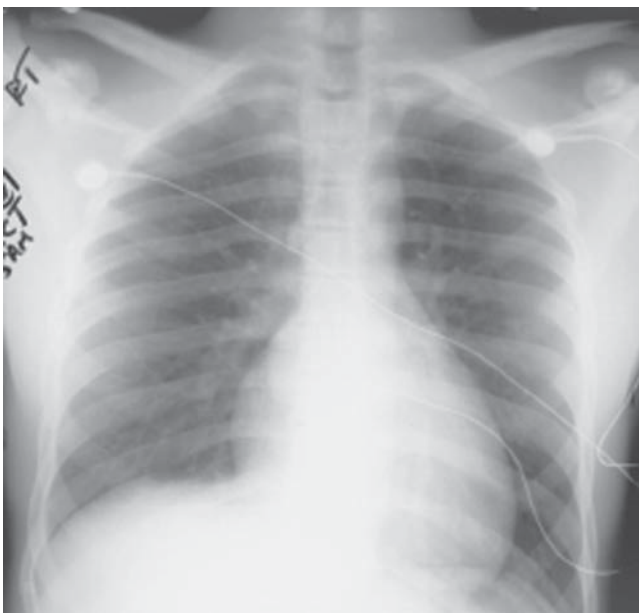
TENSION PNEUMOTHORAX

- Tension pneumothorax occurs when a pneumothorax puts pressure against the mediastinal structures, causing vascular compromise of the heart and great vessels.

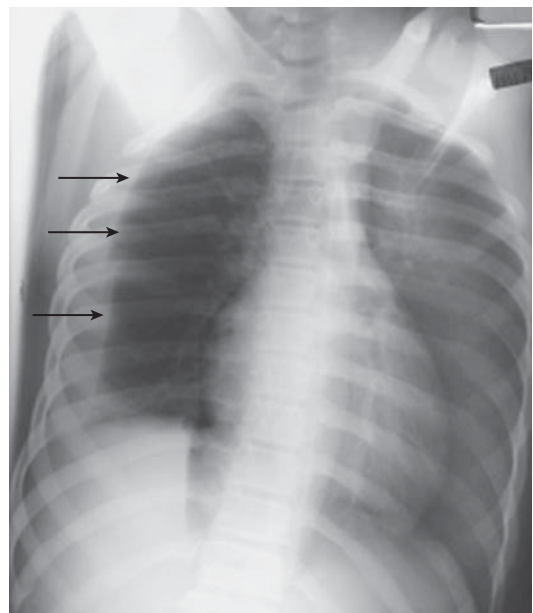
- Clinical findings include severe respiratory distress, decreased breath sounds and hyperresonance on the affected side, contralateral tracheal deviation, distended neck veins from compromised venous return, a narrow pulse pressure, and hypotension.
- A tension pneumothorax affects cardiac output and must be expeditiously decompressed; delay or failure to treat may rapidly lead to cardiovascular collapse.
- Diagnosis is made clinically and, when suspected, treatment should never be delayed to obtain confirmation by chest radiograph.
- Bedside decompression can be performed using a needle thoracostomy (see Procedures section for details). Definitive treatment is accomplished using a large-caliber thoracostomy tube (see Procedures section for details).

HEMOTHORAX AND MASSIVE HEMOTHORAX

- A hemothorax is an abnormal collection of blood in the pleural space (Fig. 31-3).
- Hemothorax is rare in children and usually a result of forceful mechanisms, such as a high-speed motor vehicle crash, a fall from a great height, or a high-powered or close-range gunshot wound.



A



B

FIG. 31-3. (A) An AP radiograph of a 14-year-old boy who was hit by a car. While there are no pathognomonic findings indicative of a hemothorax, in this clinical context the caretakers were concerned about the elevation of the right hemidiaphragm. In particular, the lateral position of the right hemidiaphragm apex makes the possibility of subpulmonic fluid even more likely. (B) The subsequent right lateral decubitus film, also done as a portable, demonstrates a significant hemothorax.

- Clinical findings include decreased breath sounds and dullness to percussion on the affected side, varying signs of respiratory distress, and signs of hemorrhagic shock
- Early signs of blood loss should be noted and treated immediately, as each hemothorax can hold up to 40% of a child's blood volume before clinical signs of decompensation. Use caution when interpreting lab evaluations; initial hemoglobin and hematocrit may not accurately reflect current blood volume due to the lack of time necessary for equilibration.
- Treatment by drainage is necessary for both evaluation of blood loss and to prevent delayed complications; an emergency thoracotomy may be necessary to control massive hemorrhage (If initial drainage is >10-15ml/kg or 2-4ml/kg/hr).
- Additional treatment includes fluid resuscitation with crystalloids and blood products, and surgical repair when ongoing blood loss is significant.

OPEN PNEUMOTHORAX

- An open pneumothorax (sucking chest wound) is created when the chest wall is sufficiently injured to create bidirectional flow of air through the wound and is most commonly associated with massive penetrating injury.
- Loss of negative intrathoracic pressure does not allow for normal expansion of the lungs leading to ineffective, paradoxical breathing.
- Small injuries can be treated by covering the chest wall defect with sterile petroleum dressing and placing a thoracostomy tube through a fresh incision.
- Prehospital treatment may consist of placing a petroleum dressing with only three sides taped to create a flutter valve to allow for ongoing chest decompression. This should be converted to a sealed dressing with thoracostomy tube placed as soon as possible.
- Consider intubation and ventilatory support with large chest-wall defects; urgent thoracotomy may be necessary to repair large chest-wall defects and underlying injuries.

TRAUMATIC TRACHEAL AND BRONCHIAL DISRUPTION

- Traumatic tracheal and/or bronchial disruption is rare in children and is associated with a high mortality rate.
- Airway injury is most often caused by penetrating trauma, but blunt injury involving high-speed shearing forces or severe compressive forces may also disrupt the tracheobronchial tree.
- Clinical findings vary depending on the level of airway disruption and range from mild respiratory

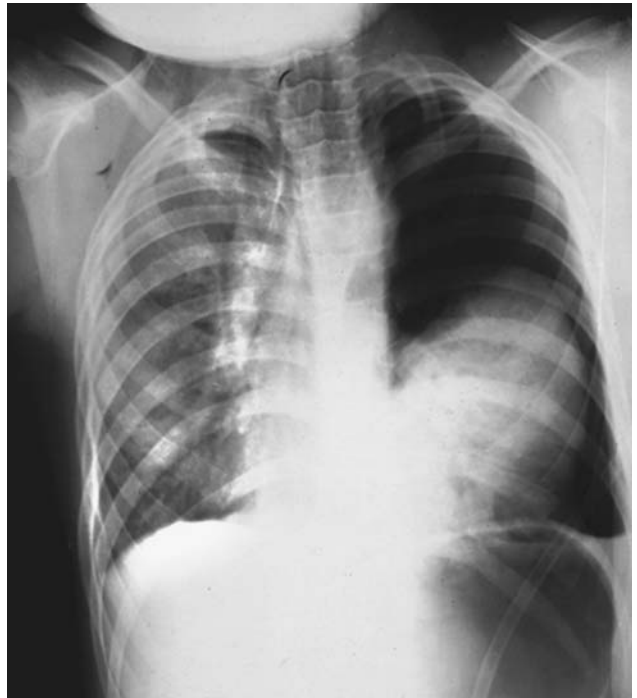


FIG. 31-4. This 13-year-old child was hit by a car. The AP chest x-ray demonstrates an obvious left-sided pneumothorax, but the position of the left lung is peculiar. Rather than collapsing toward the hilum, the lung seems to have fallen to the dependent portion of the thorax. This is called the fallen-lung sign. This is an extremely rare finding on x-ray, but when present, the abnormal position of the lung, together with the left-sided pneumothorax, is highly suggestive of rupture of the tracheobronchial tree. This was confirmed on a subsequent CT scan.

- distress to respiratory arrest secondary to a tension pneumothorax.
- Diagnosis is made both clinically and radiographically (Fig. 31-4); early bronchoscopy and chest CT enhance diagnosis.
- Treatment depends on symptoms and location of injury and varies from observation and chest thoracostomy to operative repair.
- Establishing an airway can be complicated and should be done with fiberoptic assistance; endotracheal suctioning and other blind airway interventions should be avoided.

TRAUMATIC ASPHYXIA

- Traumatic asphyxia is caused by sudden, direct compression of the elastic pediatric thoracic cage against a closed glottis causing dramatic increases in intrathoracic pressure, temporary vena cava obstruction, and transmission of the pressure into the capillaries of the head and neck.

- Patients present with cyanosis, plethora, and petechiae of the head and neck regions, subconjunctival hemorrhages, face and neck edema, and in severe cases, respiratory distress, intracranial hemorrhage, altered mental status, and seizures.
- Treatment involves removing the cause and tending to the resulting complications.

TRAUMATIC ESOPHAGEAL RUPTURE

- Traumatic esophageal rupture is extremely rare in children and occurs with severe blunt upper abdominal trauma.
- Clinical signs include pain and shock out of proportion to the apparent severity of injury. Subcutaneous emphysema may dissect into the neck or mediastinum.
- Chest radiograph often reveals mediastinal emphysema; fluoroscopy or CT scan can confirm the diagnosis.
- Treatment involves urgent surgical repair with mediastinal drainage.
- If unrecognized, this condition progresses rapidly to mediastinitis, sepsis, and death despite surgical intervention.

TRAUMATIC DIAPHRAGMATIC HERNIA

- Traumatic diaphragmatic hernia occurs when forceful blunt trauma causes a sudden increase in intra-abdominal pressure (eg, displacement of a lab belt upward into the abdomen during a motor vehicle crash).
- Clinical findings include varying degrees of respiratory distress, abnormal breath sounds on the affected side during chest auscultation (breath sounds may be decreased or obscured by bowel sounds in the chest cavity), and varying signs of external abdominal wall trauma; early respiratory symptoms are subtle, causing frequent delays in diagnosis.
- Chest radiograph may reveal bowel or stomach in the chest cavity, elevation of the hemidiaphragm, or a nasogastric tube present in the chest.
- Initial management should concentrate on adequate oxygenation, ventilation, and stabilizing other injuries; a nasogastric tube should be placed to decompress the stomach and intubation with positive-pressure ventilation may be necessary.
- Acute traumatic diaphragmatic herniation requires surgical repair.

RIB FRACTURES

- Rib fractures are uncommon in children because of their compliant, cartilaginous thoracic cage and when present, are often results of a direct blow to the chest



FIG. 31-5. This is the x-ray of an adolescent who sustained significant blunt chest trauma. Note the fractures of ribs 4 through 10 seen medially that are indicative of posterior rib fractures, and the more peripheral fractures seen in ribs 4 through 8. This patient presented with paradoxical movement of the chest with breathing, crepitus of subcutaneous emphysema, and a pneumothorax that required placement of the chest tube (also seen on the x-ray).

- or significant anterior–posterior forces seen with crush or squeezing mechanisms (Fig. 31-5).
- Rib fractures serve as important markers for potentially serious underlying injuries (eg pulmonary contusions, pneumothorax, and hemothorax).
- Child abuse should be suspected when a clear history of trauma is absent and especially if there are multiple fractures in various stages of healing; children younger than 1 year of age are at the highest risk of abuse from shaking and should have an extensive workup and a report made to local authorities when rib fractures are detected.
- Up to 50% of isolated rib fractures may be missed on the initial chest radiograph; further evaluation with chest CT scan may be necessary.
- Treatment for simple rib fractures involves optimizing the patient’s respiratory effort with aggressive pain management and breathing therapy with incentive spirometry.

FLAIL CHEST

- Flail chest occurs when the structural integrity of the chest wall is compromised by the presence of more than two fractures to the same rib, causing the ribs of the flail segment to move paradoxically.
- Signs and symptoms are the result of ineffective respirations and include varying degrees of respiratory distress, hypoxia, and paradoxical chest wall motion.
- Chest radiograph confirms the diagnosis (Fig. 31-5) and often reveals associated pulmonary contusion.
- Treatment is aimed at preventing hypoxia and respiratory failure; patients with paradoxical respirations from flail chest will need positive-pressure ventilation until operative rib fixation.

SPINE INJURIES

- Spine injuries are less frequently associated with thoracic trauma in children.
- Cervical and thoracic spine immobilization, however, should be maintained until evaluation is complete and these areas are cleared of injury.
- Spine fractures may be detected on screening or detailed radiographs and CT scan.
- Diagnosis of associated spinal cord and ligamentous injuries is facilitated by magnetic resonance imaging (MRI); some patients may have spinal cord injury without radiographic abnormality and a high index of suspicion should lead to further evaluation.

CARDIOVASCULAR INJURIES

- Cardiac and great vessel injuries are uncommon in children, but when present, increase the morbidity and mortality associated with thoracic trauma.
- Myocardial contusion is the most common injury seen with blunt chest trauma; cardiac tamponade being most common with penetrating mechanisms.
- Other injuries and complications include myocardial or pericardial laceration, valvular disruption, myocardial necrosis with subsequent aneurysm, traumatic aortic insufficiency, coronary artery injury, and cardiac conduction system injury.
- Traumatic aortic rupture is the most common great vessel injured, but underreported since more than 50% of victims succumb to this injury before reaching the hospital.

MYOCARDIAL CONTUSION

- Myocardial contusion is the most common unsuspected and under diagnosed injury after blunt thoracic

trauma and are more severe in cases of multi-system trauma.

- Contusions are most often sustained as a result of motor vehicle crashes and pedestrian injuries.
- Cardiac contusions are complicated by pulmonary contusions in half and rib fractures in a third of cases.
- Clinical findings are normal in up to half of injured patients, with no complaints of chest pain, no external evidence of trauma and a cardiac exam.
- Tachycardia is the most sensitive and important indicator of myocardial injury and should be considered significant; however, interpretation may be challenging with coexisting pain and anxiety.
- More severely injured myocardium may present with dysrhythmias, hypotension, and signs of cardiac failure.
- Diagnosis should be made using a combination of clinical suspicion, clinical examination findings, and cardiac-specific evaluation.
- ECG abnormalities are less common in children.
- Abnormalities in cardiac function on echocardiography and an elevation in cardiac-specific enzyme levels, specifically troponin I, are important indicators of cardiac contusion.
- Management of cardiac contusion is mostly supportive and when suspected, children should be admitted for observation with cardiac monitoring and serial measuring of cardiac enzymes.

CARDIAC TAMPONADE

- Cardiac tamponade is a life-threatening condition that occurs when fluid (blood or serous fluid) fills the pericardial space compromising venous return.
- Penetrating injuries, such as stab and gunshot wounds, are the most common etiologies.
- Patients often present in normovolemic shock which may rapidly progress to hypotension and cardiac arrest unless promptly treated.
- Clinical findings include presence of a precordial wound, tachycardia, narrow pulse pressure, and pulsus paradoxus. The clinical combination of muffled or distant heart sounds, hypotension, and jugular venous distention (Beck's triad) should increase suspicion.
- Diagnosis is made clinically, but confirmation is assisted with bedside echocardiography. Chest radiograph may reveal the classic water bottle cardiac silhouette and ECG often shows tachycardia with extremely low voltage.
- Treatment should be based on clinical suspicion and patient deterioration; immediate bedside needle pericardiocentesis may be life saving (see Procedures section). Definitive treatment involves thoracotomy,

pericardiotomy, and repair of the underlying injury. Repeated pericardial aspiration and aggressive blood resuscitation are reasonable alternatives until a physician with expertise in emergency thoracotomy is available.

TRAUMATIC RUPTURE OF THE GREAT VESSELS

- Rupture of the great vessels is extremely rare in children due to the higher elastin content of their connective tissue.
- Aortic disruption at the level of the ligamentum arteriosum is the most common injury in children.
- Great vessel injuries carry high morbidity and mortality with more than 50% of injured patients dying at the scene.
- Blunt aortic injury is most commonly caused by rapid deceleration, as seen in high-speed automobile crashes and falls from great heights.
- Clinical findings include chest pain localized to the anterior chest, back, or upper abdomen, and a murmur radiating to the back. Many children will have significant associated injuries to the heart, lungs, abdomen, and central nervous system.
- Diagnosis can be difficult in children; a widened mediastinum on chest radiograph should increase suspicion for aortic injury, but alone does not confirm the diagnosis. Early surgical consultation, chest CT scan, and angiography help confirm the diagnosis.
- Initial treatment should be directed toward the ABCs of trauma care and aggressive fluid resuscitation. Hemopneumothorax is universally associated with penetrating chest trauma and should be treated promptly and aggressively.
- Definitive treatment requires immediate surgical repair.

PROCEDURES

These procedures should only be performed by physicians trained in the techniques and for the appropriate indication.

NEEDLE THORACOSTOMY

Needle thoracostomy is an emergent but temporary procedure used to relieve a pneumothorax, in particular, a tension pneumothorax. Uncertainty as to the side of the tension pneumothorax should not prohibit initiation of this potentially life-saving procedure. Decompression of the alternate side should be done

if immediate improvement is not seen with the initial thoracostomy.

To perform this procedure:

- Place the patient in a supine position and prepare the site in a sterile manner.
- Identify the insertion site between the second and third rib (second intercostal space) at the midclavicular line on the affected side.
- Attach a syringe filled with 3 mL of saline to an angiocath or butterfly needle.
- While maintaining negative pressure on the syringe, insert the needle or catheter and advance it over the superior aspect of the rib to avoid the intercostal neurovascular bundle.
- When air bubbles are visualized in the syringe or an audible rush of air is detected (in cases where a syringe is not attached), the pleural space has been entered.
- If a tension pneumothorax was relieved, the catheter should remain in place and air intermittently aspirated to prevent a reaccumulation of air under pressure.
- Place a large-bore chest tube (tube thoracostomy) as soon as possible and attach it to a drainage/suction device.

TUBE THORACOSTOMY

Performing tube thoracostomy (inserting a chest tube) (Fig. 31-6) is indicated for the drainage of air (pneumothorax), blood (hemothorax), or fluid (eg, chylothorax) from the pleural space.

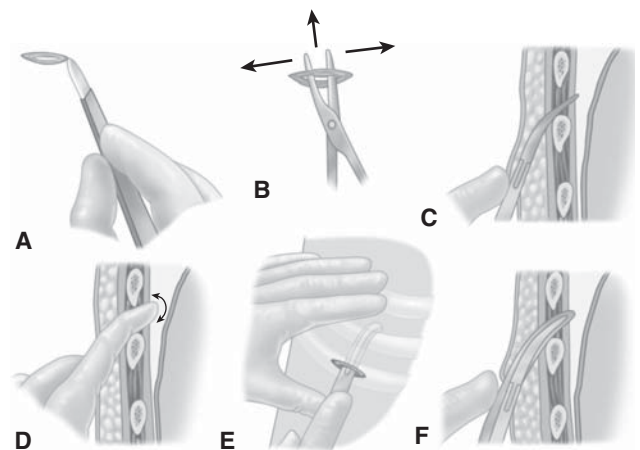


FIG. 31-6. The tube thoracostomy. (A) The skin incision is made. (B) A tract is bluntly dissected in the subcutaneous tissues. (C) The Kelly clamp is forced into the pleural cavity. (D) A finger is inserted through the tract to feel for adhesions. (E) The chest tube is held in the Kelly clamp and inserted through the tract. (F) The chest tube is guided into the pleural cavity.

TABLE 31-2 Chest Tube Sizes

WEIGHT (Kg)	CHEST TUBE SIZE (Fr)
<5	8–12
5–10	10–14
11–15	14–20
16–20	20–24
21–30	20–28
31–50	28–40
>50	32–40

To perform this procedure:

- Place the patient in a supine position and prepare a sterile surgical field.
- Select the appropriate size chest tube based on the patient's weight in kilograms (Table 31-2).
- Prepare for appropriate analgesia and sedation. Use local anesthetics at the insertion site and consider intravenous sedation and pain control in younger children who may have difficulty verbalizing pain.
- Based on clinical scenario, plan the path for tube insertion: tubes intended to drain air should be directed anteriorly; tubes to drain fluid should be directed toward the patient's back.
- Localize the insertion site at the fourth or fifth intercostal space at or just anterior to the midaxillary line.
- Make a small skin incision parallel to the intercostal space approximately one or two intercostal spaces below the projected pleural entry site. The incision should be large enough to allow for the passage of the chest tube, a hemostat attached to it, and the provider's finger.
- Using a blunt dissecting technique to create a tunnel from the skin to the intercostal space, guide the tip of the hemostats over the top of the rib, again to avoid the intercostal neurovascular bundle. Apply pressure to the tip of the hemostats to enter the pleural space. Once the tip is through, make the hole larger by spreading the instrument.
- Secure the end of the chest tube with the hemostats and using your finger as a guide, direct the chest tube through the skin and soft tissue into the pleural space and along the predetermined pathway.
- Once inserted, suture the chest tube in place and connect the tube to an underwater seal apparatus with or without suction.
- A post insertion chest radiograph should be obtained to confirm proper placement of the tube and reexpansion of the lung.

PERICARDIOCENTESIS

- Emergency pericardiocentesis should be performed for life-threatening cardiac tamponade by providers skilled in this technique.

- Prior to procedure, maximize patient airway and breathing and ensure IV access.
- Place the patient on a cardiac monitor and position patient in a slight reverse Trendelenburg position.
- If available, use ultrasonography to visualize the pericardial fluid collection.
- Prepare a sterile surgical field and use local anesthesia and sedation in the awake patient undergoing this procedure.
- Choose the appropriate equipment for the procedure:
 - For infants and young children, a 20-gauge spinal needle or a 1.5-inch, 18- to 20-gauge needle with a catheter may be used.
 - For older children, an 18-gauge spinal needle or a 1.5-3 inch, 16-gauge needle with catheter may be used depending on size of child and approach used.
- Attach a 5 or 10 mL syringe to the appropriate size needle.
- Attach an alligator clip to the hub of the needle and connect to a precordial lead on an ECG monitor. This connection will detect dysrhythmias produced when the myocardium is irritated by the advancing needle.
- Identify the needle insertion site on the anterior chest wall immediately inferior and 1 cm left of the xiphoid process.
- While applying negative pressure to the syringe, advance the needle at a 45° angle in the direction of the tip of the patient's left scapula until pericardial fluid or blood is obtained, or until ECG changes are noted.
- An alternate parasternal approach can be used for adolescents and adults. For this approach, the needle is inserted perpendicular to the skin surface at the left fifth intercostal space, just lateral to the sternum.
- Definitive treatment by a skilled surgeon may include a pericardial catheter for ongoing drainage, creation of a pericardial window, and surgical repair of any injuries.

EMERGENCY DEPARTMENT THORACOTOMY

- Survival outcomes for patients who suffer cardiopulmonary arrest after thoracic trauma are poor, with less than 5% surviving to discharge.
- Only a few select injuries may benefit from thoracotomy including ventricular laceration, cardiac tamponade, thoracic arterial injuries, and intercostal arterial injuries.
- Use of ED thoracotomy for pediatric patients is limited:
 - Children suffering trauma who have lost vital signs in the field and have not regained cardiac function in the ED do not benefit from ED thoracotomy.
 - Children who have suffered penetrating or blunt thoracic trauma associated with detectable vital

signs who then deteriorate despite maximal conventional therapy have a better chance of survival from ED thoracotomy.

ORGAN DONATION

- Victims of thoracic trauma and cardiopulmonary arrest who do not experience a return of spontaneous circulation may be eligible for organ donation.
- Continue resuscitative efforts until a decision regarding organ viability is made.

LAW ENFORCEMENT

- Most states require reporting of stab wounds, gunshot wounds, and assaults.
- Laws require suspected child abuse be reported to local authorities and child protective services.

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QUESTIONS

1. A 2-year-old child falls from the second story window of his home and lands on his back. He presents to the ED alert and awake with these vitals: HR 180, BP 98/66, RR 60, and O₂ saturation of 89%. Which of the following is the most likely injury sustained in this patient?
 - A. Simple pneumothorax
 - B. Rib fracture with pneumothorax
 - C. Pulmonary contusion
 - D. Pulmonary laceration
 - E. Tracheal disruption
2. A 6-year-old child in a motor vehicle collision presents to the ED in cardiopulmonary arrest. The likelihood of survival after prehospital traumatic arrest is which of the following?
 - A. 5%
 - B. 10%
 - C. 15%
 - D. 30%
 - E. 50%
3. A 12-year-old presents to the ED with a gun shot wound to the right inferior chest wall. The trauma team should have a high index of suspicion for which of the following associated injuries that occur in 30–40% of such cases?
 - A. Major cardiovascular injury
 - B. Spinal cord injury
 - C. Myocardial contusion
 - D. Abdominal injury
 - E. Tracheal rupture
4. Children are at increased risk of sustaining significant injuries from thoracic trauma, even when there is little to no external sign of trauma. A characteristic of pediatric anatomy that contributes to this risk includes which of the following?
 - A. Decreased elastin component of major vessels
 - B. Strong supporting musculature
 - C. Decreased mobility of mediastinal structures
 - D. Stiff, noncompressible trachea
 - E. Increased chest wall compliance
5. A 3-year-old pedestrian struck by a car arrives in the ED with external evidence of head trauma and a respiratory rate of 8 breaths/min, HR 130, and BP 70/30. Which of the following is your initial step in this patient's management?
 - A. Bag-mask ventilatory support
 - B. Assess airway patency with jaw thrust maneuver while maintaining cervical spine immobilization
 - C. Oxygenation with 100% FiO₂ by face mask
 - D. Immediate vascular access
 - E. Trauma team consultation
6. Children have a remarkable ability to compensate for significant hemorrhage and may remain in a state of compensated shock until up to 40% of their blood volume is lost. Pediatric patients maintain their

- cardiac output during compensated hypovolemic shock by which of the following mechanisms?
- Increasing their blood pressure by endogenous catecholamine secretion
 - Increasing their stroke volume through vasoconstriction
 - Increasing their respiratory rate
 - Increasing their heart rate
 - Increasing cardiac contractility by endogenous catecholamine secretion
7. An 8-year-old boy was hit by a car while riding his bicycle. On arrival to the ED, he develops acute respiratory distress, hypoxia, and hypotension. After assessment reveals absent breath sounds over the right side of the chest, your immediate treatment for this child would be which of the following?
- Fluid resuscitation with isotonic solutions to increase venous return
 - Preparation for intubation and mechanical ventilation
 - Call for “stat” bedside chest radiograph
 - Call for surgical tube thoracostomy
 - Perform bedside needle thoracostomy on the affected side
8. A 24-month-old child is evaluated after falling approximately 10 feet from playground equipment. On ED evaluation, he has normal vital signs for age and no evidence of hypoxia. On chest radiograph, he is noted to have a small, isolated pneumothorax. The recommended ED management for this child is
- Chest thoracostomy and admission for 24-hour observation
 - ED observation for at least 6 hours with repeat chest radiograph and if clinically stable discharge with plans for follow-up within 24 hours
 - Mechanical ventilation to allow for quicker resolution of the pneumothorax
 - 100% FiO₂ by face mask and admission until resolution of pneumothorax
 - Discharge from the ED after the initial evaluation is comforting for a clinically stable child without signs of respiratory distress
9. A 10-year-old male presents after being stabbed in the back. He is alert and awake with moderate respiratory distress. His airway is patent and he is experiencing some improvement with 100% FiO₂ by face mask but still breathing at a rate of 40. Chest radiograph reveals 50% opacification of the left thorax. You suspect a hemothorax. Your next step in the care of this child is which of the following?
- Left-sided chest needle thoracostomy
 - Large bore chest tube thoracostomy on the left
 - Surgical chest thoracotomy
 - Establish IV access for fluid or blood product administration
 - Intubate and provide mechanical ventilation
10. A 6-month-old infant presents to the ED with respiratory distress. Chest radiograph reveals two nondisplaced rib fractures and a small pulmonary contusion. Detailed history taking does not reveal a significant mechanism of injury capable of causing these findings. Your management of this infant includes which of the following?
- Comprehensive physical exam looking for other signs of injury
 - Treating the immediate rib fractures and pulmonary contusion with oxygen as needed and pain medications
 - Admission for observation and early detection of clinical deterioration
 - Immediate reporting of suspicion of nonaccidental trauma (child abuse) to local law enforcement and child protective services.
 - All of the above

ANSWERS

- C. Hypoxia to this degree should represent lung injury, significant hemothorax or tension pneumothorax. The other injuries should not result in these vital signs.
- A. Traumatic arrest usually results from catastrophic injuries such as severe brain injury, high spinal cord injury, or severe hemorrhage and has a low survival rate.
- D. Any of these injuries are possible but a GSW to the lower chest has at least a 30–40 % chance of injuring abdominal contents.
- E. The chest wall is very compliant and the forces are transmitted to the internal organs.
- B. While all of these will be done, the first step is open the airway, and then support respiratory efforts with oxygen and BVM while preparing to use RSI for control of the airway. Other team members start the IV and call the trauma team.
- D. Cardiac output in young children is rate dependent due to their heart size. They can increase systemic vascular resistance but heart rate is the first compensatory effort.
- E. The child is exhibiting tension pneumothorax physiology and needs immediate treatment with needle thoracostomy on the affected side.
- B. Six hours of observation is the recommended time frame to watch for expansion of a small pneumothorax and for worsening of the patient’s clinical status.

9. D. The patient can have at least 40% of blood volume accumulate in the chest and initial hemoglobin levels may be misleading. Thus, start an IV, give blood, and place a large-bore chest tube. Consider using a cell saver to return the blood from the chest back to the patient.
10. E. Rib fractures are rare in children and require severe forces. Child abuse must be considered.

32 ABDOMINAL TRAUMA

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HIGH-YIELD FACTS

- Trauma is the most common cause of death in children and abdominal trauma is the most common unrecognized cause of fatal traumatic injury in children.
- Blunt trauma accounts for 85% of pediatric abdominal trauma. Blunt trauma from motor vehicle collisions causes more than half of the abdominal injuries seen in children and is the most lethal.
- Children are susceptible to different injury patterns than adults.
- Immediate stabilization and transfer of the most severely injured children to an appropriate trauma center when indicated will result in greatly improved outcomes.

PATTERNS OF INJURY

MOTOR VEHICLE COLLISIONS (TABLE 32-1)

- Multisystem trauma, along with abdominal injury, as demonstrated by Waddell's triad is common when an automobile strikes a child.
- Waddell's triad (Fig. 32-1) includes impact first to the upper leg, and then chest and abdomen, followed by head injury.

- The head and extremity components of Waddell's triad should not divert attention from the possibility of more serious intra-abdominal injury.
- In the hypotensive child with blunt trauma, always complete a thorough investigation for potentially more serious abdominal injuries.
- The "seat belt sign" is characterized by ecchymosis, abrasion, or erythema in the pattern of a lap belt across the abdomen (Fig. 32-2) and flanks (Grey-Turner sign) and has been associated with increased risk of gastrointestinal injury.
- The *lap belt complex* in the restrained child is a bursting injury of solid or hollow viscera, and disruption of the diaphragm or lumbar spine (Fig. 32-3).

BICYCLE CRASHES, SPORTS INJURIES, AND FALLS

- Abdominal injury can occur if a child bicyclist is impacted by the handlebars or falls to the ground.
- Following handlebar injuries (Fig. 32-4) children may show no serious sign of injury for hours to days after the impact.
- Mean elapsed time to onset of symptoms is almost 24 hours and as many as one-third are discharged home initially.
- Traumatic pancreatitis, often with pseudocyst formation, is the most common handlebar injury followed by injuries to the kidneys, spleen, and liver, duodenal hematoma, and bowel perforation.
- Abdominal CT scan and observation of children with suspected handlebar injuries is recommended.
- Sports-related trauma typically produces isolated organ injury because of a direct blow to the abdomen.
- The spleen, kidney, and gastrointestinal tract are particularly vulnerable following sports injury.

CHILD ABUSE

- Significant abdominal injury represents the second most common cause of death from abuse after head injury.
- The diagnosis can be obscured by the lack of external signs of trauma in up to one-half of these patients.
- Common patterns of abusive injury are to the liver and spleen with associated rib fractures.

TABLE 32-1. Patterns of Injury by Mechanism

WADDELL'S TRIAD	LAP BELT COMPLEX	FALL FROM A HEIGHT
Pedestrian mechanism in child	Restrained occupant in MVC	
Midshaft femur fracture	Blowout diaphragm injury	Head injury
Abdominal injury	Duodenal injury	Multiple long bone fractures
Head injury	Solid organ injury; "Chance fracture" of lumbar spine occurs as a result of hyperflexion	Chest wall injury

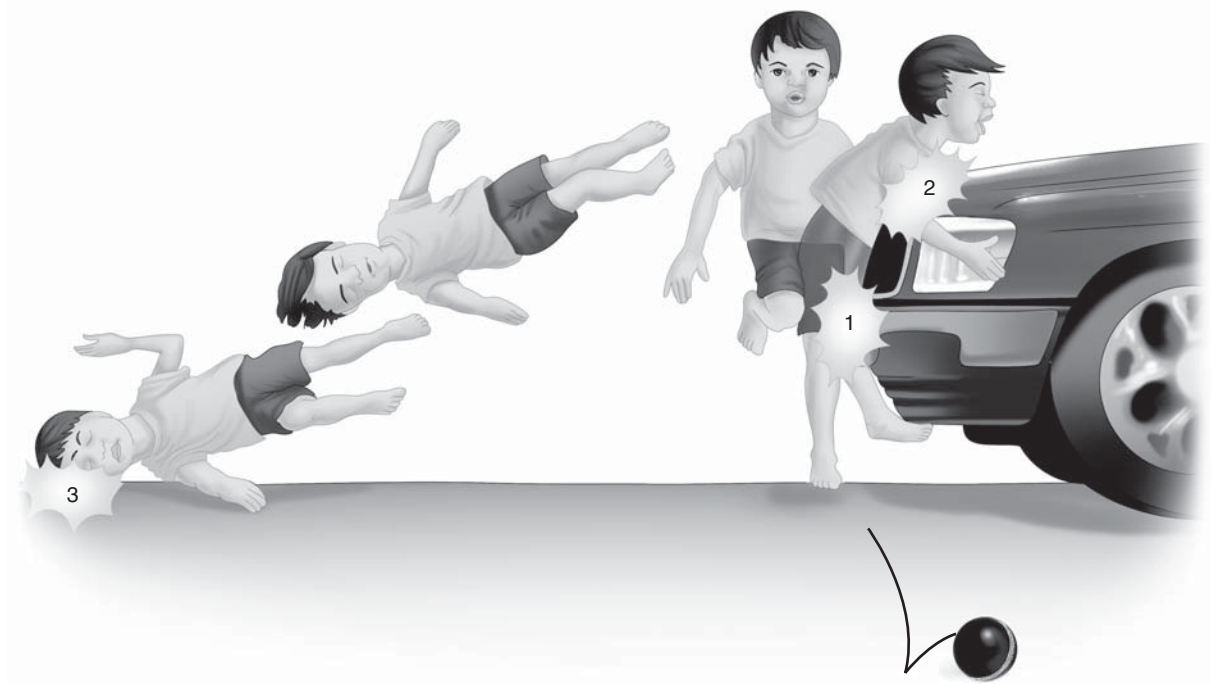


FIG. 32-1. Waddell's triad. (With permission from Susan Gilbert)

PATHOPHYSIOLOGY

- Diagnosis of major intra-abdominal hemorrhage may be delayed because children can maintain normal blood pressure and pulse rate, even with significant blood loss.
- Abdominal distention, due to gastric distention from crying and air swallowing, can confound the examination by masking serious abdominal injury or bleeding.
- Severe gastric dilation can result in respiratory compromise by interference with diaphragm motion, gastric aspiration, or vagal dampening of the tachycardic response.
- In children, the primary response to decreased cardiac output is increased heart rate; therefore, vagal dampening and hypovolemia can lead to circulatory collapse.

MANAGEMENT

GENERAL PRINCIPLES

- The basic principles of trauma evaluation and resuscitation should be followed in all cases of abdominal trauma.
- Evaluation of the abdomen is included in both primary and secondary surveys.
- Insert a nasogastric or orogastric tube, decompress the stomach, and check for blood/ bile.

- Insert an orogastric tube if there is any suspicion of head trauma or basilar skull fracture.
- Place a urinary catheter to check for blood and urinary retention, if there is no gross blood at the meatus, and obtain a urinalysis.
- Complete a rectal examination to check for blood, prostate position in males, and rectal tone.
- Do not let the child eat or drink due to the possibility of surgery or development of paralytic ileus.
- Obtain blood for type/cross-match, electrolytes, CBC, amylase, and liver transaminases.
- Log roll the patient to inspect the posterior torso for additional wounds.
- Designate a team member to take care of the child's emotional needs and to comfort them through trauma evaluation and treatment. Many pediatric trauma centers have instituted policies on family member presence for trauma and pediatric resuscitation.

PENETRATING ABDOMINAL TRAUMA

- The hollow organs are most commonly injured as a result of penetrating trauma, followed by the liver, kidney, spleen, and major vessels.
- Surgical evaluation, wound debridement, and possible exploration, along with broad-spectrum intravenous antibiotics are necessary.



FIG. 32-2. Seat belt sign. Ecchymosis, abrasion, and/or erythema across the anterior abdominal wall because of seat belt in a motor vehicle collision associated with intra-abdominal injury, disruption of the diaphragm, and chance fracture of the lumbar spine.



FIG. 32-3. Chance fracture of the lumbar spine. Because of improperly applied lap belt which rides up and compresses the child's abdomen during a motor vehicle collision. (Courtesy of Dr. James F. Holmes, Department of Emergency Medicine, University of California, Davis Medical Center.)

- Location, size, and possible trajectory of entrance and exit wounds help to identify potential underlying injuries.
- The following should be performed when there has been significant penetrating abdominal trauma:
 - Place a nasogastric or orogastric tube and a urinary catheter.
 - Upright posteroanterior chest radiograph with a lateral, if possible, supine, upright, and cross-table abdominal radiographs.
 - Obtain a CT scan of the abdomen with IV contrast for deep penetrating stab wounds and all gunshot wounds.
- Gunshot wounds to the abdomen result in high morbidity and mortality and requires immediate exploration result in high morbidity and mortality.
- Stab wounds pose the greatest threat to blood vessels. Commonly injured vessels include the aorta, inferior vena cava, the portal vein, and hepatic veins.
- Conservative management with 12–24 hour observation can be entertained in the patient with no sign



FIG. 32-4. Bicycle handle bar injury associated with pancreatic trauma. (Courtesy of Dr. James F. Holmes, Department of Emergency Medicine, University of California, Davis Medical Center.)

TABLE 32-2. Radiographic Clues in Abdominal Trauma

A ground glass appearance of the abdominal cavity may suggest intraperitoneal blood or urine
Medial displacement of the lateral border of the stomach, as evidenced by the nasogastric tube, suggests splenic laceration or hematoma as the enlarged spleen pushes the stomach aside
Obliteration of the psoas shadow or renal outline and fracture of the lower ribs suggest renal trauma
Bleeding from the short gastric vessels gives the fundal mucosa a “saw tooth” appearance
With nasogastric tube in place, the relative lack of gas in the distal small intestine suggests a duodenal or proximal jejunal hematoma
Air injected via the nasogastric tube may increase the chance of detecting a pneumoperitoneum indicative of perforated viscus

of shock or peritonitis, no blood in the stomach, rectum, or urine, no free abdominal or retroperitoneal air on x-ray.

BLUNT ABDOMINAL TRAUMA

- Both isolated abdominal and multisystem trauma present challenges in the pediatric patient because information is inherently difficult to obtain.
- Multiple other injuries may overshadow often subtle early abdominal findings and the physical examination may be accurate only 55–65% of the time.
- For the emergency physician, the key to management is suspecting the diagnosis and obtaining appropriate studies and consultation.
- In children, minor mechanisms, such as falling 2 ft to the ground from a hammock, can result in significant splenic injury with minimal symptoms.
- Laboratory and radiologic studies may be necessary depending on clinical status, mechanism of injury, and suspicion for injury on physical examination.
- Radiographs of the chest (supine or preferably upright posteroanterior plus a lateral) and supine abdomen and pelvis can give important clues to the diagnosis of abdominal injury (Table 32-2).

LABORATORY EVALUATION (DIAGNOSTIC STUDIES)

- The child with blunt trauma is at high-risk for intra-abdominal injury if any of the laboratory or physical examination findings listed in Table 32-3 are present.
- If the initial hematocrit is <30% with other signs of impending shock, this suggests significant hemorrhage.
- An initial hematocrit <24% is associated with high mortality, and transfusion should be initiated.
- A persistently distended abdomen after nasogastric tube placement, hemodynamic instability not immediately responsive to fluid resuscitation, recurrent

TABLE 32-3 Clinical Findings Predictive of Intra-Abdominal Injury in Children with Blunt Trauma

ALT >125 or AST >200 (U/L)
Urinalysis >5 RBCs/hpf
Abdominal tenderness
Hematocrit <30%
Femur fracture

hypotension, or signs of peritoneal irritation warrant immediate surgical intervention by a surgeon experienced in pediatric abdominal injuries.

COMPUTED TOMOGRAPHY

- Indications for abdominal and pelvic CT scan are listed in Table 32-4.
- CT scan is useful for evaluation of the liver, kidney, spleen, retroperitoneum, and, to a lesser extent, gastrointestinal injuries.
- CT scan identification of pancreatic injury, diaphragm injury, and bowel perforation are much less sensitive and warrant a high index of suspicion with serial abdominal examinations to rule out occult injury.
- Radiation exposure is the greatest risk associated with CT scan.
- Abdominal CT scan carries a significantly high lifetime cancer mortality risk with radiation attributable risks from a single abdominal CT scan within the first and tenth years of life estimated at 1/550 and 1/700, respectively.
- Clinical decision rules are necessary to reduce variability in medical management by providing evidence derived guidelines for clinical care, thereby, decreasing unnecessary radiation.
- Oral contrast is rarely used in the trauma setting because of the technical difficulty of administration and increased waiting time before scanning, risk of aspiration, and apparent limited value because of frequent lack of bowel opacification.

DIAGNOSTIC PERITONEAL LAVAGE

- In the pediatric patient, close observation, serial physical examinations, and abdominal ultrasound and CT scans have replaced peritoneal lavage.
- Diagnostic peritoneal lavage (DPL) may still be useful if these other modalities are unavailable or the child must undergo immediate general anesthesia for other injuries. Under these circumstances, DPL can often be performed in the operating suite, when indicated.
- The usefulness of DPL remains questionable. It is neither organ-specific nor injury-specific, and can-

TABLE 32-4 Comparison of Techniques for Evaluation of Abdominal Trauma

	ABDOMINAL CT SCAN	ABDOMINAL ULTRASOUND
Indication	Relatively stable patient Multiple trauma or major thoracic, head, or orthopedic (pelvic) injury Physical findings or a mechanism suggesting possible abdominal injury Unexplained hypotension Hematuria (gross or microscopic >20 RBC/HPF), CNS injury, spinal injury, or mental status alteration precluding serial abdominal examination Declining hematocrit or unaccountable fluid and blood requirements	May be used as a triage tool and adjunct to the physical examination Evaluation of pancreatic injury and intra-abdominal fluid (presumably blood) May also be used to identify other intra-abdominal injuries when CT scan is not readily available.
Advantage	Relatively noninvasive High sensitivity and specificity Evaluates multiple organ systems simultaneously	Available at the bedside and more readily available than CT scan in some locales Can be used at the bedside for a FAST examination to evaluate for peritoneal fluid and blood
Disadvantage	Radiation risk 1/550 lifetime cancer mortality attributable to single CT scan of abdomen in first year of life Generally requires intravenous with or without oral contrast Time delay	Not as sensitive as CT scan

not reliably assess retroperitoneal injury, and the decision to operate for liver or splenic injuries is not based on the amount of intraperitoneal blood in children.

- The introduction of air and fluid into the abdomen and the resulting peritoneal irritation following DPL make subsequent radiographic and physical examinations more difficult.

ABDOMINAL ULTRASOUND

- Bedside ultrasound (US) is more readily available and has significantly reduced the need for DPL.
- US is particularly useful in the unstable patient as an immediate triage tool and adjunct to the physical examination.
- US is best used for detecting intra-abdominal injuries that require immediate attention (such as in the setting of hypotension) rather than for a definitive diagnosis.
- US is also useful when CT scan is not available and its greatest utility is in detecting intraperitoneal hemorrhage and pancreatic injuries.
- The use of bedside US has become part of the core emergency medicine curriculum and is often taught using the FAST method.
- The FAST examination evaluates up to six areas of the abdomen with the principal objective of identifying hemoperitoneum.
- Children who are hemodynamically unstable with abdominal trauma will require laparotomy regardless of the US and those that are stable are often managed nonsurgically even with abdominal organ injury.

SOLID ORGANS

SPLEEN

- The spleen ranks first among the solid abdominal organs susceptible to major hemorrhage and second only to the liver in lethal injury following blunt abdominal trauma.
- A right-sided blow to a pedestrian or fall can cause a contrecoup splenic injury.
- Mononucleosis, common in children, can result in splenic enlargement and predispose to splenic rupture with even mild impact.
- Findings typical with splenic injury are left upper quadrant abdominal pain, radiating to the left shoulder (Kehr's sign), associated with palpable tenderness on examination.
- Significant tenderness in the left upper abdomen and/or splenic enlargement should prompt surgical consultation and consideration of a CT scan.
- Persistent unexplained leukocytosis or hyperamylasemia also suggests splenic injury.
- Abdominal radiographs may incidentally reveal a medially displaced gastric bubble secondary to the enlarged spleen.
- Conservative management includes initial hospitalization for a few days of bed rest for grade 1 or 2 injuries and longer for higher grade injuries, followed by limited activity.
- Some splenic lacerations can be embolized to reduce bleeding and need for resection.
- Delayed spontaneous rupture can occur at any time and is most common on days 3–5. Children who

develop hypotension unresponsive to volume resuscitation require surgery.

- There is an increase in infection and lethal sepsis in children with splenectomy, particularly with encapsulated organisms (*Streptococcus pneumoniae*, *Haemophilus influenzae*, *Neisseria meningitidis*, *Staphylococcus aureus*, and *Escherichia coli*). All children should receive the pneumococcal vaccination if they have a splenectomy.

LIVER

- The liver ranks second among solid abdominal organs for major hemorrhage and significant injury, but it is the most common source of *lethal* hemorrhage.
- The majority of liver injuries in children are minor and remain undetected unless discovered incidentally by abnormal liver enzymes or imaging studies.
- Serum aspartate aminotransferase (AST) greater than 200U/L and alanine aminotransferase (ALT) greater than 125U/L have been associated with IAI (Table 32-3). Significant tenderness in the right upper abdomen and/or liver enlargement should prompt surgical consultation and CT scan evaluation.
- Children with liver injuries who are not in shock or who respond to volume resuscitation rarely require surgery to control bleeding.
- Conservative management includes careful monitoring of vital signs, serial abdominal examinations, and serial hematocrit measurements.
- Some lacerations can be embolized to reduce bleeding and need for resection.
- Large stellate liver lacerations and subcapsular hematomas that have eroded through Glisson's capsule rarely stop bleeding without surgery.
- Hepatic resection and biliary tree drains are rarely indicated, and direct suturing and drainage can manage most hepatic lacerations.
- In preparation for surgery, circulating blood volume should be restored since rapid hemorrhage can occur during surgery as blood clots are evacuated during repair.

PANCREAS

- The pancreas is vulnerable to a direct blow to the upper central abdomen as seen with bicycle handlebar injury (Fig. 32-4).
- Pancreatic injuries are difficult to diagnose and signs such as elevated amylase and lipase may take up to 72 hours postinjury to present.

- Traumatic pancreatitis without major pancreatic injury is common, followed by pancreatic hematomas and, rarely, transection of the body or duct.
- Pancreatic transections can result in pancreatic pseudocyst formation within 3–5 days and chronic intermittent attacks of abdominal pain, nausea, vomiting, and weight loss.
- Acutely, the leakage of pancreatic fluid into the lesser peritoneal sac causes a chemical peritonitis and pancreatic ascites.
- The classic triad of epigastric pain radiating to the back, a palpable abdominal mass with or without acute peritonitis or ascites, and hyperamylasemia are rare in children.
- Elevated serum amylase may indicate pancreatic injury; its absence does not rule it out.
- Simple traumatic pancreatitis is treated similarly to other types of pancreatitis with bowel rest, nasogastric suction, intravenous fluids, and pain medication.
- Severe pancreatic injury will typically require surgical drainage with repair or partial resection of the pancreas.
- Pancreatic pseudocyst treatment involves 6–8 weeks of total parenteral nutrition followed by a surgical drainage procedure.

ABDOMINAL WALL

- Hematomas of any of the abdominal wall muscles can occur, as well as concomitant injury to the spine and other skeletal structures.
- The psoas muscle is particularly susceptible to hematoma, even with minor trauma, in patients with a bleeding diathesis such as hemophilia, or those on warfarin.
- Ecchymoses (such as the “seat belt sign”) are indicative of intra-abdominal injury and the onset may occur several hours after the trauma.
- Grey–Turner sign is an ecchymosis in the abdominal or flank area and may represent a retroperitoneal hematoma.
- Cullen's sign is a bluish discoloration around the umbilicus and may represent an intraperitoneal hemorrhage.
- Abdominal wall injuries are typically self-limited but determination of deeper injury can be difficult, so a low threshold for abdominal CT scan is warranted.
- At discharge patients and caregivers should watch for vomiting, increasing pain, abdominal distention, hematuria, and fever.
- Assure close follow-up for re-examination within 24 hours for any significant abdominal wall injury.

TABLE 32-5 Summary

- Blunt abdominal trauma is proportionally more common in children and results in more injuries and deaths than penetrating trauma. However, penetrating trauma is far more lethal as a sole injury.
- Management of pediatric abdominal trauma requires a coordinated effort between the emergency physician, trauma surgeon, and pediatric referral center.
- The spleen and liver are the most commonly injured organs as a result of blunt abdominal trauma. Liver injuries constitute the most common cause of death.
- Care must be taken not to let the head and extremity components of Waddell's triad divert attention from the more subtle findings of intra-abdominal injury that may include life-threatening hemorrhage.
- Computed tomography (CT) scan has eliminated much of the difficulty surrounding the diagnosis of abdominal injuries and is the procedure of choice for stable pediatric trauma patients. However, CT scan is not without risk and must be used judiciously using the ALARA standard (as low as reasonably achievable).
- The focused abdominal sonography for trauma (FAST) ultrasound examination evaluates up to six areas of the abdomen with the principal objective of identifying hemoperitoneum. The FAST examination is less sensitive in children than adults.
- Perforations of the duodenum and proximal jejunum are the most common intestinal injuries and are usually associated with lap belt or bicycle handlebar injury.

HOLLOW ORGANS

- Perforations of the duodenum and proximal jejunum are the most common hollow organ injuries and often associated with a lap belt or bicycle handlebar injury.
- Without obvious evidence of free air on radiographs, the diagnosis of a perforated viscus in blunt trauma can be difficult.
- Tenderness may initially be localized and slowly worsen over 6–12 hours, accounting for the time necessary for peritonitis or obstruction to occur.
- Abdominal CT scan is not particularly sensitive for these injuries, and repeated physical examinations remain the most reliable indicator of enteric disruption.
- Treatment of perforated abdominal viscus is laparotomy to repair the injury.
- Intramural hematomas of the duodenum or jejunum can cause symptoms of intestinal obstruction with pain, bilious vomiting, and gastric distention.
- The diagnosis can be made with US or upper GI series, which reveals the coiled spring sign.
- This problem rarely requires surgery. It may cause traumatic pancreatitis with involvement of the ampulla of Vater. Treatment is conservative and supportive including nasogastric suction and parenteral nutrition for up to 3 weeks.
- When a large abdominal wall defect is present, evisceration can be prevented by keeping the bowel moist with saline-soaked gauze and not allowing the patient to assume a dependent position that would increase edema of the bowel wall.

SUMMARY

- Evaluation and treatment of children with suspected abdominal trauma is challenging (see Table 32-5). Physiologic characteristics of children make vital signs and physical examination less predictive of

serious injury than in adults and other diagnostic clues such as mechanism of injury and maintaining a high suspicion for common injuries are paramount.

- An awareness of useful diagnostic tests such as abdominal CT scan and their risks and limitations is also important.
- When treating the multisystem traumatized child, a systematic approach will lead to identification of less obvious injuries within the abdomen.
- It is important to identify resources for treatment of pediatric trauma well in advance.
- The ability to provide definitive care in an efficient manner through trauma teams or expeditious transfer to a trauma center optimizes the chances of survival and limitation of morbidity for the injured child.

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QUESTIONS

- Waddell’s triad highlights focused attention on which of the following during the trauma evaluation of a pediatric patient?
 - Head, neck, and chest injuries
 - Head, torso, and femur injuries
 - Abdominal, chest, and back injuries
 - Head, neck, and upper extremity injury
 - Head, neck, and abdominal injuries
- A 5-year-old falls off a bicycle and is found to have a “handlebar sign”. Which of the following is true of pancreatic injuries in children?
 - Abdominal CT scan is usually diagnostic immediately postinjury
 - The triad of epigastric pain radiating to the back, palpable abdominal mass, and hyperamylasemia are common
 - Lipase elevations may not be seen until 72 hours postinjury
 - Amylase elevations occur early postinjury
 - Can be identified on ultrasound
- A 7-year-old male presents to the ED 24 hours after falling off a bicycle with diffuse abdominal tenderness and bilious emesis. He was brought into the ED yesterday immediately after the injury and had a negative abdominal CT and was discharged home. Which of the following is true regarding this patient?
 - Upright plain film of the abdomen is the initial study of choice.
 - Splenic injury is the most likely cause of this patient’s presentation.
 - Hepatic injury is the most likely cause of this child’s presentation.
 - Repeat abdominal CT is almost 100% sensitive.
 - Renal injury is the most likely cause of this child’s presentation.
- A 3-year-old female is belted in the rear seat during a high-impact motor vehicle collision. Which of the following best describes the *lap belt complex* in the restrained child?
 - Is a bursting injury of solid or hollow viscera, and disruption of the diaphragm or lumbar spine
 - Involves shoulder and pelvic injury
 - Occurs in 30% of restrained children
 - Includes injury to the cervical spine
 - Includes head injury
- A 10-year-old male falls off his bicycle and is hit in the abdomen by the bicycle handlebars. Which of the following is true regarding this type injury?
 - The most common handlebar injury is injury to the spleen.
 - Mean elapsed time to onset of symptoms is 2 hours.
 - Traumatic pancreatitis, often with pseudocyst formation, is a very rare handlebar injury.
 - Following handlebar injuries children may show no serious sign of injury for hours to days after the impact.
 - The majority of patients with handlebar injury are discharged home initially.
- A 7-year-old child falls from a second story balcony. The patient is at high-risk for intra-abdominal injury if which of the laboratory or physical examination findings are present?
 - An initial hematocrit <40%.
 - An initial hematocrit <35%.
 - ALT > 125 (U/L) or AST > 200 (U/L)
 - Urinalysis >2 RBC/hpf
 - Humeral fracture
- A 2-year-old child is a restrained passenger in a roll-over motor vehicle collision. Which of the following is true of findings on x-ray of the abdomen regarding this patient?
 - A ground glass appearance of the abdominal cavity may be seen with a shattered kidney with extravasation of urine.
 - Medial displacement of the lateral border of the stomach, as evidenced by the nasogastric tube, suggests hepatic laceration.
 - Relative lack of gas in the small intestine suggests a hepatic injury.
 - “Saw tooth” appearance of the fundal mucosa is a result of gastric perforation.
 - Obliteration of the psoas shadow suggests splenic injury.
- During a baseball game, a 9-year-old boy is hit in the torso with a baseball bat. He presents with severe abdominal tenderness and distension 7 hours postinjury. Placement of a nasogastric tube can provide diagnostic evidence by which of the following methods?
 - Inability to pass the nasogastric tube suggests hepatic injury
 - Relative lack of gas in the distal small intestine suggests a duodenal or proximal jejunal hematoma.

- C. Injection of air into the nasogastric tube is contra-indicated and has no diagnostic value.
- D. Placement of a nasogastric tube is contra-indicated in this setting.
- E. Injection of air into the nasogastric tube increases the likelihood of detecting a splenic injury.
9. During the evaluation of a 10-month-old that fell off a changing table, an ED physician decides to obtain a bedside FAST ultrasound rather than an abdominal CT. Which of the following is true in this setting?
- A. Ultrasound is more sensitive than CT
- B. Ultrasound is more specific than CT
- C. Ultrasound is good at detection of intraperitoneal blood
- D. CT is a more cost-efficient modality
- E. Risk of lifetime cancer mortality attributable to CT is 1/5000
10. A 14 year-old male is both punched and stabbed in the abdomen. Which of the following statements regarding abdominal trauma is accurate?
- A. Penetrating trauma is more common than blunt trauma in older children and adolescents.
- B. Penetrating trauma is more lethal than blunt trauma.
- C. Splenic injuries constitute the most common cause of death.
- D. Transfer of care of this child to a pediatric referral center is not indicated.
- E. Pancreatic injuries constitute the second most common cause of death.
3. A. Delayed presentation posthandle bar injury in the setting of an initial negative abdominal CT suggests bowel perforation. Abdominal CT is highly sensitive for hepatic, splenic, and renal injuries. As such, upright plain film would be indicated here and would demonstrate free air under the diaphragm.
4. A. The lap belt complex in the restrained child (bursting injury of solid or hollow viscera, and rarely disruption of the diaphragm or lumbar spine) is characterized by ecchymosis, abrasion, or erythema in the pattern of a lap belt (“seat belt sign”) across the abdomen (Fig. 32-2) and flanks (Grey–Turner sign) and occurs in up to 10% of restrained children.
5. D. Following handlebar injuries (Fig. 32-4), children may show no serious sign of injury for hours to days after the impact. Mean elapsed time to onset of symptoms is almost 24 hours and as many as one-third are discharged home initially. Traumatic pancreatitis, often with pseudocyst formation, is the most common handlebar injury followed by injuries to the kidneys, spleen, and liver, duodenal hematoma, and bowel perforation.
6. C. If the initial hematocrit is <30% with other signs of impending shock, this suggests significant hemorrhage. An initial hematocrit <24% is associated with high mortality, and transfusion should be initiated.

TABLE 32-3

ALT >125 or AST >200 (U/L)
Urinalysis >5 RBCs/hpf
Abdominal tenderness
Hematocrit <30%
Femur fracture

7. A. Discussed in Table 32-2 below:

TABLE 32-2 Radiographic Clues in Abdominal Trauma

A ground glass appearance of the abdominal cavity may suggest intraperitoneal blood or urine
Medial displacement of the lateral border of the stomach, as evidenced by the nasogastric tube, suggests splenic laceration or hematoma as the enlarged spleen pushes the stomach aside
Obliteration of the psoas shadow or renal outline and fracture of the lower ribs suggest renal trauma
Bleeding from the short gastric vessels gives the fundal mucosa a “saw tooth” appearance

8. B. With nasogastric tube in place, the relative lack of gas in the distal small intestine suggests a duodenal or proximal jejunal hematoma obstructing the passage of air distally. Air injected via the nasogastric tube may increase the chance of detecting a pneumoperitoneum indicative of perforated viscus (Table 32-2).

ANSWERS

1. B. Waddell’s triad (Fig. 32-1) seen in the pediatric pedestrian struck by a motor vehicle involves direct impact to the upper leg, and then chest and abdomen, followed by head injury (as child impacts with the bumper, is flung onto the hood, and then falls to the ground).
2. C. Following trauma to the pancreas amylase and lipase elevations may not be seen until 72 hours postinjury. Pancreatic injuries are difficult to diagnose and signs such as elevated amylase and lipase may take up to 72 hours postinjury to present. The classic triad of epigastric pain radiating to the back, a palpable abdominal mass with or without acute peritonitis or ascites, and hyperamylasemia are rarely detected in children. Ultrasound is not sensitive enough to identify injury to the pancreas (Fig. 32-4).

9. C. See the table below comparing the two diagnostic modalities:

CT	ULTRASOUND
Relatively noninvasive	Available at the bedside and more readily available than CT scan in some locales
High sensitivity and specificity	Can be used at the bedside for a FAST examination to evaluate for peritoneal fluid and blood
Evaluates multiple organ systems simultaneously	Not as sensitive as CT scan
Radiation risk 1/550 lifetime cancer mortality attributable to single CT scan of abdomen in the first year of life	
Generally requires intravenous with or without oral contrast time delay	

10. B. See the table below:

- Blunt abdominal trauma is proportionally more common in children and results in more injuries and deaths than penetrating trauma. However, penetrating trauma is far more lethal as a sole injury.
- Management of pediatric abdominal trauma requires a coordinated effort between the emergency physician, trauma surgeon, and pediatric referral center.
- The spleen and liver are the most commonly injured organs as a result of blunt abdominal trauma. Liver injuries constitute the most common cause of death.

33 GENITOURINARY TRAUMA

Joyce C. Arpilleda

INTRODUCTION

Genitourinary (GU) tract injuries occur in 10% of abdominal trauma patients. The kidney is the most commonly injured organ in the urinary tract. Blunt trauma accounts for 80–95% of all renal injuries. The most common cause of blunt trauma is motor vehicle collisions, that is, rapid deceleration. Sexual and physical abuse should be considered in patients with perineal injuries. Diagnostic studies used in genitourinary trauma are outlined in Fig. 33-1.

INITIAL ASSESSMENT AND MANAGEMENT

- Begin with the basics of advanced trauma life support.
- Kidneys may be sources for major bleeding in hypovolemic shock.

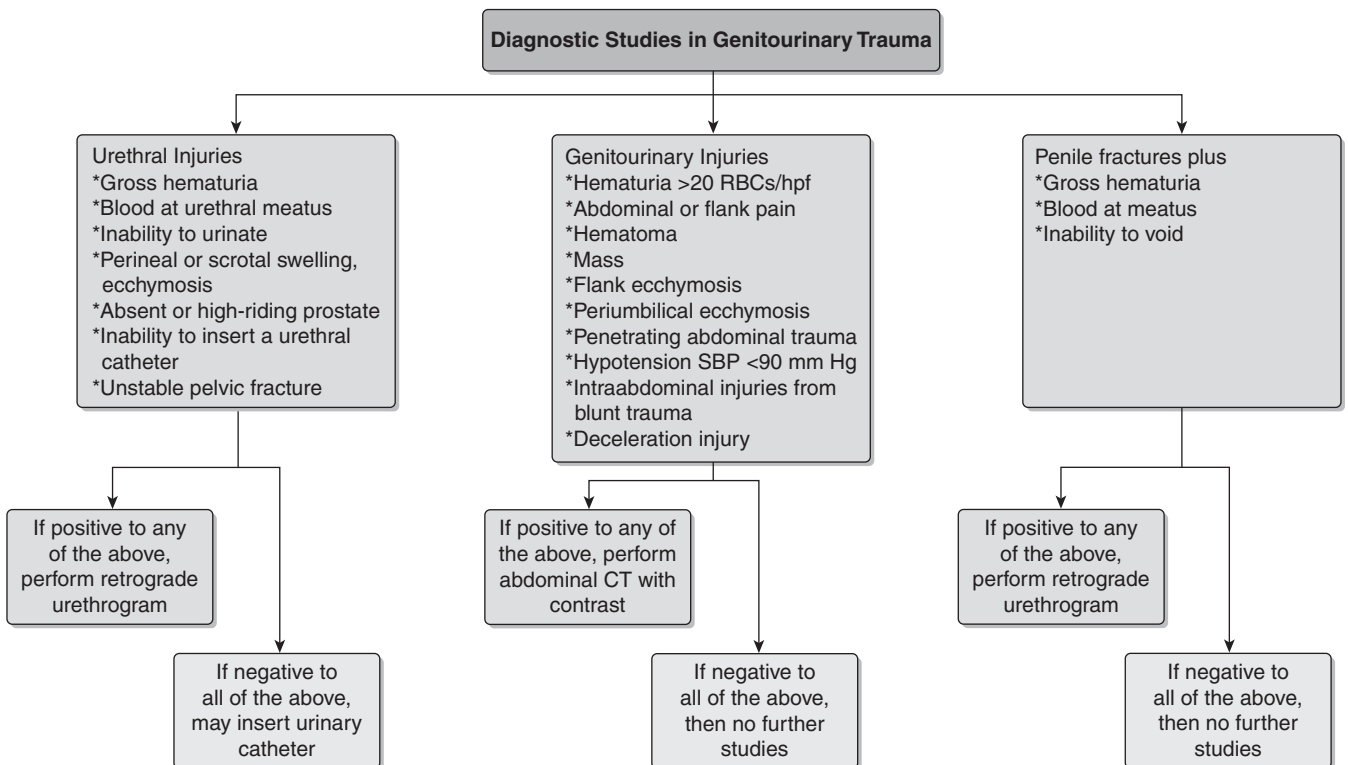


FIG. 33-1. Diagnostic studies in GU trauma.

TABLE 33-1 Initial Assessment and Management of Genitourinary Injuries

1. Inspect the back for signs of blunt and penetrating injuries and internal bleeding
2. Perineal assessment:
 - a. Contusions and hematomas
 - b. Lacerations
 - c. Urethral bleeding
 - d. Rectal examination is performed before placing a urinary catheter, i.e., a high-riding prostate is suspicious for a urethral disruption
3. Vaginal assessment:
 - a. Blood in the vaginal vault
 - b. Vaginal lacerations

- Failure to diagnose and delay in treatment of urologic injuries can lead to significant morbidity.
- Table 33-1 shows the initial assessment and management.
- Urine dipstick analysis is an initial screening test for hematuria; if positive for blood, perform microscopic urinalysis.
- Hematuria may be absent.
- Table 33-2 lists indications for further evaluation.
- Signs of GU trauma in a pelvic plain film are
 - loss of psoas shadow
 - scoliosis
 - lower rib or transverse process fractures.
- Monitor urinary output.

RENAL INJURIES

- Hematuria present in more than 75–95% of cases of renal trauma.
- Hematuria may be absent in:
 - ureteropelvic junction injuries,
 - thrombosis of the renal artery,
 - disruption of the renal pedicle,
 - in penetrating trauma, renal vessels or the ureter may be severed without hematuria.

TABLE 33-2 Indications for Further Genitourinary Evaluation

1. Gross or microscopic hematuria >20 RBCs/hpf in children
2. Abdominal or flank pain. The flank is the area between the anterior and posterior axillary lines from the sixth intercostal space to the iliac crest
3. Hematoma
4. Mass
5. Flank ecchymosis (Gray Turner sign)
6. Periumbilical ecchymosis (Cullen sign)
7. Penetrating trauma that can injure the GU system

- Indications for further evaluation are gross or microscopic hematuria with:
 - penetrating abdominal trauma,
 - hypotension with systolic blood pressure less than 90 mm Hg,
 - other intra-abdominal injuries from blunt trauma,
 - rapid deceleration injury.
 - CT scanning is the gold standard.
- The focused assessment sonography for trauma (FAST) scan cannot differentiate between blood, extravasated urine, and other types of free fluid.
- Unstable patients or major penetrating injuries to the kidneys with extravasation usually require immediate surgery.
- 95% of blunt renal injuries can be treated non-operatively.
- Most children with blunt renal trauma respond to rapid crystalloid fluid resuscitation, and then admission to the intensive care unit.
- Classification of renal injuries is shown in Table 33-3.

URETERAL INJURIES

- Less than 1% of all GU traumas.
- Penetrating trauma is the most common mechanism of injury.
- Traumatic avulsion of ureter in children due to blunt trauma.
- Most are hypotensive.
- Gross or microscopic hematuria is present.
- Hematuria absent for complete ureteral transection or adynamic segment of ureter.
- CT scan with contrast is the best initial imaging study, followed by a second scan about 10 minutes after contrast injection.
- Ureteral transection is treated with ureteropyelostomy.

BLADDER INJURIES

- 10% of those with pelvic fractures have concomitant bladder injury.
- Bladder contusions are self-limited.
- Bladder rupture requires surgical consultation due to high mortality rate, which increases with delay in diagnosis.
- Bladder rupture presents with gross hematuria, blood at the urethral meatus, inability to void, or little to no urine flow on catheterization.
- Perform cystography if bladder rupture is suspected.
- Perform retrograde urethrogram before catheterization and cystography if suspecting urethral injury.

TABLE 33-3 American Association for the Surgery of Trauma Grading System for Renal Injury

GRADE ^a	TYPE OF INJURY	DESCRIPTION OF INJURY	TYPE OF INJURY	DESCRIPTION OF INJURY
I	Contusion	Microscopic or gross hematuria; urologic imaging studies normal	Hematoma	Subcapsular, nonexpanding without parenchymal laceration
II	Hematoma	Nonexpanding perirenal hematoma confined to retroperitoneum	Laceration	<1 cm parenchymal depth of renal cortex without urinary extravasation
III	Laceration	>1 cm parenchymal depth of renal cortex or medulla without collecting system rupture or urinary extravasation		
IV	Laceration	Parenchymal laceration extending through renal cortex, medulla, and collecting system	Vascular	Main renal artery or vein injury with contained hemorrhage. Segmental infarctions by thrombosis, dissection or laceration of segmental arteries.
V	Laceration	Completely shattered kidney. Uteropelvic junction avulsion.	Vascular	Avulsion or thrombosis of renal hilum main renal artery or vein which devascularizes kidney

^aAdvance one grade for bilateral injuries up to grade III.

URETHRAL INJURIES

- Traumatic urethral injuries occur in about 10% of patients with pelvic fractures.
- Usually from blunt trauma
- Indications for a retrograde urethrogram before inserting a urinary catheter are:
 - gross hematuria,
 - blood at the urethral meatus,
 - inability to urinate,
 - perineal or scrotal swelling and ecchymosis, for example, butterfly hematoma,
 - absent or high-riding or boggy prostrate on rectal exam,
 - inability to insert a urethral catheter, and
 - unstable pelvic fracture.
- Consider suprapubic catheter.
- Early urologic consultation.

SCROTAL TRAUMA

- Scrotal trauma can cause testicular or appendage torsions, testicular dislocation, epididymitis, hematocele, hematoma, pyocele, hydrocele, and testicular rupture.
- Suspect testicular or epididymal rupture when there is a recurrence of pain and delayed onset of scrotal swelling from hours to 3 days after the injury.
- Suspect testicular dislocation when there is an empty hemiscrotum after trauma.
- Evaluate with Doppler ultrasound.
- Evaluate significant straddle injuries with a pelvic radiograph.
- Delay of treatment for 4–6 hours may result in loss of testis.
- Early urologic consult.
- If discharged home, recommend scrotal support and cold packs.

PENILE INJURIES

- Obtain urinalysis in significant penile injury.
- Retrograde urethrography is recommended.
- Repair superficial lacerations as in any other laceration.
- In zipper injuries, use bone cutters or similar device to cut the bridge of the sliding piece of the zipper.
- Evaluate scrotum if large hematocele or rupture of tunic albuginea is present.

- Consult urology for penile fractures.
- Consider scrotal ultrasound or cavernosonography in penile fractures.
- Obtain retrograde urethrogram if penile fractures are accompanied by gross hematuria, blood at the meatus, and inability to void.
- Tourniquet injuries from hair may present as balanitis, paraphimosis, or cellulitis
- Treatment of tourniquet injuries from hair usually involves removal of the band of hair and treating any infection, with urology consultation if deeper injury is suspected.

VULVAR AND VAGINAL INJURIES

- Confirm an intact urethra with a retrograde urethrogram before inserting a urinary catheter if there is gross hematuria; blood at the urethral meatus; inability to urinate; perineal swelling and ecchymosis; inability to insert a urethral catheter; or unstable pelvic fracture.
- A disrupted urethra may require suprapubic tube placement.
- Screen significant straddle injuries with pelvic radiography.
- Minor injuries are treated with rest and cold packs.
- Large or expanding vulvar hematomas may require surgical drainage and are susceptible to secondary infection.

PEDIATRIC SEXUAL ABUSE

- Evaluate the risk of child sexual abuse in all cases of genital trauma.
- Consider child abuse when the history or reported mechanism does not match the injuries.
- All developmentally capable patients with suspected sexual abuse should be referred early on for formal forensic interview.
- Perform a screening exam.
- Majority of those who have disclosed sexual abuse or sexual assault have normal exams.
- Absence of genital findings does not exclude sexual abuse.
- Progestin-only regimen, or plan B (levonorgestrel) for postexposure prophylaxis within 5 days of sexual contact.
- Involve child protective services.
- Injuries to the hymen or vagina are unusual in accidental genital trauma.

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QUESTIONS

- A 9-year-old boy is brought to the emergency department after falling off his bicycle. He is alert and cooperative on presentation. His vital signs and examination are normal except for periumbilical ecchymosis. Urinalysis reveals microscopic hematuria of 40 RBC/HPF. Appropriate evaluation and management should include:
 - Diagnostic peritoneal lavage
 - Repeat urinalysis in 2 hours
 - Abdominal CT scan
 - Discharge home
 - Outpatient follow-up
- A 10-year-old female is brought to your emergency department with seizures after being knocked down by her horse. She has been intubated and paralyzed by EMS. Despite multiple intravenous fluid boluses, she remains hypotensive. Upon examination, you note flank ecchymosis. Urine microscopy shows an RBC count of 0 per HPF. You suspect that this isolated injury is:
 - Parenchymal contusion of the kidney
 - Vascular pedicle injury
 - Ruptured bladder
 - Transection of the right ureter
 - Hematuria.
- After being in an ATV rollover, an 11-year-old girl landed on a fallen tree on the ground, sustaining vaginal lacerations from a straddle injury. Initially, she appears stable. Which of the following radiography results require further renal evaluation?
 - Femoral fracture
 - Possible torticollis on cervical spine films
 - Pneumomediastinum on chest-xray
 - Pelvic fracture
 - Tibia fracture.
- A 12-year-old boy was riding his mountain bike going fast downhill. At some point, he flew into the air with his bicycle but landed upright and continued to ride. At the end of the day he complains of pain over his genitalia. There is contusion of the penis and blood at the urethral meatus. The most helpful diagnostic test is:
 - Ultrasonography
 - The ability to pass a urinary catheter
 - CT scan
 - Pelvic radiographs
 - Retrograde urethrography.
- A 13-year-old boy complains of pain after being kicked in the scrotum by his brother. Examination reveals swelling over the left hemiscrotum and exquisite tenderness over the left testicle. Which test will be most helpful?
 - Retrograde urethrography
 - Ultrasonography
 - Intravenous pyelogram
 - CT scan
 - Urinalysis.
- A 14-year-old boy has suffered a scrotal injury after playing flag football. There is a small bruise on the scrotal sac. Appropriate management includes:
 - Magnetic resonance imaging
 - Ice packs and scrotal support
 - Operative surgical exploration
 - Drainage of the ecchymosis under local anesthesia
 - Retrograde urethrogram.
- A 14-year-old girl is brought to your ED following a sexual assault. There are no complaints of pain.

- The genitourinary exam is normal. Appropriate management includes:
- Involvement of Child Protective Services
 - A pelvic examination under sedation
 - Urethral catheterization
 - Placement of a suprapubic tube
 - Warm water soaks.
- A 5-year-old boy is brought to your emergency department after having problems with his zipper. There is a small abrasion on his penis. Appropriate management includes:
 - Intravenous antibiotics
 - Operative surgical exploration
 - Supportive care
 - Urethral catheterization
 - Placement of a suprapubic tube.
 - A 3-year-old girl is brought to your emergency department after a straddle injury. She is not cooperative for the exam. A limited examination shows significant tenderness and profuse bleeding from her vaginal area. Appropriate management includes:
 - Laceration repair under local anesthesia
 - Supportive care and warm soaks
 - Sitz baths
 - Urethral catheterization
 - Operative surgical exploration of the wound.
 - A 15-year old girl is brought to your emergency department after landing on a rock. She has an enlarging hematoma on her vulva. Appropriate management includes:
 - Surgical drainage
 - Placement of a suprapubic tube
 - Involvement of Child Protective Services
 - Ice packs and vulvar support
 - Ultrasonography.
- with hematuria and would be unlikely to cause shock. Transection of the ureter would also be unlikely to produce shock.
- D. Abnormalities on C-spine films, CXRs, or long bone films do not necessarily suggest renal injury. A pelvic fracture, but not a femur fracture, would suggest possible renal injury.
 - E. Retrograde urethrography is the test of choice for diagnosing a urethral injury. Passing a urinary catheter is contraindicated in patients with suspected urethral trauma. Pelvic radiographs, CT scan, and ultrasonography are not adequate.
 - B. Ultrasonography is the test of choice for diagnosing a testicular injury. Torsion can occur with direct injury to the scrotum. None of the other studies will identify injury to the testicle or its blood flow
 - B. With minor isolated injuries of the scrotal sac, ice packs and scrotal support are sufficient. Surgical exploration is indicated when there are large testicular hematomas or suspicion for a testicular rupture. Drainage or an MRI is not indicated. Retrograde urethrogram will help with urethral injuries.
 - A. In the case of a minor in which a sexual encounter occurred, one of the most important management is contacting Child Protective Services.
 - C. A zipper injury to the penis has been reported. If the history does not match the injury, however, sexual abuse should be considered. Treat penile abrasions as with any other abrasion. Supportive care is indicated for this injury. There is no role for the other interventions listed.
 - E. The patient has a potential for a significant laceration, which may be revealed with an optimal examination. A laceration involving deeper structures may be revealed with an examination under anesthesia and repaired in the operating room. Warm soaks and Sitz baths are indicated for minor abrasions or lacerations. Urethral catheterization is contraindicated at this point because the possibility of urethral injury has not been eliminated on a limited examination in an uncooperative child.
 - A. Large or expanding vulvar hematomas may require surgical drainage and are susceptible to secondary infection. If the history does not match the injury, however, inquire about the possibility of a sexual assault. Ultrasonography is the test of choice for diagnosing a testicular injury. There is no role for the other interventions listed.

ANSWERS

- C. Gross or microscopic hematuria > 20 RBCs/hpf in children warrants further diagnostic imaging. A diagnostic peritoneal lavage does not exclude a retroperitoneal injury. CT scan is the current study of choice in a stable patient. Only doing a repeat UA does not identify the injury and D and E are incorrect.
- B. Absence of hematuria does not exclude renal injuries. Given the significance of the fall, the presenting symptoms and signs, and the absence of hematuria, suspect a vascular pedicle injury. Parenchymal contusions are likely to produce hematuria. A ruptured bladder usually presents

34 MAXILLOFACIAL TRAUMA

Joanna York

Stephen A. Colucciello

INTRODUCTION

Accurate bony alignment is important in the growing child, and missed fractures or inappropriate treatment may result in permanent facial deformity. A child with severe maxillofacial injury requires a team approach involving emergency physicians, pediatricians, general surgeons, maxillofacial specialists, and radiologists. Emergency specialists must recognize and prioritize injuries, manage the airway, stabilize the patient, read initial radiographs, and make appropriate consultations.

INCIDENCE

- Maxillofacial trauma in children more often results in soft tissue injury than facial fractures.
- The pediatric population contributes to less than 15% of all facial fractures, and facial fractures are especially rare below the age of 5.
- Fracture site distribution is influenced by the growth and change of the pediatric skeleton. In early childhood, the skull is particularly prominent, whereas the face and mandible are small. For this reason, in children younger than 5 years, orbital and frontal skull fractures predominate, whereas in older children, maxillary and mandibular fractures become more prominent.
- The most frequently fractured bones are the nose (45%), mandible (32%), orbit (17%), and zygoma/maxilla (5%).

ETIOLOGY

- Motor vehicle crashes, including auto/pedestrian incidents, are the most frequent cause of facial injury.
- Maxillofacial injuries due to child abuse have a high incidence of associated head and neck bruising and skull fractures are particularly common.

ASSOCIATED INJURIES

- Up to 55% of seriously injured children with facial trauma also have intracranial injury, a much higher percentage than occurs with adults.
- Cervical spine fractures are rare in children younger than 8 years. If a cervical spine injury is suspected

by radiography or clinical presentation, it is recommended that one should get a CT scan.

- If the patient is unresponsive with a neurological deficit, magnetic resonance imaging (MRI) is the study of choice within 72 hours of admission and has been shown to be cost-effective.

EMERGENCY MANAGEMENT

- The most urgent complication of facial trauma is airway compromise, which is most often associated with mid- and lower face injury.
- Simple maneuvers, such as chin-lift jaw thrust, oropharyngeal suctioning, and oral or nasal airway, when appropriate provide immediate benefit.
- Infants younger than 3 months are obligate nose breathers, and nasal or midface trauma can lead to complete airway obstruction. Mandibular fractures can result in loss of support of the tongue and occlusion of the upper airway. These fractures may also produce hematomas of the floor of the mouth, which can displace tongue and obstruct the airway. In this situation, the physician should open the mouth and pull the tongue forward, either manually or with a large suture or towel clip.
- Avoid emergency cricothyroidotomy in children younger than 12 years
- Percutaneous transtracheal jet ventilation is an excellent temporizing measure in these situations.

HISTORY

- Obtain as much history as possible in regards to the mechanism, loss of consciousness, any current visual problems, pain, and facial anesthesia.

PHYSICAL EXAMINATION

- Carefully and systematically palpate the entire face, evaluating for swelling, ecchymosis, deformity, asymmetry, trismus, malocclusion, and a traumatic Bell's palsy.

EYES

- Detailed eye exam is critical, paying specific attention to assess pupillary light reflex, presence of a hyphema, subconjunctival hemorrhage, proptosis, enophthalmos, function of extraocular muscles, and palpation of the entire orbital rim.
- Raccoon's eyes secondary to basal skull fracture usually occur 4 to 6 hours after a traumatic event,

whereas direct trauma to the periorbital region may result in more immediate bruising.

- Telecanthus suggests that the medial canthal ligaments are torn or underlying bone is avulsed from the nasal orbital complex.

EARS

- Examine the pinna for the presence of subperichondrial hematoma.
- The ear canal must be examined for lacerations and cerebrospinal fluid (CSF) leak.
- A basilar skull fracture is suggested by ecchymosis over the mastoid area (battle sign), and the presence of hemotympanum.
- Tympanic membrane rupture may occur with mandibular condyle fractures.

MIDFACE

- Carefully palpate the zygomatic arches for symmetry and evaluating for flattening caused by a fracture.
- Intraoral palpation of the arch is helpful in detecting a minimally displaced fracture.

NOSE

- Carefully palpate the nose for crepitus and deformity, as edema may obscure bony anatomy.
- Examine the inside of the nose for septal hematoma, which may be recognized by a bluish, bulging mass on the septum, or by the subjective impression of an abnormally wide septum. Pressure with a cotton swab will detect the presence of a soft, doughy swelling.
- With any significant facial trauma, it is important to assess for CSF rhinorrhea.

INTRAORAL AND MANDIBULAR EXAMINATION

- Injury of the inferior orbital nerve or inferior alveolar/mental nerve produces anesthesia of the upper or lower lip, respectively.
- The emergency physician must observe movement of the patient's jaw through a full range of motion. Difficulty in jaw movement, malocclusion, and trismus all suggest injury to the mandible, temporomandibular joint (TMJ), or zygoma.
- Stress the mandible with lateral and medial pressures on the dental arches, and subsequently apply up and

down manual pressure to test for bony disruption. An older cooperative child can be asked to bite down on a tongue blade.

FACIAL LACERATIONS

- Key to evaluating facial lacerations is an understanding of the relationship between the skin and the underlying vital structures including the facial nerve, the lacrimal duct apparatus, the parotid gland, and Stenson's duct.
- Injuries to the medial third of the upper or lower eyelids may result in lacrimal apparatus disruption.
- Facial nerve injury will result in paralysis on the ipsilateral side.
- Facial nerve injuries must be surgically repaired if they are posterior to a vertical line drawn through the lateral canthus. Injuries anterior to such a line are usually not repaired.

IMAGING STUDIES

- Have a low threshold for obtaining a CT scan in a seriously injured child who you suspect facial fractures. Frequently the history and physical examination are compromised and modern CT scan images provide excellent detail of the midface structures, cranium, and mandibular condyle.

NASAL FRACTURES

- Nasal fractures are the most commonly encountered pediatric facial fracture, accounting for up to 50% of the total.
- For optimal repair of displaced nasal fractures, consultation should take place within 5 to 6 days postinjury, after which time fractures begin to unite and manipulation becomes increasingly difficult.
- If the diagnosis is unclear, the child should be rechecked in 3 to 4 days after swelling has subsided to reassess for deformity or septal deviation.
- It is critical that emergency physicians recognize and treat septal hematomas. An untreated septal hematoma results in collapse of the septum and a "saddle" deformity of the nose due to septal cartilage necrosis.
- To drain a septal hematoma, the physician should anesthetize the area and then use a No. 11 blade to make an L-shaped incision through the mucoperiosteum along the floor of the nose and extend the incision vertically. The hematoma will then be evacuated through the flap (Fig. 34-1). Subsequent packing of the nasal antrum prevents reaccumula-

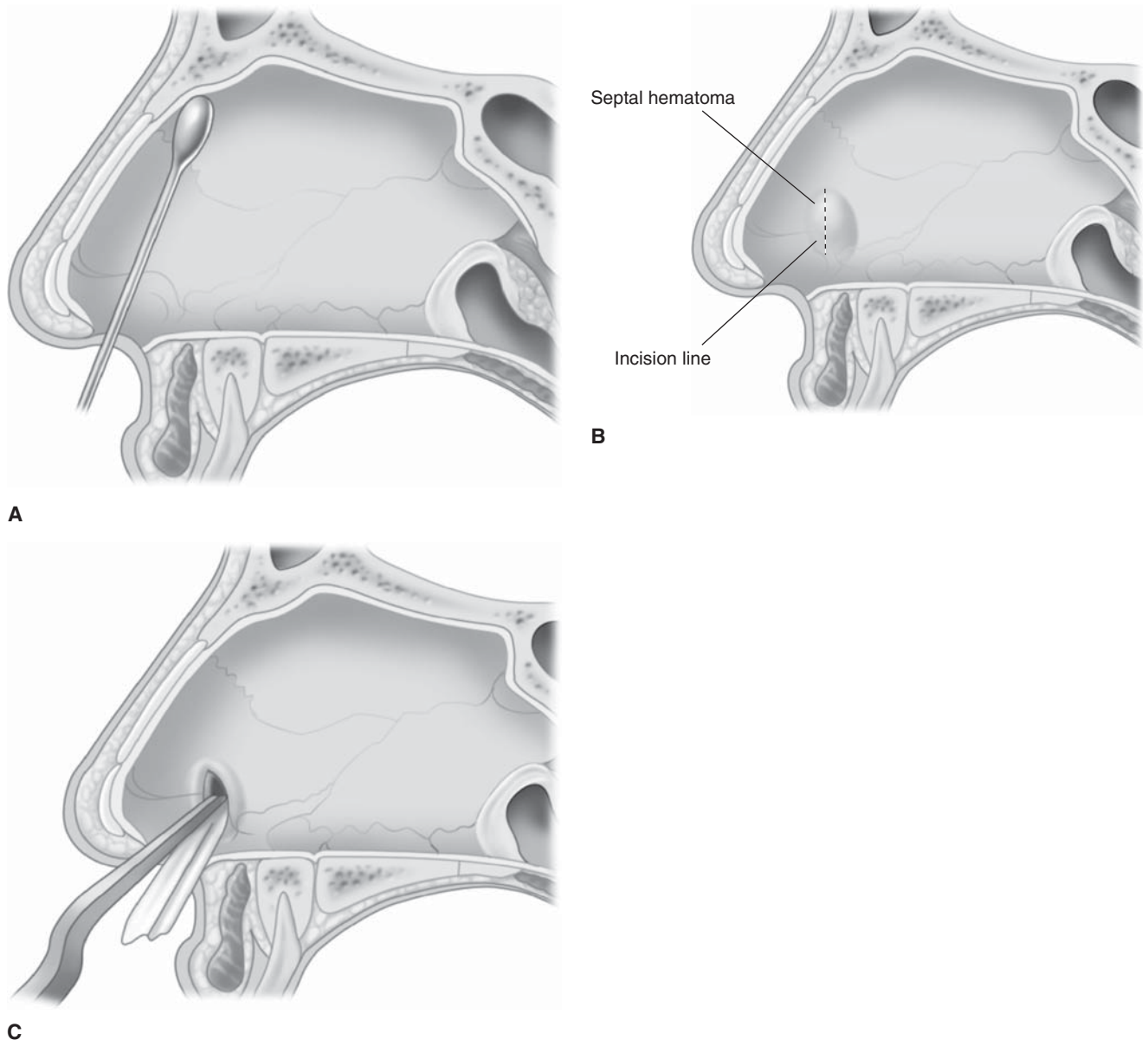


FIG. 34-1. How to drain and pack a nasal hematoma.

tion, and the child must be referred to the appropriate specialist.

NASAL–ETHMOIDAL–ORBITAL FRACTURES

- These injuries are rare in children.
- Nasal–ethmoidal–orbital (NEO) fractures occur when the bony structures of the nose are driven backward into the intraorbital space, presenting with telecanthus on exam.

ORBITAL FRACTURES

- The most common orbital fracture is the blowout fracture, which occurs when a blunt object, often a ball or fist, strikes the globe.
- The intraorbital pressures increase suddenly and contents decompress through the orbit, most commonly the floor. This may lead to entrapment of the inferior ocular muscles, with subsequent diplopia on upward gaze.
- Urgent consultation is required in the presence of exophthalmus or extraocular muscle entrapment.

- A facial CT scan can evaluate for entrapment and blowout fractures.
- Patients with NEO or orbital fractures should be instructed not to blow their nose.

FRONTAL SINUS AND SUPRAORBITAL FRACTURES

- Exophthalmus and ptosis may be present with impairment of upward gaze.
- The *superior orbital fissure syndrome* results in paralysis of extraocular muscles, ptosis, and anesthesia in the ophthalmic division of the trigeminal nerve.
- The *orbital apex syndrome* is a combination of the superior orbital fissure syndrome plus optic nerve damage and results in blindness.

MALAR FRACTURES

- The zygomatic arch itself is frequently fractured in isolation. If a zygomatic complex fracture is without displacement, diplopia, or sensory deficits, it may be managed by observation.
- The malar complex is often broken in a tripod fashion, with fractures at the infraorbital rim, across the zygomatic–frontal suture and along the zygomatic–temporal junction. Inward displacement of this fragment may result in impingement upon the mandible, giving rise to impaired mouth opening and trismus.

LEFORT FRACTURES

- LeFort I is a transverse fracture that separates the hard palate from the lower portion of the pterygoid plate and nasal septum. Traction on the upper incisors produces movement of only the hard palate and dental arch.
- LeFort II or pyramidal fracture separates the central maxilla and palate from the rest of the craniofacial skeleton. Mobilization of the upper incisors will move the central pyramid of the face, including the nose.
- LeFort III, also known as craniofacial dysjunction, separates the facial skeleton from the rest of the cranium

MANDIBLE FRACTURES

- Mandible fractures are the second most common facial fracture, following nasal bone injury.
- The most frequently injured areas are the condyle (70%), followed by the body, angle, and symphysis

- Facial CT scan has become the standard of care for suspected mandible fracture in children less than 15 years. In teenagers 16 years and older, oral pan tomogram (panorex) and posteroanterior mandible radiographs are appropriate initial tests.

SOFT TISSUE INJURIES

- Emphasis must be placed on irrigation, foreign body removal, and cosmetic approximation of important landmarks such as the vermilion border of the lip and margins of the eyebrows.
- Hematomas of the pinna must be relieved: Otherwise a chronic deformity of cauliflower deformity of the ear may result. Drain hematomas of the external ear by either needle aspiration or formal incision, and then apply a pressure dressing to prevent reaccumulation.
- Repair of lacerations to the salivary duct or to the lacrimal drainage system must be performed by a specialist. These repairs are achieved over a stent.

PAIN MANAGEMENT AND ANESTHESIA

- Local anesthesia is the primary method of achieving immediate pain control. Nerve blocks rely on the deposition of anesthetic solutions in areas of nerve blocks and are an especially helpful technique. The most useful maxillofacial nerve blocks in acute injury are mental, inferior alveolar, infraorbital, and supraorbital.
- These procedures require patient cooperation, and in the uncooperative child local wound infiltration becomes the procedure of choice.¹⁶ In the very uncooperative child, procedural sedation may be necessary.
- Systemic analgesia, either enteral or parenteral, is often necessary for adequate pain control in the acute setting. Dose-appropriate use of nonsteroidal anti-inflammatory agents, opioids, or a combination of the two will be required for pain relief. In cases of lengthy repairs, implementation of procedural sedation may be necessary for adequate patient and physician comfort and safety.

CONCLUSIONS

- Maxillofacial trauma in children more often results in soft tissue injury than facial fractures.
- When fractures do occur, associated injuries, particularly intracranial, may be present. Fractures heal rapidly over 2 to 3 weeks, and repair must be undertaken before bony union occurs.
- Conservative management is often the rule.

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QUESTIONS

- A 6-year-old boy was hit in the nose with a baseball at T-ball practice. On exam there is significant swelling to the nasal bridge. He is able to breathe out of his nose without difficulty. His parents want an imaging study to know if his nose is broken. Which of the following is the best management option?
 - Order sinus radiographs
 - Order a facial CT
 - Consult ENT to come in and evaluate the patient
 - Order plain nasal radiographs
 - Educate parents that no radiographs are needed in the ED
- What is the most common pediatric facial fracture?
 - Mandible
 - Orbit
 - Nose
 - Maxilla
 - Zygoma
- A 5-year-old boy presents after being hit in the face accidentally with a wooden board by his older brother while playing. He has soft tissue swelling and ecchymosis over his left cheek and inferior orbit. What study is the definitive diagnostic test for facial fractures in this patient?
 - MRI
 - Facial radiographs
 - Clinical exam
 - Facial CT
 - Bone scan
- A 4-year-old girl fell approximately 6 feet off a slide at school and is brought to the ED by EMS in full spinal precautions. There was no LOC, she was ambulating at the scene, and currently is following commands and moving her arms and legs on the board. She has multiple abrasions on the face and arms but no facial fractures noted. What is the most appropriate way to clear her cervical spine?
 - C-spine radiographs
 - CT of her C-spine
 - MRI of her C-spine
 - Take off her collar, palpate for tenderness, and observe her neck motion
 - Extension/Flexion radiographs of her C-spine
- A 7-year-old is brought into the ED by his parents. He had an unwitnessed fall while playing in the neighborhood. His mother states he has had difficulty answering questions. On exam, he has hemotympanum bilaterally. Hemotympanum is associated with which fracture?
 - Orbital blowout fracture
 - Basilar skull fracture
 - LeFort II fracture
 - LeFort I fracture
 - Mandible fracture
- What is the most appropriate treatment of a hematoma located on the pinna?
 - Incision and drainage followed by application of a pressure dressing
 - Nothing, it will absorb on its own and draining it increases the risk of infection in the cartilage
 - No treatment but discharge on prophylactic oral cephalixin
 - Incision following by suturing of the wound
 - Warm compresses and a pressure dressing
- A child is hit in the eye with a tennis ball and you are concerned about an orbital blowout fracture. Which of the following exam findings is most suggestive of entrapment of ocular muscles?
 - Crepitus upon palpation of the orbital rim
 - Limited upward gaze
 - A septal hematoma
 - Battle's sign
 - Hyphema
- Which of the following is typically not repaired by an emergency physician but by a specialist?
 - Hematoma of the pinna
 - Lip laceration crossing the vermilion border
 - Disruption of the lacrimal apparatus

- D. Septal hematoma
E. Large facial laceration on an infant
9. A 6-year-old was an unrestrained passenger in a MVC and has sustained significant injuries to her mid-face and jaw. Initially her GCS was 10 but she now has a GCS of 6 and needs to be intubated. There is significant swelling and blood in her oral pharynx inhibiting your ability to intubate and perform effective bag and mask ventilation. The most appropriate option for managing her airway is
A. Insert a LMA
B. Perform a needle cricothyroidotomy
C. Perform a cricothyroidotomy
D. Call ENT
E. Try a nasal intubation
10. An 8-year-old boy is brought into the ED after being punched in the nose by another child. The nasal bridge is very swollen and tender. He also is tender to palpation over the medial right orbit. You notice telecanthus on the right side, which is suggestive of:
A. Zygoma fracture
B. LeFort I fracture
C. Torn medial canthus ligaments
D. Basilar skull fracture
E. Mandible fracture
- If nothing is done, there could be a cosmetically disfiguring result.
7. B. As the orbital floor fractures, the orbital contents including the inferior rectus muscles can become trapped in the fracture leading to limited upward gaze.
8. C. Disruption of the lacrimal apparatus is associated with injuries to the medial third of the upper and or lower eyelids. These injuries are typically repaired by an ophthalmologist. All of the other injuries listed could be repaired by the emergency physician.
9. B. In children under the age of 12, a needle cricothyroidotomy with jet insufflation is the temporizing method of choice. The other options will not be successful if routine intubation and bag and mask ventilation are not possible. Cricothyrotomy is not the procedure of choice in the young child.
10. C. Telecanthus, an increased width between the medial canthi of the eyelids, with flattening of the medial canthus, is associated with nasal ethmoidal injury. In this situation, the medial canthal ligaments are torn or underlying bone is avulsed from the nasal orbital complex. The other fractures listed would not present with telecanthus.

ANSWERS

- E. Emergent imaging is not necessary in the ED as long as the patient is able to breathe. Patient should be seen within 5–6 days when the swelling is decreased but healing is not so far along as to make a reduction difficult.
- C. Nasal fractures account for 45% of pediatric facial fractures, followed by mandible, orbit, maxilla, and zygoma.
- D. Though facial fractures may be evident in other imaging modalities, a facial CT provides the best information in regards to facial fractures.
- D. C-spine injuries are very rare in children under the age of 8. Nothing by history or physical exam of this patient suggests a C-spine injury requiring imaging and exposing her to radiation. Her cervical spine can be cleared clinically.
- B. A basilar skull fracture is associated with hemotympanum. The other facial fractures usually do not result in a hemotympanum.
- A. If not drained, the hematoma will cause a permanent deformity in the cartilage known as cauliflower ear. If a pressure dressing is not applied after drainage, the hematoma will reaccumulate.

35 ORTHOPEDIC INJURIES

Greg Canty

INTRODUCTION

Orthopedic injuries are one of the most common reasons for pediatric visits to the emergency department (ED) with more than 40% of boys and 25% of girls experiencing a fracture before the age of 16. These patients often present diagnostic dilemmas because of how the growth plate affects injury patterns and radiographs as the skeleton matures. Proper recognition and diagnosis allows for optimal management and prevention of possible complications.

THE IMMATURE SKELETON

- Growing bone is unique because it contains a physis, or growth plate, that is typically the weakest link surrounding each joint (see Fig. 35-1).

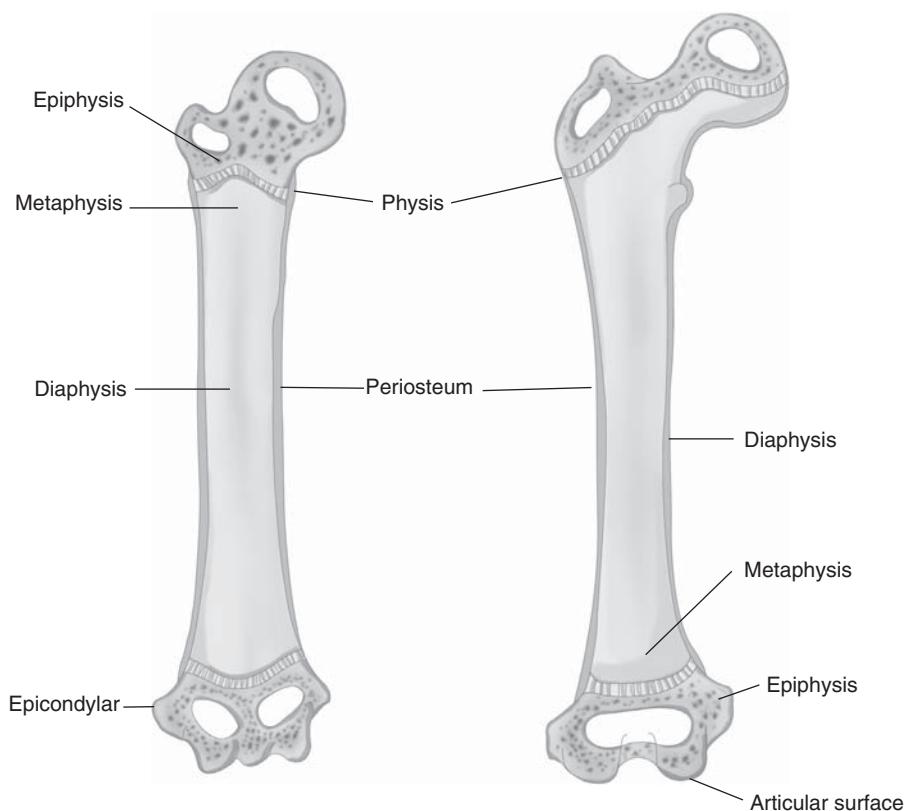


FIG. 35-1. Illustrations of the humerus and femur demonstrating specific features of the immature skeleton.

- The physis consists of radiolucent cartilage, which is not visible on radiographs, and each physis ossifies at different stages, which can lead to confusion.
- Remember there is a large portion of each bone that is cartilaginous and radiopaque, so the fracture fragment is often much bigger than it appears in radiographs.
- The growing bone is unique because of the physis and very active periosteum, which promotes faster healing and better remodeling than in adults, but this advantage can create problems if the fracture is not aligned properly or prompt follow-up is not arranged.
- The properties of the physis, along with a much more porous and flexible skeleton, lead to the many unique fracture patterns seen in pediatrics such as Salter–Harris classifications, the greenstick, the torus (buckle), and the bowing (plastic deformation) fractures.

TERMINOLOGY

- It is essential to speak the “language of orthopedics” to effectively communicate with consultants, and this requires knowledge of terminology (see Table 35-1), practice, and repetition.

- A good rule of thumb when communicating with consultants is to start with age, gender, mechanism, location, degree of soft tissue damage, and finish with an excellent radiographic description of the injury.

PHYSEAL INJURIES

- Over 20% of all injuries in children involve the physis, or “growth plate.”
- When properly recognized and treated most of these go on to heal well, but there is an increased risk for complications such as growth arrest, overgrowth, and malunion which should be communicated to the patient and family.
- The most widely used classification system for physeal fractures is the Salter–Harris (I–V) classification which provides guidance on management and outcomes (see Fig. 35-2).
- Most physeal fractures are Salter–Harris II, which can usually be managed nonoperatively with closed reduction if necessary.
- Salter–Harris III and IV fractures involve the articular surface and typically require surgical fixation for the

TABLE 35-1 Fracture Terminology**Anatomic location**

Epiphysis: articular end of each long bone; completely cartilaginous at birth (except distal femur); secondary ossification centers develop and replace cartilage over time

Physis (growth plate): cartilaginous structure between epiphysis and metaphysis responsible for longitudinal bone growth; injury may result in growth disturbance or arrest; weakest area of bone

Metaphysis: flared end of diaphysis adjacent to the physis; represents new bone; structurally weak area

Diaphysis: central shaft of long bone

Apophysis: nonarticular bony prominence where muscles or tendons attach; not directly involved in longitudinal growth but contribute to bony contour; exposed to significant traction forces

Fracture patterns

Avulsion: bony fragment pulled off by action of tendon or ligament

Transverse: fracture line at right angle to long axis of bone

Oblique: unstable fracture usually at about 30° to long axis of bone

Spiral: rotational, oblique fracture (usually due to torsion)

Comminuted: any fracture with more than two fracture fragments

Bowing (plastic deformation): bending of bone without complete fracture; most often seen in ulna or fibula

Torus (buckle fracture): compression of porous bone causes “buckle” near metaphysis

Greenstick: incomplete fracture when energy starts a fracture but cannot complete it; tension side (convex) fractures after bone bends beyond limits, but compression side (concave) bows into plastic deformation; most common fracture pattern in children

Pathologic: fracture through abnormal, weakened bone (i.e., tumors, osteomyelitis, cysts, inherited metabolic disorders)

Fracture descriptions

Alignment: refers to longitudinal relationship of one fragment to another

Displacement: deviation of fracture fragments from anatomic position; describe distal fragment in relation to proximal portion; varus displacement is toward midline of body; valgus displacement is away from midline of body

Angulation: the apex of the angle formed by the two fracture fragments; direction will be opposite to the displacement of distal fragment

Shortening: overlapping fracture fragments due to muscle contraction

Malrotation: refers to the rotational alignment of one fragment to another

Butterfly fragment: wedge-shaped fragment at apex of a fracture; caused by overload of both axial and angulation forces to the bone

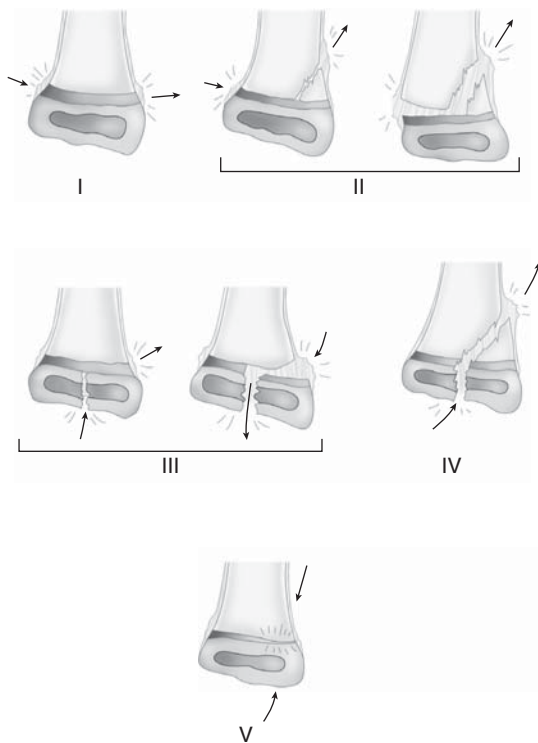


FIG. 35-2. Examples of physis injuries classified by Salter–Harris (I–V).

best outcomes. These fractures also carry a higher risk of growth deformities.

- Salter–Harris V fractures are usually unrecognized in the ED and are only diagnosed retrospectively with a history of an axial compression followed by growth arrest.

CLINICAL EVALUATION

- Since many orthopedic injuries are the result of trauma, always remember to first address life-threatening issues such as airway, breathing, circulation, intracranial, and internal injuries.
- Pain control, reassurance, and establishing patient confidence are keys to successful orthopedic evaluation.
- A thorough history is a key to establishing a mechanism of injury and is essential in detecting “red flags” that may suggest possible child abuse when the history is inconsistent with the injury pattern.
- Physical exam begins with complete exposure of injured and uninjured extremity for comparison.
- Examining closely for any swelling, deformity, or breaks in the skin suggestive of an open fracture. Always palpate joints above and below to determine point tenderness and range of motion. Neurovascular

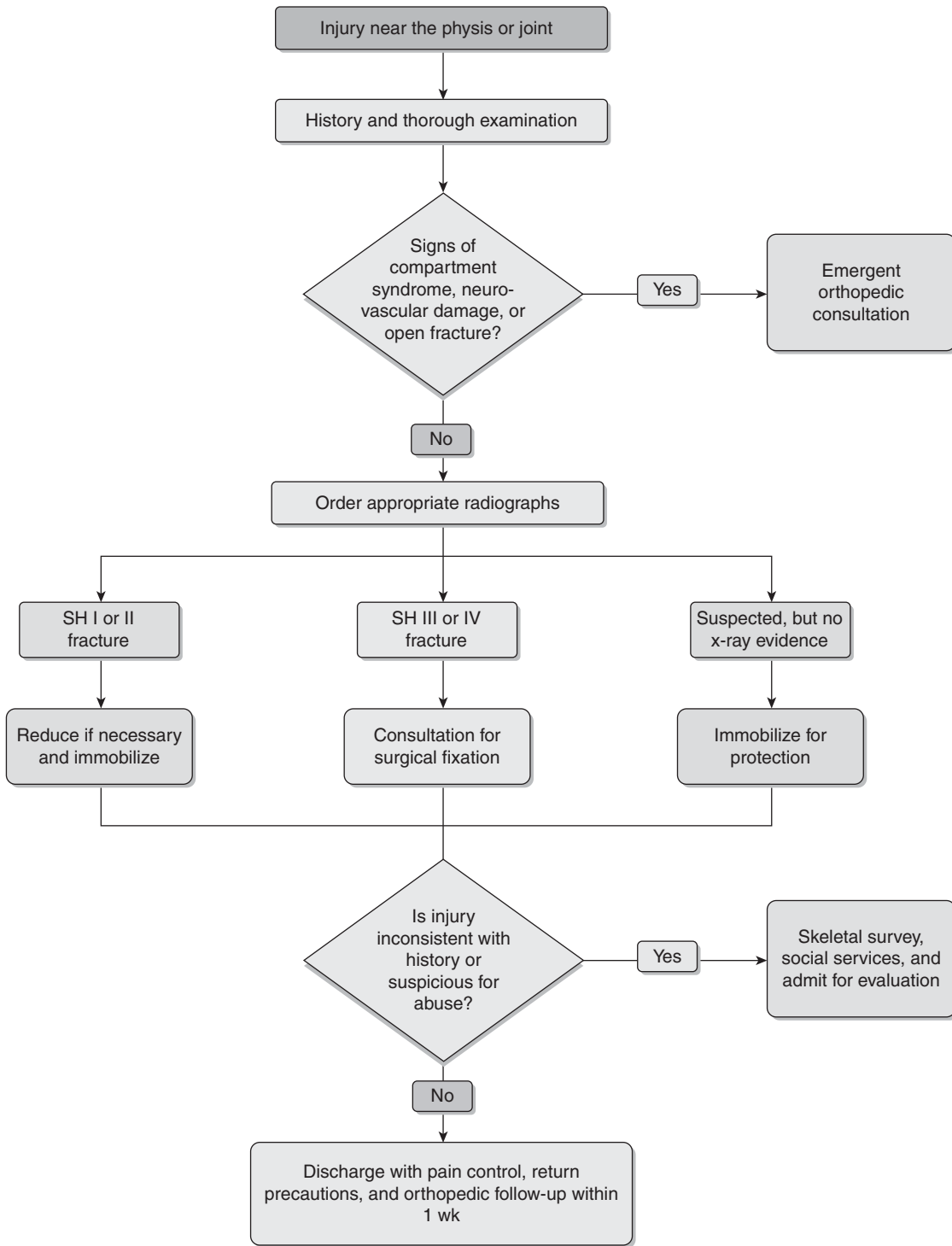


FIG. 35-3. Algorithm for the management of physis injuries in children.

assessments should be thorough and reassessed throughout the visit.

- Any abnormalities warrant appropriate radiographs which should include at least two views (anteroposterior and lateral) perpendicular to one another, and some injuries may occasionally benefit from an additional oblique view.
- Emergency physicians must insist on high-quality radiographs to make a proper diagnosis, and occasionally some dilemmas may be solved with comparison views of the uninjured extremity to determine pathology versus development.

MANAGEMENT

- Initial fracture management begins with establishing pain control and determining if management decisions are going to be operative versus nonoperative (see Fig. 35-3).
- Most pediatric fractures are managed nonoperatively with immobilization and often closed reduction.
- Fractures requiring operative management are usually Salter–Harris III or IV, tibia, femur, or open fractures.
- Beware of limb-threatening conditions such as compartment syndrome, which begin as pain out of proportion to exam findings, and may progress to pallor, paresthesias, and pulselessness.
- Open fractures need irrigation, antibiotics, and appropriate tetanus prophylaxis in the ED while awaiting orthopedic consultation.
- Proper immobilization provides protection, decreases risk for further displacement, and improves patient comfort. Immobilization may be in the form of a splint (plaster or fiberglass) or cast depending upon the nature of the injury.
- Immobilization should always take away the painful arc of motion and frequently requires immobilizing a joint above and below the injury.
- Ensure that all patients are discharged with proper pain control, cast/splint care instructions, return precautions, and follow-up plans with an orthopedist in 3–5 days.

FRACTURES FROM BIRTH TRAUMA

- Fractures seen within the first few weeks of life may be due to birth trauma and are often found accidentally on chest x-rays or in evaluation of a “bump” or “nonmoving extremity.”
- Most commonly associated fractures with birth trauma are the clavicle, femur, and humerus.
- If there is no history of a traumatic delivery or no callous formation is seen within 2 weeks, then investigate for possible nonaccidental trauma because healing from birth trauma should be rapid.

FRACTURES OF CHILD ABUSE OR NONACCIDENTAL TRAUMA

- Nonaccidental trauma is a major concern with pediatric patients because 25–50% of abused children will have a fracture and 10% of all injuries in children younger than 2 years are the result of nonaccidental trauma.
- The younger the patient with a fracture, the higher the suspicion should be for abuse.
- Injuries such as “corner” femur fractures, transphyseal distal humerus fractures, spiral fractures, and “bucket-handle” fractures always raise concerns about nonaccidental trauma, but recent evidence shows the most common fracture with abuse is the routine transverse fracture.
- The biggest clue is not the fracture type, but the often-underemphasized history, which should always raise concerns when it is inconsistent with the type or degree of injury seen on exam.
- Any suspicion of nonaccidental trauma in young children requires a skeletal survey and hospital admission for further evaluation.
- It is the law in all 50 states that physicians must report any concerns about nonaccidental trauma to the proper authorities for further investigation.

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QUESTIONS

1. A young girl falls and suffers a Salter–Harris II fracture of the distal radius. Which of the following part of the bone is most responsible for longitudinal growth in the immature skeleton?
 - A. Periosteum
 - B. Epiphysis
 - C. Metaphysis
 - D. Physis
 - E. Diaphysis
2. A 9-year-old male falls while skateboarding and sustains a both bone fracture of his forearm. Which of the following terms is best used to describe overlapping fracture fragments that result from muscle contraction?
 - A. Displacement
 - B. Angulation
 - C. Malrotation
 - D. Shortening
 - E. Alignment
3. A teenager jumps off the roof landing directly on his feet and complains of pain over his distal tibia after the axial compression injury. Which one of the following fracture types is of greatest concern even if radiographs are negative initially?
 - A. Salter–Harris I
 - B. Salter–Harris II
 - C. Salter–Harris III
 - D. Salter–Harris IV
 - E. Salter–Harris V
4. An 18-month-old child presents to the ED with multiple fractures after a “fall.” Which of the following most suggests possible nonaccidental trauma or abuse?
 - A. Spiral fracture of the tibia
 - B. Transverse fracture of the radius
 - C. “Corner” femur fractures
 - D. A history that the child had a fall on an outstretched hand from a standing position
 - E. All the above
5. A 6-year-old boy presents with a posteriorly angulated tibial fracture. In this patient the *earliest* sign of compartment syndrome you would anticipate is which of the following?
 - A. Pallor
 - B. Paresthesia
 - C. Pulselessness
 - D. Pain out of proportion to exam
 - E. Fracture blistering
6. Which of the following injuries would be expected to require surgical management for definitive care?
 - A. Salter–Harris I fracture of distal femur
 - B. Salter–Harris II fracture of distal fibula
 - C. Both bone forearm fracture of the radius and ulna
 - D. Salter–Harris III fracture of distal tibia
 - E. Torus (buckle) fracture of distal radius
7. A 1-month-old child presents to the ED for increased crying and decreased use of his right arm. Radiographs reveal a nondisplaced transverse fracture without any evidence of callous formation. The child's father states the injury is likely due to a traumatic birth. When would you expect callous formation to appear after a fracture from a traumatic birth?
 - A. 2 weeks
 - B. 3 weeks
 - C. 1 month
 - D. 6 weeks
 - E. 2 months
8. An 8-year-old boy presents with a both bone forearm fracture and is placed in a reverse sugar tong splint after reduction. Which of the following are reasons for placing a splint following reduction?
 - A. Decreases the risk for further displacement
 - B. Provides protection from further injury or accidental trauma
 - C. Improves patient comfort while decreasing pain
 - D. May allow for more swelling in acute injuries
 - E. All of the above
9. A 12-year-old boy sustains a Salter–Harris II of the distal radius with minimal displacement (<2 mm). After immobilization, when should he be seen by an orthopedist?
 - A. Immediate consultation in the ED prior to discharge
 - B. Within 3–5 days
 - C. Within 7–10 days
 - D. 2 weeks after injury for transition to a fiberglass cast
 - E. 6 weeks after injury for removal of splint and clearance
10. It is known that about 20% of pediatric fractures involve the physis, and this means there is a slight increased risk of growth arrest, overgrowth, or non-

union following the injury. Which Salter–Harris classification is the most common type of physal fracture?

- A. Salter–Harris I
- B. Salter–Harris II
- C. Salter–Harris III
- D. Salter–Harris IV
- E. Salter–Harris V

ANSWERS

1. D. The physis is the thin rim or radiolucent cartilage known as the growth plate and is the structure most responsible for longitudinal growth in the growing skeleton. It is situated between the epiphysis (the articular surface of the long bone) and the metaphysis (the flare between the physis and bone shaft). The shaft of the bone is referred to as the diaphysis. The periosteum is strong and thick in young bones, and it is responsible for the remarkable healing and remodeling that takes place in pediatric orthopedic fractures.
2. D. Shortening refers to two overlapped fracture fragments that result from muscular contraction forces following a complete fracture. Displacement refers to deviation of a fracture fragment from its anatomic position. Angulation is the apex of the angle formed by the two fracture fragments, and its direction is opposite to the displaced distal fragment. Malrotation refers to the rotational alignment of one fragment to another. Alignment refers to the longitudinal relationship between one fragment and the other.
3. E. Salter–Harris V fracture is most likely in this scenario due to the axial compression forces. These injuries may be suspected based on mechanism/history in the ED but radiographs are often negative with the final diagnosis being made retrospectively once growth arrest occurs. Salter–Harris I fractures should also be considered because they may also present with negative radiographs and only point tenderness in the ED, but the mechanism of axial/longitudinal compression suggests this scenario is much more likely a Salter–Harris V injury. Salter–Harris II, III, and IV injuries should all be detectable by abnormal radiographs and clinical exam at the time of presentation to the ED.
4. E. All of the answers listed should make one suspicious of nonaccidental trauma in the 18 month old described with multiple fractures. Remember the most suspicious finding with any pediatric fracture is a history that is not consistent with the findings on exam. A fall on an outstretched hand (FOOSH) from a standing position in an 18 month old should not result in multiple fractures though they may have an isolated upper extremity injury. Routine transverse fractures are the ones most commonly seen with nonaccidental trauma, but the other patterns are highly suspicious of abuse due to the mechanism required to create “corner” femur fractures and spiral tibia fractures.
5. D. Pain out of proportion to exam is the earliest sign of potential compartment syndrome and warrants orthopedic consultation in the ED. Pallor, paresthesia, and pulselessness are all later signs of compartment syndrome. Fracture blistering is not associated with compartment syndrome.
6. D. Remember that Salter–Harris III and IV fractures are intra-articular and typically require surgical management for definitive care. In pediatrics, most both bone forearm fractures can be successfully managed with closed reduction and immobilization. It is rare for Salter–Harris I or II fracture to require surgical management. A torus (buckle) fracture heals wonderfully with immobilization alone.
7. A. Fractures from a traumatic birth may be seen in the first few weeks following a traumatic birth, but healing is rapid with callous formation expected within 2 weeks. The most common fractures from traumatic birth are of the clavicle, femur, and humerus. The lack of callous formation 2 weeks after birth or an absent history of traumatic birth should lead one to consider investigation for nonaccidental trauma.
8. E. All of the above answers are reasons for placing a well-molded splint following an injury or reduction. Splinting is thought to allow some added protection over casting when dealing with the progressive swelling commonly seen after injuries.
9. B. Young patients with fractures need to be followed up for re-evaluation on a timely basis due to the rapid rate of healing. Once callous formation begins to take place, it is difficult to alter management plans. Due to the risk of further displacement or angular drift, most fractures should be seen within 1 week and preferably within 3–5 days.
10. B. Salter–Harris II is the most common type of physal injury and reportedly involves about 75% of all physal injuries. Salter–Harris I injuries are likely under-reported due to their benign appearance on radiographs and need for clinical correlation. Salter–Harris III and IV injuries are rare except during adolescence when the physis begins to close resulting in unique forces across the joint with specific injuries. Salter–Harris V injuries are quite rare.

36 PEDIATRIC SPORTS INJURIES IN THE ED

Greg Canty

Over 30 million youth and adolescents participate in organized sports and many more participate in unorganized sports such as skateboarding and cycling. As a result, more than 10% of pediatric ED visits involve a sporting injury. Many unique injury patterns exist in the youth athlete and recognizing them is essential to emergency medicine.

FRACTURES

- Sport injury mechanisms that result in adult sprains frequently result in physeal fractures of the younger athlete because the physis may be weaker than the attached ligament.
- The most common fractures are Salter–Harris I and II, but the physis begins to close during adolescence resulting in unique patterns like the Salter–Harris III and IV fracture (see Chapter 35 for description of Salter–Harris fractures).

AVULSIONS

- Avulsion injuries are a specific type of fracture that occur when a stronger tendon pulls off a weaker piece of bone around an apophysis or secondary ossification center.
- The most common scenario involves an adolescent athlete having a strong muscular contraction that results in an avulsion of pelvis at the iliac crest, iliac spine, or ischium.
- Most avulsions can be managed with crutches and nonweight bearing for 4–6 weeks.

DISLOCATIONS

- Dislocations are rare before adolescence but are associated with an increased risk of recurrence in young athletes.
- The most common dislocations involve the proximal interphalangeal (PIP) joint, the glenohumeral joint of the shoulder, and the patellofemoral joint of the knee.
- Reduction should be performed in a timely manner and may be preceded by radiographs if time permits.
- Neurovascular evaluation and postreduction radiographs are essential to confirm proper reduction and look for any related osteochondral fragments or fractures.

- Immobilize and arrange proper follow-up for 3–5 days.

SPRAINS AND LIGAMENTOUS INJURIES

- Much like dislocations, the risk of ligamentous sprains increase as athletes reach adolescence and the growth plates begin to close.
- The most common sports injury during adolescence is the lateral ankle sprain due to an inversion injury, and the most commonly injured ligaments of the ankle are the anterior talofibular (ATFL) and the calcaneofibular (CFL) ligaments.
- Anterior cruciate ligament (ACL) injuries are markedly increasing during adolescence, and most acute ruptures present to the ED with an acute, traumatic effusion following a twisting or a hyperextension mechanism.
- Sprains are graded I through III with III being a complete rupture.
- Most sprains can be managed with rest, ice, compression, early mobilization, and ambulation as tolerated.

OVERUSE SYNDROMES

- Overuse syndromes are epidemic in young athletes due to year-round training, sport specialization, repetitive activities, improper conditioning, and a growing skeleton.
- The ED physician must be aware of these syndromes because many will present to the ED with an acute worsening of a chronic pain.
- Overuse syndromes include stress fractures, spondylolysis, gymnast's wrist, thrower's elbow/shoulder, Osgood–Schlatter condition, jumper's knee, and shin splints.
- Overuse syndromes are managed by stopping the offending activity and encouraging proper rehabilitation.

CONCUSSION

- Growing evidence about concussions (mild TBI) tells us that young athletes recover much more slowly than professional and college athletes, so they should be managed much more conservatively.
- The majority of concussed athletes do not present with any loss of consciousness, and instead have headaches, amnesia, dizziness, and confusion following a blow to the head.
- Physical exam in concussion is typically normal except for some confusion and decreased cognitive function.
- Most concussed patients do not need any neuroimaging unless there are focal neurologic deficits.

- The severity of initial symptoms does not predict long-term outcomes and initial grading of concussions has been abandoned.
- Families should be educated on the diverse symptoms of concussion, the need for physical and cognitive rest, and the risk of second impact syndrome.
- All concussed patients need follow-up with a physician in 3-5 days and should never be cleared for return to sports or activities by the ED physician.

BRACHIOPLEXUS INJURIES

- Brachioplexus injuries, or “burners” and “stingers,” are the result of traction to the plexus when the head and shoulder are stretched in opposite directions, and often seen in American football.
- The nicknames come from the unilateral numbness, paresthesias, and weakness experienced in the injured upper extremity.
- Beware of any cervical spine tenderness, bilateral symptoms, or radicular symptoms into the legs because these suggest more serious injury and require cervical spine immobilization.
- All patients with brachioplexus injuries need follow-up with a physician in 3–5 days and should not be cleared for return to sports or activities by the ED physician.

RETURN-TO-PLAY

- Athletes and their families are often interested in when they can return-to-play following an injury, but this conversation cannot be properly determined in the ED.
- In order to return-to-play, proper follow-up should be emphasized to the athlete since this decision is best based on return of functional ability, cognitive function, range of motion, and adequate strength after the injury.

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QUESTIONS

1. A 15-year-old hockey player presents to the ED complaining of dizziness after hitting his head on the ice, but his exam is nonfocal. He has not had a head injury before. The family is traveling from out-of-town for a 3 day tournament, so they want to know when you recommend he can return-to-play.
 - A. Tomorrow since his exam is nonfocal
 - B. 3 days because this is a “mild concussion”
 - C. 1 week
 - D. Never
 - E. Not until after a follow-up visit with his physician
2. A 14-year-old basketball player presents to the ED after injuring her ankle playing basketball. She has significant lateral ankle swelling but is able to bear weight with point tenderness just inferior to the lateral malleolus. She most likely injured her
 - A. Anterior talofibular ligament (ATFL)
 - B. Deltoid ligament
 - C. Calcaneofibular ligament (CFL)
 - D. Tibiofibular syndesmosis
 - E. Fibular physis
3. A 12-year-old has recently started track and field and presents to the ED complaining of chronic knee pain that worsened during practice today. On exam he has point tenderness along the distal pole of the patella but no effusion and no fracture seen on radiographs. His most likely diagnosis is which of the following?
 - A. Osgood–Schlatter apophysitis
 - B. Anterior cruciate ligament rupture
 - C. Tibial plateau fracture
 - D. Sindig–Larsen–Johansson syndrome (jumper’s knee)
 - E. Medial tibial stress syndrome
4. A 15-year-old volleyball player presents to the ED after injuring her right knee while “twisting on a bent knee.” She had a noticeable deformity of her right knee with excruciating pain that seemed to “correct” when she extended her knee. In the ED you diagnosis a patellofemoral dislocation with spontaneous reduction and recommend follow-up imaging due to the common complication of which of the following conditions?
 - A. Popliteal artery dissection
 - B. Tibial plateau fracture

- C. Transverse patellar fracture
 D. Osteochondral fragments
 E. Femoral-tibia dislocation
5. A 16-year-old football player presents to the ED after tackling another player and experiencing some transient symptoms lasting 15–20 minutes that are now improved. Which of the following would be most consistent with a brachioplexus injury or “stinger?”
- A. Midline cervical spine tenderness
 B. Bilateral loss of sensation in the arms
 C. Unilateral deltoid weakness
 D. Limited range of motion of the cervical spine
 E. Transient paralysis of all 4 extremities lasting 10 minutes

ANSWERS

1. E. Patients should not be cleared from the ED for return-to-play following a concussion. We now know that initial symptoms and former grading scales are not predictive of long-term outcomes, and patients must be followed for persistent symptoms. Patients should be re-evaluated by a physician in 3–5 days to assess if symptoms persist. Once symptoms have resolved, there is a step-wise progression of activity before athletes are cleared for return-to-play. Answers A–C are all incorrect because they do not direct the family and patient to follow up to evaluate for ongoing symptoms. D is incorrect as the patient did not have more than one concussion.
2. C. The best answer is calcaneofibular ligament (CFL) because it is commonly involved in lateral ankle sprains with tenderness usually just inferior to the lateral malleolus and increased with inversion stress. A is also an excellent choice because the ATFL is commonly injured in lateral ankle sprains, but the point tenderness should be more anterior to the tip of the distal fibula. B is the medial ligament complex, which is rarely injured. D is often referred to as a “high ankle sprain” with pain anteriorly over the distal tibia and fibula. E is another excellent choice, but this patient scenario describes pain inferior to the fibula rather than along the distal fibular physis.
3. D. The patient in this scenario has chronic knee pain that has recently been exacerbated which suggests an overuse syndrome. Pain at the inferior pole of the patella suggests Sindig–Larsen–Johansson syndrome or “jumper’s knee.” This is an apophysitis injury of the distal patella worsened by traction from the patella tendon. The same mechanism may also cause pain over the tibial tubercle when it is referred to as A. B and C are unlikely because the patient does not have any swelling, normal radiographs, and history suggestive of chronic pain. E. Medial tibial stress syndrome is also known as shin splints and presents with pain over the posteromedial portion of the tibia with running.
4. D. Osteochondral fragments may be seen in over 50% of patellofemoral dislocations as the patella and femoral condyles are exposed to shearing forces during dislocation and spontaneous reduction. These fragments may be seen on radiographs or may require follow-up MRI if symptoms remain suggestive. Popliteal artery dissection is more common with femoral-tibial dislocations, which are rare. Tibial plateau and transverse patellar fractures are not associated with patellofemoral dislocations. Femoral-tibia dislocations are rare and more associated with motor vehicle trauma.
5. C. Unilateral deltoid weakness is common following brachioplexus injuries and may last minutes to days. Permanent nerve damage is very uncommon. Any signs of midline cervical spine tenderness, bilateral symptoms, decreased cervical range of motion, or transient paralysis should lead to immediate cervical immobilization and radiographic investigation for possible fracture, intervertebral disc injury, cervical cord neuropraxia, or more significant injury.

37 INJURIES OF THE UPPER EXTREMITIES

Jim R. Harley

PHYSICAL

- Look for lacerations, puncture wounds, soft-tissue swelling, deformity, and color.
- The resting hand normally has increasing flexion of the fingers and joints from the index through little finger and the DIP through MCP joints. This is disrupted when there is a laceration to an extensor or flexor tendon.
- Table 37-1 shows the muscles associated with each upper extremity peripheral nerve and their sensory distribution, and do a careful exam to assess wrist and hand injuries. Table 37-2 gives the appropriate test for each muscle.

TABLE 37-1 Guide to Upper Extremity Neurologic Examination

NERVE	MUSCLE	EXAMINATION
Motor Function		
Radial	Extensor carpi radialis longus	Wrist extension
Ulnar	Flexor carpi ulnaris	Wrist flexion and adduction
Median	Interosseous	Finger spread
	Flexor carpi radialis	Wrist flexion and abduction
	Flexor digitorum profundus	Flexion of fingers at PIP
	Opponens pollicis	Opposition of thumb to little finger
Anterior interosseous	Flexor digitorum profundus	Flexion distal phalanx of index finger
Sensory		
Nerve	Sensory	
Radial	Dorsal web space between thumb and index finger	
	Ulnar aspect palm and dorsum hand	
Ulnar	Ulnar aspect palm and dorsum hand	
Median	Radial aspect palm and hand	
	Thumb, index, middle, radial aspect ring finger	

TABLE 37-2 Flexor Tendons of the Hand and Wrist

TENDON	TEST OF FUNCTION
Flexor digitorum superficialis	Hold MCP in neutral and flex PIP
Flexor digitorum profundus	Hold MCP and PIP in neutral and flex DIP
Flexor carpi radialis	Flex and radially deviate wrist
Flexor carpi ulnaris	Flex and ulnarly deviate wrist
Extensor digitorum communis	Extension of the fingers at the MCP joints
Flexor pollicis longus	Hold MCP straight, flex thumb across palm
Palmaris longus	No specific test

CLAVICLE FRACTURES

- The clavicle is the most commonly fractured bone during delivery and is the fourth most commonly fractured bone in older children, usually from a fall or blow to the shoulder.
- The vast majority of injuries involve the area between the middle and distal third of the clavicle (>90%).

MEDIAL CLAVICLE FRACTURES

- Fractures of the medial clavicle are rare in children.
- CT scan is the best diagnostic if the diagnosis is not clear after obtaining plain films.

**FIG. 37-1.** Midshaft clavicle fracture.

DIAPHYSEAL CLAVICLE FRACTURES

- The most common mechanism for a midshaft clavicle fracture is a fall on the shoulder.
- Decreased or painful movement of the arm with point tenderness over the medial clavicle, localized swelling, crepitus, and tenting of the overlying skin is seen.
- Physical examination is very accurate and usually diagnostic; if plain films are needed, obtain one 30° cephalad (Fig. 37-1).
- Most heal without complication, and reduction is rarely necessary. Injuries due to birth trauma only require careful handling of the infant.
- Place young children in a sling or a shoulder strap; sling and swathe older patients.
- These fractures can be very painful; do not neglect adequate pain control.
- Educate parents that a bump from callus formation may appear about a month later as the bone heals.

LATERAL CLAVICLE INJURIES

- Direct trauma to the lateral clavicle produces metaphyseal fractures in young children rather than true acromioclavicular joint separations, as seen in adolescents and adults.
- The fracture heals well with use of sling and swathe, and surgery is only rarely indicated.
- Acromioclavicular separation is very rare before age 16.

SHOULDER DISLOCATIONS

- The same forces that result in shoulder dislocation in adults usually cause displaced Salter–Harris type II fracture separation of the proximal humerus in young children.

- When pediatric shoulder dislocations occur, anterior dislocations are much more common than posterior or inferior.
- Physical examination of an anteriorly dislocated shoulder reveals loss of the normally rounded contour, creating a squared-off appearance. The arm is held in slight abduction and external rotation, and the humeral head may be palpated anterior to the glenoid fossa.
- Obtain anteroposterior (AP) x-ray of the shoulder and a true scapular lateral or a transaxillary view.
- Provide adequate analgesia and relaxation before reduction with either traction–countertraction, scapular manipulation, or external rotation techniques.
- Obtain x-rays after reduction to look for occult fractures.
- Immobilize the joint after reduction for 3 to 6 weeks then begin rehabilitation therapy.
- Axillary nerve damage may accompany shoulder dislocation; assess sensation over the deltoid muscle before and after reduction. Other complications include greater tuberosity fractures, damage to the glenoid labrum, the Hill–Sachs deformity (a compression fracture of the posterolateral humeral head), rotator cuff injury, and recurrent dislocation.

HUMERUS FRACTURES

PROXIMAL HUMERUS FRACTURE

- The normal proximal humerus growth plate is often mistaken for a fracture (Fig. 37-2). A comparison view of the uninjured shoulder may be helpful.
- Salter–Harris type I fractures and proximal metaphyseal injuries, including greenstick and torus fractures, occur in children aged 5 to 11 years. Children of age 11 to 15 years suffer the majority of proximal humerus fractures, usually type II injuries.
- Most fractures of the proximal humerus heal well with a sling and swathe. Closed reduction attempts or operative treatments depend upon age of the patient, degree of angulation and amount of displacement. The younger the child, the more likely the fracture will heal without intervention.

HUMERUS SHAFT FRACTURES

- Proximal and distal humerus fractures are much more common than diaphyseal injuries. Most humeral shaft fractures are the result of a direct blow to the area.
- Torsional forces from falls or severe twists may result in a spiral diaphyseal fracture. Consider nonaccidental



FIG. 37-2. Normal proximal humerus epiphyseal plate.

trauma in patients under 3 years with a spiral humerus fracture.

- Midshaft fractures heal well even with angulation of up to 15° to 20° and as much as 2 cm of overriding, due to bony remodeling and longitudinal overgrowth that occurs in response to the fracture. Use a sling and swathe to young children and a sugar-tong splint for adolescents.
- Fractures involving the junction of the middle and distal thirds of the humerus may be associated with injury to the radial nerve. Motor and sensory functions should be assessed initially and following any manipulation. Acute radial nerve palsy has an excellent long-term prognosis, with reports of 80% to 100% recovery of function.

THE ELBOW

RADIOGRAPHS

- Good radiographs are essential in assessment of elbow injury other than a radial head subluxation, and must include an AP view with the joint in extension and a true lateral view with the elbow flexed at a right angle. Provide adequate pain control to flex the elbow fully.
- The anterior fat pad is deep in the coronoid fossa and normally appears as a small lucency just anterior to the fossa on a true lateral x-ray of the elbow (Fig. 37-3). Joint space fluid collection may also cause the anterior fat pad to be pushed away from the joint and appear as a wind-blown sail—the “sail sign.”
- The posterior fat pad deep in the olecranon fossa is normally not visible. When seen on a true lateral x-ray, it suggests blood within the joint capsule



FIG. 37-3. Normal elbow with a thin anterior fat pad.

and provides evidence of occult fracture of the distal humerus, proximal ulna, or radius (Fig. 37-4), which may only be detected with the elbow fully flexed at 90°.

- The anterior humeral line, drawn along the anterior cortex of the distal humerus on a true lateral view of the elbow, should normally intersect the middle third of the capitellum distally. Posterior displacement of the capitellum may be consistent with an otherwise radiographically inapparent supracondylar fracture.
- The radiocapitellar line is drawn down the axis of the proximal radius on the true lateral view of the elbow and should bisect the capitellum regardless of



FIG. 37-4. Note the posterior fat pad sign, signifying the presence of blood within the joint space.

TABLE 37-3 Supracondylar Fracture Types: Description

Type I	No displacement
Type II	Moderate displacement but contact between fragments remains
Type III	Complete displacement, fragments are not touching

the degree of flexion or extension present. Failure to do so suggests the presence of an occult radial neck fracture or radial head dislocation.

- Any question about the anatomic relationships can be further investigated using comparison views of the uninjured elbow.

SUPRACONDYLAR HUMERUS FRACTURES

- Supracondylar fractures account for 50% to 60% of all elbow fractures in children 3 to 10 years of age (Table 37-3).
- Falls onto an outstretched hand cause violent hyperextension of the elbow; this is the usual mechanism of injury.
- On examination, there will be pain, swelling, deformity, and functional impairment. A careful neurovascular examination is crucial to determine if there is an associated injury.
- Obtain AP and lateral x-rays (Fig. 37-5). Oblique views may identify occult fractures.
- Use of the anterior humeral line may be useful in determining whether the fracture is a type I or II (Fig. 37-5).



FIG. 37-5. Type II supracondylar fracture. Note the anterior humeral line that intersects the anterior portion of the capitellum.

- Prompt reduction is needed when there are signs of significant distal ischemia such as pallor and cyanosis of the fingers, prolonged capillary refill, or absence of radial pulse.
- Patients are at risk of developing a forearm compartment syndrome. Unrecognized, this will lead to Volkmann's ischemic contracture and a nonfunctional hand and wrist.
- Forearm pain with passive flexion or extension of the fingers or distal paresthesias is an ominous early sign of compartment syndrome.
- Nerve impairment occurs in 10% to 20% of children with supracondylar fractures, yet the prognosis for return of function is good. Radial, median, and ulnar nerve injuries, in descending order of frequency, have all been reported.
- A late complication of supracondylar humerus fractures is cubitus varus, a change in the carrying angle of the elbow.
- Type I supracondylar fractures can be managed with casting but type II and III fractures require operative reduction and internal fixation.
- Most children with type II or III are admitted for 24 to 48 hours of observation so that the neurovascular status of the extremity can be reassessed frequently. Open reduction and internal fixation may be necessary (Fig. 37-6).

THE MEDIAL AND LATERAL CONDYLES

- Fractures involving the articular surface of the lateral condyle (capitellum) comprise 15% of all pediatric elbow fractures.



FIG. 37-6. Comminuted supracondylar fracture with large joint effusion (type III). The patient required fasciotomy and skin grafting due to neurovascular compromise.



FIG. 37-7. Nondisplaced fracture of the medial condyle in a 5-year-old.

- Peak incidence is 6 years of age and the most likely mechanism is a fall on the outstretched arm with forearm supination or elbow flexion.
- Lateral condyle fractures are not usually associated with acute neurovascular injuries, but require aggressive intervention to prevent later complications such as nonunion, loss of elbow mobility, and growth arrest of the lateral condylar physis leading to eventual cubitus valgus and tardy ulnar palsy.
- Management is usually operative in all but nondisplaced fractures (Fig. 37-7).
- Nonunion and ulnar nerve neuropraxia are the most frequent complications.

MEDIAL EPICONDYLAR FRACTURE

- Epicondyles are located just proximal to the articulating surface of the distal humerus.
- The medial epicondyle is a traction apophysis to which the forearm flexors are attached.
- Fractures of the medial epicondyle are rarely encountered in children younger than 4 years, occurring most commonly in children aged 7 to 15 years. The vast majority are associated with elbow dislocations, occurring approximately 50% of the time (Fig. 37-7).
- The medial epicondyle may dislocate and then block reduction or become entrapped intra-articularly.
- A more insidious injury to the medial epicondyle may occur with repetitive traction stress by the forearm flexors (Little Leaguer's elbow). Treatment is usually nonoperative, while severe displacement or ulnar



FIG. 37-8. Posterior elbow dislocation with avulsion of the medial epicondyle.

nerve dysfunction considered relative indications for operative management.

ELBOW DISLOCATIONS

- Pediatric elbow dislocations occur infrequently, since most forces that would result in dislocations in adults usually cause fractures in children (Fig. 37-8).
- As with adults, most dislocations are posterior.
- Associated fractures are the rule and most commonly involve fracture of the medial epicondyle, coronoid process, radial head, or olecranon.
- Most dislocations can be reduced after providing adequate analgesia and muscle relaxation. The elbow should be flexed to 60° to 70° and the forearm placed in supination. The proximal humerus is then stabilized by an assistant while longitudinal traction is applied at the wrist.
- Upon successful reduction, gently flex the elbow, immobilize it and assess the neurovascular status of the arm. Obtain postreduction radiographs to verify location of the medial epicondyle as extra-articular.
- Reduction may be difficult when there are occult fracture fragments in the elbow joint.

RADIAL HEAD SUBLUXATION

- This is the most common pediatric elbow injury called “nursemaid’s elbow” or “pulled elbow,” and occurs when abrupt axial traction is applied to the wrist or hand of the extended, pronated forearm of a child younger than 5 years, causing the annular

- ligament to slip free of the radial head and become entrapped between the radial head and capitellum.
- There may be a history of the patient being lifted by the arm though the precipitating event is often neither witnessed nor recognized.
- The child appears comfortable yet refuses to reach for objects with the affected arm.
- The forearm is held in pronation with the elbow in slight flexion or fully extended.
- There is a remarkable lack of swelling and only mild tenderness over the radial head. If there is elbow swelling, a different diagnosis should be considered.
- Pain is worse with supination or pronation.
- Radiographic evaluation is not necessary in the presence of a clear history of precipitating arm traction.
- There are two methods of reduction, supination, and pronation.
- In the supination method, face the patient, placing your thumb over the radial head, and your opposite hand around the wrist. Supinate the forearm flexing at the elbow.
- In the pronation method, which is typically less painful, extend the arm at the elbow, place a finger over the radial head, and pronate the forearm. A palpable or audible “pop” usually signals successful reduction.
- Typically, the patient again reaches for objects with the affected arm within 5 to 10 minutes of reduction. No further treatment is necessary.
- Educate parents not to lift their children by the wrists and that recurrence may be as high as 30%.
- Several attempts at reduction may be needed before normal use of the arm resumes. Occasionally, radial traction prior to supination and flexion is necessary. If the subluxation occurred several hours earlier, it may be longer before normal function of the arm is observed.
- If normal use does not follow reduction attempts, alternative diagnoses should be considered. Immobilization with prompt orthopedic follow-up is indicated.

FRACTURES OF THE RADIUS AND ULNA

- The radius and the ulna are the second most frequently injured bones during childhood.
- Three-quarters of all injuries involve the distal third of the forearm.
- For this reason, forearm x-rays should always include the wrist and elbow.
- Most fractures of the radius and ulna heal without significant complications.

PROXIMAL RADIUS AND ULNA FRACTURES

- Falls onto extended, supinated arm with valgus stress can result in fracture of the radial head or neck.
- Most proximal radius fractures in young children involve the narrow metaphyseal neck since the head is cartilaginous until ossification begins at age 5 years.
- Salter–Harris type I and type II radial neck fractures are most common. Salter–Harris type IV radial head fractures may be encountered in older children.
- An abnormal fat pad sign or abnormal radiocapitellar line on x-ray points to the presence of an occult radial head or radial neck fracture.
- Minimally displaced or nondisplaced fractures can be treated in a posterior splint with the elbow flexed at 90°.
- Olecranon fractures occur commonly in combination with other elbow injuries, such as radial head dislocations, radial neck fractures, and fractures of the medial epicondyle.

DIAPHYSEAL RADIUS AND ULNA FRACTURES

- Most forearm diaphyseal fractures are either greenstick or bowing injuries. Both bones may suffer greenstick or bowing injuries, or one bone may have a greenstick fracture while the paired bone is bowed (Fig. 37-9).
- Overriding of fracture fragments in the presence of an isolated fracture of one of the forearm bones suggests either a Monteggia or Galeazzi fracture. An isolated fracture of the proximal ulna may be associ-

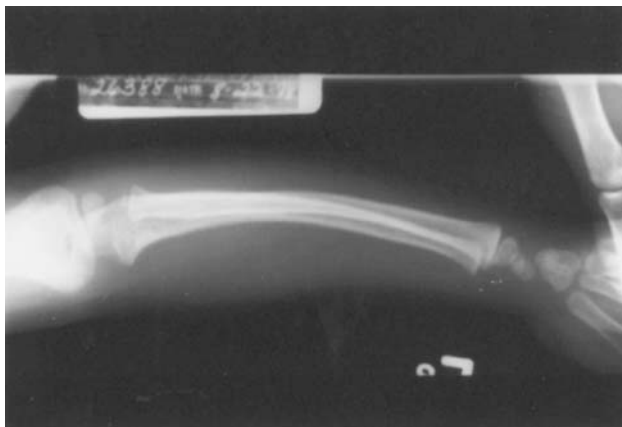


FIG. 37-9. Radiographic appearance of midshaft bowing injury of both the radius and ulna.

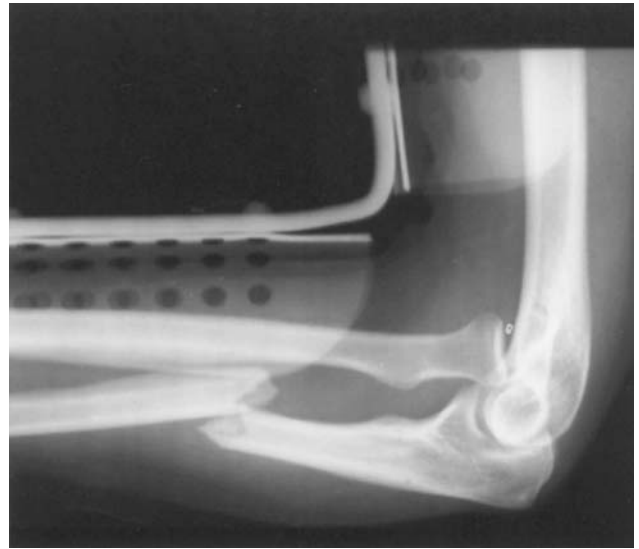


FIG. 37-10. Fracture of the proximal ulna with radial head dislocation (Monteggia fracture). A line bisecting the proximal radius completely misses the capitellum.

ated with concomitant dislocation of the radial head (Monteggia fracture).

- This combined injury may be inadvertently overlooked initially because attention is focused on the obvious ulnar fracture. An aberrant radiocapitellar line on plain x-ray is evidence of the accompanying radial head dislocation (Fig. 37-10). Closed reduction is usually successful.
- A fracture at the junction of the middle and distal thirds of the radius in association with distal radioulnar joint dislocation is called a Galeazzi fracture and is rare in children (Fig. 37-11).

DISTAL RADIUS AND ULNA FRACTURES

- Fractures of the distal third of the radius and ulna are among the most common orthopedic injuries in children 6 to 12 years of age, often occurring after a fall onto an outstretched hand.
- Torus fractures of the distal radius and ulna are frequently encountered and can be treated in a removable splint (Fig. 37-12).
- These fractures are often very subtle. Have a high index of suspicion for a radius fracture when the patient has point tenderness over the distal radius.
- Tenderness over the physis with a normal radiograph suggests nondisplaced Salter–Harris type I injury; splinting with orthopedic follow-up is appropriate.
- The capacity for angular remodeling after forearm fracture is great, but rotational remodeling does not



FIG. 37-11. Galeazzi fracture in a 16-year-old.

occur and rotational abnormalities must be accurately corrected.

- Complications are uncommon, but vascular compromise or compartment syndrome can develop with any forearm fracture.

HAND AND WRIST INJURIES

- In younger children, the most common mechanism is to get the hand caught in a closing door resulting in a crush injury to the distal phalanxes.
- Commonly encountered injuries include distal radius fractures, ligamentous injuries to the wrist and fingers, dislocations of the metacarpophalangeal (MCP),



FIG. 37-12. Torus fracture of the distal radius.

proximal interphalangeal (PIP), and distal interphalangeal (DIP) joints.

FRACTURES OF THE PHALANXES

DISTAL PHALANX—CRUSH INJURIES

- Pediatric distal phalanx fractures are either a crush or hyperextension injuries.
- Crush injuries are quite common, usually resulting from entrapment in a closing door
- There is frequently a partial amputation of the fingertip.
- Wound care must be meticulous, the nail bed approximated with absorbable suture, and the finger splinted. Some authors recommend replacing the nail plate in the nail fold; however, there is no evidence yet to suggest that this improves outcome.
- There is often an associated tuft fracture associated with nail bed injuries (37-13).
- Some authors recommend prophylactic antibiotics when a tuft fracture is accompanied by a nail bed



FIG. 37-13. Partial fingertip amputation and tuft fracture.

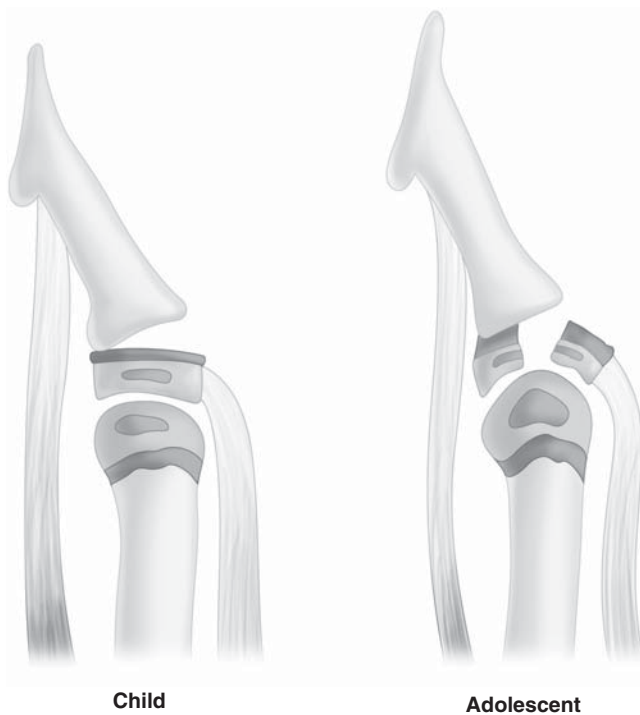


FIG. 37-14. Mallet injuries are caused by avulsion of the extensor tendon from the distal phalanx or from a fracture of the dorsal base of the distal phalanx. An extension lag at the DIP joint is present, and the patient is unable to actively extend the DIP joint. In children, a Salter–Harris type I or II fracture is seen. In the adolescent, a Salter–Harris type III fracture occurs. These fractures may be difficult to detect radiographically since the epiphysis is not fully ossified in children. Inability to actively extend the DIP joint and a mallet deformity reveal the extent of injury.

laceration but there is no evidence to suggest a difference in outcome.

DISTAL PHALANX—OTHER INJURIES

- A hyperflexion force applied to the tip of the finger may result in one of two types of pediatric injuries (Fig. 37-14).
- In the preadolescent, a Salter–Harris type I or II fracture occurs with a mallet deformity. A mallet finger is the result of an avulsion of the extensor tendon from the base of the distal phalanx. If a child has a mallet finger and inability to extend the distal phalanx, this injury should be assumed even if a fracture cannot be identified on radiographs.
- In the adolescent hyperflexion injury, a displaced Salter–Harris type III fracture occurs. These injuries are treated by wound care, closed reduction, and splinting in slight hyperextension.

- If adequate reduction is not obtained, then open reduction and Kirschner-wire fixation is performed. It is important to recognize the mallet deformity present in both of these injuries and treat, and refer appropriately.

MIDDLE AND PROXIMAL PHALANX

- Middle and proximal phalangeal fractures commonly occur at the physis.
- These fractures are usually Salter–Harris type II fractures.
- A common fracture of the little finger proximal phalanx results in an “extra octave finger” with ulnar displacement of the little finger.
- They are reduced by flexion of the MCP joint and adduction of the finger.

METACARPAL FRACTURES

- The neck is the most common area to be fractured.
- Most metacarpal fractures are undisplaced or minimally displaced and are treated initially by splint immobilization.
- Displaced fractures can generally be treated with closed reduction and splinting, but occasionally Kirschner-wire fixation is required.
- The most common hand fracture is of the fifth metacarpal. Typically, it results from striking someone or a solid object with a closed fist (Boxer’s fracture). Closed reduction is indicated if there is more than 30° to 40° of angulation. These fractures can be treated with an ulnar gutter splint with orthopedic referral.

CARPAL FRACTURES

- Fractures of the carpal bones in children are exceedingly rare.
- Scaphoid fractures are the most commonly encountered of the carpal bone fractures.
- The ossification center of the scaphoid appears by age 5 to 6 years.
- The usual mechanism of injury is a fall on an outstretched hand.
- Physical examination may reveal limitation of range of motion from pain and swelling and tenderness in the “anatomic snuffbox.”
- As in the adult, initial x-rays often appear normal.
- If there is suspicion of a scaphoid fracture, apply a short-arm thumb spica splint and refer for an early orthopedic evaluation.

- Nonunion and avascular necrosis is rare in children because most injuries are avulsions or nondisplaced fractures through the distal third of the bone rather than fractures through the waist, as in adults. Other carpal bone fractures are very rare in children and are treated as in adults with splinting and orthopedic referral.

DISLOCATIONS

DISTAL INTERPHALANGEAL JOINT

- DIP dislocations result from a hyperextension force.
- This dislocation generally displaces dorsally and is often open, due to the tight adherence of skin to bone in this area.
- Reduction is usually uncomplicated and consists of traction countertraction followed by flexion.
- Test active motion to ensure that the extensor and flexor tendons are functioning and that the volar plate is not interposed in the joint.

PROXIMAL INTERPHALANGEAL JOINT

- Dorsal dislocations are the most common. These dislocations occur as a result of an axial load with concomitant hyperextension.
- Reduction is accomplished through slight hyperextension and longitudinal traction applied to the middle phalanx while correcting the ulnar or radial deformity.
- The finger is then gently flexed into position.
- Active range of motion is tested, and stress testing of the collateral ligaments and volar plate is performed.
- The finger is placed in a splint immobilizing the PIP (20–30° of flexion) and MCP joints (60–70° degrees of flexion).
- Orthopedic referral is recommended.
- Volar dislocations are rare and may be irreducible due to entrapment of the proximal phalangeal condyle between the central tendon and lateral band. These dislocations may result in avulsion of the central slip of the extensor tendon leading to a late boutonnière deformity. Orthopedic referral is required.

METACARPOPHALANGEAL JOINT

- MCP joint dislocations sometimes occur in the pediatric age group. As in the adult, the thumb MCP dislocation is the most common (Fig. 37-15). It occurs as a result of a hyperextension force, usually from a fall on an outstretched hand.



FIG. 37-15. Carpometacarpal dislocation of the thumb.

- Dislocations may be simple reducible or complex irreducible.
- In the simple reducible dislocation, the proximal phalanx assumes a dorsal point at a 90° angle to the metacarpal. This dislocation can be reduced by gentle traction countertraction. Joint stability is assessed, the finger splinted, and the patient referred.
- The complex irreducible dislocation has the same mechanism of injury, but the proximal phalanx assumes a bayonet position parallel to the metacarpal. The volar plate is interposed in the joint, and the metacarpal head may also be trapped in the substance of the intrinsic muscles. Closed reduction is impossible.
- This dislocation can only be reduced by open reduction. Some authors recommend one attempt at gently performed closed reduction. Vigorous traction is to be avoided since this may convert a simple reducible dislocation to a complex irreducible one.

CARPOMETACARPAL JOINT

- Carpometacarpal dislocations are rare in the pediatric age group. These injuries are generally a result of violent trauma that results in multiple dislocations

or multiple fracture dislocations. These injuries require prompt orthopedic consultation for surgical intervention.

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QUESTIONS

- A 10-year-old boy has sustained a laceration to his hand and you suspect a nerve injury. To test the motor function of the anterior interosseous nerve the patient should be asked to do which of the following?
 - Flex the wrist.
 - Extend the wrist.
 - Flex the index finger PIP joint.
 - Extend the index finger PIP joint.
 - Flex the index DIP joint.
- A 7-year-old boy presents after falling off his bicycle. He has pain with movement of his right arm and swelling and tenderness over his clavicle. Pediatric clavicle fractures occur most frequently at which of the following locations?
 - Acromial-clavicular joint
 - Distal third of the clavicle
 - Middle third of the clavicle
 - Proximal third of the clavicle
 - Sterno-clavicular joint.
- A 2-year-old girl is brought to the emergency department (ED) by her parents when she refuses to use her right arm. The mother states this occurred after she was picked up by her older brother and swung around playing airplane. She is otherwise doing well and is happy but unwilling to move her right arm. Which of the following would not be consistent with a radial head subluxation in this patient?
 - The mechanism of injury.
 - The age of the patient.
 - Refusal to reach for objects.
 - Marked swelling on physical examination.
 - Pain increased with supination or pronation.
- A Monteggia fracture is a proximal ulna fracture in association with which of the following injuries?
 - Radial head dislocation
 - Radial head fracture
 - Proximal radius fracture
 - Distal radioulnar dislocation
 - Distal radius fracture
- A 7-year-old girl presents to the ED with elbow pain and swelling after a fall on an outstretched hand. Radiographs are ordered. Which of the following is not a true statement in the evaluation of elbow radiographs?
 - The radio-capitellar line should intersect the middle of the radial head.
 - The anterior humeral line should intersect the middle of the capitellum.
 - The anterior humeral head should intersect the anterior 1/3 of the capitellum.
 - A thin anterior fat pad is normal.
 - A posterior fat pad is abnormal.
- Which of the following best describes a mallet finger?
 - Avulsion of the extensor tendon from the base of the distal phalanx resulting in flexion at the DIP joint.
 - Injury to the central slip of the extensor tendon causing flexion of the PIP joint and extension at the DIP joint.
 - Avulsion of the flexor tendon from the base of the distal phalanx.
 - Injury to the central slip of the flexor tendon.
 - A crush injury to the distal phalanx.
- Crush injuries to fingertip are associated with all of the following *except* which of the following?
 - A tuft fracture of the distal phalanx.
 - Injury to the nail bed.
 - Partial amputation of the fingertip.

- D. Frequently need Kirschner-wire fixation.
E. Result from getting a finger caught in a door.
8. A 12-year-old girl presents with wrist pain after a fall. On examination, you suspect a scaphoid fracture. True statements about pediatric scaphoid fractures include all of the following *except*:
- A. It is usually associated with a fall on an outstretched hand.
B. They are rare in children under 10 years of age.
C. They are associated with anatomic “snuff box” tenderness.
D. Are usually evident on initial radiographs.
E. Require close follow up with orthopedics.
7. D. Most crush injuries to the fingertip can be managed in the emergency department.
8. D. Scaphoid fractures are frequently not seen on initial radiographs.

38 INJURIES OF THE PELVIS AND LOWER EXTREMITIES

Greg Canty

ANSWERS

1. E. The anterior interosseous nerve innervates the flexor digitorum profundus and is responsible for flexion at the distal phalanx of the index finger. Refer to Table 37-1.
2. C. Clavicle fractures occur most commonly at the middle third of the clavicle and the clavicle is the most commonly fractured bone during delivery and is the fourth most commonly fractured bone in older children, usually from a fall or blow to the shoulder.
3. D. If there is marked swelling of the elbow; there is likely a fracture present. Radial head subluxations occur with abrupt axial traction applied to wrist or hand as when being lifted in children younger than 5 years of age as in this child. Children tend to be comfortable but will refuse to reach for objects with the affected arm. Pain will be worse with supination or pronation.
4. A. A Monteggia fracture is a proximal ulna fracture associated with a radial head dislocation.
5. C. The anterior humeral line and the radiocapitellar line are two reference lines that are useful in assessing elbow radiographs and helping to identify occult injury. The anterior humeral line, drawn along the anterior cortex of the distal humerus on a true lateral view of the elbow, should normally intersect the middle third of the capitellum distally. Posterior displacement of the capitellum may be consistent with an otherwise radiographically inapparent supracondylar fracture. The radiocapitellar line is drawn down the axis of the proximal radius on the true lateral view of the elbow and should bisect the capitellum regardless of the degree of flexion or extension present.
6. A. Avulsion of the extensor tendon from the base of the distal phalanx with a flexion deformity is a mallet finger. B is a Boutonniere deformity. C is an avulsion of the volar plate.

PELVIC FRACTURES

- Unlike the adult pelvis, the young pelvis is quite pliable and absorbs a tremendous amount of energy, so fractures of the pelvis are quite rare in children.
- Almost all pelvic fractures in children are part of a multisystem trauma from high-energy trauma such as motor vehicle crashes or significant falls.
- Pelvic ring fractures are classified as either stable from a single break in the ring or an unstable fracture (see Fig. 38-1) due to two breaks within the ring or single break with an associated sacro-iliac dislocation.
- Acetabular fractures are rare in children and almost all are associated with a hip dislocation.
- Avulsion fractures of the pelvis (Fig. 38-2) result from lower energy mechanisms and are commonly associated with sports. These avulsion fractures are not associated with multiple injuries and are generally managed nonoperatively.
- Since nearly all pelvic fractures in children are associated with multiple injuries, special attention must

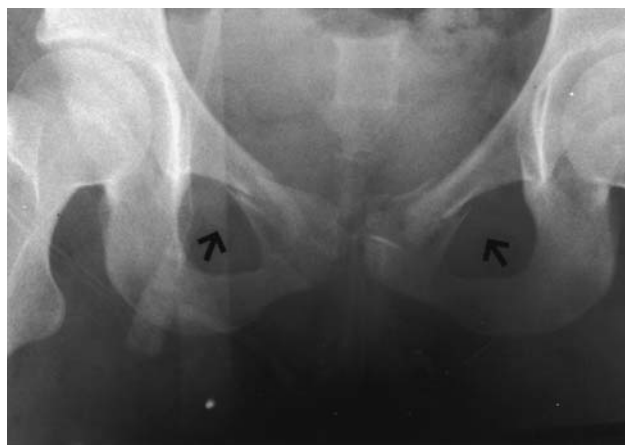


FIG. 38-1. Bilateral pelvic rami fractures which are unstable and at high risk for accompanying visceral injuries.



FIG. 38-2. Right anterior inferior iliac spine (AIIS) avulsion fracture in a young track and field athlete. There also appears to be a previous injury to the ischial apophysis.

be paid to the abdomen, genitourinary system, and rectum during exam.

- AP radiographs of the pelvis are part of most trauma protocols, but these may be omitted if a patient is also getting a CT of the abdomen/pelvis since it is more sensitive in detection.
- The majority of children with pelvic fractures have good outcomes with conservative, nonsurgical management, and the overall morbidity and mortality are more closely tied to the severity of their accompanying injuries.

HIP DISLOCATIONS

- Hip dislocations are rare events in pediatric emergency medicine, and they account for less than 5% of all dislocations in this age group.
- Young children may dislocate their hip with trivial falls or sporting injuries due to a shallow acetabulum and increased joint laxity, but adolescents require much greater forces like adults because the acetabulum deepens and ossifies.
- Hip dislocations usually present with severe discomfort, a shortened leg, and a flexed hip in internal rotation.
- Posterior dislocations without accompanying fractures should be reduced within 6 hours by flexing the hip to 90° and providing axial traction to the thigh.
- Postreduction x-rays and suspicious radiographs often require a CT scan to better look for associated intra-articular fractures and bony fragments.

PROXIMAL FEMUR FRACTURES

- Similar to hip dislocations with the high-energy forces required and their rarity, proximal femur fractures account for less than 1% of fractures around the hip.
- These fractures are complicated by a high risk of premature physeal closure and avascular necrosis (AVN) of the femoral head, so any suspicion should lead to orthopedic consultation in the ED.
- Another similarity to the pelvis is the risk of lower energy avulsion fractures from sporting activities, which may be seen around the apophyses of the greater and lesser trochanter.

FEMORAL SHAFT FRACTURES

- The majority of femur fractures in children occur along the middle third of the femoral shaft and management depends upon the patient's age.
- Unlike adult femur fractures, the pediatric patient rarely experiences hypotension or shock-like symptoms due to an isolated femur fracture, and physicians should always look for accompanying life-threatening injuries to the chest, abdomen, and pelvis if these symptoms are present.
- When a femur fracture is suspected, initial evaluation should include assessment of neurovascular status, pain control, and splinting before radiographic evaluation. Radiographs should include the femoral shaft, hip, and knee to look for possible dislocations and accompanying injuries.
- Orthopedics should be involved in the management of all femur fractures in the ED. Current management strategies range from a hip spica cast in the young toddler to an intramedullary rod in older children and adolescents, but the trend is to manage the majority of these injuries surgically.

FRACTURES OF THE KNEE AND PATELLA

- The knee region is commonly injured in sports, falls, and motor vehicle accidents.
- Fractures of the knee region are especially concerning because two-thirds of the lower extremity length comes from the physes of the distal femur and proximal tibia; so there is a very high risk of growth arrest or progressive deformity when either of these regions are involved.
- Most common fractures of the knee involve the distal femur as a result of high-energy trauma. The most common fracture pattern is the Salter–Harris II,



FIG. 38-3. Tibial spine avulsion fracture after hyperextension in a young athlete.

which requires reduction and occasional pinning to ensure anatomic alignment. Despite proper management, a large number of these fractures will go into growth arrest.

- The proximal tibial physis is rarely fractured because it is “protected” by the fibula and insertion of the medial collateral ligament distal to the physis. The tibia does have its own challenges though as two unique avulsion fractures appear in adolescence.
- The first unique injury is the tibial spine avulsion (Fig. 38-3), which occurs frequently after falling from a bike or hyper-extending the knee during sports. The mechanism is similar to the anterior cruciate ligament (ACL) injury in adults, but either ligamentous or bony injury can occur in children and adolescents.
- The second unique injury involves avulsion of the tibial tubercle (Fig. 38-4) which occurs in adolescence following a forceful contraction of the quadriceps, and those forces are transmitted to the tibial tubercle that may be in varying stages of maturation.
- Fractures of the patella are rare in children due to the large amount of cartilaginous “padding,” and fractures should not be confused with the bipartite patella seen in 5% of the population along the superolateral corner.



FIG. 38-4. Tibial tubercle avulsion fracture in a basketball player requiring surgical fixation because of intra-articular extension.

- A thorough examination of the knee should include palpation of the bony prominences, physes, collateral ligaments, and distal pulses due to the risk of injury near the popliteal artery.
- Acute radiographic series of the knee should include anterior–posterior (AP), lateral, and patella views.
- Any acute traumatic effusion or tenderness along the physis warrants immobilization, nonweight bearing, and follow-up with an orthopedist or sports medicine physician in 3–5 days.

OSGOOD–SCHLATTER APOPHYSITIS

- Osgood–Schlatter apophysitis is due to traction along the tibial tuberosity from the extensor mechanism, and it is a common cause of knee pain in active adolescents. It may occur bilaterally in up to 25% of patients.
- Management is supportive with activity modification, ice, and NSAID’s with the vast majority being self-limited conditions that fluctuate in severity over a 1–2 year period.

KNEE AND PATELLA DISLOCATIONS

- Femur-on-tibia knee dislocations are usually associated with vehicular trauma and are extremely rare in children. They must be strongly considered though in

any traumatic knee effusion because of the high risk of popliteal artery damage (40%) and peroneal nerve injury (33%) which can be limb-threatening. Prompt reduction and admission for frequent neurovascular checks are essential.

- Patellar dislocations are quite common in adolescents and nearly all are lateral dislocations. Most reductions occur spontaneously with knee extension prior to examination, but reduction can be performed prior to radiographs with classic presentations.
- After reduction, repeat radiographs are important along with arranging close follow-up because many patients will have osteochondral fragments from shearing of the patella or femoral condyle during dislocation.

FRACTURES ALONG THE SHAFT OF THE TIBIA AND FIBULA

- Fractures of the tibia and fibula are the most common long bone fractures of the lower extremity in pediatrics. Tibial fractures require special attention because of their risk for compartment syndrome and ischemia, especially if the fracture is more proximal where the tibial artery is easily damaged.
- Fracturing both the tibia and fibula requires a high-energy mechanism, but they are frequently injured together. Beware of the isolated fibula fracture because it is often seen in conjunction with a distal tibial physeal injury.
- Fractures of the tibia are at a high risk for a valgus deformity, and greenstick fractures require completion of the fracture under anesthesia along with ensuring proper alignment when immobilized.
- The toddler's fracture is an occult tibia fracture that commonly presents in a young child with a limp or refusal to walk with an otherwise unimpressive exam. Oblique radiographs are occasionally helpful, and these patients should be managed with nonweight bearing and immobilization until they can be seen in follow-up by an orthopedist.

FRACTURES OF THE FOOT AND ANKLE

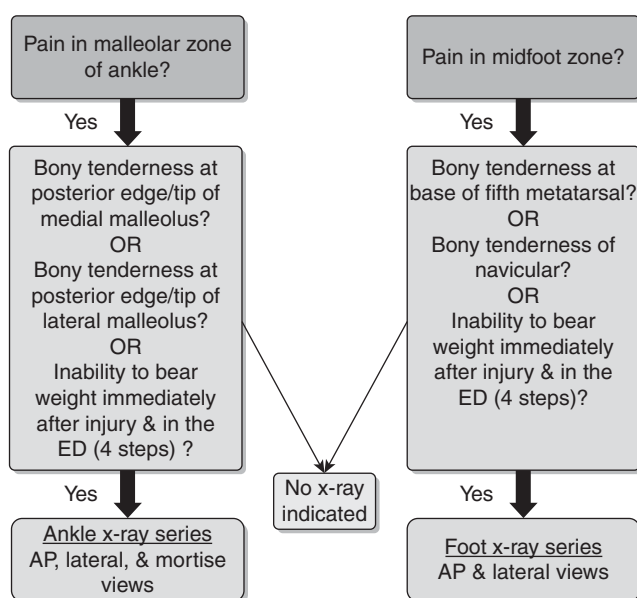
- The foot and ankle are injured frequently in sporting activities similarly to adults, but the big difference is that the physis in children allows for a broad range of injuries from sprains to physeal fractures. The younger the patient, the more likely an injury will result in a fracture due to the immaturity of the physis.
- The most common mechanism for fractures of the ankle is inversion and supination which places a



FIG. 38-5. Mortise view of the ankle demonstrating a Salter–Harris III fracture of the tibia commonly referred to as a Tillaux fracture.

great deal of stress on the distal fibular physis. The most common fracture is a Salter–Harris I of the distal fibula with tenderness and swelling on exam with radiographs possibly being normal otherwise. Most fractures of the distal fibular physis are non-displaced and can be managed with an ankle stirrup (sugar-tong) splint.

- The distal tibial physis is most commonly injured in a Salter–Harris II pattern requiring reduction and may be accompanied by a greenstick of the fibula. The tibia in adolescents may also be fractured in two distinct patterns known as the Tillaux, a Salter–Harris III (see Fig. 38-5), and the Triplane, a Salter–Harris IV. It is important to recognize both of these fractures as they normally require surgical fixation to maintain the articular surface.
- Radiographs of the ankle should always include at least three views (AP, lateral, mortise), and the foot should also be examined closely with any ankle injury due to the high likelihood of concomitant injuries.
- The Ottawa Ankle Rules have been validated in children and found to be very sensitive in children older than 5 years of age (Fig. 38-6).

Acute ankle injury*

*Use with caution in children younger than 5 years

FIG. 38-6. Ottawa ankle rules for use in adults and school-aged children.

FOOT FRACTURES

- The vast majority of foot injuries involves the fore-foot and result from numerous mechanisms including inversions of the ankle, direct blows, twisting forces, and falls.
- Metatarsal fractures are the most common foot fracture and are usually associated with an impressive amount of soft tissue swelling. The most common metatarsal fracture is the avulsion off the base of the 5th metatarsal (see Fig. 38-7) seen during ankle inversion injuries. The famous “Jones” fracture is rare in pediatrics and usually involves chronic findings along the metaphyseal/diaphyseal junction. Most metatarsal fractures can be managed with reduction, when necessary, immobilization in a posterior splint, and nonweight bearing.
- Phalangeal fractures are also usually easily recognized on clinical exam and radiographs. Management consists of possible reduction, buddy-taping, and immobilization in a hard sole cast shoe or splint.
- Rare injuries to the midfoot include the Lisfranc dislocation/tarsometatarsal fracture (see Fig. 38-8) with marked tenderness and swelling of the mid-foot. Oblique and weight bearing views may be helpful when evaluating possible Lisfranc injuries. Recognition is important because most of these injuries require surgical fixation.

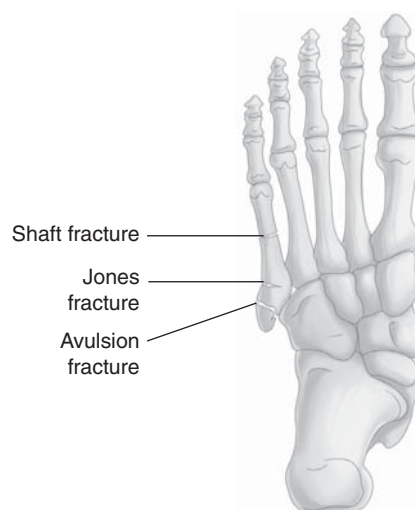


FIG. 38-7. Fractures of the fifth metatarsal are much more likely to be avulsions or shaft fractures in pediatrics. The Jones fracture occurs at the metaphyseal–diaphyseal junction and is rare in children.

- The calcaneus is rarely injured in children, but may be associated with falls from a significant height. AP, lateral, and axial views should be obtained when a fracture is suspected. The Bohler angle is much less reliable in children. Any calcaneal fracture from axial loading requires close inspection of the thoracolumbar vertebrae for associated injuries.
- Fractures of the talus usually involve the talar neck after a mechanism of forced dorsiflexion, and patients present with anterior ankle pain, swelling, and the inability to ambulate. A unique talar fracture seen in some parts of the country is the

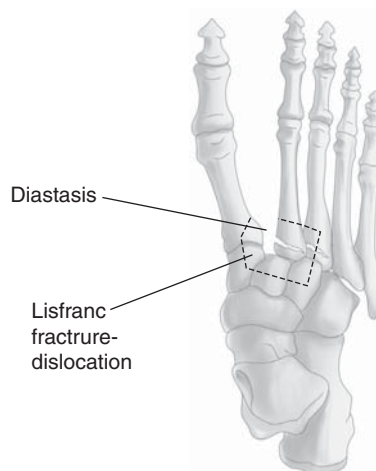


FIG. 38-8. The rare Lisfranc injury consists of a tarsal–metatarsal dislocation. A fracture of the second metatarsal with dislocation is the most common pattern.

“snowboarder’s fracture” where the lateral process of the talus is injured during dorsiflexion of the ankle and inversion of the hindfoot. Fractures of the talus can be managed with a posterior splint and nonweight bearing unless there is significant displacement. All talar fractures should be followed-up closely due to the risk of osteonecrosis.

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QUESTIONS

- Upon starting your shift in the ED, you assume the care of a 10-year-old male involved in a motor vehicle accident who suddenly becomes hypotensive. Which of the following unstable fractures is most likely to be found in this patient?
 - Avulsion of the anterior superior iliac spine (ASIS)
 - Isolated femoral shaft fracture
 - Bilateral pelvic rami fractures
 - Unilateral fracture of the ischium
 - Fracture of the iliac wing
- An 8-year-old young girl is jumping on a trampoline when she suddenly lands awkwardly and screams out in pain. She is in severe discomfort, unable to ambulate, and presents to the ED via ambulance. Radiographs confirm a femoral acetabular hip dislocation. Which of the following statements are TRUE concerning hip dislocations in the pediatric patient?
 - Hip dislocations in children require high-energy trauma like a motor-vehicle accident.
 - Most patients present with the leg shortened, extended at the hip, and externally rotated.
 - The majority of dislocations are posterior and may be complicated by fractures of the acetabulum or proximal femur.
 - The best method for reduction is flexion followed by gentle external rotation.
 - It is appropriate to arrange for an unreduced hip dislocation to undergo a prolonged ground transport to a facility with a pediatric orthopedist available.
- A 12-year-old girl is in the ED with an isolated femur fracture along the shaft. Which of the following would you expect to find as part of her ED presentation and hospital management?
 - A history of injury due to a sporting injury
 - Shock-like symptoms due to her femur fracture
 - Posterior long leg splint and nonweight bearing discharge
 - Hospitalization for pain control, observation, and surgery
 - Fracture location most likely proximal shaft near lesser trochanter
- A 13-year-old basketball player hyper-extends his knee during a game and presents to the ED with an acute traumatic effusion. His dad says the mechanism looked just like when his dad ruptured his ACL a few years ago. Which of the following fractures would be most suspected with this mechanism?
 - Salter–Harris II of the distal femoral physis
 - Tibial spine (tibial plateau) avulsion
 - Transverse patella fracture
 - Tibial tubercle avulsion
 - Midshaft femur fracture
- A 15-year-old softball player presents to the ED with right knee pain and swelling following an injury where she planted her right foot and then “twisted” her bent knee to make a throw. She immediately fell to the ground in pain and felt a “pop” when she tried to straighten her knee. The most likely diagnosis based on this history is which of the following?
 - ACL rupture
 - Medial collateral ligament sprain
 - Femur-on-tibia knee dislocation

- D. Patellar tendon rupture
 - E. Patella dislocation
6. A 3-year-old child presents to the ED because he refuses to bear weight on his left leg. The family states he was doing well earlier in the day, and they did not directly observe any injury prior to finding him crying and refusing to walk. His exam is otherwise unimpressive, but you suspect a toddler's fracture. Which radiograph is likely to BEST show evidence of a fracture?
- A. AP view of tibia
 - B. Lateral view of tibia
 - C. Oblique view of tibia
 - D. Follow-up radiographs of the tibia in 2 weeks
 - E. Mortise view of the ankle
7. A 14-year-old boy presents to the ED with marked ankle pain, swelling, and the inability to bear weight after sliding into second base. The radiologist calls over to say this is a triplane fracture. What classification of Salter–Harris (SH) injury to the physis is this type injury?
- A. Salter–Harris I
 - B. Salter–Harris II
 - C. Salter–Harris III
 - D. Salter–Harris IV
 - E. Salter–Harris V
8. A 14-year-old basketball player lands on an inverted foot and presents to the ED complaining of foot pain over the lateral border of the foot. Which of the following is the most likely injury?
- A. Sprain of the anterior talofibular ligament (ATFL)
 - B. Avulsion fracture of the 5th metatarsal
 - C. Jones fracture
 - D. Iselin “disease” (apophysitis of the 5th metatarsal)
 - E. Lisfranc fracture/dislocation
9. A 15-year-old snowboarder presents to the ED with persistent anterior ankle pain following an ankle injury yesterday. He states initial radiographs at the ski clinic were negative, but there is still too much pain to bear weight. He's concerned and wants to know what you think is the most likely diagnosis?
- A. Talar neck fracture
 - B. Calcaneal fracture
 - C. Lateral ankle sprain
 - D. Fracture of the lateral process of the talus
 - E. Phalangeal fracture
10. The Ottawa ankle rules have been validated in which of the following groups following an acute ankle injury?
- A. Adults
 - B. Adolescents

- C. Children younger than 5 years
- D. All the above
- E. A and B only

ANSWERS

1. C. Bilateral pelvic rami fractures are frequently unstable and may require external fixation or open reduction/internal fixation. These injuries are usually the result of very high-energy trauma such as motor vehicle accidents and associated with significant abdominopelvic injuries. Avulsions of the ASIS are stable injuries that frequently occur in adolescent athletes. Isolated femoral shaft fractures in young patients are stable, and not usually the source of hypotension like in adults. A unilateral fracture of the ischium would be expected to be a stable injury that can be managed nonoperatively. Fractures of the iliac wing are rare, but not traditionally associated with severe abdominopelvic injury.
2. C. The overwhelming majority of dislocations are posterior in nature and are frequently complicated by fractures surrounding the hip joint. A CT scan is often helpful following reduction in order to look for any intra-articular fractures or bony fragments. Hip dislocations in children do not require high-energy trauma, and they may result from trivial falls or sporting injuries due to increased joint laxity and a shallow, nonossified acetabulum. With posterior dislocations being most common, patients usually present in severe pain with the leg shortened, flexed at the hip, and internally rotated. The best method for reduction of a posterior hip dislocation is flexing the hip to 90° and applying axial traction to the thigh. Hip dislocations have an increasingly serious risk of AVN and osteoarthritis along with being more difficult to reduce if the reduction is delayed more than 6 hours. Attempts at reduction should be prompt and reduction confirmed by radiographs or CT.
3. D. Most patients with isolated femur fractures have a tremendous amount of pain requiring hospital admission, observation, and usually surgical management with a rod or flexible nail. Fractures of the shaft usually involve high-energy mechanisms such as motor vehicle accidents or significant falls. Isolated femur fractures in younger patients do not regularly result in hypotension or shock-like symptoms, and if these are present one should strongly investigate for associated injuries to internal organs. A posterior long leg splint and discharge would be inappropriate management for most femur fractures in a 12-year-old girl. The fracture location is most commonly along the mid-shaft or middle third of the femur.

4. B. The tibial spine (plateau) fracture occurs in a similar mechanism to the ACL rupture in adults. In adolescents, the bone where the ACL inserts onto the tibia may actually be weaker than the ACL resulting in a fracture. It has also been increasingly noted that ACL ruptures may also occur at this age, so clinicians should consider each possibility with a hyperextension, or twisting, injury of the knee. A Salter–Harris II fracture of the distal femur is less likely with the described mechanism. Transverse patella fractures are less common in youths and the mechanism is usually a direct fall. Tibial tubercle avulsions do start to occur around this age, but the mechanism is a forceful contraction of the quadriceps. Midshaft femur fractures are rarely the result of a sporting injury and are associated with higher energy trauma.
5. E. Patella dislocations commonly present in adolescent athletes with a twisting-type mechanism on a bent knee with a planted foot. Ninety percent will spontaneously reduce with knee extension prior to presentation in the ED. The most helpful examination maneuver is the apprehension test. ACL ruptures classically occur with a hyperextension and twisting mechanism, but they do not “pop” with knee extension. Medial collateral ligament sprains occur when there is a valgus force to the knee. Femur-on-tibia dislocations are rare and result from high-energy trauma. Patellar tendon ruptures are also rare and patients cannot extend the knee following this injury.
6. D. Follow-up radiographs of the tibia in 2 weeks offer the BEST opportunity to show callous formation and confirm your suspicion of a toddler’s fracture. Other tibia views (AP, lateral, oblique) may potentially show the toddler’s fracture and the oblique is often helpful in acute situations. The diagnosis is often suspected based on the acute history and lack of significant findings (fever, swelling, etc) other than refusal to bear weight and tenderness with palpation or external rotation of the foot while the ankle is held in a neutral position. The mortise view is not traditionally helpful in diagnosing toddler’s fractures.
7. D. The triplane fracture is a Salter–Harris (SH) IV injury that exists in three planes of the tibia as it crosses the epiphysis, travels along the physis, and then exits out through the metaphysis. This injury involves the articular surface of the ankle and requires orthopedic consultation for surgical repair. CT scanning is often helpful to better delineate the fracture fragments and plan for surgery. SH I injuries of the ankle are most common in the distal fibula with some degree of tenderness and swelling over the physis but otherwise normal radiographs. SH II injuries may be present in the tibia or fibula and involve fractures through the physis exiting into the metaphysis. SH III fractures are unique to adolescents and known as “Tillaux” fractures where the fracture extends across the physis and exits onto the articular surface through the epiphysis. Both SH III and IV of the tibia warrant orthopedic consultation since they are intra-articular fractures. SH V fractures are due to axial loading injuries and best diagnosed in retrospect following growth arrest.
8. B. Foot pain following an acute ankle injury is most likely an avulsion fracture of the 5th metatarsal where the peroneus brevis attaches. The foot should always be palpated closely following an acute ankle injury. A sprain of the ATFL is common with the described mechanism, but it would not present as pain over the lateral border of the foot. The Jones fracture is rare in younger patients and presents as more of a chronic injury with a fracture at the metaphyseal–diaphyseal junction. Iselin apophysitis of the 5th metatarsal is also where the peroneus brevis attaches, but it presents as more of a chronic problem and may be bilateral. Lisfranc fracture/dislocations are rare and the most common pattern is a fracture at the base of the 2nd metatarsal with subsequent dislocation of the tarsal–metatarsal joint.
9. D. The “snowboarder’s fracture” is a fracture of the lateral process of the talus due to forced inversion of the hindfoot on a dorsiflexed ankle. The pain is anterolateral along the joint line, and the fracture may be difficult to detect on initial radiographs. Maintain a high degree of suspicion for this injury in select patients, and consider a CT if initial radiographs are negative. Talar neck fracture is the most common fracture of the hindfoot, but it would not be expected in the described scenario. A calcaneal fracture is very rare and classically results from a fall from significant height. A lateral ankle sprain is a common sporting injury, but along with the ability to bear weight, pain is typically described as along the lateral ankle. A phalangeal fracture is a very common injury of the forefoot, but these typically result from a direct traumatic force that is easy to recognize.
10. E. The Ottawa ankle rules have been validated in adults and youth past 5 years of age. Caution was recommended in utilizing the rule in children younger than 5 years of age due to the high likelihood of fracture following an ankle injury. Physicians should be familiar with the Ottawa ankle rules and understand when to order the appropriate foot or ankle radiographs.

39 SOFT TISSUE INJURY AND WOUND REPAIR

D. Matthew Sullivan

HIGH-YIELD FACTS

- Lacerations and soft tissue injuries are the most common reasons for children to present to the ED.

SKIN AND SOFT TISSUE ANATOMY AND BIOMECHANICS

- Cosmetically pleasing scars result when the long axis of the wound is in the direction of maximal static skin tension (Fig. 39-1)
- A wound crossing a joint may result in a significant contracture, since scars do not have the elasticity of uninjured tissue.
- It is essential to warn the child and parent of possible adverse cosmetic outcomes.

CLASSIFICATION OF MINOR INJURIES

LACERATIONS

- Lacerations are cuts through the skin and, after contusions, are the most common type of soft tissue injury seen in the ED.
- The three main classes of lacerations are shear, tension, and compression.

ABRASIONS, CONTUSIONS AND HEMATOMAS

- Management of all contusions involves elevation of the injured area, application of ice packs intermittently for the first 24 to 48 hours, and careful monitoring of circulation and neurologic function.

PREHOSPITAL CARE

- Use direct manual pressure or a pressure dressing to control blood loss.

HISTORY AND PHYSICAL EXAMINATION

- The management of soft tissue injuries can be compartmentalized by addressing *host factors*, *wound factors*, and *after-care*.

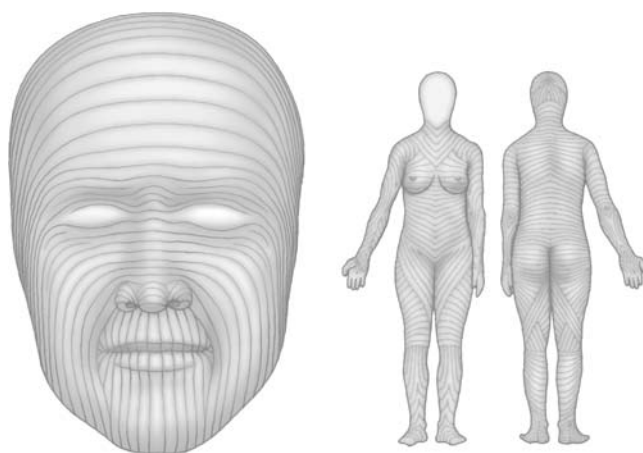


FIG. 39-1. Lines of skin tension.

- The history of the injury should include timing, mechanism, extent, and the potential for possible contamination of the wound.
- Obtain a detailed history of the patient's *host factors* with particular attention to the child's immunization status, past medical history, allergies, medications, and what prehospital wound care was performed prior to arrival.
- Address *wound factors* to assess the length and depth of the injury, circulatory status, motor and sensory function, the presence of foreign bodies and contaminants, and the involvement of underlying structures (nerves, tendons, muscles, ligaments, vessels, bones, joints, and ducts).
- Whereas the sensory examination must precede the administration of anesthesia, the remainder of the examination should be performed with adequate anesthesia.
- Use a toy or light pen that requires manipulation by the child to help in evaluating motor function. Involve Child Life providers and observe the child at play to evaluate wounds that might involve underlying motor structures.

MANAGEMENT

INSTRUMENTS, SUTURES, STAPLES, TAPE, AND TISSUE ADHESIVES

- Most wound repairs can be accomplished with basic instruments and supplies (Fig. 39-2).
- Wound repair can be accomplished with reverse precision point cutting needles that are manufactured in various sizes, curvatures, and paired with differing thread types.



FIG. 39-2. Standard laceration equipment.

- Each suture type has inherent characteristics that are suited for specific uses. Strong consideration should be given to the use of absorbable suture externally in order to eliminate the need for return visit for suture removal.
- Surgical staples are a useful alternative to suturing for selected wounds.
- Steri-strips are effective for the closure of small linear lacerations that are under minimal tension (Fig. 39-3). Adhesives have slightly less tensile strength across wound edges and are therefore suited for use in low-tension wounds.
- With the right choice of wound, noninvasive repair can be faster, less painful, require no suture removal follow-up, and results in patient and parental satisfaction.
- Adhesives should not be used in wounds with high mobility.
- Use a ribbon of petroleum as a barrier to protect against inadvertent instillation of the adhesive into the eye.
- Care should be taken to ensure that adequate anesthesia, wound irrigation, and wound exploration occur regardless of the method of closure.

ANALGESIA, LOCAL ANESTHESIA, NERVE BLOCKS, AND SEDATION

- Most wounds are adequately anesthetized using local infiltration of lidocaine, 1% to 2% with or without epinephrine. Commercial epinephrine containing anesthetics are safe and their use in regions supplied by end-arteries is safe.
- For plain lidocaine and lidocaine with epinephrine, 4.5 and 7 mg/kg are the recommended maximum doses, respectively.

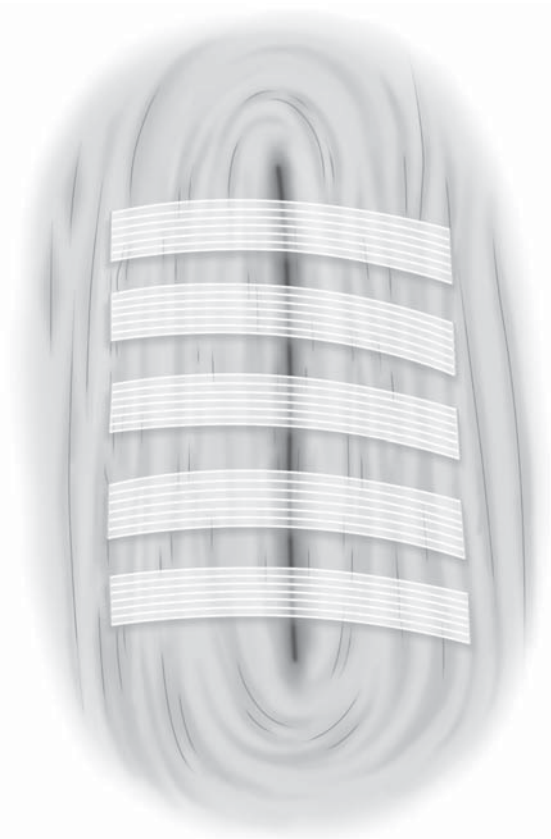


FIG. 39-3. Skin-closure tapes should be applied perpendicular to the wound edges and spaced so that the edges do not gape.

- Bicarbonate added to lidocaine (1:10) may also reduce the pain of injection.
- Topical anesthetics prior to injectable anesthetics should be strongly considered.
- When necessary, use chemical sedation to permit adequate wound management.
- You may immobilize a child using a folded sheet or a commercially available papoose board after a thorough discussion with the parents.

WOUND CLEANING AND PREPARATION

HEMOSTASIS

- Hemostasis is usually achieved by applying direct pressure for 10 to 20 minutes

ANTISEPSIS AND SCRUBBING

- Avoid mechanical scrubbing of the wound unless there is gross contamination.

HAIR REMOVAL

- Hair removal is not necessary unless it is directly within the wound edges.

IRRIGATION

- Irrigation with 5–8 psi is the method for removing bacteria and debris from most wounds.
- Low-pressure irrigation does not adequately remove bacteria and debris from a wound.
- The pressure delivered by a simple assembly consisting of an 18- to 20-gauge plastic catheter or needle attached to a 30-mL syringe is 6 to 8 psi.
- With irrigation pressures in the 5 to 8 psi range, tap water has been shown to be equivalent to normal saline.
- A consequence of irrigation is splatter, which should be minimized using one of many techniques.

FOREIGN BODY EVALUATION

- The failure to diagnose foreign bodies in wounds is a frequent cause of litigation against emergency physicians.
- Parents and patients should always be warned of potential residual contaminants in dirty wounds.

DEBRIDEMENT

- Debridement is necessary in the management of contaminated wounds or wounds with nonviable tissue.
- Subcutaneous tissue of the wound may be undermined to avoid excess tension on the wound.

PRIMARY WOUND CLOSURE

- Primary closure should be performed on lacerations that have been recently sustained (<24 hours on the face and <12 hours on other areas of the body), are relatively clean, and have minimal tissue devitalization.
- Older wounds and wounds with significant host factors should be managed with delayed primary closure.
- Identify all the injured layers, such as fascia, subcutaneous tissue, muscle, tendon, and skin.
- A laceration closed in layers usually does not need large or tight skin sutures to complete the closure.
- In the hands and feet, placement of deep sutures increases the risk of infection, and should be avoided.
- Use 3–0 suture for tissues with the need for strong tension, such as fascia in an extremity, and 6–0 suture

for tissues with light tension, such as the subcutaneous tissue of the face.

BURIED STITCH

- Deep (buried) sutures provide additional support to the wound after the skin sutures are taken out, minimize dead space within the wound, and help deter the development of pitting in the injured region caused by inadequate healing of the deep tissues.
- The subcuticular stitch is a running buried suture at the dermal–epidermal junction that is used for skin closure (Fig. 39-4) and avoids skin suture marks.

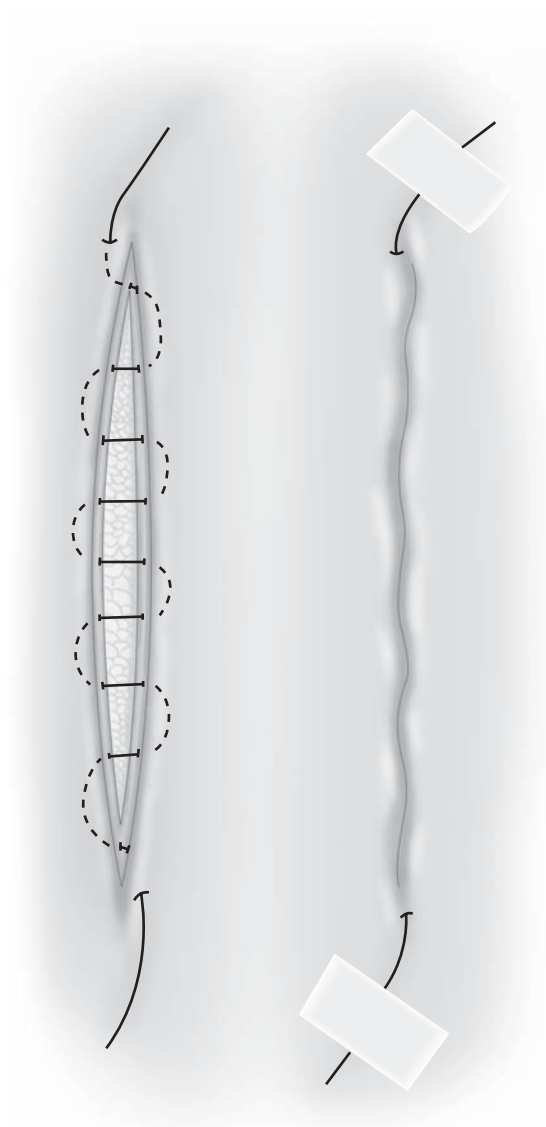


FIG. 39-4. Subcuticular stitch (see the text for discussion).

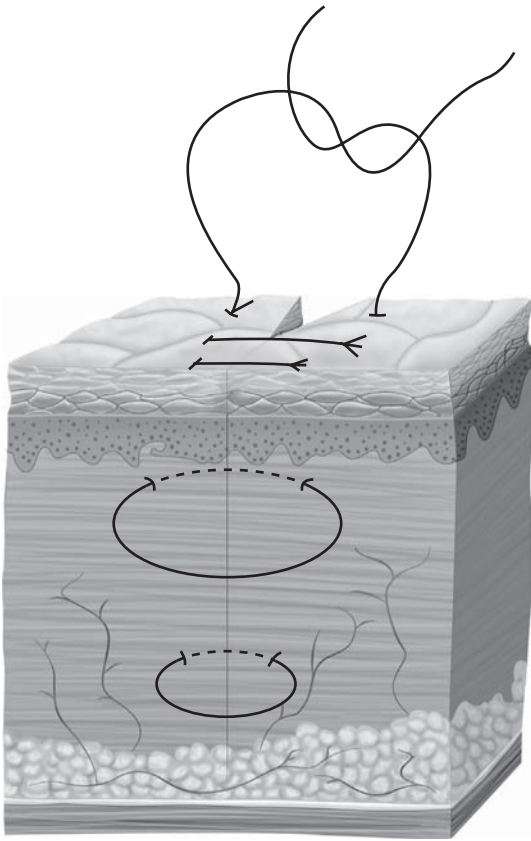


FIG. 39-5. Simple interrupted stitch (with buried subcutaneous stitch) (see text for discussion).

SKIN CLOSURE

- Place sutures such that the same depth and width is entered on both sides of the incision.
- A key to cosmetically acceptable closure is edge eversion obtained by entering the skin at a 90° angle.

SIMPLE INTERRUPTED STITCH

- The simple interrupted stitch is used most frequently for skin closure (Fig. 39-5).

RUNNING STITCH

- The running or continuous stitch is rapid, removal is easier, provides effective hemostasis, and distributes tension evenly along its length (Fig. 39-6).

MATTRESS STITCHES

- The horizontal mattress stitch can be used for single-layer closure of lacerations under tension (Fig. 39-7).
- The half-buried horizontal mattress stitch (corner stitch) is the suture of choice for closure of complex wounds with angulated (V-shaped) flaps (Fig. 39-8).

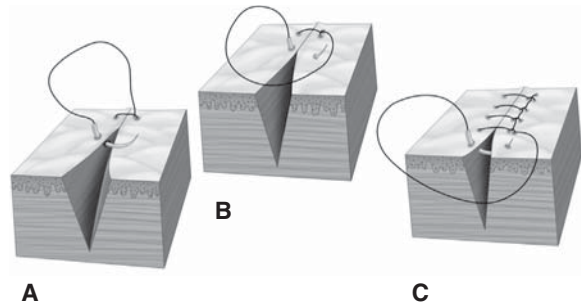


FIG. 39-6. Continuous single lock stitch (see the text for discussion).

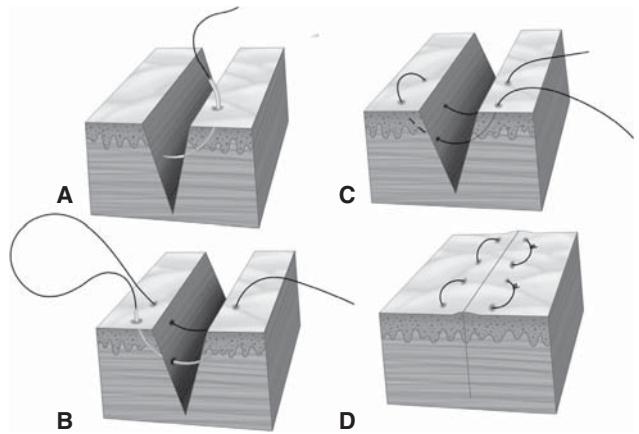


FIG. 39-7. Horizontal mattress stitch. (A) The needle is passed 0.5–1 cm away from the wound edge deeply into the wound. (B) The needle is then passed through the opposite side and re-entered the wound parallel to the initial suture. (C) One must enter the skin perpendicularly to provide some eversion of the wound edges and must enter and exit both the wound and skin at the same depth, otherwise “buckling” and irregularities occur in the wound margin. (D) The suture loop is then tied as shown.

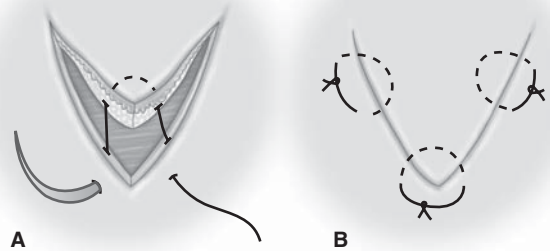


FIG. 39-8. (A, B). Half-buried horizontal mattress stitch. This minimizes the vascular compromise at a corner flap (see the text for discussion).

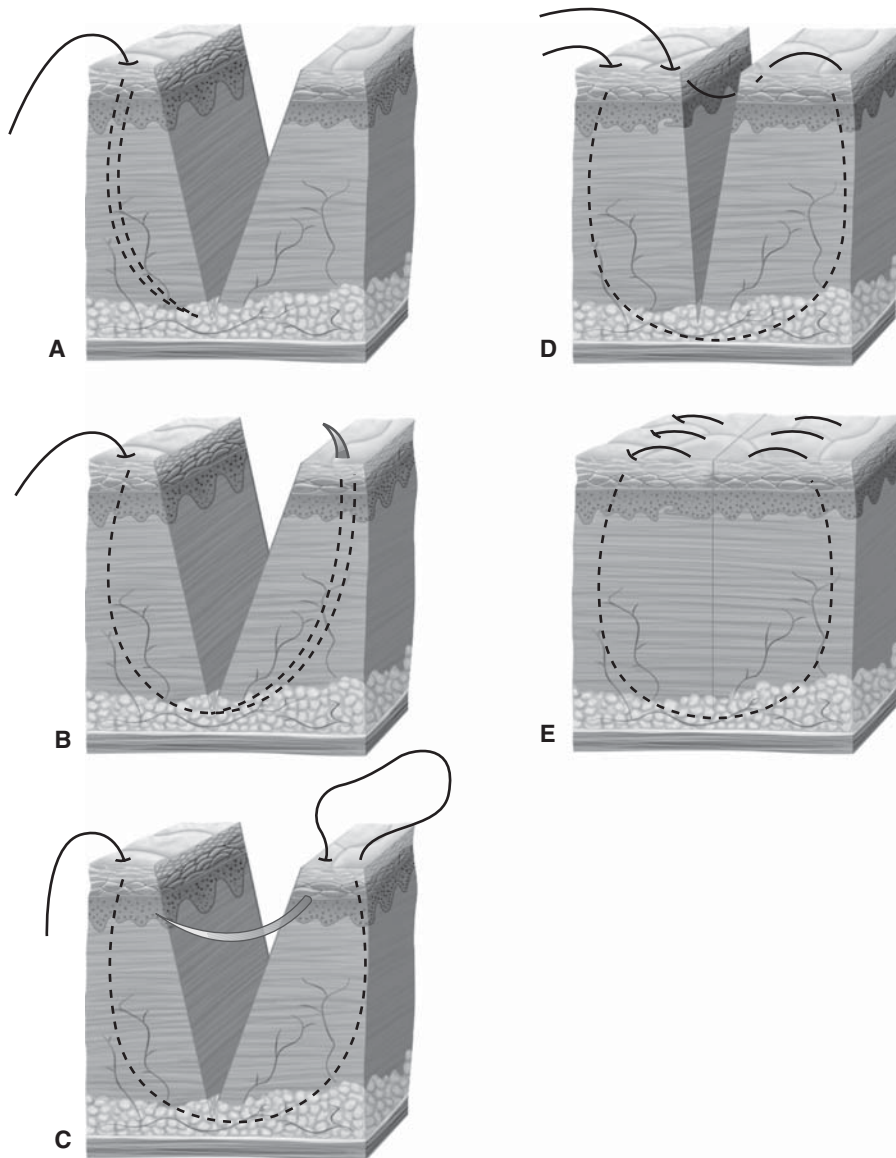


FIG. 39-9. (A–E). Vertical mattress stitch (see the text for discussion).

- The vertical mattress stitch is helpful to evert skin edges (Fig. 39-9).

KNOTS

- The knot used most commonly in the ED repair of lacerations is the surgeon's knot followed by one to four half-knots, usually formed as instrument ties.

CORRECTION OF DOG-EARS

- A dog-ear can be corrected using the following technique (Fig. 39-10).

SECONDARY CLOSURE

- Secondary closure is a technique that allows wounds to heal by granulation and reepithelialization.

DELAYED PRIMARY (TERTIARY) CLOSURE

- Delayed primary closure with sutures is a repair option for wounds that are deemed too contaminated to close at the initial visit and is performed 3 to 5 days after they have been initially cleansed, debrided, and dressed appropriately.

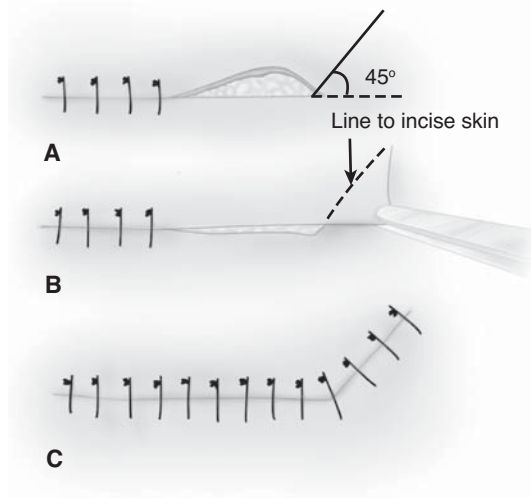


FIG. 39-10 (A–C). The dog-ear (see the text for discussion).

WOUND DRESSING, DRAINS, AND IMMOBILIZATION

- Sutured and stapled lacerations heal best in a moist environment.
- Topical antibiotic ointment has been shown to be clinically less effective after about 3 hours.
- Use a layer of sterile gauze or adhesive bandage (Band Aid) to cover the ointment or nonadherent dressing.
- If there is potential for the formation of a hematoma apply a pressure dressing, taking care to avoid compression of arterial, venous, and lymphatic circulations.
- Splint a wound overlying a joint in the position of function for 7 to 10 days.

ANTIBIOTIC USE AND TETANUS PROPHYLAXIS

- More than 95% of wounds treated in the ED heal without complications with appropriate wound care alone.
- When antibiotics are indicated, their effectiveness depends on early administration.
- Antibiotics should be given to patients who present with
 - a wound infection
 - intraoral lacerations
 - wounds that are more than 24 hour old
 - contaminated or devitalized wounds
 - immunocompromise (sickle cell disease, diabetes, steroid use, or lymphoma).
 - involvement of cartilage, joint spaces, tendon, or bone

- wounds of the hand
- all noncanine bites
- Wounds that are heavily contaminated allow them to close by secondary intention or undergo delayed primary closure.
- Most infections are caused by sensitive staphylococci and streptococci that will respond to penicillin, first-generation cephalosporins, or erythromycin for penicillin-allergic patients. Consideration of methicillin-resistant *Staphylococcus aureus* infection as a pathogen should be given, particularly to those patient who worsen or do not improve with standard therapy.
- Dog bites in normal hosts with low-risk wounds will do well with meticulous wound care, closure, splinting, elevation, and close follow-up without antibiotics.
- If the child has a tetanus-prone wound and was not immunized or only partially immunized give human tetanus immune globulin (HTIG) 250 U IM and complete or initiate primary immunization. If a child has completed primary immunization and has received appropriate boosters, then HTIG is not required.

POSTOPERATIVE WOUND CARE AND SUTURE REMOVAL

- Inform patients that all wounds heal with scars, regardless of the quality of care.
- Inform patients about the possibility of an infection and residual foreign body in the wound.
- Keep a clean dressing in place and the wound clean for 24 to 48 hours followed by 24 hour wound check and daily dressing changes.
- Suture removal should be done at an appropriate time to prevent suture track marks and stitch abscesses resulting from late removal (Table 39-1).
- Instruct your patients to avoid sun exposure to the scar and use sunscreen to avoid a hyperpigmented scar.
- To prevent scar formation, the mechanical action of rubbing may have positive effects.
- Vitamin E preparations should be avoided because of potential local hypersensitivity reactions.

MANAGEMENT OF SELECTED INJURIES

ABRASIONS

- Clean abrasions and dress them with a nonadherent dressing.
- Remove any foreign bodies (eg, gravel, dirt, or tar) to avoid infection or tattooing (“road rash”) of the wound.

TABLE 39–1 Repair of Soft Tissue Injuries by Body Location

LOCATION	TYPE OF CLOSURE	SUTURE REMOVAL
Scalp	Single tight layer with simple interrupted, vertical mattress, or horizontal mattress for hemostasis; galea requires close approximation, but preferably with single-layer closure	7–10 d
Pinna (ear)	Simple interrupted; stent dressing	4–6 d
Eyebrow	Layered closure	4–5 d
Eyelid	Horizontal mattress	3–5 d
Lip	Three layers (mucosa, muscle skin) if through and through, otherwise two layers	3–5 d
Oral cavity	Simple interrupted or horizontal mattress	7–8 days or allow to dissolve
Face	If full-thickness, layered closure	3–5 d
Neck	Two-layered closure	4–6 d
Trunk	Single or layered closure	7–12 d
Extremity	Single or layered; splint if over joint	10–14 d (joint) 7–10 d (other)
Hands and feet	Single-layer closure with simple interrupted or horizontal mattress; splint if over joint	10–14 d (joint) 7–10 d (other)
Nail beds		Allow to dissolve

SCALP LACERATIONS

- The presence of a rich vascular supply and vessels that tend to remain patent when cut are responsible for the profuse bleeding associated with scalp injuries.
- Bleeding is stopped by direct pressure, epinephrine infiltration, and rapid closure.
- Scalp wounds are best closed with a single layer of sutures that incorporates the skin, the subcutaneous fascia, and the galea.
- In order to find the ends of the tied sutures these should be left longer than usual, and the use of blue nylon may also facilitate removal.

FOREHEAD LACERATIONS

EYELID LACERATIONS

- A thorough eye examination must to be performed whenever there is a laceration of the eyelid or peri-orbital region.
- Ensure that the levator palpebrae muscle and its tendinous attachment to the tarsal plate are intact, or ptosis may result.
- A laceration to the medial aspect of the lower lid often involves the lacrimal duct, which requires specialized repair by an ophthalmologist.
- If consultation is not required, close lid lacerations in a single layer with 6–0 suture, taking care to avoid skin inversion.

EAR LACERATIONS

- Anesthesia of the external ear is simply accomplished with a field block around the ear.

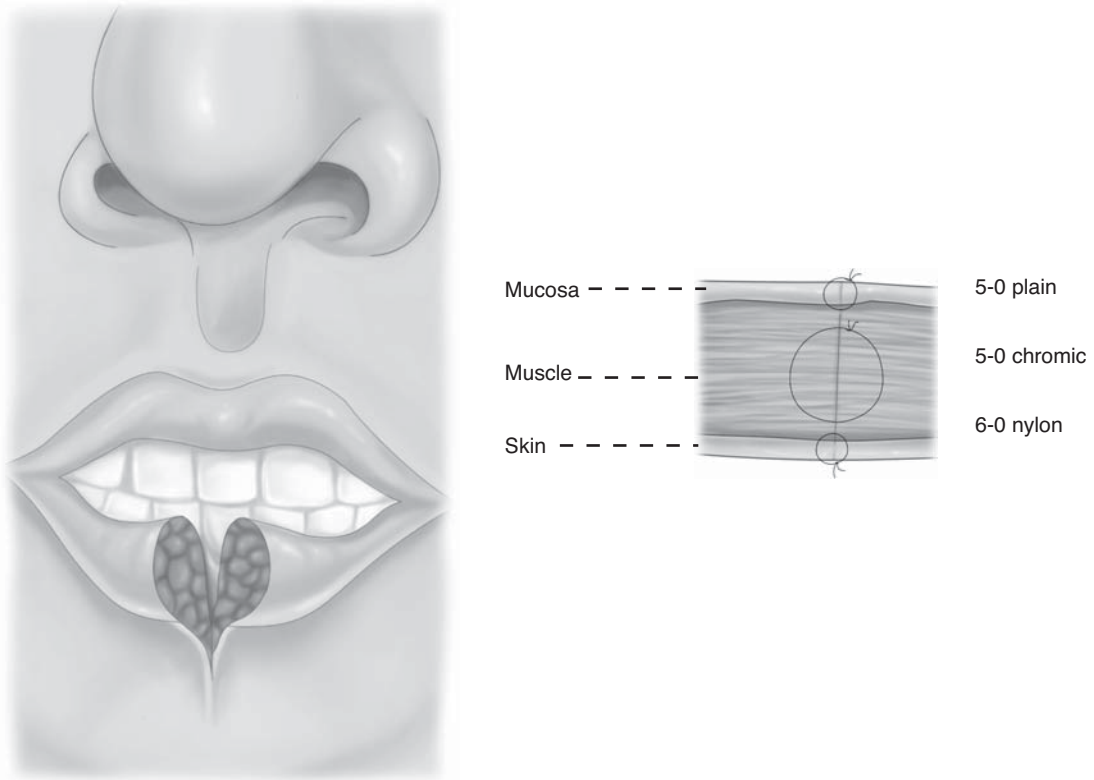
- Approximate cartilage with 5–0 absorbable suture material placed through the posterior and anterior perichondrium.
- Approximate the visible surface of the ear using 5–0 or 6–0 suture, ensuring approximation of landmarks and full coverage of all cartilage.
- After repair, dress the ear with an anterior and posterior compression dressing. This prevents accumulation of a perichondral hematoma, which can lead to necrosis of cartilage and subsequent deformity (“cauliflower ear”).

LIP LACERATIONS

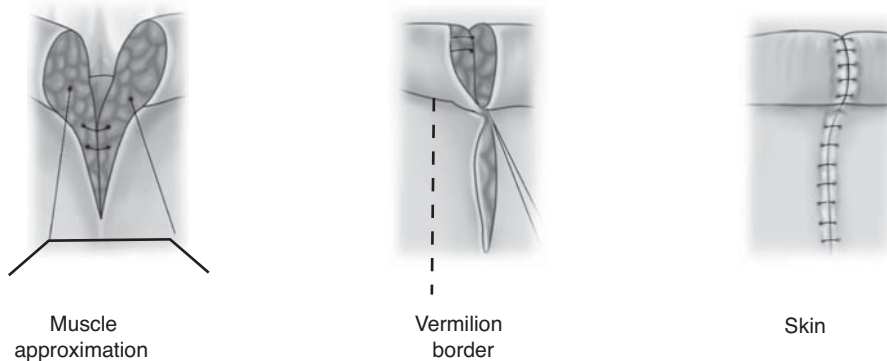
- Lip lacerations require careful attention to visual landmarks to ensure a good cosmetic result (Fig. 39-11).
- As local infiltration of anesthetic obscures the lip’s landmarks, perform a mental nerve block for the repair of lower lip or an infraorbital nerve block for upper lip lacerations.
- Through-and-through lip lacerations often require three-layer closure.

FINGERTIP INJURIES

- Prognosis of distal amputations depends on how much of the tip is lost.
- More proximal amputations uniformly require consultation with a hand surgeon.
- For fingers, an extensive procedure is rarely required for treatment of a paronychia.
- Paronychias of the toes are often caused by ingrown toenails.



A



B

FIG. 39-11. Repair of through-and-through lip laceration.

- If the hematoma involves less than 25% to 50% of the nail bed, trephinate the nail using one of a number of techniques.
- For subungual hematomas involving >50% of the nail bed, repairing the underlying nail bed laceration has minimal benefit.
- If there is an underlying fracture of the distal phalanx, splint the finger and prescribe antibiotics appropriate for an open fracture.

PUNCTURE WOUNDS TO THE FOOT

- Puncture wounds, most often to the foot, have a very low potential to result in significant morbidity.
- Patients who present quickly after puncture wounds, have normal host factors, and no evidence for retained foreign body may be discharged without antibiotics and re-evaluated after 48 hours of elevation, nonweight bearing, and meticulous wound care (Fig. 39-12).



FIG. 39-12. Puncture wound through tennis shoe.

CONSULTATION GUIDELINES

- Specialty consultation should be considered for the following:
 - Complex or extensive wounds which exceed your resources or skill set.
 - Wounds with large tissue defects not amenable to repair in the ED.
 - Wounds in which there is tendon, nerve, joint, or critical vessel involvement.
 - Lacerations involving the parotid or lacrimal ducts.
 - Lacerations of the eyelid tarsal plates.
 - Lacerations over fractures.
 - Facial lacerations in which cosmetic results are a concern.
 - Wounds about which there is physician uncertainty.

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QUESTIONS

1. A child presents to the ED with a laceration to her extremity. The parents are asking about whether or not the wound needs to be sutured and if other possibilities are available. Wound adhesives should not be used for wounds in which of the following locations?
 - A. Face
 - B. Scalp
 - C. Over a joint
 - D. Lower extremity
 - E. Hand
2. A 2-year-old child presents with a linear laceration of the right index finger. Which of the following agents is appropriate for local anesthesia?
 - A. Lidocaine, 1%
 - B. Lidocaine, 2%
 - C. Lidocaine, 1% with epinephrine
 - D. Lidocaine, 2% with bicarbonate
 - E. All of the above
3. A 5 year-old boy presents with a laceration to the chin. He is very anxious during examination. Which of the following is true regarding the use of a topical anesthetic prior to using an injectable anesthetic in this patient?
 - A. Effective but results in delayed healing and should not be used
 - B. It is ineffective
 - C. Is controversial
 - D. Effective and should be strongly considered
 - E. Minimally effective and result in unacceptable delays
4. Which of the following statements regarding wound cleaning is correct?
 - A. Low pressure irrigation (<5 PSI) is satisfactory for most wounds.
 - B. Tap water has been shown to be equivalent to sterile saline.
 - C. The pressure delivered by an 18-gauge catheter attached to a 30-mL syringe is 10-12 PSI.
 - D. The only effective way to remove bacteria and debris from wounds is debridement.
 - E. High pressure irrigation should not be used due to an unacceptable degree of splatter.
5. Primary wound closure should be performed for the laceration in which of the following circumstances?
 - A. Clean wound of the face that occurred 36 hours prior to arrival.
 - B. Relatively clean wound of the lower extremity that occurred 10 hours prior to arrival.
 - C. Wound of the lower extremity contaminated with dirt and gravel but which is clean after irrigation and debridement.
 - D. Wound that is clean but has significant tissue devitalization.
 - E. Clean wound of the hand that occurred 18 hours prior to arrival.
6. A 6 year-old girl presents with a grossly contaminated wound of her leg. The wound is irrigated and debrided and managed with delayed primary closure. When should closure occur?
 - A. 24–36 hours
 - B. 48–72 hours
 - C. 3–5 days
 - D. 7–10 days
 - E. 14 days
7. A 13 year-old male has a scalp laceration repaired with staples and a facial wound repaired with sutures. The parents ask how they should best care for these wounds at home. Sutured and stapled lacerations have been shown to heal best in what kind of environment?
 - A. Moist and covered
 - B. Dry and covered
 - C. Open
 - D. Moist and dry are equivalent
 - E. Open but covered when in dirty environment

8. Prophylactic antibiotics are indicated in which of the following laceration cases?
 - A. Lip laceration
 - B. Wounds > 12 hour old
 - C. Wound in a patient with sickle cell trait
 - D. Wound in a patient on steroids
 - E. Wound over a joint space
 9. A 2 year-old boy from Mexico presents with a leg laceration from a fall. The parents state he has never gotten any of his childhood immunizations. Tetanus immunoprophylaxis for a previously unimmunized child who presents with a clean laceration consists of which regimen in the ED?
 - A. Tetanus toxoid
 - B. Human tetanus immune globulin
 - C. Tetanus toxoid and human tetanus immune globulin
 - D. Tetanus toxoid and human tetanus immune globulin and referral for completion of the tetanus toxoid series
 - E. Antibiotics, tetanus toxoid, human tetanus immune globulin and referral for completion of the tetanus toxoid series
 10. A 10 year-old presents to the ED with a laceration to her arm. Which of the following statements is appropriate for use in discharge instructions for this patient and her parents after the laceration is repaired?
 - A. For clear lacerations, you can reassure patients and parents that the wound will heal with little or no scar.
 - B. You can reassure that no foreign body was present and that there is little possibility of infection.
 - C. Instruct to avoid sun on the scar and to use sunscreen to avoid a hyperpigmented scar.
 - D. Instruct to apply vitamin E preparation to augment healing and decrease scar formation.
 - E. Instruct parents to regularly rub a scar-detering preparation into the wound.
3. D. Topical anesthetics are effective and underused. They should be strongly considered.
 4. B. Tap water is equivalent to sterile saline. Low pressure irrigation is not sufficient to clean any wound. The pressure delivered by an 18-gauge catheter attached to a 30-mL syringe is 6–8 PSI. High pressure irrigation is effective in removing bacteria and debris from wounds. Splatter is a problem but can be ameliorated with various types of shield devices.
 5. B. Clean wounds can have primary closure if done within 12 hours, 24 hours on the face. Contaminated wounds should not be primarily closed even if they can be cleaned. The same is true for wounds with devitalized tissue.
 6. C. Delayed primary closure should be done in 3–5 days.
 7. A. Sutured and stapled lacerations heal best when maintained in a moist environment.
 8. D. Prophylactic antibiotics should be used for contaminated wounds, intraoral wounds, wounds greater than 24 hours old and for patients with certain host factors that make them susceptible to infection, such as sickle cell disease (not trait), diabetes, and immunocompromise. Steroid use is another of those host factors. Wounds that extend into a joint space would require antibiotics, but simply being located over a joint is not an indication for antibiotics.
 9. D. Unimmunized patients should receive tetanus toxoid and human tetanus immune globulin in the ED and should be referred for completion of the tetanus toxoid series. There is no need for antibiotics unless there are other indications.
 10. C. Sun exposure should be avoided to deter development of hyperpigmented scars. All lacerations heal by scarring and it is unwise to downplay this to patients or parents. Likewise, it is wise to advise all patients and parents that initial exploration of wounds cannot detect all foreign bodies and that retained foreign body as well as infection is always a possibility. Vitamin E preparations should not be used due to unacceptable rate of hypersensitivity reactions. There is inadequate evidence to recommend scar-detering preparations, although some believe that the mechanical act of rubbing may decrease scar formation.

ANSWERS

1. D. Adhesives should not be used in wounds which will be subjected to high mobility, such as those over joints. There is no specific contraindication for use in the other locations listed.
2. E. Lidocaine, 1% or 2 %, lidocaine with epinephrine and buffered lidocaine are all appropriate

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Section 5

RESPIRATORY EMERGENCIES

40 UPPER AIRWAY EMERGENCIES

Richard M. Cantor
Linnea Wittick

PATHOPHYSIOLOGY—UPPER AIRWAY CONSIDERATIONS

- The small caliber of the child's upper airway makes it vulnerable to occlusion resulting in greater baseline airway resistance.
- Any process narrowing the airway causes an exponential rise in airway resistance and work of breathing. As the child perceives distress, an increase in respiratory effort leads to increased turbulence and resistance.
- Young infants are primarily nasal breathers and any obstruction to the nasopharynx results in increased work of breathing.
- The large tongue of infants and young children can obstruct the oropharynx, especially in children with mental status changes.
- Maneuvers such as a head tilt or chin lift or the insertion of an oropharyngeal or nasopharyngeal airway assist in alleviating respiratory distress.
- The pediatric trachea is easily compressible secondary to incomplete closure of cartilaginous rings so overextension of the neck should be avoided.
- The cricoid ring is the narrowest portion of the upper airway.

PATHOPHYSIOLOGY—LOWER AIRWAY CONSIDERATIONS

- Pediatric patients possess a decreased number of alveolar and bronchiole subunits and a diminished pulmonary vascular bed, predisposing them to respiratory distress even after partial airway occlusion.

- Immaturity of the musculoskeletal system and the central nervous system contributes to the development of apnea and respiratory failure.
- Abdominal distention can worsen respiratory status by interfering with diaphragmatic excursion.
- Infants are less sensitive to hypoxemia secondary to the poor development of central respiratory control leading to poor response to disease states.

SIGNS OF RESPIRATORY DISTRESS

- Respiratory distress occurs when there is increased work of breathing or increased respiratory rate in order to meet the body's oxygenation and ventilation demands.
- Respiratory failure ensues when efforts cannot maintain adequate respiratory function.
- Tachypnea, the most common response to increased respiratory needs, is caused by hypoxia, hypercarbia, metabolic acidosis, pain, or CNS dysfunction.
- Intercostal, subcostal, sub- or supersternal and supraclavicular retractions, and nasal flaring are signs of respiratory distress.
- Grunting, caused by closure of the glottis at the end of expiration in order to generate positive end-expiratory pressure to prevent alveolar collapse, is an ominous sign.
- A "position of comfort" in upper airway disease includes an upright position while leaning forward, open mouth with jaw thrust forward and the inability to swallow.
- Pulsus paradoxus of >20 mm Hg is secondary to excessive negative intrathoracic pressure causing an increased venous return to the heart and left ventricular volume compromised.
- Cyanosis results from inadequate oxygenation within pulmonary vascular beds or inadequate oxygen delivery by the cardiovascular system.

- Mental status changes secondary to hypoxia include somnolence, agitation, irritability, and decreased feeding in infants.

GENERAL MANAGEMENT PRINCIPLES

- Provide supplemental oxygen using nasal prongs, mask with or without rebreather apparatus, face tent, or oxyhood.
- Prevent agitation by keeping the young child in the mother's arms or position of comfort.
- Avoid noxious stimuli and unnecessary procedures.
- Maintain normothermia and hydration.
- Continue to reassess the patient's status at appropriate intervals.

RESPIRATORY FAILURE

- Signs include decreased level of consciousness, progressive fatigue, increased work of breathing, increased respiratory rate, poor color, diaphoresis, retractions, grunting, nasal flaring, decreased air movement on auscultation, hypoventilations, and apnea.

ASSESSMENT AND MANAGEMENT OF SPECIFIC CLINICAL SCENARIOS

- Upper airway compromise results from inspiratory turbulence transmitted against a narrowed lumen.
- The narrowing may originate anywhere in the upper airway from the anterior nares to the subglottic area.
- Stridor in young infants most commonly originates from a congenital anomaly including macroglossia, laryngomalacia, or tracheomalacia.
- In the ED, the most common acute causes of upper airway obstruction are croup, foreign body aspirations, epiglottitis, tracheitis, peritonsillar abscess, and retropharyngeal abscess.

CROUP

- Accounts for 90% of stridor and fever.
- Results from viral infections including parainfluenza type 1, 2 and 3, adenovirus, respiratory syncytial virus, and influenza causing a swelling in the subglottic region.
- Seen in children aged 1–3 years most commonly in winter.
- Presents with brassy or barking cough, moderate temperature elevation, and stridor after several days of nonspecific upper respiratory infection symptoms.

- Symptoms worsen for 3–5 days followed by resolution over several days.
- Physical exam reveals a child with hoarse voice, coryza, and slight increase in respiratory rate.
- PA and Lateral neck radiographs may demonstrate the “steeple sign” with narrowing of the subglottic airway.
- Patients with mild croup, mild respiratory distress, and stridor only with activity or agitation may be treated as outpatients if well hydrated.
- Patients with stridor at rest benefit from nebulized racemic epinephrine (0.5 mg) and corticosteroids (dexamethasone 0.6 mg/kg po).
- Patients receiving racemic epinephrine deserve 2–3 hours of observation to watch for “rebound” stridor.
- Children with severe croup deserve admission and treatment with oxygen, mist, racemic epinephrine, and steroids.
- If intubation must be performed, an endotracheal tube 1 size smaller than that calculated for age should be used due to subglottic edema.

EPIGLOTTITIS

- A true airway emergency.
- In the past, it traditionally affected children 2–5 years, but with the advent of the Hib vaccine it is now more common in young infants and adults.
- Patients demonstrate acute onset of fever, sore throat, dysphagia, with progression to respiratory distress.
- The child assumes a position of comfort, sitting upright with mouth open and head, neck, and jaw extended.
- Symptoms include stridor, muffled voice, “toxic” appearance, and profound drooling.
- *Streptococcus pneumoniae*, *Staphylococcus aureus*, and group A-hemolytic *Streptococcus* have overtaken *Haemophilus influenzae* type B as causative agents.
- Noninfectious causes include trauma, burns, leukemia, and angioneurotic edema.
- Avoid all maneuvers that agitate the child including separating from parents, alteration of optimal posture, fearful procedures, and gagging.
- Radiographs will demonstrate a thickened epiglottis or “thumb sign.”
- Be prepared to provide supplemental ventilation at all times including BVM, ET intubation, needle cricothyrotomy, and tracheostomy.
- Ideally, intubate the patient in the operating room and obtain epiglottic cultures.
- After intubation, provide IV antibiotics such as a third-generation cephalosporin.

BACTERIAL TRACHITIS

- An infection of the subglottic region causing subglottic edema and pseudomembrane formation in the trachea and bronchi.
- More common than epiglottitis.
- Polymicrobial etiology—most commonly *S aureus*, *S pneumoniae*, *H influenzae*, *Pseudomonas*, and *Moraxella*.
- Average age 3 years of age, boys more often than girls, in the winter.
- Symptoms begin with a mild upper respiratory infection lasting 1–2 weeks with rapid deterioration with respiratory distress, high fevers, and toxic appearance.
- Physical exam shows stridor, tachypnea, retractions, barking cough, and often, wheezing.
- Radiographs show subglottic and tracheal narrowing, a ragged tracheal border. secondary to pseudomembrane formation, and possibly a concurrent pneumonia.
- Definitive diagnosis occurs with direct visualization of the normal epiglottis with pus and inflammation and pseudo membrane upon intubation.
- During tracheal intubation, use one size smaller ETT.
- Treat with broad spectrum antibiotics with a third-generation cephalosporin and vancomycin, frequent suctioning, and ICU monitoring.

RETROPHARYNGEAL ABSCESS

- Seen predominantly in children 1–3 years of age secondary to cervical lymphadenitis.
- Secondary to penetrating trauma of the posterior oropharynx in older children.
- Causative organisms include Group A hemolytic *Streptococcus*, *S aureus*, and anaerobes.
- Symptoms include high fever, muffled voice, difficulty swallowing, drooling, and occasionally stridor.
- Presentation with stiff neck and torticollis will often prompt a work-up for meningitis.
- May note swelling of the posterior pharynx.
- Lateral radiographs demonstrate prevertebral soft tissue swelling >7 mm at the level of the second cervical vertebrae or >14 mm at the sixth cervical vertebrae and normal epiglottis and aryepiglottic folds.
- CT may identify soft tissue swelling of abscess formation.
- Children without abscess formation are treated with IV antibiotics such as clindamycin and third-generation cephalosporins.
- Management should be coordinated by an ENT consultant.

- Definitive treatment involves intraoperative drainage of abscess.

PERITONSILLAR ABSCESS

- Most common deep infection of the head and neck.
- Affects children >8 years of age.
- Polymicrobial- Group A *Streptococcus*, *Peptostreptococcus*, *Fusobacterium*, and anaerobes.
- Present with dysphagia, drooling, ipsilateral ear pain with progression to trismus, dysarthria, and toxicity.
- “Hot Potato” voice results from splinting of the palatine muscles during speech.
- Pharynx will be erythematous with unilateral tonsillar swelling and displacement of the uvula toward the unaffected side and fluctuance felt above the tonsil.
- Complications include sternocleidomastoid spasm, torticollis, fasciitis, mediastinitis, and airway obstruction
- Throat cultures and serologic testing for EBV virus should be obtained.
- CT scan with contrast should be done if doubt exists after physical exam.
- Experienced otolaryngologists should perform direct tonsillar needle aspiration.
- Some patients require admission for drainage, IV hydrations, and antibiotics (nafcillin and third-generation cephalosporin).

FOREIGN BODY ASPIRATION

- Most occur in children <5 years of age.
- In >50% of cases, there is no history of foreign body aspiration or choking spells
- May present multiple times for respiratory-like illness or recurring pneumonia or lung abscess.
- Radiographs are frequently normal.
- Radiographs should include inspiratory and expiratory chest films or, in younger children, bilateral decubitus views to detect unilateral hyperinflation secondary to a ball valve effect of a foreign body.
- Treatment of acute complete obstruction for children <1 year of age, use four back blows followed by chest thrusts
- For children >1 year of age repetitive abdominal thrusts.
- If unsuccessful, use Magill forceps under direct laryngoscopy to remove foreign body, vigorous BVM ventilation, and bronchoscopy.
- Treatment for incomplete obstruction includes supplemental oxygen, position of comfort, decreased stimuli, and arrange for rigid bronchoscopy in the OR.

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QUESTIONS

1. A 6-month-old infant is brought to the emergency department for evaluation of increased work of breathing. The parents report that the infant has been less active and refusing her bottle. Which of the following signs is most consistent with impending respiratory failure in infants?
 - A. Tachypnea
 - B. Tachycardia
 - C. Grunting
 - D. Hypertension
 - E. Hypoxemia
2. A 2-month-old child presents with a chief complaint of “noisy breathing” for 2 weeks. Birth history is normal. The child appears well, with normal vital signs. When agitated, there is inspiratory and expiratory stridor. Which of the following is most likely the cause of this infant’s stridor?
 - A. Macroglossia
 - B. Vocal cord paralysis
 - C. Tracheomalacia
 - D. Croup
 - E. Epiglottitis
3. A 2-year-old presents with stridor at rest. You administer PO steroids and provide aerosolized vaponephrine. The stridor resolves and the child maintains normal vital signs and oximetry for 2 hours. After this observation period, the child is without stridor. Which of the following should be done next?
 - A. Provide home vaponephrine
 - B. Admit to the inpatient service
 - C. Administer ceftriaxone
 - D. Discharge
 - E. Discharge on 3 days of steroids
4. A toddler is brought to the emergency department for respiratory distress. The child is toxic appearing. The mother reports the acute onset of fever and sore throat. The immunization status is not up to date. Which of the following is true regarding epiglottitis?
 - A. It most commonly affects children from 2 to 5 years of age
 - B. It is gradual in onset
 - C. Dysphagia rarely occurs
 - D. Blood cultures are often positive
 - E. Most patients are afebrile
5. A 6-year-old child arrives with drooling, dysphonia, and stridor at rest. He is unimmunized. He has a toxic appearance. Which of the following interventions is indicated?
 - A. Visualization of the pharynx with a tongue blade
 - B. Lie the patient down
 - C. Immediately notify ENT and the OR
 - D. Send the child to radiology for lateral neck radiographs
 - E. Suction the pharynx
6. A 3-year-old presents with fever and sore throat of 2 days duration. The child now refuses to swallow and has nuchal rigidity, accompanied by torticollis. The pharyngeal exam is unremarkable. There is no lymphadenopathy. Which of the following diagnoses is most likely?
 - A. Peritonsillar abscess
 - B. Streptococcal pharyngitis
 - C. Herpangina
 - D. Retropharyngeal abscess
 - E. Herpetic gingivostomatitis

7. A 14-year-old presents with a 5 day history of worsening sore throat and fever. He is febrile, with dysphagia and drooling. His voice is muffled. The uvula is displaced laterally by an enlarged right tonsil, with exudate. Which of the following laboratory studies are indicated?
- Monospot
 - CBC
 - ESR
 - Sedimentation rate
 - Procalcitonin

ANSWERS

- C. Grunting is a form of end expiratory pressure generated by infants in marked respiratory distress. It is usually in response to varying degrees of alveolar collapse or hypoventilation. The other choices are nonspecific responses to work of breathing.
- C. Neonatal stridor is almost always associated with an upper airway congenital anomaly, laryngomalacia, or tracheomalacia. Macroglossia is readily visible on physical exam. A normal cry rules out vocal cord paralysis. Croup and epiglottitis are infectious etiologies, associated with fever.
- D. Many studies have shown that if a child with croup is asymptomatic for 2 hours after Vaponephrine, he or she may be discharged safely. Antibiotics are not indicated in croup. Prolonged steroid therapy is not warranted.
- D. With the advent of childhood immunization, epiglottitis most often occurs at the extremes of age. It is acute in presentation and patients appear toxic in most cases. In some instances, a sore throat may be the only complaint.
- C. This child has epiglottitis. Any invasive oral intervention may lead to full upper airway obstruction. The clinician should never leave the child unattended (as in the x-ray suite). Immediate notification of the OR and appropriate subspecialists (ENT or anesthesia) is always indicated.
- D. This is a classic case of retropharyngeal abscess. It occurs in preschoolers, and is accompanied by drooling and nuchal rigidity or torticollis. The other four choices are readily diagnosed by a careful pharyngeal exam.
- A. This young man has a peritonsillar abscess. Many patients will also have infectious mononucleosis. The other lab tests are nonspecific and will unlikely add to the diagnostic accuracy.

41 ASTHMA

Kathleen M. Brown

HIGH-YIELD FACTS

- Asthma is the most common chronic disease of childhood. The current prevalence of asthma in the United States is estimated at 8.9%.
- It is the third most common reason for hospitalization of children in the United States, exceeded only by injuries and pneumonia.
- The National Heart Lung and Blood Institute (NHLBI) expert panel guidelines on asthma define it as: a common chronic disorder of the airways that is complex and characterized by variable and recurring symptoms, airflow obstruction, bronchial hyperresponsiveness, and an underlying inflammation.
- The interaction of these features of asthma determines the clinical manifestations and severity of asthma and the response to treatment.

ETIOLOGY/PATHOPHYSIOLOGY

- The major mechanisms thought to contribute to the pathophysiology of asthma are increased airway responsiveness, inflammation, mucus production, and submucosal edema.
- Airway inflammation plays a critical role in both the development of obstruction and the degree of hyperresponsiveness.
- The early bronchospastic response is a classic antigen antibody reaction. When a sensitized cell is re-exposed to the specific antigen, mediators are released, including histamine, leukotrienes, and chemotactic factors that attract inflammatory cells to the area (Fig. 41-1).
- Convergence of inflammatory cells correlates with the late asthmatic response. These inflammatory cells release a number of products that cause damage to the bronchial wall.
- Airway inflammation produces an alteration of the sensory nerve endings that may lead to bronchial hyperreactivity.
- Once bronchial hyperactivity is present, nonspecific triggers may produce acute bronchospasm. The most common trigger is an upper respiratory infection. Other common triggers include inhaled allergens, exercise, and cold air.
- A vicious cycle can develop, in which continuous or repeated exposure to allergens, in sensitized persons,

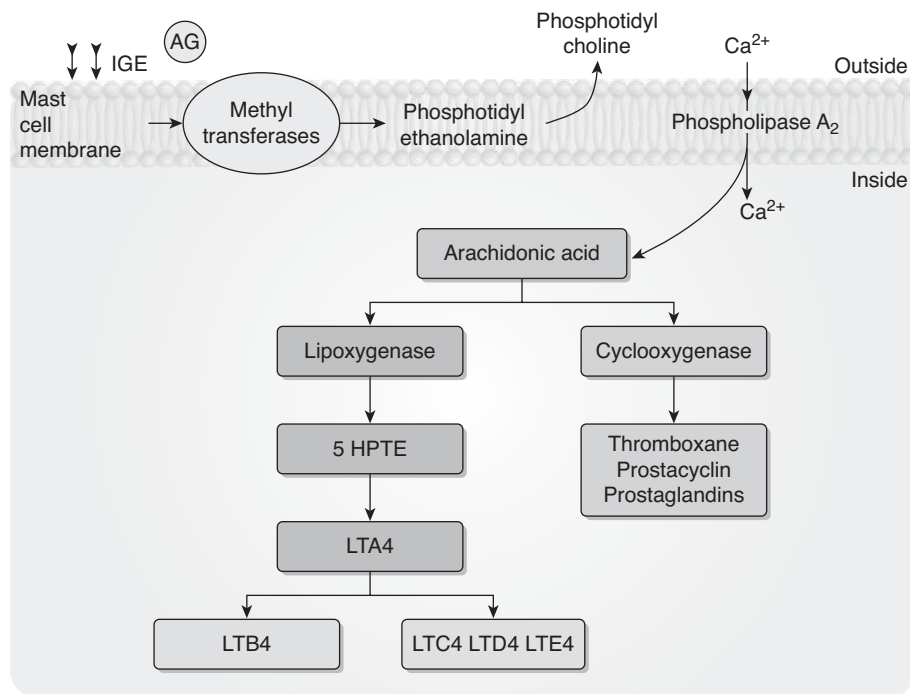


FIG. 41-1. Pathophysiology of asthma.

increases airway responsiveness. This is the chronic stage of asthma.

- There are several differences in the anatomy and physiology of a child compared to an adult, which makes them more prone to obstruction and more vulnerable to respiratory failure.
- Viral infections, particularly respiratory syncytial virus (RSV), are the most common cause of acute wheezing illness in infants.

CLINICAL PRESENTATION

- Acute asthma exacerbations are characterized by decreases in expiratory airflow that can be documented and quantified by simple measurement of lung function (spirometry). These objective measures more reliably indicate the severity of an exacerbation than does the severity of symptoms (Fig. 41-2).
- Tables 41-1 and 41-2 illustrate the classification of asthma severity recommended by the NHLBI expert panel report.
- Numeric asthma scores can also be used to classify severity and to measure effectiveness of treatment.
- The brief history should assess
 - Time of onset and any potential causes of current exacerbation.
 - Severity of symptoms especially compared with previous exacerbations, and response to any treatment given before arrival to the ED.
 - Note all current medications and time of last dose, especially of asthma medications.
 - Estimate of number of previous unscheduled office visits, ED visits, and hospitalizations for asthma, particularly within the past year.
 - Any prior episodes of respiratory insufficiency due to asthma.
 - Other potentially complicating illness.
- A family history of asthma, atopy, or allergic disease is common in patients with asthma.
- A recent history of an upper respiratory infection or exposure to a specific trigger is usually obtained.
- The initial physical examination should focus on the severity of the exacerbation (see Table 41-2).
- Wheezing may be localized and may shift in location with time as the relative degree of obstruction may vary with location and time. If airway obstruction is severe, there will be little airflow and the chest may be quiet.
- Lung examination may also reveal diffuse or localized rales, or a persistent cough with a clear lung examination.
- The use of accessory muscles is a more reliable indicator of degree of obstruction.

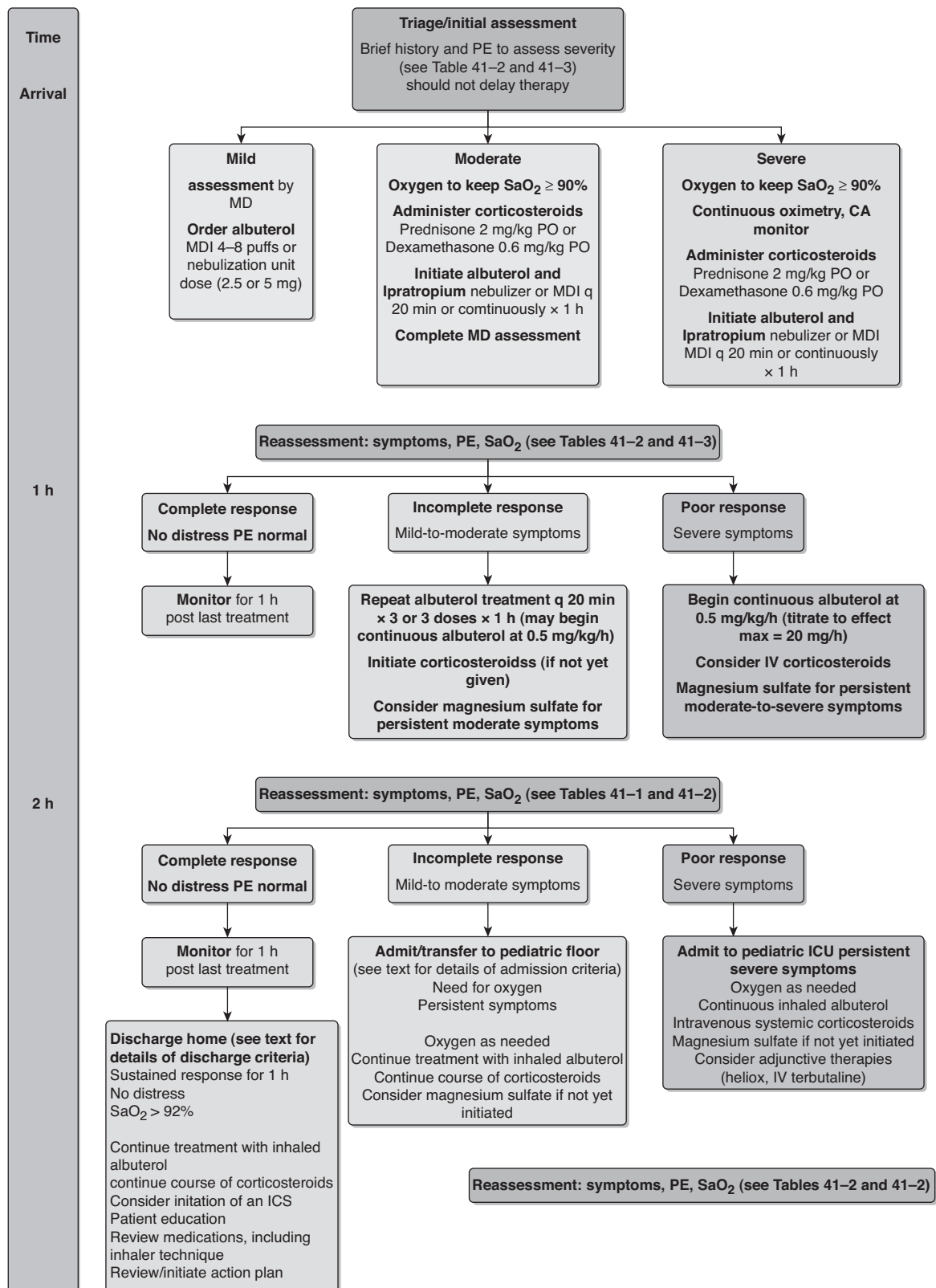


FIG. 41-2. ED management of a child with an acute asthma exacerbation.

TABLE 41-1 Classifying Severity of Asthma Exacerbations in the Urgent or Emergency Care Setting

	SYMPTOMS AND SIGNS	INITIAL PEF (%) (OR FEV ₁ ^b)	CLINICAL COURSE
Mild	Dyspnea only with activity (assess tachypnea in young children)	PEF ≥70 predicted or personal best	<ul style="list-style-type: none"> • Usually cared for at home • Prompt relief with inhaled SABA • Possible short course of oral systemic corticosteroids
Moderate	Dyspnea interferes with or limits usual activity	PEF 40–69 predicted or personal best	<ul style="list-style-type: none"> • Usually requires office or ED^a visit • Relief from frequent inhaled SABA • Oral systemic corticosteroids; some symptoms last for 1–2 d after treatment is begun
Severe	Dyspnea at rest; interferes with conversation	PEF <40 predicted or personal best	<ul style="list-style-type: none"> • Usually requires ED visit and likely hospitalization • Partial relief from frequent inhaled SABA • Oral systemic corticosteroids; some symptoms last for >3 d after treatment is begun • Adjunctive therapies are helpful
Subset: life threatening	Too dyspneic to speak; perspiring	PEF ^d <25 predicted or personal best	<ul style="list-style-type: none"> • Requires ED/hospitalization; possible ICU^c • Minimal or no relief from frequent inhaled SABA^e • Intravenous corticosteroids • Adjunctive therapies are helpful

^aED, emergency department.

^bFEV₁, forced expiratory volume in 1 second.

^cICU, intensive care unit.

^dPEF, peak expiratory flow.

^eSABA, short-acting β₂-agonist.

Patients are instructed to use quick-relief medications if symptoms occur or if PEF drops below 80% predicted or personal best. If PEF is 50%–79%, the patient should monitor response to quick-relief medication carefully and consider contacting a clinician. If PEF is below 50%, immediate medical care is usually required. In the urgent or emergency care setting, the following parameters describe the severity and likely clinical course of an exacerbation.

LABORATORY AND RADIOGRAPHIC FINDINGS

- Chest radiographs were shown to change the course of treatment in only 10% of asthmatics.
- A chest x-ray may be important for any child that presents with wheezing for the first time, as there are many illnesses that can present with wheezing.
- Specific indications for a chest radiograph in a known asthmatic child include clinical suspicion of consolidation, effusion, pneumothorax, or impending respiratory failure.
- Typical chest radiograph findings are hyperinflation, peribronchial cuffing, and areas of subsegmental atelectasis (Fig. 41-3). These findings are nonspecific and usually add little to the clinical assessment.
- The simplest spirometry test, peak expiratory flow rate (PEFR), can usually be performed in children older than 5 years. A PEFR of less than 30% to 50% of predicted or of the patient's personal best indicates severe airway obstruction.
- An initial pulse oximetry in infants and young children might be useful for assessing exacerbation severity but not for predicting the need for hospital admission.

- Blood gases may help assess the status of severe asthmatics but are not necessary for management of most acute asthma exacerbations. A “normal” or slightly elevated PCO₂ in a patient with an asthma exacerbation may be a sign of muscle fatigue and impending respiratory failure.

DIFFERENTIAL DIAGNOSIS

- The diagnosis of asthma is made by demonstrating episodic and reversible airway disease.
- The diagnosis in small children is usually made on a clinical basis. The diagnosis of asthma should be considered in all children with recurrent wheezing and symptom-free intervals, especially if there is a family history of asthma, atopy, or allergies.
- In an infant with wheezing, it is often impossible to clinically differentiate between bronchiolitic wheezing and asthma. The most important clue to infantile asthma is a history of recurrent episodes of wheezing or persistent cough.
- A list of other possible etiologies for wheezing in an infant or child is provided in Table 41-3.

TABLE 41-2 Formal Evaluation of Asthma Exacerbation Severity in the Urgent or Emergency Care Setting

SYMPTOMS	MILD	MODERATE	SEVERE	SUBSET: RESPIRATORY ARREST IMMINENT
Breathlessness	While walking	While at rest (infant—softer, shorter cry, difficulty in feeding)	While at rest (infant—stops feeding)	
Talks in Alertness	Can lie down Sentences May be agitated	Prefers sitting Phrases Usually agitated	Sits upright Words Usually agitated	Drowsy or confused
Signs				
Respiratory rate	Increased	Increased Guide to rates of breathing in awake children: <i>Age</i> <2 mo 2–12 mo 1–5 y 6–8 y	Often >30/min Often >30/min <i>Normal rate</i> <60/min <50/min <40/min <30/min	
Use of accessory muscles; suprasternal retractions	Usually not	Commonly	Usually	Paradoxical thoracoabdominal movement
Wheeze	Moderate, often only end expiratory	Loud; throughout exhalation	Usually loud; throughout inhalation and exhalation	Absence of wheeze
Pulse/minute	<100	100–120 Guide to normal pulse rates in children: <i>Age</i> 2–12 mo 1–2 y 2–8 y	>120 Normal rate <160/min <120/min <110/min	Bradycardia
Pulsus paradoxus	Absent <10 mm Hg	May be present 10–25 mm Hg	Often present >25 mm Hg (adult) 20–40 mm Hg (child)	Absence suggests respiratory muscle fatigue
Functional Assessment				
PEF percentage predicted or percentage personal best	≥70%	Approx. 40%–69% or response lasts <2h	<40 percentage	<25 percentage cPEF testing may not be needed in very severe attacks
^a PaO ₂ (on air) and/or ^b Pco ₂	Normal (test not usually necessary) <42 mm Hg (test not usually necessary)	≥60 mm Hg (test not usually necessary) <42 mm Hg (test not usually necessary)	<60 mm Hg: possible cyanosis >42 mm Hg: possible respiratory failure (see pages 393–394, 399)	
^d SaO ₂ percentage (on air) at sea level	>95% (test not usually necessary)	90%–95% (test not usually necessary)	<90%	
Hypercapnia (hypoventilation) develops more readily in young children than in adults and adolescents				

^aPaO₂, arterial oxygen pressure.

^bPco₂, partial pressure of carbon dioxide.

^cPEF, peak expiratory flow.

^dSaO₂, oxygen saturation.

The presence of several parameters, but not necessarily all, indicates the general classification of the exacerbation.

Many of these parameters have not been systematically studied, especially as they correlate with each other. Thus, they serve only as general guides.

The emotional impact of asthma symptoms on the patient and family is variable but must be recognized and addressed and can affect approaches to treatment and follow-up.

TREATMENT

- The choice and intensity of therapy depends on the severity of the exacerbation and the patient's response to initial treatment.
- Recommended doses for asthma therapies are summarized in Table 41-4.
- Therapies that should be considered in all ED patients with an acute exacerbation of their asthma include oxygen and fluids.
- Hypoxia can lead to hypoventilation and acidosis, which can cause pulmonary vasoconstriction, pulmonary hypertension, and right heart failure.
- Asthmatic patients are also often dehydrated due to decreased intake or vomiting and may require IV fluids. However, acute asthma is associated with increased secretion of antidiuretic hormone and with an increase in capillary permeability and interstitial fluid, thus overhydration may result in pulmonary edema (Table 41-5).

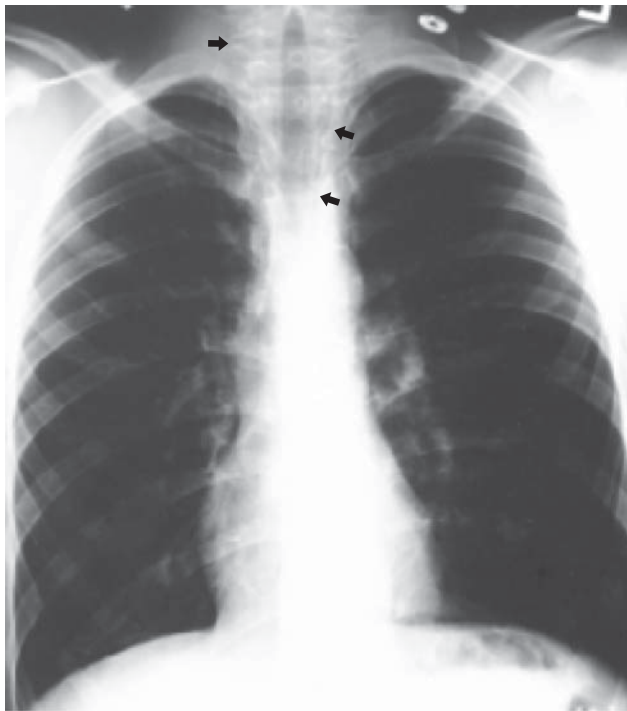


FIG. 41-3. Chest x-ray of a child with an acute asthma exacerbation shows hyperinflation (abnormally lucent lungs). The diaphragm is flattened and relatively small and air is present within the mediastinum.

TABLE 41-3 Differential Diagnosis in a Wheezing Infant

Anaphylaxis
Aspiration
Bronchiolitis
Bronchopulmonary dysplasia
CHF
Cystic fibrosis
Extrinsic airway compression
Foreign body aspiration
Immotile cilia
Immune deficiency
Mediastinal masses
Pneumonia
Vascular rings

- Antibiotics should be used in asthma only if evidence of concurrent infection exists. Chronic sinusitis, in particular, is thought to cause persistent asthmatic exacerbations.

β-ADRENERGIC AGONISTS

- Aerosol therapy is the most commonly used and recommended form of β-adrenergic agents. It has been shown to be as effective as IV or SC therapy and more effective than oral therapy. Comparable efficacy of metered dose inhalers and jet nebulization has been demonstrated.

TABLE 41-4 Medications for an Acute Asthma Exacerbation

MEDICATION	ROUTE	DOSE
β-adrenergic agents		
Albuterol solution for nebulization	Nebulizer	0.15 mg/kg (min. 2.5 mg, max. 5 mg) q 15–20 min × 3 May mix 3 doses with ipratropium in appropriate size holding chamber and run over 1 h Children <35 kg (7.5 mg albuterol and 500 μg ipratropium) Children >35 kg (15 mg albuterol and 1000 μg ipratropium)
90 μg/puff	Continuous nebulization	0.3–0.6 mg/kg/h up to 20 mg/h
Epinephrine (1:1000 solution)	MDI	4–8 puffs q 20 min × 3
Terbutaline (0.1%)	SC	0.01 mg/kg (max. 0.3 mg)
	SC	0.01 mg/kg (max. 0.3 mg) q 20 min. × 3
	IV	Loading dose 10 μg/kg Infusion: 0.4 μg/kg/min may titrate up to 6 μg/kg/min
Corticosteroids		
Methylprednisolone	IV	2 mg/kg (max. 125 mg)
Prednisone/prednisolone	PO	2 mg/kg as loading dose in ED Discharge: 1–2 mg/kg/d (max. 60 mg) × 5 d
Dexamethasone phosphate	PO	0.6 mg/kg in ED (max. dose 16 mg) Discharge: 0.6 mg/kg in 24 h
Anticholinergics		
Ipratropium bromide (500 μg/2 mL)	Nebulizer	250–500 μg q 20 min × 2–3 doses (usually with albuterol)
18 μg/puff	MDI	4–8 puffs as needed
Magnesium sulfate	IV	50–75 mg/kg (max. 2.5 g) over 20 min.
Ketamine	IV	Induction: 1–2 mg/kg

TABLE 41-5 Asthma Facts

- Asthma is the most common chronic disease of childhood and is associated with significant morbidity and mortality.
 - Inhaled albuterol remains the first line therapy for acute asthmatic exacerbations. Delivery of albuterol by metered dose inhaler has been shown to be superior to delivery by nebulization.
 - The addition of nebulized ipratropium to the first 2 to 3 albuterol doses has been associated with a decreased need for hospitalization in pediatric patients with moderate-to-severe asthma exacerbations.
 - Administration of oral corticosteroids in the emergency department has been shown to enhance recovery from an acute asthma exacerbation and decrease rates of hospitalization.
 - Oral dexamethasone (1–2 doses) has been shown to be as efficacious as a 5-day course of oral prednisone.
 - Magnesium sulfate may be of benefit in patients with moderate-to-severe exacerbations who do not respond to initial bronchodilator therapy.
 - Heliox has been suggested to be of benefit for patients with severe asthma exacerbations in small clinical trials, but convincing evidence of its benefit is not available.
-
- One method for enabling younger children to use an MDI more effectively is the use of an aero chamber or spacer, which provides a reservoir of particles for inspiration requiring less coordination of MDI activation with inhalation.
 - Doses of 0.5 puffs/kg with a maximum of 6 to 8 puffs are recommended for treatment of an acute exacerbation.
 - Doses of 7.5 mg of albuterol mixed with 500 μ g ipratropium for patients less than 35 kg and 15 mg albuterol with 1000 μ g ipratropium in children greater than 35 kg can be mixed in the holding chamber and run over 1 hour.
 - Continuous nebulization with albuterol is usually started at approximately 0.5 mg/kg/h and titrated up or down as needed.
 - Levalbuterol (xopenex) has been promoted as an alternative to racemic albuterol with the rationale that eliminating the isomer decreases side effects. However, no studies have demonstrated a benefit of this medication in the treatment of an acute asthma exacerbation.
 - Epinephrine SC injection is more toxic and no more effective than inhalation of a β_2 selective drug. Parenteral administration (0.01 mL/kg up to 0.3 mL of the 1:1000 solution SC) should be reserved for those patients who are unable to generate adequate tidal volume to deliver aerosolized drug to the bronchial tree.
 - Subcutaneous terbutaline (0.01 mg/kg up to 0.25 mg), which is more β_2 specific, may be used as an alternative to subcutaneous epinephrine. It is preferred to SC epinephrine in the pregnant patient as SC epinephrine

has been associated with fetal malformations from decreased uterine blood flow.

- Intravenous (IV) terbutaline has been shown to be safe and effective in two small studies of pediatric patients with severe asthma exacerbations. However, IV β -agonists have not been shown to have benefit over inhalational therapy
- Side effects associated with all β -adrenergic agonists are largely due to sympathomimetic effects and include tremors, anxiety, nausea, headache, vomiting, tachycardia, arrhythmia, hypertension, and hypotension. Nonsympathomimetic side effects include decreased oxygen saturation (secondary to altered V/Q matching), which is common, and paradoxical bronchospasm, which is rare.
- Metabolic side effects include hypokalemia, hypophosphotemia, hyperglycemia, and lactic acidosis.
- These side effects are often related to dose and route of administration and rarely require cessation of therapy. However, all patients receiving β -prolonged adrenergic therapy should have their oxygen saturation, heart rate, blood pressure, and serum electrolytes monitored closely.

ANTICHOLINERGICS

- Ipratropium in combination with β -agonists in pediatric patients with an acute exacerbation of their asthma is helpful in reducing rates of hospitalization.
- It is most efficacious when given as two to three doses in combination with the initial two to three albuterol treatments in patients with moderate-to-severe exacerbations.
- Reported side effects include dry mouth and a metallic taste.

CORTICOSTEROIDS

- Demonstrated benefits of early initiation of corticosteroids include increased rate of improvement, decreased duration of symptoms, decreased hospitalization rates, decreased relapse rates, and decreased need for β -agonists.
- Oral and parenteral corticosteroids are known to be equally efficacious. Oral dosing is preferred in children when possible as it is less invasive. IV dosing should be reserved for those patients who cannot tolerate oral medications, are requiring continuous β -agonist therapy, or have impending respiratory failure.
- Traditionally, 1 to 2 mg/kg of prednisone or an equivalent dose of another corticosteroid should be given

as an initial bolus and then continued for another 4 days as a bid dose (total of 1–2 mg/kg for 5 days with a maximum single dose of 60 mg).

- Clinical trials have shown that 1–2 doses of 0.6 mg/kg of dexamethasone phosphate (maximum single dose of 16 mg) have similar clinical responses such as risk for hospitalization, return for additional care, or persistence of symptoms at 10 days, with a much lower (10-fold) rate of vomiting of the medication when compared to a 5-day course of prednisolone.
- These trials used the parenteral form of dexamethasone phosphate administered orally and not a commercially available elixir of dexamethasone.
- The use of inhaled steroids in the acutely ill asthmatic patient has been investigated but a consistent significant benefit has not been reported.
- Initiation of inhaled corticosteroids should be considered for ongoing management of patients who have moderate or severe persistent asthma.

MAGNESIUM

- A systematic review of the literature on IV magnesium for asthmatics of all ages did demonstrate a decrease in admission rate for those with severe acute asthma exacerbations.
- Doses of 25 to 75 mg/kg IV over 20 minutes are recommended for patients with a moderate or severe acute asthma exacerbation who do not respond to initial therapy with albuterol and ipratropium.

HELIOX

- Inhalation of a blend of helium and oxygen has also been suggested to be helpful for severe asthmatic patients unresponsive to other therapies.
- Current published literature on this subject is inconclusive as to its benefit and has not shown any significant complications with its use.

INTUBATION/MECHANICAL VENTILATION

- Indications for intubation of an asthmatic patient include decreased level of consciousness, apnea, exhaustion, rising PaCO₂ after treatment, PaO₂ <60 mm Hg, and pH <7.2.
- An asthmatic may not immediately improve with intubation, since intubation does nothing to change lower airway obstruction. Intubation and mechanical ventilation may also put the patient at risk for serious complications.

- When intubating an asthmatic patient, the largest diameter tube appropriate for the patient's size is used to avoid increasing resistance even further.
- The dissociative anesthetic ketamine is known to have bronchodilatory properties and is a good choice for a sedative for intubation.
- Once intubated, patients with asthma will require sedation and paralysis to maintain effective ventilation. They also require a long expiratory time to avoid air trapping due to airway obstruction.
- Intrinsic PEEP may cause an increase in intrathoracic pressure that leads to decreased venous return to the heart and can cause hypotension.
- Air trapping also puts the patient at risk for the development of air leaks. Intubated asthmatic patients need to be watched carefully for the development of pneumothorax, or pneumomediastinum.
- The use of permissive hypercapnea (PCO₂) levels (as high as 70–90 mm Hg) has been associated with decreased morbidity and mortality rates in intubated asthmatic patients.

DISPOSITION/OUTCOME

- Despite appropriate therapy, approximately 10% to 25% of ED patients who have acute asthma will require hospitalization.
- There is a high relapse rate for patients discharged after treatment, and many patients return to the ED requiring further therapy or hospitalization.
- Clinical examination and scoring systems perform poorly in identifying patients requiring hospital admission. However, these assessments may help to determine which patients should be admitted after an initial 1- to 2-hour period of treatment, leaving the ED resources for those who are more likely to go home after extended ED treatment and observation.
- Initial pulse oximetry is a poor predictor of need for hospitalization. Pulse oximetry of <92% to 94% at 1 hour was a better predictor of need for hospitalization.
- Various spirometric parameters have also been proposed but have not proved to have adequate sensitivity and are often difficult to obtain in children.
- The following risk factors have been identified as being associated with mortality: previous intubation (greatest predictor of subsequent death), two or more hospitalizations in the last year, three or more ED visits in the last year, use of systemic steroids, rapid progression of attacks, hypoxic seizures, severe nighttime wheezing, barotrauma, self-weaning from medications, lack of perception of the severity of the disease, poor medical management, poor access to medical care, and smoke exposure.

- The NHLBI guidelines urge that on discharge from the ED patients receive: referral for follow-up asthma care within 1 to 4 weeks, an ED asthma discharge plan with instructions for medications prescribed at discharge and for increasing medications or seeking medical care if asthma worsens, review of inhaler technique whenever possible, and consideration of initiating inhaled corticosteroids.

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QUESTIONS

1. A 8-year-old African-American boy is brought to the ED by his mother for evaluation of wheezing

and shortness of breath. You learn that this is his fourth ED visit in the last 2 months. None of the ED visits resulted in hospitalizations. The mother is concerned that his “asthma symptoms keep recurring.” In your discussions with the mother, it will be important to stress the key components of the pathophysiology of asthma, which include airflow obstruction, bronchial hyper-responsiveness, and

- A. Reversibility
 - B. Bronchospasm
 - C. Underlying inflammation
 - D. Hyperventilation
 - E. Emotional triggers
2. A 6-year-old boy with a history of being hospitalized twice for wheezing and once for pneumonia, last when he was 3 years old, now presents to your ED with a complaint of difficulty breathing. He has received albuterol MDI with a spacer at home four times today with no improvement. You note that He appears anxious, prefers to sit up. His vital signs are temperature of 38.5 C, RR 40, HR 120 BP 90/p, and RA oximetry of 93%. He is using accessory muscles. On auscultation, you appreciate moderately decreased air movement throughout and expiratory wheezing that is more apparent on the left than the right side of his chest. Which of the following would be highest on your differential diagnosis list?
 - A. Bacterial Pneumonia
 - B. Bronchiolitis
 - C. Mild acute asthma exacerbation
 - D. Moderate acute asthma exacerbation
 - E. Severe acute asthma exacerbation
 3. Your initial therapy for the child in question one would most appropriately include
 - A. 0.6 mg/kg Oral dexamethasone, 7.5 mg albuterol, 500 mcg ipratropium inhaled over 1 hour.
 - B. 2.5 mg albuterol inhaled and ceftriaxone.
 - C. Intramuscular epinephrine.
 - D. 2 mg/kg oral prednisolone and IV magnesium.
 - E. 2 mg/kg IV methylprednisolone, and 7.5 mg albuterol, 500 mcg ipratropium inhaled over 1 hour.
 4. Appropriate initial ancillary testing on this child would include
 - A. Pulse oximetry and stat portable chest x-ray
 - B. ABG and stat chest x-ray
 - C. Pulse oximetry and PEFr
 - D. RSV PCR
 - E. CBC and Blood culture
 5. After 4 hours of ED therapy the child now appears much improved. He is active and eating chips. His vital signs are T-38 C, RR 30, HR 125, and BP 90/p

- and pulse oximetry of 95%. On auscultation, you hear good air movement and some diffuse rhonchi but no wheezing. His last therapy was 90 minutes ago. The most appropriate disposition at this time is
- Admission for continued treatment.
 - Discharge with albuterol MDI, a prescription for oral and inhaled corticosteroids and follow up with his primary care provider.
 - Continued observation and treatment in the ED.
 - Discharge with a nebulizer machine, a prescription for amoxicillin and follow up with his primary care provider.
 - Reassessment after CXR and laboratory testing results are available.
- A 5-year-old girl with a history of asthma presents to the ED with a moderate acute asthma exacerbation. The most appropriate initial therapy would include
 - 0.5 puff/kg of an albuterol MDI and reassessment
 - 2 mg /kg oral prednisolone 0.5 puffs/kg of an albuterol MDI and reassessment
 - 2 mg/kg IV methylprednisolone 7.5 mg nebulized albuterol and reassessment
 - 7.5 mg nebulized albuterol and reassessment
 - 2.5 mg nebulized albuterol and reassessment
 - An 11-months-old presents with a 2-day history of low-grade fever and cough. Today her family noted she was having difficulty breathing and not feeding well. She has a history of having a febrile illness with wheezing when she was 6-months old. No other known PMH.
On physical examination, you see a smiling drooling infant. Her vital signs are temperature of 38.5C, HR 150, RR 60, BP 85/p Pulse oximetry of 93%. She has mild intercostal retractions. On auscultation, you here diffuse expiratory wheezing. You most likely diagnosis for this child is
 - Bacterial Pneumonia
 - Bronchiolitis
 - Mild acute asthma exacerbation
 - Moderate acute asthma exacerbation
 - Foreign body aspiration
 - A 10-year-old boy with a known history of asthma presents to the ED with a moderate-severe acute asthma exacerbation. He has received 30 mg albuterol over 2 hours, 1 mg Ipratropium and 2 mg/kg oral prednisone. On reassessment, his VS are T 37.8 C, P 125, RR 32, pulse oximetry 91% on room air. He has intercostal retractions, diffuse expiratory wheezing and scattered rales. The most appropriate management at this time is
 - Continuous albuterol at 0.5 mg/kg/hr, 50 mg/kg IV magnesium, reassessment.
 - IM epinephrine, IV terbutaline, inpatient admission.
 - CXR, Heliox therapy, IV methylprednisolone, ICU admission.
 - Continuous albuterol at 0.5 mg/kg/hr, ceftriaxone, inpatient admission.
 - IV terbutaline, and continuous albuterol at 0.5 mg/kg/hr.
 - A 9-year-old child with a known history of asthma has been receiving continuous albuterol for 4 hours. Other than tremors, anxiety, nausea, headache, vomiting, and tachycardia, what other side effects would you most expect to see in this patient?
 - Hyperkalemia and hypoxia (due to VQ mismatching)
 - Hyperglycemia and hypoxia (due to VQ mismatching)
 - Hypokalemia and hypoglycemia
 - Hypomagnesemia and hypokalemia
 - Hypotension and ventricular arrhythmias
 - A 12-year-old known asthmatic presents to the ED via EMS. On initial examination, you note that she is very lethargic, with the following vital signs: T 38.5 C, P135, RR 12, BP 80/p. You are unable to measure her pulse oximetry. On auscultation, she has a “quiet chest.” The most appropriate initial management for this child is
 - Continuous albuterol at 0.5 mg/kg/hr, oral prednisone, ceftriaxone, ICU admission
 - Continuous albuterol at 0.5 mg/kg/hr, IV methylprednisolone, ceftriaxone, ICU admission
 - Continuous albuterol at 0.5 mg/kg/hr, IV methylprednisolone, 50 mg/kg IV magnesium
 - Bag Valve mask ventilation, 3 mg IM epinephrine, intubation facilitated with 2 mg/kg ketamine
 - Heliox, IV terbutaline, intubation facilitated with 2 mg/kg ketamine

ANSWERS

- C. Inflammation plays a key role in both the development of obstruction and the degree of hyperresponsiveness. Pathologic specimens from patients demonstrate inflammation of the airways even in the mildest forms of the disease. Increased mucus production and submucosal edema add to the obstruction that occurs secondary to bronchospasm and inflammation.

Reversibility: Chronic asthma is not always reversible. During the immune response, proliferating

fibroblasts deposit extensive networks of collagen that can lead to fibrosis, remodeling of the bronchioles, and irreversible airway disease.

Bronchospasm is related to airway responsiveness. Airway responsiveness is defined as the ease with which airways narrow in response to various nonallergic stimuli. These stimuli include inhaled pharmacologic agents, such as histamine and methacholine, and physical stimuli, such as exercise. Hyperventilation is a symptom or response to the pathophysiologic mechanisms described above. Emotional triggers may play a role in triggering or worsening an acute asthma exacerbation but is not a key pathophysiologic mechanism.

2. D. This case describes a typical presentation of a child with known asthma and an acute asthma exacerbation. Table 41.2 describes clinical features that can be used to classify an acute asthma exacerbation.

While bacterial pneumonia is a possible component of this child's acute exacerbation an acute asthma exacerbation is more commonly triggered by a viral infection or noninfectious allergen exposure. Because viral infections are a very common trigger in children, it is common that patients will present with fever at the time of their acute asthma exacerbation. It is also common that patients present with focal and or shifting wheezing patterns due to changing levels of obstruction of airflow. Bronchiolitis is a disease with similar pathophysiology that is seen almost exclusively in children <2 years of age.

3. A. 0.6 mg/kg Oral dexamethasone, and 7.5 mg albuterol, 500 mcg ipratropium inhaled over 1 hour.

Initial ED treatment in a patient with a moderate acute asthma exacerbation should include oral corticosteroids and inhaled beta agonists. Studies have shown that when ipratropium bromide is added to this therapy that there is a decrease in the rate of hospitalization. Oral corticosteroids have been shown to be as efficacious as IV corticosteroids. Some studies have shown that oral dexamethasone is better tolerated (decreased rate of emesis) than oral prednisolone in children. In order to avoid the morbidity associated with repeated use of parenteral medications in a chronic disease, they should be reserved for those who cannot take or absorb oral medications.

Empiric antibiotic therapy is not recommended for treatment of a moderate acute asthma exacerbation even in patients with fever as the majority of these patients do not have a bacterial infection.

Magnesium is recommended for patients with a moderate to severe acute asthma exacerbation who do not have a satisfactory response to initial therapy with B agonists.

IM epinephrine offers no advantage and increased adverse effects over inhaled beta agonists in patients who able to take inhaled medications.

4. C. Pulse oximetry and pulmonary function testing can be useful in assessing the severity of an acute asthma exacerbation (see Tables 41-1 and 41-2), monitoring response to therapy and determining disposition. PEFR can usually be measured in children 5 years of age and older. Younger children may have more trouble with performing this task.

Blood gases may help assess the status of severe asthmatics but are not necessary for management of most acute asthma exacerbations. Chest radiographs were shown to change the course of treatment in only 10% of asthmatics. Obtaining a chest x-ray is important for any child that presents with wheezing for the first time, as there are many illnesses that can present with wheezing. Thereafter, specific indications for a chest radiograph in a known asthmatic include clinical suspicion of consolidation, effusion, pneumothorax, or impending respiratory failure.

Testing for occult bacteremia is not recommended for immunocompetent children over age 3. As noted above, children with an acute asthma exacerbation often have fever secondary to a concurrent viral infection. RSV is a virus that commonly causes wheezing in infants. This syndrome is called bronchiolitis. RSV and other respiratory viruses may trigger an acute asthma exacerbation in older children or adults. However, testing for this or other viruses in children with an acute asthma exacerbation does not contribute their acute management in the ED.

5. B. Numerous studies have been published attempting to establish objective criteria for admission. In general, individual clinical exam findings and scoring systems perform poorly in identifying patients requiring hospital admission. However, these assessments may help to determine which patients should be admitted after an initial 1- to 2-hour period of treatment, leaving the ED resources for those who are more likely to go home after extended ED treatment and observation. The following risk factors have been identified as being associated with mortality: previous intubation (greatest predictor of subsequent death), two or more hospitalizations in the last year, three or more ED visits in the last year, use of systemic steroids, rapid progression of

attacks, hypoxic seizures, severe nighttime wheezing, barotraumas, self-weaning from medications, lack of perception of the severity of the disease, poor medical management, poor access to medical care, and smoke exposure.

Patients discharged from the ED after an acute asthma exacerbation should be instructed to continue β -agonist use and be placed on a short course of oral corticosteroids. The most recent NHLBI guidelines suggest referral for follow-up asthma care within 1 to 4 weeks, an ED asthma discharge plan with instructions for medications prescribed at discharge and for increasing medications or seeking medical care if asthma worsens, review of inhaler technique whenever possible, and consideration of initiating inhaled corticosteroids.

6. B. As noted in question 2 initial ED therapy of a moderate acute asthma exacerbation should include oral corticosteroids and inhaled beta agonists. Multiple studies have demonstrated that inhalation of beta agonists using an MDI is at least equivalent efficacy to inhalation through a jet nebulizer.
7. B. Not all children who present with wheezing have asthma. Other diagnoses should be considered particularly in infants. However many children with asthma have their first asthmatic episode prior to 6 months of age. In infants, as in older children, viral infections are the most common trigger for asthma. Both infants who have asthma and those who do not may become infected with RSV or other viruses, and develop bronchiolitis as their first or only episode of wheezing. Therefore, in an infant with wheezing, it is often impossible to clinically differentiate between bronchiolitic wheezing and asthma. The most important clue to infantile asthma is a history of recurrent episodes of wheezing or persistent cough.

Fever is common in children with bronchiolitis. Its presence in this setting does not increase the risk of a bacterial infection. Foreign body aspiration most commonly occurs in the toddler age group but is possible in infants. Further clues to this diagnosis may be found in the history- choking, or gagging episode or in the physical exam such as unequal breath sounds on auscultation.

8. A. Continuous nebulization of albuterol has been shown to be safe and effective. Continuous nebulization is usually started at approximately 0.5 mg/kg/h and titrated up or down as needed.

Parenteral epinephrine is more toxic and no more effective than inhalation of a β_2 selective drug.

Parenteral administration should be reserved for those patients who are unable to generate adequate tidal volume to deliver aerosolized drug to the bronchial tree. Parenteral terbutaline, which is more β_2 specific, may be used as an alternative to IM epinephrine. IV terbutaline has been shown to be safe and effective in two small studies of pediatric patients with severe asthma exacerbations. However, IV β -agonists have not been shown to have benefit over inhalational therapy.

In patients who have not responded to therapy with β -agonists, ipratropium, and corticosteroids, additional therapies may be considered. A systematic review of the literature on IV magnesium for asthmatics of all ages demonstrated a decrease in admission rate for those with severe acute asthma exacerbations.

9. B. Side effects associated with all β -adrenergic agonists are largely due to sympathomimetic effects and include tremors, anxiety, nausea, headache, vomiting, tachycardia, arrhythmia, hypertension, and hypotension. Nonsympathomimetic side effects include decreased oxygen saturation (secondary to altered V/Q matching), which is common, and paradoxical bronchospasm, which is rare. Metabolic side effects include hypokalemia, hypophosphotemia, hyperglycemia, and lactic acidosis. These side effects are often related to dose and route of administration and rarely require cessation of therapy. However, all patients receiving prolonged β -adrenergic therapy should have their oxygen saturation, heart rate, blood pressure, and serum electrolytes monitored closely.
10. D. This patient needs acute intervention and assisted ventilation. Indications for intubation of an asthmatic patient include decreased level of consciousness, apnea, exhaustion, rising PaCO₂ after treatment, PaO₂ <60 mm Hg, and pH <7.2. A modified rapid sequence induction should be used. The dissociative anesthetic ketamine is known to have bronchodilatory properties and therefore is a good choice for a sedative. An asthmatic may not immediately improve with intubation, since intubation does nothing to change lower airway obstruction. Therefore, continued asthma specific therapy is needed.

Although IM epinephrine has increased adverse effects and comparable efficacy to inhaled Beta agonists, it should be used in patients who are not able to generate adequate tidal volume to inhale aerosolized Beta agonists.

42 BRONCHIOLITIS

Kathleen M. Brown

HIGH-YIELD FACTS

- Bronchiolitis is a disease of the very young and occurs almost exclusively in children younger than 2 years. It is most common between the ages of 2 and 6 months.
- In the United States, it is the leading cause of hospitalization in infancy.
- It is more common in males than females and has a seasonal pattern, being most common in the winter and spring.
- Bronchiolitis is an acute inflammatory disease of the lower respiratory tract that is characterized by acute inflammation, edema, and necrosis of epithelial cells lining small airways, increased mucus production, and bronchospasm.
- The clinical syndrome is characterized by rapid respiration, chest retractions and wheezing, and, frequently, hypoxia.

ETIOLOGY

- The most common etiologic agent in bronchiolitis is respiratory syncytial virus (RSV). RSV is responsible for 70% of all bronchiolitis cases and for 80% to 100% of cases in winter months. Parainfluenza, adenovirus, and influenza account for most of the remaining cases.
- Infection with RSV does not grant permanent or long-term immunity. Reinfections are common and may be experienced throughout life. Other viruses that are known to cause bronchiolitis are mumps, echovirus, and rhinovirus.
- *Mycoplasma* has been shown to be the principal agent in school-age children with bronchiolitis.
- Adenovirus is associated with a particularly severe form of bronchiolitis that can lead to a chronic condition known as bronchiolitis obliterans.

PATHOPHYSIOLOGY

- Infection produces inflammation of the bronchiolar epithelium, which leads to necrosis, sloughing, and luminal obstruction.
- Increased mucus production and edema contribute further to airway obstruction.

- Obstruction is not uniform throughout the lungs. This leads to ventilation/perfusion mismatching, and resultant hypoxia.
- The hypoxia leads to compensatory hyperventilation. If the obstruction is severe, hypercapnia may occur.
- Distal to the obstructed bronchiole, air trapping or atelectasis may occur.

CLINICAL PRESENTATION

- Typically, a child with bronchiolitis will have a prodrome of an upper respiratory tract infection.
- Respiratory rates of 70 to 90 per minute, or greater, are not uncommon.
- Flaring of the nasal alae and use of intercostal muscles may also be present.
- Wheezing, prolonged expiration, and musical rales are common.
- The chest is often hyperexpanded and hyperresonant due to the air trapping. The liver and spleen may be displaced downward because of the hyperinflation and flattening of the diaphragm. Thoracoabdominal asynchrony may be present with breathing and correlates with the degree of obstruction.
- Fever is present in two-thirds of children with bronchiolitis.
- Respiratory fatigue may occur since the broncholytic infant may increase his work of breathing up to sixfold.
- Apnea is not uncommon (18–20% of those hospitalized with RSV bronchiolitis), especially in very young and premature infants. It generally occurs early in the illness often prior to the onset of other respiratory symptoms.

LABORATORY AND RADIOGRAPHIC FINDINGS

- A chest radiograph will reveal hyperinflation in the majority of patients with bronchiolitis. Peribronchial cuffing (thickening of the bronchiole walls) will be seen in approximately half. There may be areas of subsegmental atelectasis that can be difficult to differentiate from pneumonia.
- A chest radiograph may be useful in ruling out the other disease processes in the differential diagnosis of bronchiolitis. However, there is controversy about whether all patients with a first-time episode of bronchiolitis should undergo a chest radiograph.

- The American Academy of Pediatrics (AAP) practice guidelines for bronchiolitis state that the current evidence does not support routine radiography in children with bronchiolitis.
- Laboratory studies are generally not helpful and not indicated in the acute management of bronchiolitis.
- Virologic tests for RSV, if obtained during peak RSV season, demonstrate a high predictive value. However, the knowledge gained from such testing rarely alters management decisions or outcomes for the vast majority of children with clinically diagnosed bronchiolitis (Fig. 42-1).
- Young febrile infants with clinical bronchiolitis are less likely to have SBI than febrile infants without bronchiolitis.

- Hypoxia is common and the patient should have oxygen saturations assessed with a pulse oximeter. Hypercarbia will be present in those with more severe obstruction.

DIFFERENTIAL DIAGNOSIS

- The differential diagnosis for bronchiolitis is essentially the same as for asthma. Bronchiolitis may be very difficult to differentiate from infantile asthma.
- Response to bronchodilators does not exclude bronchiolitis since some children with bronchiolitis may have some degree of bronchospasm.

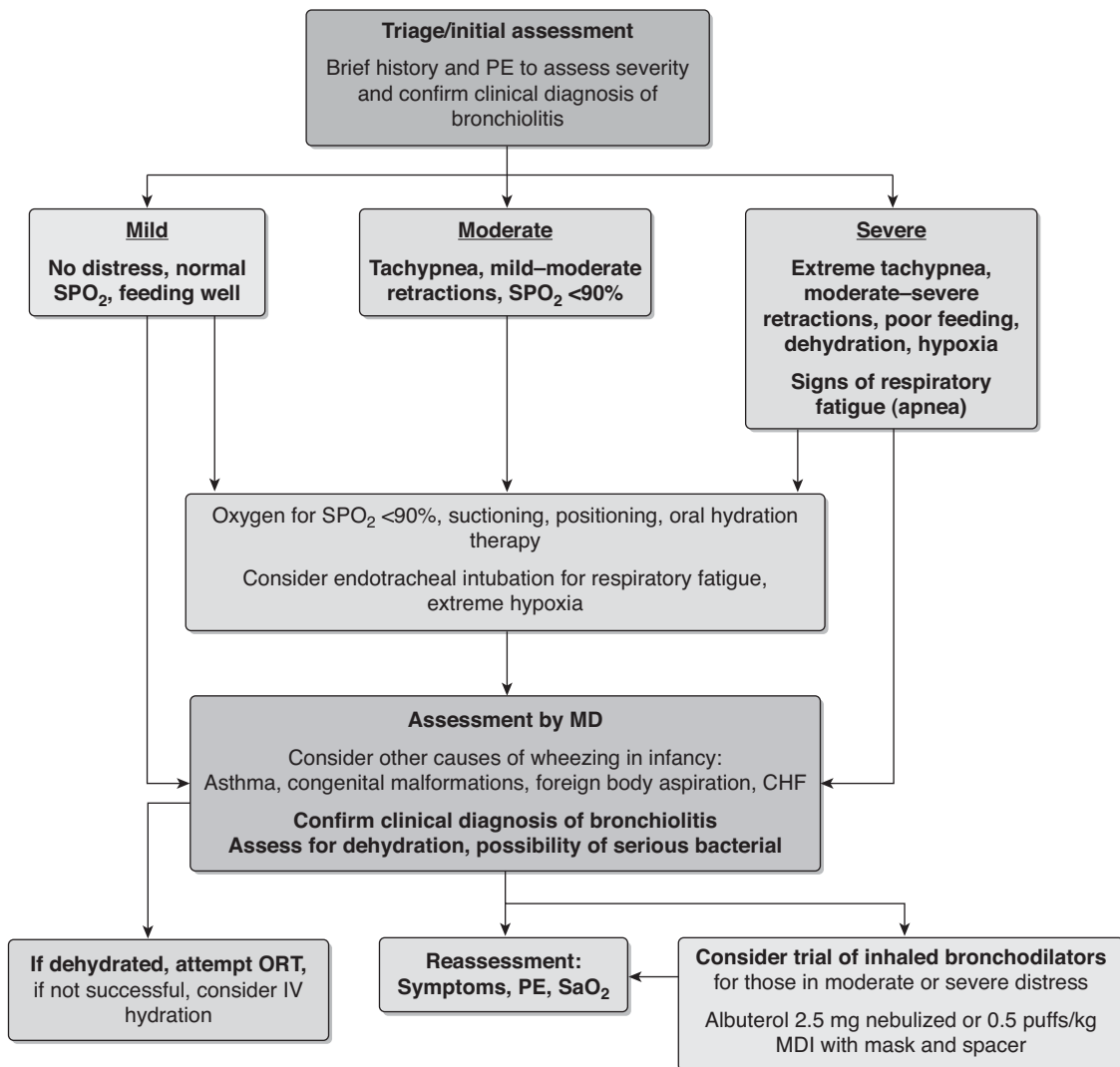


FIG. 42-1. Management of a child with bronchiolitis.

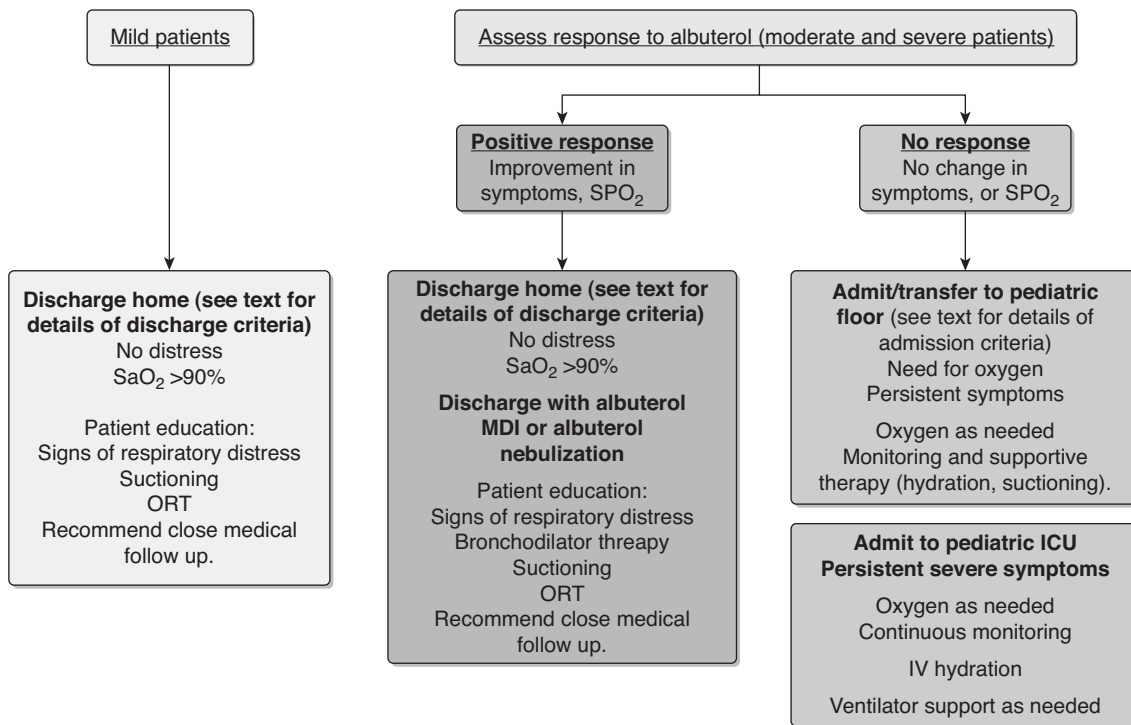


FIG. 42-1. (Continued)

- Pay particular attention to other processes that may mimic bronchiolitis and present in infancy. Congenital heart disease, cystic fibrosis, vascular rings, and other congenital anomalies may all mimic the findings of bronchiolitis.
- Infants and toddlers are particularly prone to foreign body aspiration, and this possibility must be considered.

TREATMENT

- Therapy is supportive. No medication has been shown to be beneficial in patients with bronchiolitis.
- The AAP practice guidelines on the diagnosis and management of bronchiolitis state that the supplemental oxygen is indicated if SpO₂ falls persistently below 90% in previously healthy infants.
- Intravenous hydration should be considered if the patient cannot take adequate oral fluids.
- No significant benefit has been demonstrated from routine antibiotic usage in patients with a clinical diagnosis of bronchiolitis.
- A multicenter randomized-controlled trial of the use of dexamethasone in patients younger than 1 year presenting to an ED with a first episode consistent with

bronchiolitis (no previous history of wheezing) did not demonstrate a reduction in rate of hospitalization or any other clinical benefit.

- The use of oral bronchodilators (albuterol) has not been shown to be of benefit in most of these patients. The use of inhaled bronchodilators in bronchiolitis remains controversial.
- A systematic review, assessing the use of any bronchodilator therapy in bronchiolitis, demonstrated a modest improvement in clinical scores that was of questionable clinical significance. No difference was found in oxygenation or rates of hospitalization.
- Despite the lack of proven benefit, most authors in this country recommend that patients with bronchiolitis, especially those with a past history of wheezing, should be given at least a trial of adrenergic bronchodilators. If there is no response to the trial dose, then therapy should be discontinued.
- Inhaled bronchodilators should be continued only if there is a documented positive clinical response to the trial using an objective means of evaluation.
- Some authors have suggested that the use of nebulized epinephrine therapy is superior to nebulized albuterol therapy in these patients.
- Two to five percent of infants hospitalized for bronchiolitis will go on to develop respiratory failure and

require mechanical support. There are no absolute criteria for endotracheal intubation. Suggested indications include PCO₂ greater than 60 to 65 mm Hg, recurrent apneic spells, decreasing mental status, and hypoxia despite oxygen therapy.

- Once intubated, these infants have many of the same problems that intubated asthmatics have and are at risk for air trapping and the development of air leaks and barotrauma.
- A mixture of helium and oxygen (heliox) has also been shown to be of benefit by some authors. However, no large-scale clinical trial of this therapy has been conducted.
- Continuous positive airways pressure (CPAP) has also been used as an adjunctive therapy in patients with severe symptoms.

DISPOSITION/OUTCOME

- Bronchiolitis is a short-lived, self-limited disease lasting a few days.
- Most patients do not require admission.
- The disease is characterized by a high degree of variation. The clinical examination may vary depending on the infant's state of agitation, whether they have been recently suctioned and other factors.
- A recent multicenter trial attempted to identify factors associated with safe discharge from the ED. Of 1456 enrolled patients, 57% were discharged to home from the ED. The following factors predicted safe discharge to home: Age of ≥ 2 months, no history of intubation, no history of eczema, age-specific respiratory rates (< 45 breaths/min for 0–1.9 months, < 43 breaths/min for 2–5.9 months, and < 40 breaths/min for 6–23.9 months), no/mild retractions, initial oxygen saturation of $\geq 94\%$, fewer albuterol or epinephrine treatments in the first hour, and adequate oral intake.
- Other suggested criteria for admission include age (adjusted for prematurity) less than 6 weeks, hypoxemia, persistent respiratory distress, and children with underlying conditions that put them at a greater risk (congenital heart disease, bronchopulmonary dysplasia (BPD), underlying lung disease, compromised immune function)
- Follow-up within 24 hours is recommended for those who are discharged.
- 15% to 30% of infants who are hospitalized with bronchiolitis will require admission to an ICU or need ventilatory support.
- Up to 50% of infants with RSV bronchiolitis will go on to have recurrent wheezing. The only factor shown to increase the likelihood of subsequent wheezing is a family history of asthma, or atopy.

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QUESTIONS

1. Which of the following statements is true regarding the etiology of bronchiolitis?
 - A. RSV causes up to 50% of all cases of bronchiolitis
 - B. Rhinovirus and enteroviruses account for the majority of the remainder of cases
 - C. Infants are unlikely to contract RSV bronchiolitis more than once in a season
 - D. During flu season influenza is the most likely etiologic agent in an infant with bronchiolitis
 - E. *Mycoplasma* has been shown to be the principal agent in school-age children with bronchiolitis
2. The key pathophysiologic components of bronchiolitis are:
 - A. Inflammation, necrosis, and luminal obstruction
 - B. Inflammation, hyper-responsiveness, and luminal obstruction

- C. Bronchospasm, necrosis, and luminal obstruction
- D. Inflammation, necrosis, and dehydration
- E. Hyper-responsiveness, inflammation, and bronchospasm

3. An 8-month-old infant girl presents with a 2 day history of low grade fever and cough. Today her family noted she was having difficulty breathing and not feeding well. She has a history of having a febrile illness with wheezing when she was 6 months old. She has no other PMH.

On physical exam you note a smiling, drooling infant. Her vital signs are temperature of 38.5°C, HR 150, RR 60, BP 85/p Pulse oximetry of 94%. She has mild intercostal retractions. On auscultation you here diffuse expiratory wheezing. You most likely diagnosis for this child is:

- A. Upper respiratory tract infection
 - B. Pneumonia
 - C. FB aspiration
 - D. Cyanotic congenital heart disease
 - E. Bronchiolitis
4. The most evidence based testing strategy in this child would include
- A. No additional studies
 - B. CXR, RSV PCR, and CBC
 - C. CXR and RSV PCR
 - D. CXR CBC, blood culture, urinalysis, and culture
 - E. ABG, CXR, RSV PCR, CBC, blood culture, urinalysis, and culture
5. A 6-month-old infant presents with a 2 day history of low grade fever and cough, one day of difficulty breathing and not feeding well. She was a full term infant with no neonatal history.

On physical exam you see an active playful infant. Her vital signs are temperature of 38.3°C, HR 160, RR 60, BP 85/p Pulse oximetry of 88%. She has mild intercostal retractions. On auscultation you hear diffuse expiratory wheezing.

Optimal initial management in the ED should include

- A. Oxygen, 2.5 mg albuterol v by nebulization, oral corticosteroids, and reassessment
 - B. Oxygen, racemic epinephrine by nebulization, amoxicillin, and reassessment
 - C. Oxygen, 7.5 mg albuterol and 500 mcg ipratropium by nebulization over 1 hour and reassessment
 - D. Nasal suctioning, oxygen, albuterol nebulization if symptoms persist and reassessment
 - E. Oxygen, CXR, and reassessment
6. The patient is treated with nebulized albuterol with some initial improvement. His CXR reveals mild hyperinflation and some subsegmental atelectasis.

His exam now reveals a “happy wheezer.” He is active alert and drinking a bottle. Vital signs are: temp 38.2°C, RR 45, HR 128, RA oximetry 94%. On auscultation you hear mild end expiratory wheezing.

Further management of this child should include

- A. Discharge home with supportive care and amoxicillin
 - B. Discharge home with nebulized albuterol and follow up with primary care physician
 - C. Discharge home with nebulized albuterol, oral corticosteroids, and follow up with primary care physician
 - D. Inpatient admission for continued oxygen and albuterol therapy
 - E. Inpatient admission for continued oxygen albuterol and corticosteroid therapy
7. A 3-month-old infant, who was a 34 week preemie, presents with difficulty breathing for one day. Her vital signs are temp 39°C, P 185, RR 80 BP 80/P RA sat 85%. She has moderate accessory muscle use. On auscultation you hear moderately decreased breath sounds in all lung fields and diffuse rales.
- Your initial management of this child would include
- A. Oxygen, 2.5 mg albuterol by nebulization, and reassessment
 - B. Oxygen, racemic epinephrine by nebulization, ceftriaxone, and reassessment
 - C. Oxygen, 7.5 mg albuterol and 500 mcg ipratropium by nebulization and reassessment
 - D. Oxygen, albuterol nebulization, IV corticosteroids, and reassessment
 - E. Oxygen, stat CXR, and reassessment
8. This child does not respond to initial therapy. On reassessment she is becoming lethargic and despite oxygen her pulse oximetry occasionally dips to <70%. This is accompanied by frequent periods of bradycardia that respond to tactile stimulation
- Appropriate therapy at this point would include
- A. Endotracheal intubation, IV corticosteroids, IV antibiotics
 - B. Trial of CPAP, IV hydration, endotracheal intubation if not improved
 - C. Heliox, contiguous albuterol nebulization and intubation if not improved
 - D. Continuous albuterol, IV hydration and intubation if not improved
 - E. IM epinephrine or IV terbutaline and endotracheal intubation
9. CXR findings in a 6 week old who presents with wheezing, tachypnea, retractions, and mild hypoxia,

that would suggest a diagnosis other than bronchiolitis include all of the following except

- A. Right-sided aortic arch
- B. Atelectasis
- C. Lobar infiltrate
- D. Localized hyperinflation
- E. Enlarged cardiac silhouette

ANSWERS

1. E. *Mycoplasma* has been shown to be the principal agent in school-age children with bronchiolitis. RSV is responsible for 70% of all bronchiolitis cases and for 80% to 100% of cases in winter months. Parainfluenza, adenovirus, and influenza account for most of the remaining cases. Infection with RSV does not grant permanent or long-term immunity. Reinfections are common and may be experienced throughout life. *Mycoplasma* has been shown to be the principal agent in school-age children with bronchiolitis.

2. A. Inflammation, necrosis, and luminal obstruction. Infection produces inflammation of the bronchiolar epithelium, which leads to necrosis, sloughing, and luminal obstruction. Increased mucus production and edema contribute further to airway obstruction. The absence of ciliated epithelium prevents adequate mobilization of secretions and debris. Histologic sections of the tracheobronchial tree of patients with bronchiolitis are very similar to those in asthmatics. The bronchioles and small bronchi are obstructed secondary to the submucosal edema, peribronchiolar cellular infiltrate, mucous plugging, and intraluminal debris. Bronchospasm and hyperresponsiveness can occur during bronchiolitis but do not represent the key pathophysiology. Dehydration is also common in infants with bronchiolitis but is a secondary to the underlying pathophysiology.

3. E. Bronchiolitis. This is a typical presentation of an infant with bronchiolitis. The majority of infants of 2–12 months of age who present to the ED with respiratory distress or wheezing, will have diagnosis of bronchiolitis. In the United States, it is the leading cause of hospitalization in infancy.

However, it must be remembered that all wheezing infants do not have bronchiolitis and other diagnosis must be considered in the differential. Congenital heart disease, cystic fibrosis, vascular rings, and other congenital anomalies may all mimic the findings of bronchiolitis. Infants and toddlers are particularly prone to foreign body aspiration, and this possibility must be considered. History is often the most important factor in making the diagnosis.

4. A. No additional studies. There is controversy about whether all patients with a first-time episode of bronchiolitis should undergo a chest radiograph. The American Academy of Pediatrics (AAP) practice guidelines for bronchiolitis state that the current evidence does not support routine radiography in children with bronchiolitis. Laboratory studies are generally not helpful and not indicated in the acute management of bronchiolitis. The use of complete blood counts has not been shown to be useful in either diagnosing bronchiolitis or guiding its therapy. Virologic tests for RSV, if obtained during peak RSV season, demonstrate a high predictive value. However, the knowledge gained from such testing rarely alters management decisions or outcomes for the vast majority of children with clinically diagnosed bronchiolitis. Young febrile infants with clinical bronchiolitis are less likely to have an occult bacterial infection than febrile infants without bronchiolitis.

Hypoxia is common and the patient should have oxygen saturations assessed with a pulse oximeter. Hypercarbia will be present in those with more severe obstruction. Respiratory rates greater than 60 breaths/min correlate well with carbon dioxide retention noted on blood gas analysis.

5. D. Nasal suctioning, oxygen, albuterol nebulization, if symptoms persist, and reassessment.

No medication has been shown to be beneficial in patients with bronchiolitis. Therapy is mainly supportive. Nasal suctioning has been shown to improve respiratory distress scores in children with bronchiolitis. The AAP practice guidelines on the diagnosis and management of bronchiolitis state that the supplemental oxygen is indicated if SpO₂ falls persistently below 90% in previously healthy infants.

A chest radiograph often reveals areas of opacity suggestive of atelectasis or pneumonia. However, no significant benefit has been demonstrated from routine antibiotic usage in patients with a clinical diagnosis of bronchiolitis. There is no evidence that the addition of ipratropium to B agonist therapy benefits children with bronchiolitis.

The use of inhaled bronchodilators in bronchiolitis remains controversial. Most clinicians believe that bronchodilators produce clinical improvement in some patients with bronchiolitis. However, systematic reviews looking at studies of the efficacy of β-agonists in bronchiolitis have not demonstrated a significant benefit of their routine use. The AAP practice guidelines state that a carefully monitored

trial of α -adrenergic or β -adrenergic medication is an option.

6. B. Discharge home with nebulized albuterol and follow up with primary care physician. Inhaled bronchodilators should be continued only if there is a documented positive clinical response to the trial using an objective means of evaluation. As noted above other medications such as corticosteroids and antibiotics have not been shown to be of benefit. The following factors predict safe discharge to home: Age of ≥ 2 months, no history of intubation, history of eczema, age-specific respiratory rates (<45 breaths/min for 0–1.9 months, <43 breaths/min for 2–5.9 months, and <40 breaths/min for 6–23.9 months), no/mild retractions, initial oxygen saturation of $\geq 94\%$, fewer albuterol or epinephrine treatments in the first hour, and adequate oral intake. The importance of each factor varied slightly according to age. Other suggested criteria for admission include age (adjusted for prematurity) less than 6 weeks, hypoxemia, and persistent respiratory distress. Follow-up within 24 hours is recommended for those who are discharged.
7. A. Oxygen, 2.5 mg albuterol by nebulization, and reassessment. Supplemental oxygen is indicated if SpO_2 falls persistently below 90%. As discussed above a trial of B agonist therapy may be warranted in infants with bronchiolitis. Some authors have suggested that the use of nebulized epinephrine therapy is superior to nebulized albuterol therapy in these patients. However, as noted above, empiric antibiotic therapy is not indicated in patients with a clinical diagnosis of bronchiolitis. Ipratropium and corticosteroids have not been shown to be of benefit.
8. B. Trial of CPAP, IV hydration, endotracheal intubation if not improved. Continuous positive airways pressure (CPAP) has been used successfully as an adjunctive therapy in patients with severe symptoms. Two to five percent of infants hospitalized for bronchiolitis will go on to develop respiratory failure and require mechanical support. There are no absolute criteria for endotracheal intubation. Suggested indications include PCO_2 greater than 60 to 65 mm Hg, recurrent apnea spells, decreasing mental status, and hypoxia despite oxygen therapy. Once intubated, these infants have many of the same problems that intubated asthmatics have and are at risk for air trapping and the development of air leaks. A mixture of helium and oxygen (heliox) has also been shown to be of benefit by some authors. However, no large-scale clinical trial of this therapy has been conducted.

Inhaled bronchodilators should be continued only if there is a documented positive clinical response to the trial using an objective means of evaluation. Parenteral or oral B agonists have not been shown to be of benefit in these patients. As noted above, empiric antibiotics and corticosteroids are also not indicated.

9. B. Atelectasis. A chest radiograph will reveal hyperinflation in the majority of patients with bronchiolitis. Peribronchial cuffing (thickening of the bronchiole walls) will be seen in approximately half. There may be areas of subsegmental atelectasis that can be difficult to differentiate from pneumonia. A chest radiograph may be useful in ruling out the other disease processes in the differential diagnosis of bronchiolitis. A right-sided aortic arch may be suggestive of a vascular abnormality. A lobar infiltrate would suggest pneumonia/localized hyperinflation may represent foreign body aspiration of congenital emphysema. An enlarged cardiac silhouette suggests congenital heart disease of myocarditis each of which may present with wheezing, respiratory distress and hypoxia.

43 PNEUMONIA

Sharon Mace

OVERVIEW

- Pneumonia is an inflammation of lung parenchyma, most commonly due to an infection, occasionally from a noninfectious etiology (physical or chemical).
- Pneumonia is determined by an abnormal chest radiograph (CXR) showing pulmonary infiltrates although a presumptive clinical diagnosis of pneumonia is based on clinical signs and symptoms.
- In the United States, pneumonia is the No. 1 cause of death from an infectious disease in all ages of patients. Worldwide, pneumonia is the No. 1 killer of children.
- Respiratory tract infections are the No. 1 diagnosis for ED visits in children (under 15 years). Respiratory illnesses account for 20% of all pediatric hospital admissions and 10% of all pediatric ED visits. There are greater than 200,000 pediatric hospital admissions each year in the United States for pneumonia with an average length of stay = 5.2 days.

CLINICAL PRESENTATION

- Wide clinical spectrum of disease from mild illness to life-threatening disease
- Attack rates are highest in the youngest patients.
- Triad (fever, cough, and rales) is often present in adolescents/adults and is rarely present in the infant or young child.
- Symptoms: cough, chest pain; nonspecific symptoms in infant and young child: lethargy, irritability, apnea, poor feeding; gastrointestinal symptoms: vomiting, diarrhea, abdominal pain.
- Nonspecific signs in infant and young child: abnormal vital signs (fever, hypothermia, tachycardia, bradycardia, and tachypnea); respiratory distress: hypoxia, retractions, flaring, grunting, tachypnea; fever without source, poor muscle tone, sepsis, shock.
- Clinical presentation depends on many factors including age, comorbidity, risk factors, disease severity, and causative microorganisms.
- Two clinical presentations overlap. “Typical” pneumonia: presumed due to a bacterial pathogen, sudden onset of high fever, chills, pleuritic chest pain, productive cough, toxic appearance, and rales. “Atypical” pneumonia usually due to a virus, mycoplasma or Chlamydia; has a gradual onset, low grade fever, nonproductive cough, malaise, headache, and physical examination with \pm wheezing, \pm viral enanthem, \pm upper respiratory infection (URI): \pm rhinitis, \pm pharyngitis, \pm conjunctivitis.
- Determination of the etiologic agent based solely on clinical presentation, laboratory data, or radiographic findings is not 100% sensitive or specific although clinical presentation and other data (including CXR and laboratory data) may suggest a likely pathogen.
- Clinical presentation and risk factors indicate severity and need for admission: dehydration, apnea, respiratory distress, hypoxia, toxic appearance, altered mental status, sepsis, and shock, risk factors: immunosuppression or a chronic disease.

CAUSES OF PNEUMONIA

- Bacteria—common: “typical” *Streptococcus pneumoniae* (“pneumococcal pneumonia”). “Atypical” *Mycoplasma pneumoniae*, chlamydia (formerly *Chlamydia pneumoniae*).
- Bacteria—less common: “typical” staphylococcus, group β streptococcus, listeria, klebsiella, pseudomonas, hemophilus: type B/nontype B, *Moraxella catarrhalis*. “Atypical”: Legionella, Bordetella pertussis.

- Viral—common: respiratory syncytial virus (RSV), human metapneumovirus, parainfluenza, influenza.
- Viral—less common: coronavirus, adenovirus, rhinovirus.
- Other: fungal, rickettsial, protozoa.
- Noninfectious causes:
 - Inflammation from physical agents: inhalation injury (smoke, toxic inhalants), lipoid pneumonia, kerosene pneumonia.
 - Inflammation from chemical agents: drugs: chemotherapeutic agents (eg, bleomycin), antibiotics (eg, nitrofurantoin).
 - Iatrogenic lung disease: graft versus host disease.

PNEUMONIA SYNDROMES: PRESENTATION BASED ON ETIOLOGIC AGENT

- A given patient may have all, some, or none of the classic features of pneumonia and there is overlap between the clinical presentations.
- Bacterial: any age especially neonates, sudden onset, about 50% occur after URI, may have sputum if >10 years, \pm pleuritic chest pain, toxic appearance, usually high fever ($>39^{\circ}\text{C}$), rales/decreased breath sounds, may obtain blood culture, CXR: consolidation, pleural effusion.
- Viral: any age, especially toddlers/preschoolers, gradual onset, dry nonproductive cough, sometimes coryza/sore throat/rash, nontoxic appearance, usually low grade fever, rales/wheezing/rhonchi, normal or slight increase WBC, consider nasal aspirate for RSV, CXR: hyperinflation, interstitial infiltrates.
- Afebrile pneumonia of infancy—chlamydia: age 1–4 months, gradual onset, dry cough (may be only symptom), conjunctivitis in $\approx 50\%$ (may precede or occur with pneumonia), nontoxic appearance, usually afebrile, rales/wheezing, normal or slight increase WBC, eosinophilia, CXR: hyperinflation, interstitial infiltrates.
- Mycoplasma: school age/adolescent, gradual onset, usually dry cough, sometimes sore throat/rash/bullous otitis/media, headaches/myalgias, nontoxic appearance, usually low grade fever, sometimes rales, atypical lymphocytosis, sometimes positive cold agglutinins, CXR: interstitial infiltrates.
- Tuberculosis: any age >4 months, onset: usually gradual/may be acute, usually productive cough, sometimes hemoptysis, weight loss, night sweats, usually nontoxic, variable fever/lung auscultation/WBC, diagnosis: sputum, gastric aspiration, skin test, blood test (recently available), CXR: hilar adenopathy, infiltrates, cavitation, apical location.

RISK FACTORS FOR PNEUMONIA

- Age, prematurity (attack rates highest in youngest patients)
- Malnutrition
- Acute respiratory infections
- Chronic disease (respiratory, cardiac, neurologic, neuromuscular, gastrointestinal, immunologic), malignancy
- Immunosuppression includes HIV
- Congenital anatomic abnormalities (eg, trachso-phageal fistula)
- “Iatrogenic”: intubation, s/p tracheostomy, drugs (impair reflexes, cause immunosuppression), radiation, surgery, invasive pulmonary procedures
- Environmental: cigarette smoking (second hand smoke), daycare, crowded living conditions, inhaled pollutants, low socioeconomic status
- Microorganism: virulence factors

PNEUMONIA: ETIOLOGIC AGENT BASED ON AGE/PREVALENCE

- Neonate: usually from aspiration of maternal genital organisms during labor/delivery, most common: group β streptococcus, then gram-negative bacilli (*Escherichia coli*, Klebsiella), others: listeria, other streptococci, *S aureus*, *Bordetella pertussis*, anaerobic bacteria, viruses.
- Infants 1–3 months: same organisms as for neonates, chlamydiophilia (formerly chlamydia, “afebrile pneumonia of infancy”), viruses (eg, RSV, parainfluenza).
- Most common (in order of prevalence).
 - Infants—3–24 months: viruses (RSV, human metapneumovirus, parainfluenza, others), *S pneumoniae*, *H influenza* (non-type B), *M catarrhalis*, *M pneumoniae*, group A streptococci, *H influenza* type B (if nonimmunized).
 - Toddler/preschooler: viruses (RSV, parainfluenza, adenovirus, influenza, others), *S pneumoniae*, *H influenza* (non-type B), “atypicals”: *M pneumoniae*/Chlamydiophilia, *S aureus*, group A streptococci, *M catarrhalis*.
 - School age/adolescents: Atypicals (*M pneumoniae*, less commonly: chlamydiophilia), *S pneumoniae*, viruses (RSV, parainfluenza, influenza, adenovirus, rhinovirus).
 - Severe life-threatening pneumonia (excluding newborns): *S pneumoniae*, *S aureus*, group A streptococcus, *H influenza* B, adenovirus, *M pneumoniae* and these patients will frequently need intensive care unit admission.

FACTORS ASSOCIATED WITH SPECIFIC ETIOLOGIES OF PNEUMONIA

- Severe acute respiratory syndrome (SARS): exposure to infected individual and/or in the endemic region.
- Tuberculosis: exposure to infected individual or visited endemic area.
- Coccidiomycosis—recent travel to southwestern United States: San Joaquin Valley, Southern California, New Mexico, Southern Arizona, Southwest Texas
- Histoplasmosis: Mississippi and Ohio River Valley.
- Hantavirus: southwestern United States (four corners area of Arizona, New Mexico, Nevada, and Utah).
- Melioidosis: South and Central America, West Indies, Australia, Southeast Asia, Guam.
- Legionnaire’s Disease: contaminated water supply, air coolers, any open water source.
- Psittacosis: exotic birds (parrots, cockatoos, etc), turkeys, pigeons.
- Anthrax: cattle, horses, swine, animal hides.
- Brucellosis: unpasteurized dairy products, cattle, goats, pigs.
- Bubonic plague: rats, lagomorphs (squirrels, rabbits, chipmunks, etc).
- Hantavirus: rodent excretions, urine, saliva, droppings.
- Histoplasmosis: soil with bird or bat droppings.
- Leptospirosis: water contaminated with animal urine, wild dogs, rodents, cats, cattle, horses, pigs.
- Pasteurella multocida: infected dogs, cats.
- Q fever: secretions of infected animals or contact with animals (cattle, sheep, goats, other domestic animals).
- Tularemia: hunting, trapping, skinning of infected animals.

SPECIFIC PATHOGENS

- *Streptococcus pneumoniae*
 - Most common cause of bacterial pneumonia in all ages of pediatric patients excluding neonates although this may change in the future due to the introduction of the pneumococcal vaccines.
 - Classic presentation of pneumococcal pneumoniae: sudden onset fever, chills, cough, may have blood tinged sputum in adolescents/adults, pleuritic chest pain, dyspnea.
 - Increased risk if asplenic (anatomic or functional, eg, sickle cell patients).
 - Decreased incidence in vaccine serotypes since introduction of pneumococcal vaccines.
- *Staphylococcus aureus*
 - Uncommon, rapidly progressive fulminant illness.
 - Primary or secondary infection.

- Blood cultures positive in 20–30%.
- CXR: pleural effusions/pneumatoceles in 45–60%.
- *Haemophilus influenzae*
 - Occurs most often in young children.
 - Extrapulmonary infections are common.
 - Marked decrease incidence since introduction of HIB vaccine.
 - *H influenzae* nontype B infections also occur.
- *Mycoplasma pneumoniae*
 - Symptoms: low grade fever, nonproductive cough, sometimes: myalgias, headache, abdominal pain, vomiting.
 - Lung exam: normal, rales, or decreased breath sounds.
 - Lab: normal WBC, positive cold agglutinins, elevated transaminases.
 - Neurologic complications (occurs in 7% of patients): meningoencephalitis, transverse myelitis, Guillain–Barre syndrome, ataxia.

LABORATORY STUDIES

- May not be needed in every pneumonia patient but may be helpful in some patients in identifying underlying pathogen/complications.
- WBC (usually): normal or slight increase (<15,000) if viral, normal; if *M pneumoniae* or *C pneumoniae*, elevated if bacterial or β pertussis.
- Differential (usually): lymphocytosis if viral, chlamydia or pertussis, marked lymphocytosis if pertussis, eosinophilia if chlamydia or parasite, bandemia with leukocytosis (WBC >15,000) if bacterial, leucopenia suggests poor prognosis
- Consider occult pneumonia in febrile patients with a WBC $\geq 20,000$.
- Blood cultures are recommended in seriously ill patients or those with complications although the yield is only 10–15% in hospitalized pediatric patients, 1–16% yield with community acquired pneumonia in children/adults, 10–30% in adults with pneumococcal pneumonia. Blood cultures may not be needed in all patients hospitalized with pneumonia.
- Sputum cultures are rarely obtainable in children <10 years of age although are recommended for tuberculosis and in selected patients.
- Rapid antigen testing for various viruses (eg, RSV, influenza, parainfluenza) and fluorescent antibody testing for β pertussis and *C trachomatis* gives a faster result than culture.
- Serologic testing with acute and convalescent sera may be useful in some patients.
- Positive cold agglutinins occur in 70–90% of patients with *M pneumoniae* but false negatives (eg, young children) and false positives (some viruses) can occur.

- Invasive diagnostic tests (such as transtracheal aspiration, bronchoalveolar lavage, lung biopsy (percutaneous or open) is generally reserved for critically ill patients unresponsive to therapy.

RADIOGRAPHIC EVALUATION

- CXR is useful in making the diagnosis, eliminates other etiologies, may suggest the likely pathogen (allowing more specific antimicrobial therapy), and detects complications.
- CXR is noninvasive, painless, easily obtained, readily available, fairly inexpensive, low risk of radiation, can give valuable information, indicated in most pneumonia patients.
- “Round” pneumonia is a large solitary consolidated spherical shaped or round lesion typically due to *S pneumoniae*.
- Three main patterns of pneumonia on CXR: (1) lobar (alveolar) usually bacterial; (2) bronchopneumonia (lobular)—small fluffy infiltrates, peribronchial markings, patchy infiltrates, often due to *Mycoplasma* or bacteria (gram-negative bacteria, *S aureus*); (3) interstitial: hyperinflation, peribronchial cuffing (thickening), increased bronchovascular markings.
- Viral pneumonias usually have an interstitial pattern, but can have bilateral bronchoalveolar or peribronchial infiltrates.
- A patient with clinical signs/symptoms of pneumonia may have a negative CXR, and later, after treatment, the repeat CXR is positive for pneumonia (eg, there may be a time lag between clinical finding and the CXR).
- It is not possible to make a definitive diagnosis based on one study (CXR or lab alone); however, the CXR and lab may suggest an etiology (bacteria or viral).

MANAGEMENT OF PNEUMONIA

- Supportive care, appropriate antimicrobial use, hospitalizations in some patients
- Indications for hospitalization: neonates, young infants (≤ 3 –6 months), dehydration, inability to take medications or fluids, failure of outpatient therapy, psychosocial concerns, comorbidity, respiratory distress, hypoxia, complications (such as empyema, abscess, fistula, etc), bacteremia, sepsis, immunocompromised, suspected virulent pathogen (eg, *S aureus*, *P carinii*)

EMPIRIC THERAPY FOR COMMON BACTERIAL CAUSES OF PNEUMONIA*

- Neonate: ampicillin plus third-generation cephalosporin** or gentamycin for group β streptococcus/gram-negative enteric bacteria/listeria, inpatient.
- Infant (1–3 months): third-generation cephalosporin (such as ceftriaxone or cefuroxime)*** plus a macrolide for *S pneumoniae*, *H influenzae*: type B or nontype B, chlamydia; usually inpatient.
- Infant/young child (3 months–5 years): third-generation cephalosporin plus a macrolide or if outpatient: Augmentin for *S pneumoniae*, *H influenzae*, group A streptococcus; use azithromycin for chlamydia—infant and *M pneumoniae*—toddler
- School age/adolescent (>5 years–18 years): third-generation cephalosporin plus macrolides or if outpatient, macrolide for *S pneumoniae*, *M pneumoniae*.
- Any age patient who is critically ill: third-generation cephalosporin plus macrolide plus vancomycin (or clindamycin), for all of the above-mentioned plus staphylococcus; Intensive Care Unit.

INDICATIONS FOR HOSPITAL ADMISSION FOR PNEUMONIA

- Age: neonate (≤ 30 days), young infant (≤ 3 –6 months) (usually admitted).
- Inability to take oral fluids or medications, dehydration.
- Lack of response to outpatient therapy.
- Psychosocial issues: compliance with therapy, follow-up.
- Comorbidity: chronic lung disease of infancy, sickle cell disease, cystic fibrosis, others.
- Respiratory failure or distress, hypoxia, apnea.
- Complications: lung abscess, empyema, fistula, other pulmonary complications; bacteremia, sepsis.
- Immunocompromised: malignancy, HIV, others.
- Suspicion of virulent pathogen: *S aureus*, *P carinii*, others.

*Common bacterial pathogens for each age group are listed. Options for empiric therapy are suggestions that may change based on local and future resistance patterns and epidemiologic factors. If other specific pathogens such as gram-negative bacteria are suspected, other antibiotics such as piperacillin tazobactam may be indicated.

**Ceftriaxone is not recommended in neonates because of concern for hyperbilirubinemia. Use cefotaxime instead.

***If Staphylococcus is a possibility and vancomycin or clindamycin is contraindicated because of allergies or resistance, the “antistaphylococcal cillins” (eg, nafcillin, oxacillin, or methicillin) or other antistaphylococcal antibiotics.

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QUESTIONS

1. An 8-year-old girl is brought to the Emergency department for evaluation of cough, rhinorrhea, and fevers. Her symptoms have been present for 1 week. On examination, the child has decreased breath sounds on the right base, and a pulse ox is 92% on RA. You suspect pneumonia. Which of the following is correct regarding respiratory infections and/or pneumonia?
 - A. In children <15 years of age, acute upper respiratory infections are the third most common diagnosis for emergency department visits
 - B. Respiratory illnesses account for 5% of all pediatric hospital admissions

- C. Respiratory illnesses account for 1% of all pediatric emergency department visits
 - D. Worldwide, pneumonia is the No. 1 killer of children
 - E. In all ages of patients, pneumonia is the No. 3 cause of death from an infectious disease
2. Which of the following applies to the clinical presentation of pneumonia?
 - A. The highest attack rates occur in adolescents
 - B. The classic triad: fever, cough, and rales, and is almost always present in infants
 - C. Nonspecific signs and symptoms are common in infants and young children
 - D. Symptoms of “typical” pneumonia may include sudden onset of high fever, chills, chest pain, cough, and rales, and is presumed to be caused by a virus
 - E. “Atypical” pneumonia is presumed due to a bacteria (such as *H influenza*) and presents with sudden onset of high fever, productive cough, and rales
 3. Which of the following applies to causes of pneumonia in immunocompetent pediatric patients (excluding neonates)?
 - A. Common pathogens include protozoa and rickettsia
 - B. Common pathogens include fungi and protozoa
 - C. Common viral pathogens include the corona virus and herpes virus
 - D. A common bacterial pathogen is listeria
 - E. Common bacterial pathogens (excluding neonates) are *S pneumoniae*.
 4. Which of the following is the correct etiologic agent based on age and prevalence (common cause of pneumonia) in immunocompetent pediatric patients?
 - A. Neonate: pneumococcal pneumonia (*S pneumoniae*)
 - B. Infants 1–3 months: chlamydia (formerly chlamydia)
 - C. Toddlers: *E coli*
 - D. Adolescents: listeria
 - E. School age: klebsiella
 5. Of the following, the correct location associated with the specific pathogen of pneumonia is
 - A. Coccidiomycosis: southeastern United States (eg, Florida, Georgia)
 - B. Histoplasmosis: southwestern United States
 - C. Meliodosis: Eastern Europe
 - D. Histoplasmosis: Mississippi/Ohio River Valley
 - E. Hantavirus: northwestern United States (Pacific Coast: Oregon/Washington State)
 6. Of the following, the correct association of pneumonia with an animal is
 - A. Psittacosis: exotic birds, turkeys, pigeons
 - B. Brucellosis: rats
 - C. Hantavirus: dogs
 - D. Histoplasmosis: dog excretions
 - E. Anthrax: cats
 7. Of the following, the correct association of pneumonia with an animal is
 - A. Q fever: secretions of infected rodents
 - B. Pasteurella species: infected cattle
 - C. Hantavirus: infected lagomorphs (squirrels, rabbits, chipmunks)
 - D. Tularemia: hunting, trapping or skinning of infected animals
 - E. Bubonic plague: infected cats and dogs
 8. Of the following, which is a suggested empiric therapy for common bacterial causes of pneumonia based on age group (in a noncritically ill, immunocompetent pediatric patient)
 - A. Neonate: ceftriaxone and gentamycin
 - B. Infant (1–3 months) penicillin and a third-generation cephalosporin
 - C. Infant/young child (3 months–5 years) ampicillin and tetracycline
 - D. School age/adolescent: nafcillin
 - E. Adolescent: macrolides
 9. Which of the following is an indication for hospital admission for pneumonia?
 - A. Time of year (winter, specifically January)
 - B. Decreased appetite for solids but with adequate po fluid intake
 - C. Fever ($\geq 38.0^{\circ}\text{C}$)
 - D. Pulse oxygen saturation 85% on room air (no comorbidity, previously healthy)
 - E. 3-year-old toddler
 10. Based on the following scenario in children with pneumonia, which would be appropriate management?
 - A. A 10 year-old with a Hickman Catheter has a $T = 104^{\circ}\text{F}$ and a cellulitis of the forearm. Oral macrolides as an outpatient.
 - B. A 15-year old with no medical problems, $T = 101^{\circ}\text{F}$, vancomycin IV
 - C. A 20-day-old infant, $T = 103.5^{\circ}\text{F}$, ceftriaxone only
 - D. A 5-year old with leukemia on chemotherapy, $T = 102^{\circ}\text{F}$, $\text{WBC} = 1000$ with 10% polymorphonuclear leukocytes: oral macrolides
 - E. A 12-year old with no previous medical problems and a pulse oxygen saturation of 86% on room air macrolides and ceftriaxone, inpatient

ANSWERS

1. D. Pneumonia ranks No. 1 as the cause of death in children worldwide. In children <15 years of age, acute respiratory infections are the most common (No. 1 diagnosis) for emergency department visits. Respiratory illnesses account for 1 out of 5 or 20% of all pediatric hospital admissions. Respiratory illnesses account for 1 out of 10 or 10% of all pediatric emergency department visits. In the United States, for all ages of patients, the leading cause (#1) of death from an infectious disease is pneumonia
2. C. Infants often present with nonspecific signs and symptoms, such as lethargy, irritability, poor feeding, vomiting, isolated fever, or tachypnea. The highest attack rates for pneumonia occur in the youngest patients, infants and then decreases for each pediatric age group with the lowest attack rates in adolescents. Attack rates from highest to lowest are: infants, preschoolers, school age, then adolescents. Infants rarely present with the classic triad. It is more likely that adolescents (and adults) present with the triad: fever, cough, and rales. “Typical” pneumonia is presumed due to a bacteria (such as *H influenzae* or *S. pneumoniae*) and classically presents with the sudden onset of high fever, chills, cough, chest pain (if verbal patients), and rales. “Atypical” pneumonia is presumed due to a virus, mycoplasma, or chlamydia; and typically presents with a low grade fever and a nonproductive cough.
3. E. *S pneumoniae* is the most common bacterial pathogen in all ages of pediatric patients (except neonates) although this may change with the introduction of the pneumococcal vaccines. Protozoa and rickettsia are uncommon causes of pneumonia in all ages of immunocompetent pediatric patients. Fungi and protozoa pathogens are uncommon causes of pneumonia in all ages of immunocompetent pediatric patients. The common viral pathogens of pneumonia in pediatric patients are respiratory syncytial virus (RSV), human metapneumovirus, parainfluenza, and influenza. The corona virus and herpes virus are **not** common viral pathogens in immunocompetent pediatric patients of all ages. *Listeria* is a common pathogen only in neonates but not in other ages of immunocompetent pediatric patients.
4. B. “Afebrile pneumonia” of infancy, the most common Pneumonia in this age group, is characterized by a well-appearing afebrile infant with a dry cough who may have (or had) conjunctivitis. Conjunctivitis occurs in about half of the patients. Group β *Streptococcus*, then gram-negative bacilli (*E coli*, *Klebsiella*) are the most common bacterial pathogens in neonates. Pneumococcal (*S pneumoniae*) is unusual (rare) in neonates. The etiologic agents in the toddler/preschooler are viruses, *S pneumoniae*, *H influenza*, *M pneumoniae*, *Chlamydia*, and others. Gram negatives, such as *E coli* or *Klebsiella*, are more common pathogens in neonates and are unusual etiologic agents in toddlers. In adolescents, *M pneumoniae* followed by *S pneumoniae* then viruses are the common pathogens. *Listeria* is a common pathogen in neonates but not in adolescents. In adolescents, the atypicals (*M pneumoniae* and less commonly: *chlamydia*) are the most common pathogens causing pneumonia followed by *S pneumoniae* then viruses. Gram negatives such as *Klebsiella* are unusual causes of pneumonia in school age immunocompetent children.
5. D. The correct location is paired with the specific pathogen. Histoplasmosis: Mississippi/Ohio River Valley. Coccidiomycosis: southwestern United States—San Joaquin Valley, southern California, New Mexico, southern Arizona, southwest Texas. Melioidosis: South America, Central America, West Indies, Australia, Guam, Southeast Asia Hantavirus: southwestern United States—four corners area of New Mexico, Arizona, Nevada, and Utah
6. A. Psittacosis is associated with (transmitted by) exotic birds, turkeys, and pigeons. Brucellosis is associated with unpasteurized dairy products, cattle, goats, and pigs. Hantavirus is associated with rodent excretions, urine, saliva, droppings. Histoplasmosis is associated with soil contaminated with bird or bat droppings. Anthrax is associated with cattle, horses, swine, animal hides.
7. D. Tularemia is often found in hunters or individuals who process the meat/skins of animals such as deer. Q fever is transmitted by contact with infected domestic animals or their saliva (eg, cattle, sheep, goats, other domestic animals). The *Pasteurella* species is an organism found in the mouth flora of cats and dogs. Hantavirus is associated with rodent excretions, urine, saliva, droppings. Bubonic plague is transmitted via infected rats, and lagomorphs (squirrels, rabbits, chipmunks, etc).
8. E. A macrolide would be appropriate outpatient empiric therapy in an adolescent and would cover: *S pneumoniae* and *M pneumoniae*. A third-generation cephalosporin such as cefotaxime or cefuroxime plus gentamycin would be appropriate for a neonate. Ceftriaxone (rocephin) plus gentamycin would give appropriate antimicrobial coverage but ceftriaxone is not recommended

in neonates because of concern for hyperbilirubinemia. Another third-generation cephalosporin such as cefuroxime would be a better choice. A third-generation cephalosporin plus a macrolides would provide appropriate antimicrobial coverage for 1–3 month old infants. Penicillin is not recommended for the causative organisms at this age. A third-generation cephalosporin plus a macrolides (augmentin is an alternative in outpatients) would provide appropriate antimicrobial coverage for those in the 3 months to 5 year age group. Tetracycline is not recommended in children ≤ 8 years because of the side effect of discoloration of the teeth. Nafcillin provides coverage for *S. aureus*. Appropriate antimicrobial coverage for school age/adolescent patients would be a third-generation cephalosporin plus a macrolide. In an outpatient, a macrolide would cover the common bacterial pathogens *S pneumoniae* and *M pneumoniae*.

9. D. Hypoxia is one criterion for admission. A low pulse oxygen saturation in a previously healthy child indicates hypoxia. Although the precise oxygen saturation cut off point has been debated, most clinicians use a pulse oxygen saturation $<92\%$ as an indicator for admission. In the Northern hemisphere, pneumonia occurs more during the winter than the summer months. But this is not a criterion for admission. Dehydration from decreased/inadequate oral fluid intake and/or increased fluid losses (from fever, tachypnea, any associated vomiting or diarrhea, etc) may be an indication for admission. Decreased solid intake with adequate oral fluid intake should not cause dehydration and is not a criterion for admission. Fever, by itself, is not a criterion for admission. In general, most clinicians would admit a neonate (age <30 days) with pneumonia and some would extend this to include infants 2–3 months of age. A 3-year old who looks well with no indications for admission could be sent home from the emergency department, while a 2-week old also with no other indications for admission would probably be admitted even if he/she looked well.
10. E. Because he is previously healthy with no risk factors, he likely has community-acquired pneumonia that a third-generation cephalosporin and macrolides would provide adequate coverage. He is hypoxic so he has criteria for admission. The forearm cellulitis and the presence of a central venous catheter (eg, Hickman) are predisposing factors for a Staphylococcal infection so coverage should include an antistaphylococcal antibiotic, such as vancomycin, methicillin or oxacillin, or other antistaphylococcal drugs. A previously

healthy adolescent with a low-grade fever who looks well with no indications for admission could be treated as an outpatient on oral macrolides. Inpatient therapy for a staphylococcal pneumonia with IV vancomycin is not indicated. Coverage for neonates would be a third generation cephalosporin (but not ceftriaxone because of concern for hyperbilirubinemia, cefuroxime could be used), plus gentamycin. This child has neutropenic fever, needs cultures, intravenous antibiotics (such as ceftriaxone, azithromycin and vancomycin) and admission. Single oral antibiotic coverage as an outpatient is inappropriate and dangerous.

44 PERTUSSIS

Sharon Mace

OVERVIEW/SIGNIFICANCE

- An acute bacterial, highly contagious respiratory infection.
- Has a significant morbidity and mortality especially in infants.
- Incidence has been increasing especially in adolescents and adults.
- Reasons for increased incidence include waning immunity after childhood immunization, decreased immunization in specific patient populations (such as immigrants, those unimmunized for religious/other reasons), increased awareness of the disease, better diagnostic tests.
- In the prevaccine era, pertussis was No. 1 cause of death in children (≤ 13 years old) in the United States from a communicable disease.
- Asymptomatic cases are much greater (4–22 times) than symptomatic cases.
- Under-reported so number of actual cases is much greater (40–160 times) than reported cases.
- After introduction of vaccine in 1940s, the incidence declined precipitously but has been increasing in the United States and worldwide since the mid-1970s.

PATHOPHYSIOLOGY

- Transmitted in aerosolized droplets during coughing.
- Spread is limited to respiratory epithelium.
- It is almost never recovered from bloodstream.
- Bacteremia is not a feature of the disease, thus blood cultures are not helpful.

- Substances and toxins elaborated by pertussis increase its virulence and is responsible for lymphocytosis (thus, increased WBC count) and increased insulin secretion (that may cause hypoglycemia).
- Highly contagious, humans are the only reservoir and attack rates at close proximity are up to 100%.
- Primary reservoirs are adolescents/adults with a cough who are often not recognized as having pertussis.
- In one study, 21% of adolescents/adults with a prolonged cough (>1 week) had pertussis (confirmed by laboratory diagnosis).

CLINICAL PRESENTATION

- Three phases: catarrhal, paroxysmal, convalescent; they occur over 6 weeks.
- Catarrhal phase: nonspecific URI signs/symptoms such as low grade fever, congestion, rhinorrhea, lacrimation, sneezing, conjunctival suffusion for 7–10 days.
- Paroxysmal phase: episodes of dry hacking intermittent cough, lasts 2–4 weeks.
- Paroxysms of coughing may be accompanied by the characteristic “whoop”, a forceful inspiratory gasp “whooping cough.”
- Classic presentation: well-appearing toddler, suddenly becomes anxious then successive series of rapid uninterrupted coughs.
- Post-tussive emesis is characteristic and suggests the diagnosis.
- Convalescent phase: symptoms gradually decrease except in infants in whom cough and whoop become worse.
- Atypical presentations without classic findings are common.
- Young infants often lack three-phase presentation, have a short catarrhal phase, lack the severe cough/whoop, may have prolonged convalescent phase.
- Lung findings such as rales are usually absent, may have upper body petechiae and conjunctival hemorrhage from coughing.

DIAGNOSTIC EVALUATION

- Consider in anyone with chief complaint of cough, especially if they lack the usual signs/symptoms such as rales, wheezing, fever, myalgias, and sore throat.
- In one study, 21% adults/adolescents with cough >1 week had pertussis.
- Consider in infants with chief complaint of apnea or cyanosis.
- ↑ WBC (catarrhal stage) usually 15,000–40,000 may be up to 100,000.
- Lymphocytosis: normal size, not atypical lymphocytes of mononucleosis.

- ↑ polymorphonuclear leukocytes implies secondary infection or wrong diagnosis.
- Marked leukocytosis and/or thrombocytosis associated with ↑ severity, ↑ mortality.
- Hypoglycemia occurs infrequently.
- Chest x-ray: patients can have a pneumonmediastinum, pneumothorax, or subcutaneous emphysema from severe cough, if consolidation consider secondary pneumonia.

LABORATORY DIAGNOSIS

- Pertussis is caused by *Bordetella pertussis*.
- β pertussis is a small gram negative fastidious pleomorphic coccobacilli that is very difficult to grow (especially if in paroxysmal phase or received antibiotic), requires special culture media, takes several days to grow.
- Culture is gold standard for diagnosis but of low sensitivity (only 15–45%) so negative culture does not rule out the disease.
- PCR testing: quick results, better sensitivity than culture.
- Serology: must have two specimens—acute and convalescent samples, needs two blood draws and is the most sensitive test in immunized patients.

DIFFERENTIAL DIAGNOSIS

- Cough: any respiratory infection; noninfectious causes include asthma, reflux, foreign body.
- Coinfections with other respiratory pathogens can occur.
- 1/3 of infants hospitalized with pertussis also had RSV infection.

COMPLICATIONS

- Occur most often in youngest patients especially infants <6 months.
- Infants have highest morbidity and mortality.
- Vomiting, apnea, pneumonia, respiratory failure, dehydration.
- Rarely, patients may have seizures; death and SIDS can occur from apnea.
- Pneumonia: primary from pertussis or secondary bacterial.

TREATMENT

- Supportive care, avoid triggers for coughing including cold ambient temperature, strenuous activity.

- Hospitalize if any complications, need rehydration, and for young infants.
- Isolate hospitalized patient and monitor oxygen saturation, respiratory rate, and heart rate.
- Treat with antibiotics based on clinical presentation (even if negative laboratory tests). Treat with antibiotics if positive laboratory test (even if asymptomatic).
- Antibiotics decreases symptoms of the disease, duration, and the transmission (eliminates nasopharyngeal carriage).
- Antibiotics of choice are the macrolides: erythromycin, clarithromycin, azithromycin, alternative: trimethoprim–sulfamethoxazole (avoid in late pregnancy).
- Azithromycin may be preferred: fewer side effects, convenient dosing.
- In neonates, use azithromycin.
- Newer macrolides not FDA approved for ages <6 months but use anyway (CDC and AAP Red Book recommendations).
- An association between hypertrophic pyloric stenosis and the two macrolides: erythromycin and azithromycin has been reported.

PREVENTION

- Antimicrobial prophylaxis: same dose as for treatment can prevent symptoms. Prophylaxis for close contacts (irregardless of immunization status) and high risk individuals.
- Close contacts include household members, childcare workers, healthcare providers (in direct contact with secretions), face to face exposure within 3 feet or in confined space for ≥ 1 hour with a symptomatic patient.
- High risk individuals: young age (<1 year), immunosuppressed, significant comorbidity, women late in pregnancy.
- Vaccination is effective: two types: killed whole cell and acellular. Acellular: fewer reactions, more expensive, used in United States.
- Isolation includes standard precautions and droplet precautions (mask).
- Post-exposure immunization does not prevent infection.
- Booster vaccination for adolescents and adults, and healthcare workers.
- Natural disease does **not** confer immunity and Vaccination does not confer life-long immunity.
- Immunization (e.g. antibodies to pertussis) starts to wane 3–5 years after immunization and is gone by 12 years.
- This is why outbreaks have occurred in college campuses, residential facilities, in the elderly, healthcare facilities, nursing homes.

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QUESTIONS

1. A 10-year-old is brought to the emergency department by his mother for evaluation of a persistent cough. The mother reports that symptoms have been present for several weeks. The cough is described as a dry hacking intermittent cough. While in the waiting room, the patient has post-tussive emesis following the coughing spell. Which of the following is correct concerning the presumed diagnosis of pertussis?
 - A. The incidence has been decreasing in the United States in the last three decades (since 1976)
 - B. The number of symptomatic cases is greater than the asymptomatic cases
 - C. The number of cases is over-reported
 - D. In the United States in the prevaccine era, pertussis was the number one cause of death in children (≤ 13 year old) from a communicable disease
 - E. Vaccination confers lifelong immunity

2. A 4-year-old is brought to the emergency department for nonspecific URI signs/symptoms such as low grade fever, congestion, rhinorrhea, lacrimation, sneezing, conjunctival suffusion for 7–10 days. Based upon the history and physical, you suspect that the patient is in Catarrhal phase of pertussis. Which of the following applies to pertussis?
 - A. Pertussis is mildly contagious
 - B. Pertussis survives for long periods in the environment. It is spread primarily by contact with fomites.
 - C. It is spread by exposure to aerosolized droplets
 - D. In the United States, adults have adequate antibody to pertussis
 - E. The primary reservoir is toddler age children
3. You suspect pertussis in a 12-year-old girl brought to the emergency department with a cough that the parents describe as “whooping.” Which of the following laboratory test would be helpful based upon the pathophysiology of pertussis?
 - A. Blood cultures because of the high incidence of bacteremia
 - B. Serum glucose because of the high incidence of hypoglycemia
 - C. White Blood cell count because of high incidence of neutropenia
 - D. Urinalysis because of the association with pyuria
 - E. Serum acetone because of the incidence of diabetic ketoacidosis in these patients
4. You are doing beside teaching rounds in the emergency department with 2nd year students. Which of the following patients is more likely to present with characteristic picture suggestion of pertussis?
 - A. Toxic appearing school age child
 - B. Toxic appearing adolescent with a productive cough
 - C. Well-appearing toddler with a successive series of rapid uninterrupted coughs
 - D. Infant with wheezing
 - E. Infant with bilateral rales and a high fever
5. You have a 5-year-old nontoxic appearing child brought by his parents for cough and rhinorrhea. The symptoms have been present intermittently for 1 week. The cough is described as congestion based upon the duration of symptoms. You exclude pertussis from your diagnosis. Which of the following would apply to the clinical presentation of pertussis?
 - A. There is one phase denoted the catarrhal phase that lasts 6 weeks
 - B. The catarrhal phase is characterized by episodes of a dry hacking cough
 - C. The characteristic whoop of pertussis occurs during the catarrhal phase
 - D. Typical features of the paroxysmal phase are nonspecific URI signs and symptoms such as low grade fever, rhinorrhea, lacrimation
 - E. The paroxysmal phase lasting 2–4 weeks is characterized by episodes of a dry hacking intermittent cough
6. A 6-month-old ex-premie born at 32 weeks is brought to the emergency department from his daycare program by the program director. The caregiver notes that the infant has had post-tussive emesis and recurrent coughing episodes. The caregiver has received written authorization from the parents to seek medical attention for the infant. Which of the following is correct regarding infants with pertussis?
 - A. During the convalescent phase in infants, the symptoms (e.g. the cough and whoop) become worse
 - B. Infants always present with classic findings
 - C. Infants always have the typical three phase presentation
 - D. During the convalescent phase in infants, symptoms gradually improve
 - E. Pertussis in infants is usually mild
7. A teenager who has just returned from a 1 ½ week long camping trip with his school. The local health officials have notified your ED of an outbreak of pertussis in the area. Which of the following if present would support the diagnosis of pertussis in this patient?
 - A. A productive cough
 - B. A cough lasting more than 1 week
 - C. A generalized maculopapular rash
 - D. A low hemoglobin
 - E. A past medical history of asthma
8. A 7-year-old is brought by her parents after returning from a family vacation in overseas. While on vacation, the child develops a dry hacking cough. Mother states that the coughing episodes became so intense that the child’s eyes “turned red” last night. The mother is an EMT and is concerned about pertussis. Since your suspicion is reasonably high, you agree to proceed with a diagnostic evaluation. Which of the following is true regarding diagnosis of pertussis?
 - A. Is easy to grow on usual culture media
 - B. Culture is the gold standard for diagnosis
 - C. Culture has high sensitivity and specificity
 - D. A negative culture rules out the disease
 - E. PCR testing is less sensitive than culture and takes several days
9. The laboratory test confirms the diagnosis of pertussis in your patient. Which of the following is

correct regarding the treatment or prevention of pertussis?

- A. Treat only symptomatic patients with a positive laboratory test
- B. Penicillin is the antibiotic of choice
- C. The newer macrolides are not FDA approved but should be used in infants less than 6 months of age (CDC and AAP Redbook recommendation)
- D. Immunization confers lifelong immunity
- E. Prophylaxis is not indicated for exposure to pertussis

ANSWERS

1. D. In the prevaccine era, pertussis accounted for more infant deaths than deaths from diphtheria, poliomyelitis, measles, and scarlet fever combined. After the vaccine was introduced in the 1940s, the incidence decreased precipitously until 1976 when there were slightly over 1000 cases in the United States. However, since then (eg, 1976) the incidence has been steadily increasing. The number of asymptomatic cases is much greater than the symptomatic cases (asymptomatic cases estimated at 4–22 times greater than symptomatic). This is important because humans serve as a reservoir of the disease. The number of cases is greatly underreported. The rates of reported pertussis are much less (10- to 100-fold less) than reported cases. Pertussis is often undiagnosed. Pertussis should be in the differential of patients with a persistent cough. Patients may have pertussis, serve as a reservoir for the disease, and spread the disease to vulnerable individuals but not be diagnosed or know they have the disease. There is no lifelong immunity: Within 3–5 years of the vaccination, immunity starts to wane and by 12 years, immunity is gone. Thus, adolescents and adults who may have been immunized as a child are susceptible to the disease. It has recently been recommended to vaccinate adolescents, adults, and healthcare providers.
2. C. Pertussis is spread by aerolized droplets. Pertussis is highly contagious with attack rates up to 100% for individuals exposed to aerosol droplets with close proximity (a distance ≤ 5 feet). Pertussis does not survive long in the environment so it is not spread primarily by fomites. It is spread by exposure to aerosolized droplets. Because immunity starts to fall off within 3–5 years of vaccination and is gone within 12 years, adults (and adolescents) are susceptible to pertussis and it has recently been recommended that adults and adolescents be revaccinated. Because antibodies to pertussis starts to fall within 3–5 years

of an immunization and are gone by 12 years, adults and adolescents who often are unaware they have pertussis (and may just have a persistent cough) can get and spread the disease and are the main reservoir for the disease.

3. B. β pertussis produces various substances and toxins that increase its virulence and are responsible for some of the complications. The toxins are responsible for causing an increase in insulin secretion, which may lead to hypoglycemia.

The spread of β pertussis is limited to the respiratory epithelium so bacteremia is not a feature of the disease. β pertussis is almost never recovered from the bloodstream so blood cultures are not helpful. The pertussis toxin leads to an increase in lymphocytes (not in polymorphonuclear leukocytes) which in turn leads to an increase in the white blood cell count. Any increase in polymorphonuclear leukocytes suggests a diagnosis other than pertussis or a secondary bacterial infection. There is an increase in the white blood cell count due to an increase in lymphocytes.

4. C. This is the classic presentation: a well appearing toddler with a paroxysmal nonproductive cough. The classic presentation is a well appearing toddler. Pertussis may occur in any age but patients are usually well appearing not toxic.

Adolescents with pertussis are generally well appearing and often asymptomatic except for a persistent nonproductive cough. A productive cough or sputum is not a feature of pertussis. Wheezing is not a feature of pertussis. In an infant with wheezing, consider other diagnoses such as bronchiolitis or asthma. Infants with pertussis generally have a low-grade (not high) fever and the lung examination is clear unless there is a complication such as a secondary pneumonia.

5. E is true. The paroxysmal phase lasting 2–4 weeks is characterized by episodes of a dry hacking intermittent cough. There are three phases of pertussis: catarrhal, paroxysmal, and convalescent that occur over six weeks. The second phase, the paroxysmal phase is characterized by episodes of a dry hacking intermittent cough lasting 2–4 weeks. The first phase, the catarrhal phase lasts 7–10 days and is characterized by nonspecific upper respiratory infection (URI) signs and symptoms including low-grade fever, congestion, rhinorrhea, lacrimation, and sneezing, conjunctival suffusion.

The nonspecific URI signs and symptoms occur during the catarrhal phase (first phase). The characteristic whoop and the dry hacking intermittent

cough occurs during the paroxysmal phase (second phase).

6. A. Patients, except infants, get better gradually during the convalescent phase. Infants are the only age group that can get worse (eg increase in the cough and whoop) during the convalescent phase. Infants commonly have an atypical presentation without classic findings. Infants often lack the three-phase presentation.

Symptoms gradually worsen in infants during the convalescent phase. This is unlike all other age groups in whom symptoms decrease during the convalescent phase

Pertussis is generally most severe in infants. Infants have the greatest morbidity and mortality

7. B. In addition to the local outbreak and the recent camping trip, a prolonged cough should increase your suspicion for pertussis. The cough associated with pertussis is typically described as dry, hacking cough. About 1 in 5 (21%) of adolescent/adult patients with a cough >1 week had pertussis. Pertussis should be considered in patients with upper body petechiae and/or conjunctival hemorrhage from coughing. A rash is not a feature of pertussis and suggests another diagnosis. Anemia is not a feature of the disease. Pertussis should be considered in patients with upper body petechiae and/or conjunctival hemorrhage from coughing. A rash is not a feature of pertussis and suggests another diagnosis.

8. B. Culture is the “gold standard.” Pertussis is very difficult to grow and requires a special culture media to grow. Culture has a low sensitivity (only 15–45%).

Because the culture has a low sensitivity, a negative culture does **not** rule out the disease. PCR is more sensitive than culture and has a more rapid turnaround time than culture.

9. C. The newer macrolides are not FDA approved but should be used anyway because they are recommended by the CDC and AAP Redbook.

Macrolides (erythromycin, clarithromycin, and azithromycin) are the antibiotics of choice with trimethoprim–sulfamethoxazole as an alternative (except in late pregnancy). Treat with antibiotics based on clinical presentation even if the laboratory tests are negative. Treat with antibiotics if the laboratory tests are positive even if the patient is asymptomatic. Immunizations do **not** confer lifelong immunity. Immunizations start to wane within 3–5 years and are gone by 12 years so revaccination of adolescents and adults is recommended. Prophylaxis is indicated for close contacts regardless of immunization status and in high-risk individuals.

45 BRONCHOPULMONARY DYSPLASIA

Madeline Matar Joseph

HIGH-YIELD FACTS

Despite advances in the prevention and management of neonatal acute respiratory illness, bronchopulmonary dysplasia (BPD) persists as one of the major complications in surviving very-low-birth weight infants. BPD is a form of chronic lung disease of infancy that follows preterm neonatal lung disease treated with oxygen and positive-pressure ventilation (PPV). The original insult may be hyaline membrane disease, apnea, persistent fetal circulation, complex congenital heart disease, or any illness requiring prolonged mechanical ventilation as a neonate.

BACKGROUND

- The overall rate of BPD is approximately 15% for premature infants requiring mechanical ventilation. The incidence of BPD, defined as oxygen dependence at 36 weeks postmenstrual age (PMA), in infants with birth weights between 500 and 1500 g, ranges between 3% and 43% in different centers.
- A June 2000 National Institute of Child Health and Human Development/National Heart, Lung, and Blood Institute Workshop proposed a severity-based definition of BPD for infants younger than 32 weeks' gestational age (GA) (Table 45-1).
- Oxygen requirement at 36 weeks corrected for post-gestational age has been shown to be an excellent predictor of the development of BPD in this group of infants.

ETIOLOGY/PATHOPHYSIOLOGY

- The pathogenesis of BPD remains complex and not fully understood. BPD results from a variety of toxic factors that can injure small airways and

TABLE 45-1 Severity of Bpd

SEVERITY OF BPD	O ₂ SUPPLEMENT >28 WEEKS PMA	O ₂ SUPPLEMENT >36 WEEKS PMA
Mild	Yes	None
Moderate	Yes	Yes, <30% O ₂
Severe	Yes	Yes, >30% O ₂ and/or PP

PMA, postmenstrual age; PP, positive pressure.

that can interfere with alveolarization (septation), leading to reduction in the overall surface area for gas exchange and clinically significant pulmonary dysfunction.

- The major host susceptibility factor associated with BPD is immature lungs secondary to prematurity (22–32 weeks' gestational age) followed by the presence of symptomatic patent ductus arteriosus (PDA).
- Factors involved in secondary injury include mechanical ventilation, oxygen toxicity, infection, and inflammatory mediators.

MECHANICAL VENTILATION

- Trauma secondary to PPV is generally referred to as barotrauma. Volutrauma suggests the occurrence of lung injury secondary to excessive tidal volume from PPV.
- Many modes of ventilation and many ventilator strategies have been studied to potentially reduce lung injury. Infants weighing < 1000 g who received synchronized intermittent mechanical ventilation (SIMV) had less BPD than did other infants.
- Several investigators comparing primary HFOV or HFJV with conventional ventilation suggested that high-volume strategies that effectively recruit alveoli may prevent BPD.
- PPV with various forms of nasal continuous positive airway pressure (CPAP) has been reported to decrease injury to the developing lung, and it may reduce the development of BPD. Centers that used more CPAP and less intubation, surfactant, and indomethacin had the lowest rates of BPD.

OXYGEN THERAPY

- Activity of antioxidant enzymes (superoxide dismutase, glutathione peroxidase, and catalase) tend to increase during the last trimester of pregnancy.
- Pre-term birth exposes the neonate to high oxygen concentrations without having the antioxidant enzymes, leading to an increase in the risk of injury due to oxygen free radicals.
- Supplementation with superoxide dismutase, in ventilated preterm infants with RDS, substantially improved their clinical pulmonary status and markedly reduced readmissions among treated subjects compared with the placebo-treated control group.
- Ideal oxygen saturation for term or preterm neonates of various gestational ages has not been definitively determined. In practice, many clinicians have

adopted conservative oxygen saturation parameters (ie, 88–92%).

- A delicate balance exists to optimally promote neonatal pulmonary (alveolar and vascular) and retinal vascular homeostasis.

INFLAMMATION

- Elevated levels of interleukin-6 and placental growth factor in the umbilical venous blood of preterm neonates are associated with increased incidence of BPD.
- Activation of leukocytes after cell injury caused by oxygen free radicals, barotrauma, infection, and other stimuli may begin the process of destruction and abnormal lung repair that results in acute lung injury then BPD.
- Activation of transcription factors, such as nuclear factor-kappa B in early postnatal life, is associated with BPD or death.
- Increased activity and decreased function of α 1-proteinase inhibitor may worsen lung injury in neonates. A decrease in BPD and in the need for continued ventilator support is found in neonates given supplemental α 1-proteinase inhibitor.

INFECTION

- Infection—either antenatal chorioamnionitis and funisitis or postnatal infection—may activate the inflammatory cascade and damage the preterm lung, resulting in BPD.
- In fact, any clinically significant episode of sepsis in the vulnerable preterm neonate greatly increases the risk of BPD, especially if the infection increases the baby's requirement for oxygen and mechanical ventilation.

MALNUTRITION

- Malnutrition is frequently seen in sick neonates. It may have profound effects on lung defenses and repair capabilities.
- Vitamins A and E are nutritional antioxidants that may help prevent lipid peroxidation and maintain cell integrity. However, supplementation of vitamin E in preterm neonates does not prevent BPD.
- Data from meta-analyses reported in a Cochrane Database review of vitamin A supplementation suggest that vitamin A supplementation reduces the risk of BPD in neonates born prematurely and death at 36 weeks.

CLINICAL PRESENTATION

- BPD causes not only significant complications in the newborn period, but is associated with continuing mortality, cardiopulmonary dysfunction, rehospitalization, growth failure, and poor neurodevelopmental outcome after hospital discharge.
- The clinical spectrum of infants with chronic lung disease ranges from mild symptomatic disease to crippling cardiopulmonary dysfunction.
- Follow-up of pediatric patients with BPD to adulthood demonstrated that patients had airway hyperreactivity, abnormal pulmonary function, and hyperinflation, as noted on chest radiography.
- Patients who are likely to present to the ED are those who have been discharged home from the neonatal intensive care unit, typically at about 3 to 6 months of age. These children are often on home oxygen, bronchodilators, apnea monitors, and other medications.
- They will present to the ED with an exacerbation of their chronic lung disease most often secondary to a viral upper respiratory infection as manifested by increased respiratory distress, poor feeding, lethargy or irritability, and an increased oxygen requirement.
- On physical examination, infants with chronic lung disease will usually be small for age and have hyperinflated chests (increased anteroposterior diameter). They will have tachypnea, rales, wheezes, or areas of decreased breath sounds. They may also have signs of an upper respiratory infection, including fever.

LABORATORY AND RADIOGRAPHIC FINDINGS

- A chest radiograph will reveal variable degrees of hyperinflation with areas of “scarring” (cystic or fibrotic areas). Comparison with old films is required to differentiate these areas from acute processes.
- Oximetry should be checked on all patients with BPD. The results should be interpreted in light of the baseline level of hypoxia and usual need for oxygen therapy.
- Blood gas results can be helpful in assessing the more severely symptomatic patient. The results will also need to be compared with previous results, as these children will often have hypercarbia and hypoxia at baseline.

DIFFERENTIAL DIAGNOSIS

- In most cases, the diagnosis of BPD will be evident from the history. As many BPD exacerbations are triggered by upper respiratory infections, and often

involve reactive airways as part of the pathology, the signs and symptoms of an exacerbation may overlap considerably with that of pneumonia, asthma, or bronchiolitis.

- Often these problems are coexistent and are the cause of the exacerbation.
- A chest radiograph can be helpful in identifying pneumonia if the radiograph can be compared to previous films.
- Testing for RSV will help identify those patients who may need hospitalization.

TREATMENT

- The treatment of an exacerbation in a patient with BPD is mainly supportive.
- The most effective treatments for ameliorating symptoms or preventing exacerbation in established BPD patients include oxygen therapy, inhaled glucocorticoid therapy, and vaccination against respiratory pathogens.
- Many other strategies for the prevention or treatment of BPD have been proposed, but have weaker or conflicting evidence of effectiveness.
- Oxygen is provided, if indicated. The patient’s fluid status is assessed and intravenous fluids provided, if indicated.
- Mechanical ventilation may be necessary for recurrent apnea spells (most commonly with RSV infections), worsening hypercarbia, or refractory hypoxemia.
- Bronchodilators are often effective in these patients and should be used as they are for asthma.
- Systemic corticosteroids are thought to be effective in acute exacerbations, but their chronic use is associated with many side effects, most notably the recently recognized adverse effects on neurodevelopment, which precludes their routine use for the prevention or treatment of BPD. Inhaled steroids have been shown to increase the ability to extubate children who are chronically ventilated but its efficacy in nonventilated patients has not been demonstrated. A review by Shah et al. found no evidence that early-inhaled steroids confer important advantages over systemic steroids in the management of ventilator-dependent preterm infants. Neither inhaled steroids nor systemic steroids can be recommended as a part of standard practice for ventilated preterm infants. Because they might have fewer adverse effects than systemic steroids, further randomized-controlled trials of inhaled steroids are needed, which address risk/benefit ratio of different delivery techniques, dosing schedules, and

long-term effects, with particular attention to neurodevelopmental outcome.

- Parenteral diuretics have been shown to improve lung function and survival in some patients. Studies have looked at the effectiveness of aerosolized furosemide for patients with BPD, but no conclusive benefit has been demonstrated.

DISPOSITION

- Children with BPD often have a very fragile respiratory status and can become very sick with relatively minor insults.
- Indications for inpatient management include increased respiratory distress, increasing hypoxia or hypercarbia, new pulmonary infiltrates, or inability to keep up with appropriate fluid intake.
- Patients with BPD and RSV infections are at high risk for complications of RSV and need to be hospitalized. Parents may have difficulty coping with an exacerbation. This factor should be considered when making a decision to discharge a patient for home care.
- Four years of retrospective and prospective data on the use of palivizumab in clinical practice has accumulated a wealth of “real life” information on the clinical effectiveness of RSV immunoprophylaxis in a large cohort of high-risk infants.

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QUESTIONS

A 9-month-old patient, with a history of BPD, was brought to the emergency department with the chief complaint of increase work of breathing, wheezing and decrease in feeding.

1. What is the most likely cause of his symptoms?
 - A. Pneumonia
 - B. Asthma
 - C. Bronchiolitis
 - D. BPD exacerbation
 - E. All the above
2. You immediately start nebulizer treatments on the above mentioned patient. Following the second treatment, the symptoms begin to improve. What additional diagnostic testing (if any) could help you in the management of this patient?
 - A. CBC
 - B. Blood cultures
 - C. Chest radiograph
 - D. Sputum culture
 - E. RSV

3. A preterm neonate is brought to the ED for respiratory distress. The neonate is on home oxygen however the parents report increases oxygen demands at home. In obtaining at perinatal and postnatal history you learn that the mother was treated for an “infection” Which type of maternal infection would be associated with an increase rate of BPD?
 - A. Neonatal sepsis
 - B. Antenatal chorioamnionitis
 - C. Rubella
 - D. Group B streptococcus
 - E. TORCH complex
 4. You notice an increasing oxygen requirement at 36 weeks corrected for postgestational age in a preterm neonate. You are aware of the relationship between oxygen requirement and risk of developing BPD. Supplementation of which vitamin is suggested to reduce the risk of BPD in preterm neonates?
 - A. Vit A
 - B. Vit C
 - C. Vit B12
 - D. Vit E
 - E. Vit K
 5. A 7-month-old ex-premie at 32 weeks is brought to the ED for increased work of breathing. You suspect a BPD exacerbation. There is no history of fevers or sick contacts. The infant has a marginal response to treatments provided in the emergency departments. Which of the following would be the most compelling reason for admission?
 - A. Baseline hypoxia
 - B. Fever
 - C. Positive RSV
 - D. Decreased oral fluid intake
 - E. Baseline hypercapnia
- compared to previous films. Testing for RSV will help identify those patients who may need hospitalization.
3. B. Infection—either antenatal chorioamnionitis and funisitis or postnatal infection—may activate the inflammatory cascade and damage the preterm lung, resulting in BPD. In addition, any clinically significant episode of sepsis in the vulnerable preterm neonate greatly increases the risk of BPD, especially if the infection increases the baby’s requirement for oxygen and mechanical ventilation.
 4. A. Malnutrition is frequently seen in sick neonates, which may have profound effects on lung defenses and repair capabilities. Vitamins A and E are nutritional antioxidants that may help prevent lipid peroxidation and maintain cell integrity. However, supplementation of vitamin E in preterm neonates does not prevent BPD. Data from meta-analyses reported in a Cochrane Database review of vitamin A supplementation suggest that vitamin A supplementation reduces the risk of BPD in neonates born prematurely and death at 36 weeks.
 5. C. Patients with BPD and RSV infections are at high risk for complications of RSV and need to be hospitalized. Children with BPD often have a very fragile respiratory status and can become very sick with relatively minor insults. Other indications for inpatient management include increased respiratory distress, increasing hypoxia or hypercarbia, new pulmonary infiltrates or inability to keep up with appropriate fluid intake. Most infants with BPD have a baseline hypoxia and hypercapnia.

ANSWERS

1. E. As many BPD exacerbations are triggered by upper respiratory infections, and often involve reactive airways as part of the pathology, the signs and symptoms of an exacerbation may overlap considerably with that of pneumonia, asthma, or bronchiolitis. In addition, these problems are often coexistent and are the cause of the exacerbation.
2. E. CBC could be within normal limits despite the absence or presence of pneumonia. Extremely low number of bacteremia occurs with pneumonia complicating BPD. Chest radiograph can be helpful in identifying pneumonia if the radiograph can be

46 CYSTIC FIBROSIS

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HIGH YIELD FACTS

- Cystic fibrosis (CF) is the most common, life-limiting, autosomal recessive disease among Caucasians in the United States. It occurs in approximately 1/3500 white births.
- Most patients with CF have the classic triad of manifestations: chronic pulmonary disease, malabsorption due to pancreatic insufficiency, and elevated concentrations of sweat sodium and chloride.

ETIOLOGY/PATHOPHYSIOLOGY

- CF is caused by mutations in the gene that encodes the cystic fibrosis transmembrane conductance regulator (CFTR) protein. The CFTR gene is located on the long arm of chromosome 7.
- The most common mutation that causes CF is F508del. More than 1500 different mutations have been identified.
- Organs that express the CFTR gene—particularly the sinuses, lungs, pancreas, liver, gastrointestinal (GI) tract, and reproductive system—are the ones affected by the mutations.
- The most important pathophysiologic consequence of these CFTR “defects” is diminished water in mucus and most exocrine secretions (along with associated electrolyte and other abnormalities). Mucus and exocrine secretions are more viscid, and they are difficult to clear, causing airway and ductal obstruction.
- Airway obstruction is a prominent feature of CF, leading to atelectasis, enlarged air spaces, and ventilation–perfusion mismatching.
- Chronic airway infection and inflammation, along with repeated exacerbations, result in destruction of the airway. Bronchiolitis develops initially, accompanied by bronchitis later. With prolonged disease, bronchiolar obliteration, bronchiolectasis, and bronchiectasis ensue.
- Patients with progressively worsening lung disease may eventually develop pulmonary hypertension and right ventricular hypertrophy (cor pulmonale).

CLINICAL PRESENTATION

- Patients with undiagnosed CF may present to the ED for a variety of complaints. Failure to thrive with a history of chronic GI and/or respiratory problems is a typical presentation. CF should be considered in any patient with chronic cough, recurrent respiratory infections, atypical asthma, or chronic diarrhea.
- Sweat salt and water losses, with associated renal compensation, can lead to electrolyte depletion and dehydration. These occur most commonly in infants with gastroenteritis and poor oral intake, or during the hot summer months, especially in arid climates.
- Patients with known CF may present to the ED with pulmonary exacerbations. These are thought to be due to more active airway infection caused by *Staphylococcus aureus*, *Hemophilus influenzae*, *Pseudomonas aeruginosa*, or *Burkholderia cepacia*.
- Pneumothorax must be excluded in any patient who deteriorates suddenly.

- A pulmonary exacerbation may precipitate congestive heart failure.
- CF patients frequently have hemoptysis, especially during exacerbations. Bleeding is usually minor and not dangerous. Massive hemoptysis (acute bleeding >240 mL/d or >100 mL/d over several days) is not rare, however, and can cause life-threatening asphyxia or hypotension.
- Hemoptysis must be distinguished from hematemesis. Hematemesis is due to bleeding esophageal varices secondary to portal hypertension from advanced obstructive biliary tract disease and cirrhosis.
- Acute GI complications of CF also include meconium ileus (in the neonatal period), distal intestinal obstruction syndrome (formerly called meconium ileus equivalent), and rectal prolapse.
- As patients approach adulthood, they may develop diabetes mellitus and obstructive azospermia.

LABORATORY AND RADIOLOGIC FINDINGS

- DNA mutation analysis (for many of the common mutations) is available for detection of patients and carriers. (Many states will perform a limited DNA mutation analysis when the newborn immunoreactive trypsinogen screen is positive for CF.)
- Traditionally, the pilocarpine iontophoresis sweat test has been a part of the diagnostic evaluation in any patient with suspected CF. This test measures the chloride concentration in sweat. Values >60 mEq/L are considered positive (however, this cutoff is not 100% sensitive for diagnosing CF).
- During pulmonary exacerbations, oxygen saturation should be measured via pulse oximetry initially. Blood gas determinations may be useful in managing patients with respiratory failure, particularly those who have had good lung functions and suffer acute decompensations likely to respond to therapy.
- Sputum culture and gram stain should be obtained to guide antibiotic therapy.
- Chest x-rays should be compared with the most recent previous ones. Common chest x-ray findings (as well as acute changes) include hyperinflation, diffuse peribronchial thickening, and areas of atelectasis and fluffy infiltrates. Any patient with a sudden deterioration in pulmonary status must have a pneumothorax excluded by chest x-ray.
- Patients with meconium ileus or distal intestinal obstruction syndrome may have dilated loops of bowel on abdominal films. A bubbly granular density in the lower abdomen, representing the meconium or fecal mass, may also be seen.

- In patients with significant hemoptysis or hematemesis, blood should be sent for complete blood count; type and crossmatch; and prothrombin time.
- Hyponatremic, hypokalemic, or hypochloremic alkalosis is the classic electrolyte abnormality, and can be a diagnostic clue, particularly when discovered in a patient with other findings suggestive of CF.

DIFFERENTIAL DIAGNOSIS

- CF may be confused with a variety of disorders.
- These include: asthma; bronchopulmonary dysplasia; congenital bronchial or pulmonary abnormalities; foreign body aspiration; immotile cilia syndrome; immunodeficiency disorders; and intrathoracic neoplasms can cause protracted or recurrent respiratory symptoms.
- Non-CF causes of exocrine pancreatic insufficiency or elevated sweat electrolyte concentrations are relatively rare in children.

TREATMENT FOR PULMONARY EXACERBATION

- Oxygen should be administered if indicated by pulse oximetry or arterial blood gas.
- Inhaled β_2 -adrenergic bronchodilators are used by those who respond clinically.
- Decisions regarding specific antibiotics should be made in consultation with the patient's CF specialist. Until current sputum culture results are known, prior results can help guide antibiotic selection. If recent results are unavailable, empiric therapy should be aimed at the most common organisms seen in CF patients: *S aureus* and *H influenzae* in infants and young children, and *P aeruginosa*, which is often present by the end of the first decade of life. Exacerbations due to *P aeruginosa* should be treated with combination antibiotics, because treatment with a single antibiotic has been associated with rapid pseudomonal resistance. For patients with massive hemoptysis, anti-staphylococcal therapy should be considered.
- Pulmozyme (recombinant human deoxyribonuclease) is a "mucolytic" medication, which is administered via nebulization. It reduces sputum viscosity by enzymatically degrading the excessive DNA from dead neutrophils and other cells in airway mucus. Although pulmozyme is generally employed for chronic management, it can be continued during an acute exacerbation as well.
- Chest physiotherapy and postural drainage techniques are generally accepted (although not as well proven)

and widely used for clearing mucus secretions in CF patients.

- Anti-inflammatory medications may be beneficial in the long-term—and, possibly, acute—care of certain CF patients. Macrolides (eg, azithromycin), which possess both an anti-inflammatory and antibacterial properties, and high-dose ibuprofen seem promising. Some CF centers administer intravenous steroids for acute exacerbations.
- Before deciding to intubate and mechanically ventilate a CF patient in respiratory failure, the ED physician must consider the baseline pulmonary function, disease course, and patient and parental expectations.

TREATMENT FOR PNEUMOTHORAX

- Pneumothoraces greater than 10% of the hemithorax on AP chest x-ray should be evacuated by tube thoracostomy (via a chest tube or pleural drain), at least initially.
- A tension pneumothorax should be aspirated first (through a large-bore IV catheter), followed by tube thoracostomy.

TREATMENT FOR HEMOPTYSIS

- Significant hemoptysis (>30–60 mL) is an indication for inpatient observation.
- Vitamin K should be administered if the prothrombin time is prolonged.
- Guidelines for replacement with packed red blood cells, fresh frozen plasma, etc are the same as for bleeding from other sources.
- For massive hemoptysis (acute bleeding >240 mL/d or >100 mL/d over several days), ligation or embolization of the bleeding vessel may be necessary.

TREATMENT FOR COR PULMONALE

- Cor pulmonale may require treatment with oxygen and diuretics, in addition to treatment for the underlying lung disease.

TREATMENT FOR MECONIUM ILEUS OR DISTAL INTESTINAL OBSTRUCTION SYNDROME

- Contrast enemas (eg, diatrizoate [Gastrografin]) can be used to treat meconium ileus and distal intestinal obstruction syndrome. In older patients, polyethylene

glycol–electrolyte solution (GoLYTELY), administered orally, may be an alternative.

- Laparotomy is indicated for bowel perforation or volvulus, or if the enema is unsuccessful in relieving the obstruction or reducing an intussusception.

TREATMENT FOR DEHYDRATION AND ELECTROLYTE LOSSES

- Infants with significant dehydration and electrolyte losses should receive a 20 mL/kg normal saline bolus initially. More than one bolus may be necessary.
- Once urine output is established (and after the bolus [es]), potassium chloride should be added to the fluids.
- Serum electrolytes should be monitored frequently to help guide fluid therapy.

OUTCOME

- Many more CF patients now survive to adulthood, with their median cumulative survival exceeding 35 years of age.
- Factors contributing to this improved survival include earlier diagnosis, better nutrition, aggressive treatment of pulmonary infections, and prompt recognition and treatment of the numerous life-limiting and life-threatening complications of CF.
- A number of patients with CF have undergone double lung (or heart-lung) transplants, although further study is required to determine whether these truly lengthen survival.

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QUESTIONS

1. A 2-year-old boy with CF is brought to the ED for recurrent episodes of abdominal pain, which began 8 hours ago. He appears lethargic between these episodes. There is no fever, vomiting, or diarrhea. On exam, you palpate a mass in the right upper quadrant of his abdomen. Shortly thereafter, he passes a grossly bloody stool. Which of the following studies will both confirm and treat the diagnosis?
 - A. Abdominal ultrasound
 - B. Abdominal x-rays, including a prone view
 - C. Diatrizoate (Gastrografin) enema
 - D. Rectal exam
 - E. Upper gastrointestinal series with oral contrast
2. A 7-year-old girl with CF is admitted to the hospital for intravenous antibiotic therapy for a pulmonary exacerbation. Her last respiratory culture was performed 4 days ago and is her first one positive for *Pseudomonas aeruginosa* (rough strain). Which of the following antibiotics is an appropriate choice?
 - A. Ticarcillin/clavulanate (Timentin) only
 - B. Tobramycin only
 - C. Vancomycin only
 - D. Ticarcillin/clavulanate and tobramycin
 - E. Ticarcillin/clavulanate and vancomycin

3. A 5-month-old girl is brought to an ED in Arizona in August because of poor oral intake and fewer wet diapers over several days. She seems to sweat profusely from the summer heat, but has not had a fever. There is no cough, vomiting, or diarrhea. Prenatally, her mother tested positive for “one of the CF genes;” however, the father, who is the mother’s cousin, refused testing to ascertain his CF status – and the baby has not been tested for CF yet either, despite not passing meconium until day 3 of life. On exam, the baby appears listless and mottled, with dry mucous membranes. Her heart rate is 190 and blood pressure 70/42. She weighs less than she did at her 4 month exam. Which of the following should you order first?
 - A. Basic metabolic panel (BMP) and await results before proceeding
 - B. Chest x-ray
 - C. DNA mutation analysis
 - D. Pilocarpine iontophoresis sweat test
 - E. 20 cc/kg normal saline bolus
4. A 16-year-old male with CF presents to the ED complaining of abdominal cramps and constipation for 1 week. His appetite is diminished, and he “feels full” soon after eating. On exam, you palpate a mass in the right lower quadrant, but are unable to elicit any tenderness, guarding, or rebound. Abdominal x-rays reveal a large amount of stool near the cecum as well as a few air-fluid levels proximally in the small bowel. You diagnose distal intestinal obstruction syndrome. What should you order next?
 - A. Gastrointestinal promotility agent
 - B. Intestinal lavage with polyethylene glycol-electrolyte solution (GoLYTELY)
 - C. Morphine
 - D. Music therapy, ideally a Beethoven Symphony
 - E. Surgical consultation
5. You are evaluating a 4-month-old girl for failure to thrive. According to her father, the baby has always had large, greasy, smelly stools. She has never had any respiratory complaints. The baby’s caloric intake is approximately 150 kcal/kg/day. Despite her voracious appetite, she weighs less than the fifth percentile for age, although her head circumference and height are at the 50th percentile. Otherwise, the physical exam is unremarkable. You consider CF in the differential diagnosis, as well as other etiologies of intestinal malabsorption. Which of the following studies should NOT be considered at this point?
 - A. Computed tomography (CT) scan of the chest
 - B. Pilocarpine iontophoresis sweat test
 - C. Prothrombin time (PT)
 - D. Stool analysis for fecal fat
 - E. Vitamin D levels

ANSWERS

1. C. Children with CF may develop intussusception. An intussusception is characterized by recurrent episodes of abdominal pain—and, eventually, grossly bloody stools. Children with intussusception may become lethargic, or have intervening periods of lethargy, possibly mistaken for sleep. An abdominal radiograph, with the child lying prone, may reveal a paucity of gas in the right lower quadrant, raising one’s suspicion for intussusception. Although a rectal exam should be performed on most patients presenting to the ED with abdominal pain, it usually does not lead to a definitive diagnosis. Abdominal ultrasound can be a good test for intussusception, but its accuracy is operator experience-dependent. Only the diatrizoate (Gastrografin) enema will both confirm and (generally) treat intussusception, and is the best of these choices when intussusception is the likely diagnosis. An upper gastrointestinal series with oral contrast helps diagnose malrotation and midgut volvulus.
2. D. Initially, recent sputum culture results can help with antibiotic selection. Either ticarcillin/clavulanate (Timentin) or tobramycin may seem a reasonable choice, since each alone is usually effective against rough strain *P aeruginosa*. However, because treatment of pulmonary exacerbations due to *P aeruginosa* with a single antibiotic has been associated with the development of rapid resistance to that antibiotic, combination therapy with two anti-pseudomonal antibiotics—such as ticarcillin/clavulanate (Timentin) and tobramycin—is recommended. The goal is to avoid the organism evolving and becoming resistant to multiple antibiotics over time. Also, ticarcillin and tobramycin demonstrate synergistic activity against *P aeruginosa*. Vancomycin may be appropriate when methicillin-resistant *S aureus* is present, but *S aureus* did not grow from this girl’s most recent respiratory culture.
3. E. For any child with such severe dehydration, rapid rehydration (with a 20 cc/kg normal saline bolus [or boluses]) is the most important intervention. Although a BMP will help estimate the degree of dehydration—and in this case will probably show the classic electrolyte abnormalities of CF—rehydration should be begun emergently while awaiting its results. Given the mother’s carrier status, the parental consanguinity, and the delayed passage of meconium, this child is certainly at risk of having CF, but a sweat test or DNA mutation analysis can wait until after stabilization. A CXR may not be necessary since the infant has no respiratory symptoms.

4. B. Distal intestinal obstruction syndrome (DIOS) is characterized by partial or complete blockage of the intestines from abnormally viscid, mucofeculent material (which is usually retained in the terminal ileum or ascending colon). DIOS occurs in 10–20% of CF patients. Causes include pancreatic enzyme insufficiency and dehydration. Besides abdominal cramps, constipation, and anorexia, there may be vomiting or diarrhea. Intestinal perforation can occur, and mandate emergency surgery; however, this patient demonstrated no signs of perforation. Intestinal lavage with polyethylene glycol–electrolyte solution (GoLYTELY) is a good choice for him initially. Gastrointestinal promotility agents are not recommended for DIOS. Agents that slow intestinal motility, like narcotics, are not recommended either. Although listening to a Beethoven Symphony might be an expunging—as well as an undoubtedly

sublime—experience, Beethoven therapy has not been studied for relief of DIOS yet (to our knowledge).

5. A. Failure to thrive—associated with a history of large, greasy, smelly stools—is a common presentation in CF. (It is worth noting, however, that some children with CF gain weight adequately, because of increased caloric intake.) CF can be diagnosed with a pilocarpine iontophoresis sweat test. Malabsorption in CF is due to pancreatic exocrine insufficiency as well as abnormal biliary secretions and intestinal mucous. When fat malabsorption (from CF or another cause) is suspected, the patient’s feces can be analyzed for excess fat. Fat malabsorption may lead to vitamin deficiencies—particularly of fat-soluble vitamins, such as vitamins A, D, E, and K—and vitamin K deficiency can result in a prolonged prothrombin time. A CT scan of the chest will not be helpful in this infant who has not had any respiratory symptoms.

Section 6

CARDIOVASCULAR EMERGENCIES

47 CONGENITAL HEART DISEASE

Timothy Horeczko

Kelly D. Young

EPIDEMIOLOGY

- Congenital heart lesions occur in approximately 8 in 1000 live births in the United States, with about 10% of cases attributed to genetic causes.
- Many syndromes (eg, the trisomies, connective tissue disorders) and teratogens (eg, congenital rubella infection) are associated with an increased risk of specific congenital heart lesions.
- Most patients present during infancy; however, presentation and chief complaints differ by age.

PHYSIOLOGY

FETAL CIRCULATION

- Oxygenated blood from the placenta enters the fetus through the single umbilical vein; half bypasses the liver via the *ductus venosus*, flowing directly into the inferior vena cava (IVC) (Fig. 47-1).
- Blood is then directed from the IVC across the *foramen ovale* into the left atrium, bypassing the right heart and pulmonary circulation.
- This highly oxygenated blood in the left atrium mixes with pulmonary venous return, enters the left ventricle and the ascending aorta, and perfuses the cerebral circulation.
- Deoxygenated blood from the cerebral circulation drains into the superior vena cava, entering the right atrium, right ventricle, and pulmonary artery.
- Since pulmonary vascular resistance is high in the fetus, most of this blood bypasses the pulmonary

circulation by way of the *ductus arteriosus* and enters the descending aorta.

NEONATAL CIRCULATION

- At birth the newborn's pulmonary arterioles dilate, pulmonary vascular resistance falls, and pulmonary blood flow increases.
- The smooth muscle of the ductus arteriosus constricts in response to increased blood PO_2 ; it is functionally closed by 15 hours of life.
- The foramen ovale closes by 3 months of age.
- The neonatal myocardium is unable to increase its contractility in response to demand. When an increased cardiac output is needed, the neonate responds by increasing heart rate.
- The young infant has rate-dependent cardiac output, increased oxygen consumption, and lower systolic reserve, predisposing children with congenital heart disease to congestive heart failure (CHF).
- Neonates and young infants who are slow to transition from their *in utero* physiology may still shunt blood via the foramen ovale or a patent ductus arteriosus (PDA). This may present as high pulmonary vascular resistance, which is responsive to oxygen (ie, oxygen decreases the resistance).

EVALUATION

HISTORY

- General health, including growth, development, and susceptibility to respiratory illnesses, should be assessed.
- Pregnancy, birth, and family history may provide valuable clues to specific genetic or teratogenic etiologies.

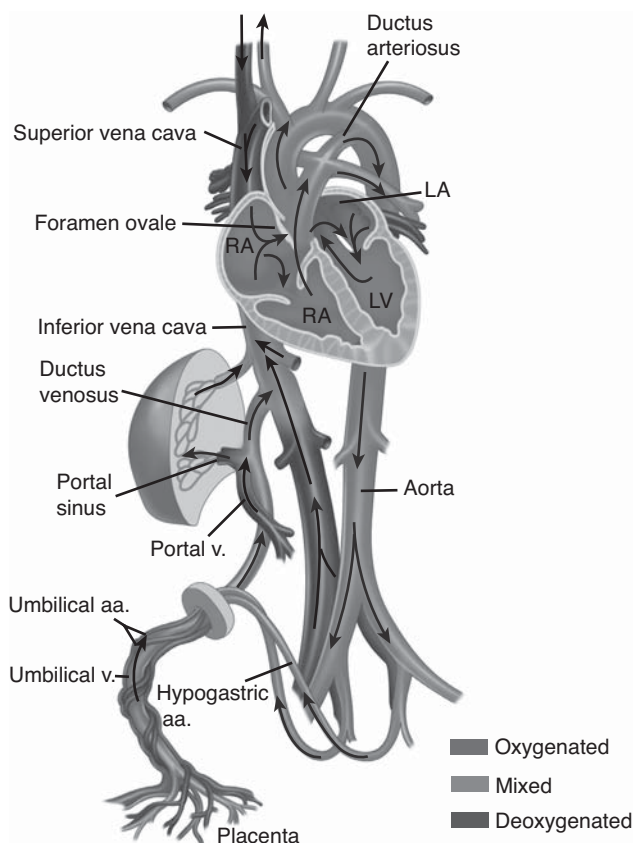


FIG. 47-1. Fetal circulation.

- Symptoms of CHF should be sought: poor feeding, longer feeding times than the average infant, poor growth or failure to thrive, sweating with feeding, irritability or lethargy, weak cry, increased respiratory effort, dyspnea, tachypnea, and coughing.

Ask about cyanosis or cyanotic episodes, which may be more noticeable during crying or exercise (Table 47-1).

PHYSICAL EXAMINATION

- Check vital signs, including four-extremity blood pressures and pulse quality in the upper and lower extremities, in the sick infant.
- Color and general appearance may be significant clues for classifying the lesion into one of three categories:
 - Pink: congestive heart failure, L → R shunt
 - Blue: cyanotic heart disease, R → L shunt
 - Gray: outflow obstruction, hypoperfusion, and shock.
- Peripheral cyanosis, acrocyanosis, and mottling may be seen in normal newborns; mucous membranes are the best locations to assess for central cyanosis.
- Auscultate for murmurs, S1 and S2, and extra sounds.
- Generally speaking, diastolic, late systolic, or pansystolic murmurs are never normal.

- Examine the abdomen for hepatomegaly; a normal infant's liver is palpable 1 to 3 cm below the right subcostal margin.
- Examine the child for dysmorphic features suggestive of a genetic or teratogenic syndrome (Table 47-2).

HYPEROXIA TEST

- The single most sensitive and specific test of a neonate with suspected congenital heart disease (in the absence of readily available echocardiography) is the *hyperoxia test*.
- An arterial blood gas is sampled with the patient on room air (if tolerated), and repeated after a few minutes of high-flow oxygen.
- When a child breathes high-flow oxygen ("100%" O₂), an arterial Po₂ of greater than 250 torr virtually excludes hypoxia due to congenital heart disease ("passed" hyperoxia test).
- An arterial Po₂ of less than 100 torr in a patient without obvious lung disease is indicative of a right-to-left shunt and extremely predictive of cyanotic congenital heart disease ("failed" hyperoxia test).
- A value of 100 to 250 torr may indicate structural heart disease with complete intracardiac mixing.
- Ideally, blood is sampled from both preductal (right upper extremity) and postductal sites (any other extremity) and carefully labeled as to site and FiO₂. When done in both sites, valuable information about the possible lesion may be obtained (eg, differential cyanosis due to aortic arch obstruction).
- The hyperoxia test should be conducted on all neonates with suspected congenital heart disease, not just those who appear cyanotic.
- Pulse oximetry is not an appropriate substitute for an arterial blood gas; it is not sensitive enough to determine "pass or fail" of the test, since a child breathing high-flow O₂ and registering 100% on pulse oximetry may actually have an arterial Po₂ of anywhere from 80 to 680 Torr.
- A neonate who fails the hyperoxia test should be presumed to have critical congenital heart disease and should receive prostaglandin E₁ immediately until a definitive anatomic diagnosis is made.

OTHER ANCILLARY TESTS

- Obtain a chest radiograph (CXR) to evaluate for cardiomegaly, chamber enlargement, and pulmonary vascularity.
- Evaluate an electrocardiogram (ECG) for conduction and rhythm disturbances, chamber forces, and rare ischemic changes.

TABLE 47-1 Common Presentations by Age

Common cardiovascular presentations by age			
0–2 wk	2–6 wk	6 wk–6 mo	Child–adult
Cyanosis Tetralogy of fallot → Eisenmenger’s syndrome Truncus arteriosus → Tricuspid atresia → Transposition of the great vessels Total anomalous pulm venous return			
Ductal-dependent circulatory failure Hypoplastic left heart Aortic coarctation Aortic stenosis	Murmur VSD → ASD Patent ductus arteriosus → Aortic stenosis Pulmonic stenosis		
	Congestive heart failure VSD Patent ductus arteriosus Atrioventricular canal defects ASD	Syncope/exercise intolerance Aortic stenosis Pulmonic stenosis Eisenmenger’s syndrome HCM	
			Arrhythmia ASD
			Hypertension Aortic coarctation

ASD – Atrial septal defect
 VSD – Ventricular septal defect
 HCM – Hypertrophic cardiomyopathy

- Neonates and young children have natural right-axis deviation (RAD), which may obscure the typical findings of right-sided disease; the addition of leads V3R or V4R increases the yield in detecting right atrial or ventricular hypertrophy.
- Neonatal right-sided forces such as RAD and right ventricular hypertrophy will transition gradually to adult form by age 3 to 4 years.
- Look for bradycardia (rate <60–80) or supraventricular tachycardia (rate >220–240).
- Analyze intervals for evidence of drug effects, electrolyte disturbances, and for long Q–T syndrome.
- T-wave changes, especially T-wave inversions are common in children and are rarely ischemic (juvenile T waves).

- Chronically cyanotic children usually compensate with polycythemia.
- A cyanotic child’s oxygen-carrying capacity will be further compromised by anemia, and a “normal” hematocrit by usual standards may be inadequate for the cyanotic child.

CLASSIFICATION

Lesions are usually classified as **cyanotic** or **acyanotic** and further subclassified according to whether or not the lesion is ductal-dependent (dependent on a patent ductus arteriosus to deliver partially oxygenated blood to the systemic circulation, or for pulmonary flow).

Cyanotic lesions: “The Six Terrible (Turquoise) Ts”

- Truncus Arteriosus
- Transposition of the Great Arteries^b
- Total Anomalous Pulmonary Venous Return
- Tetralogy of Fallot^b
- Tricuspid Abnormalities^b (Tricuspid Atresia, Ebstein’s Anomaly)
- “Tons” of Others (“*Tiny Heart*”—Hypoplastic Left Heart Syndrome,^a “*Terminated Aorta*”—Interrupted Aortic Arch^a)

Acyanotic lesions: “PiCk A Very Powerful Approach to Acyanosis”

- Pulmonic Stenosis^b
- Coarctation of the Aorta (may be^a)
- Atrioseptal Defect,
- Ventriculoseptal Defect
- Patent Ductus Arteriosus
- Aortic Stenosis (may be^a)
- Atrioventriculoseptal defects

^aLesions ductal-dependent for systemic flow.

^bLesions ductal-dependent for pulmonary flow.

TABLE 47-2 Selected Syndromes Associated with Congenital Heart Disease

SYNDROME	INCIDENCE	COMMON FEATURES	CONGENITAL HEART LESIONS
Down syndrome (trisomy 21)	1 in 1000	Decreased tone, epicanthal folds, hypothyroidism, esophageal, and duodenal atresia	Atrial septal defect, ventricular septal defect
Klinefelter syndrome (47 XXY)	Boys only 1 in 1000	Gynecomastia, hypogonadism	Mitral valve prolapse, left ventricular dysfunction
Noonan syndrome (AD or new mutation)	1:1000 to 1:2500	Webbed neck, wide-set eyes, epicanthal folds, short stature, lymphedema, blood dyscrasias	Pulmonic valve abnormalities, pulmonic stenosis, hypertrophic cardiomyopathy
Turner syndrome (XO syndrome)	Girls only 1:2000	Web neck, lymphedema, lack of secondary sexual characteristics, musculoskeletal and renal defects	Coarctation of the aorta, bicuspid aortic valve, secondary hypertension
DiGeorge syndrome (deletions, mutations)	1:3000	Hypocalcemia, immunodeficiency, hypoparathyroidism, thymic aplasia	Conotruncal anomalies, interrupted aortic arch, tetralogy of Fallot, truncus arteriosus, ventricular septal defect
Edwards syndrome (trisomy 18)	Girls >> Boys 1 in 3000	Coloboma, pectus carinatum, clenched hands/crossed legs, renal disease	Atrial septal defect, ventricular septal defect, patent ductus arteriosus
Marfan syndrome (fibrillin mutation)	1:5000 30% FamHx	Arched palate, scoliosis, ectopia lentis, thoracic cage defects; tall; thin, long fingers, arms, legs	Aortic aneurysm, mitral valve prolapse, risk of endocarditis
Patau syndrome (trisomy 13)	1 in 10 000	Cleft palate, close-set eyes, polydactyly, omphalocele, seizures, apnea	Dextrocardia, atrial septal defect, ventricular septal defect, patent ductus arteriosus
Williams syndrome (elastin deletions)	1:20 000	Hearing/visual problems, hypercalcemia, cystic kidneys, diverticula, sudden death	Supravalvular aortic stenosis, pulmonic stenosis, ventricular hypertrophy
Holt–Oram syndrome (heart–hand syndrome)	1:100 000	Upper limb bone aplasia or hypoplasia, heart block	Atrial septal defect, ventricular septal defect
Fetal alcohol syndrome	0.2 in 1000 to 1.5 in 1000	Microcephaly, smooth philtrum, short stature, developmental delay, risk of neonatal withdrawal	Valvular defects (various), cardiomyopathies
Lithium exposure	1:10 during treatment	Hypertonicity, hypothyroidism	Ebstein’s anomaly, atrial septal defect
Rubella, congenital	Mostly during outbreaks	Deafness, ophthalmic defects, “Blueberry muffin baby,” sepsis	Atrial septal defect, ventricular septal defect, pulmonic stenosis, patent ductus arteriosus

COMMON PRESENTATIONS

UNDIFFERENTIATED SICK INFANT

- There is significant similarity in presentation between congenital heart disease and other life-threatening disorders of infancy; therefore, a systematic approach is needed for the wide differential diagnosis.
- The mnemonic “THE MISFITS” outlines the broad and varied causes of acute illness in very young children.

T—Trauma (accidental and nonaccidental)

H—Heart disease and hypovolemia

E—Endocrine (congenital adrenal hyperplasia and thyrotoxicosis)

M—Metabolic (electrolyte abnormalities)

I—Inborn errors of metabolism

S—Sepsis (meningitis, pneumonia, and pyelonephritis)

F—Formula problems (over- or under-dilution)

I—Intestinal disasters (intussusception, necrotizing enterocolitis, and volvulus)

T—Toxins

S—Seizures

- As with any sick infant, assessment and intervention in airway, breathing, and circulation is critical, with close attention to vital sign abnormalities such as tachycardia and tachypnea.
- Always check blood glucose and keep the infant warm during the course of evaluation and treatment.
- Neonates who present in shock in the first few weeks of life are presumed to have ductal-dependent systemic flow until proven otherwise. Resuscitation depends on opening the ductus arteriosus with PGE₁. It is appropriate to start PGE₁ before a definitive anatomic diagnosis is made (Fig. 47-2).

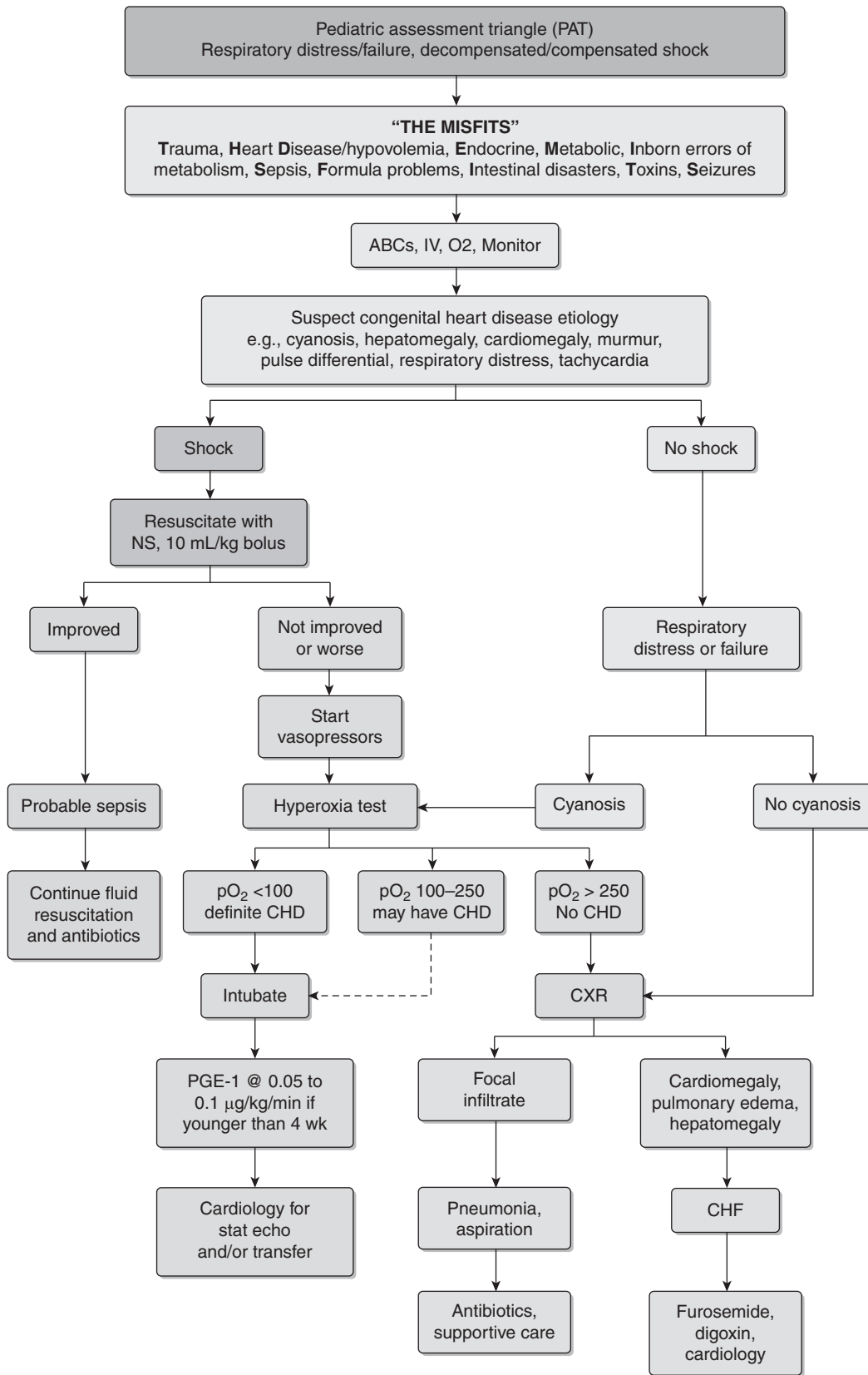


FIG. 47-2. Approach to the infant with suspected cardiogenic shock.

DUCTAL-DEPENDENT LESIONS AND CARDIOGENIC SHOCK

- Lesions which are completely dependent on a patent ductus arteriosus for systemic blood flow present with acute onset circulatory failure and shock when the ductus closes, typically within the first week of life.
- Such lesions include hypoplastic left heart syndrome (HLHS), severe aortic coarctation, interrupted aortic arch, and lesions such as pulmonary atresia or transposition of the great arteries (TGA) without a mixing lesion such as a VSD.
- Ductal-dependent cardiac failure should be suspected in any infant in the first week of life with sudden-onset circulatory collapse leading to hypoperfusion, hypotension, severe acidosis, and cyanosis.
- Infants may present in the second week of life, but rarely present beyond 4 weeks of age.
- The mainstay of therapy for suspected ductal-dependent cardiogenic shock is PGE₁ infusion to maintain the patency of the ductus arteriosus. Therapy should be begun as soon as the possibility is recognized, even before echocardiographic confirmation of the specific lesion.
- PGE₁ is infused initially at 0.05 to 0.1 µg/kg/min and advanced to 0.2 µg/kg/min if necessary; adverse effects include hyperthermia, apnea (be prepared to provide advanced airway management), hypotension (may require inotropic medications for circulatory support), rash, tremors, focal seizures, and bradycardia.
- Consult a pediatric cardiologist immediately and admit the patient to the pediatric or neonatal intensive care unit.

CONGESTIVE HEART FAILURE

- Causes other than congenital heart lesions include myocardial dysfunction (eg, cardiomyopathies) and dysrhythmias.
- Symptoms are gradual in onset and may be subtle: poor feeding (increased time to feed) or sweating while feeding; poor growth; irritability, lethargy, or a weak cry; increased respiratory effort, dyspnea, tachypnea, chronic cough, or wheezing; and increased frequency of respiratory infections.
- Physical examination may reveal tachypnea, retractions, nasal flaring, grunting, wheezing, or rales (although less commonly than in adults).
- Look for tachycardia, gallop rhythm, hyperactive precordium, murmur, poor peripheral pulses with delayed capillary refill, or hepatomegaly (a cardinal sign of CHF in infants).

- Jugular venous distension and peripheral edema, often seen in adults with CHF, are rarely seen in young children. If present, edema is best appreciated in the eyelids, sacrum, and legs. More commonly, children will present with hepatic congestion or hepatomegaly, as the relatively pliable liver becomes congested with venous blood.
- CXR shows cardiomegaly and increased pulmonary vascularity.
- Treatment includes oxygen to keep saturations about 95%. Over-oxygenating the patient may lead to pulmonary vascular dilation and worsened failure. Keep the infant in a semireclining position (such as when in an infant car seat) if possible.
- Fluid and sodium restriction is necessary, and furosemide should be given at 1 mg/kg intravenously. Nitrates are not first-line emergent therapy in children. In consultation with a pediatric cardiologist, the patient should be started on digoxin.
- If a patient's respiratory status allows, a trial of continuous positive airway pressure (CPAP) may be applied nasally in an attempt to avoid the need for intubation.
- If the patient is in shock, fluids must be used cautiously (boluses of only 5 mL/kg, if at all), and inotropic support with dobutamine may be more appropriate.

B-TYPE NATRIURETIC PEPTIDE (BNP) ASSAY

- BNP levels spike at birth with the physiologic transition from fetal to neonatal circulation, reaching a plateau at day 3 to 4, and subsequently fall to a constant level during infancy.
- No established BNP "cutoff" values in children exist to support definitively or to rule out CHF in children. Currently, BNP values can only be interpreted as either consistent with the normal mean value or not.
- Clinical presentation will often provide an adequate basis to make the diagnosis of CHF. BNP level may help to exclude the diagnosis if very low and support the diagnosis if markedly high.

HYPOXEMIC "TET" SPELLS

- Sudden-onset spells of increased cyanosis may occur in young children with tetralogy of Fallot; the classic teaching is a sudden "clamping down" of the pulmonary infundibular right ventricular outflow tract, leading to increased right-to-left shunting.
- Increased right-to-left shunting leads to hypoxemia, cyanosis, and acidosis.
- The child attempts to compensate by hyperventilating (to improve oxygenation) and squatting or

- drawing the knees to the chest (to increase systemic vascular resistance (SVR) and left heart pressure, which decreases right-to-left shunting of deoxygenated blood, and therefore improves oxygenation).
- Symptoms include restlessness, irritability, or lethargy. Signs include a sudden increase in cyanosis and occasional syncope. Hyperpnea is a cardinal sign of hypoxemic spells. Spells must be differentiated from seizures, CHF, and respiratory disease.
 - Keep the child as calm as possible, in a position of comfort with a parent present. Avoid unnecessary painful or stressful procedures. Provide oxygen, although it will have little effect in hypoxemia due to shunting. Place the child in a knee-chest position (to simulate squatting) to increase SVR.
 - Morphine, 0.05 to 0.2 mg/kg intravenous or intramuscular is the traditional first-line medical therapy. Mechanism of action is unknown—may relax pulmonary infundibulum.
 - Administer a fluid bolus of 10 mL/kg normal saline intravenously to counteract the vasodilatory effects of morphine and to ensure adequate preload, on which pulmonary flow is dependent.
 - If the above therapies are unsuccessful, propranolol 0.1 to 0.25 mg/kg by a slow intravenous route may be given and repeated once after 15 minutes.
 - Phenylephrine 5 to 20 μ g/kg/dose (alpha agonist that increases SVR) intravenously may be used and repeated every 10 to 15 minutes as necessary. Propranolol and phenylephrine are customarily given in consultation with a cardiologist.
 - If these interventions fail, general anesthesia may be necessary.

ISOLATED CORONARY ARTERY ANOMALIES

- Congenital lesions of the coronary arteries occur uncommonly, in less than 1% of the population.
- Abnormalities may affect the number (duplication of artery), site of origin (eg, from pulmonary trunk), anatomic course (eg, between the aorta and pulmonary trunk), termination (eg, fistula formation), or structure (stenosis and atresia) of the coronary arteries, and virtually any coronary artery may be affected.
- Neonates with an isolated coronary artery anomaly (ICAA) may demonstrate anginal symptoms with irritability, episodic diaphoresis and/or color change when symptomatic. Older infants may have poor feeding, dyspnea, failure to thrive, or unexplained episodes of pallor. Diaphoresis during feeding is an ominous sign, reflecting both a decreased “exercise tolerance” and a splanchnic steal syndrome.
- Older children and young adults typically present with more familiar ischemic symptoms such as angina and dyspnea.
- Anomalous origin of the left coronary artery arising from the pulmonary artery (ALCAPA) is a rare but serious lesion that is also called Bland–White–Garland syndrome. Most children present within the first few months of life with nonspecific complaints such as irritability, and they are often misdiagnosed with colic. Signs and symptoms of CHF may ensue though the physical examination may be completely normal or show signs of CHF. ECG may show an anterolateral infarct pattern with deep and wide Q waves laterally and absent Q waves inferiorly. CXR may be consistent with CHF. Echocardiography with Doppler flow is often diagnostic, especially if retrograde flow from the left coronary artery to the pulmonary trunk is visualized.
- Surgical correction is necessary to restore blood flow.

PRESENTATIONS IN OLDER CHILDREN AND ADULTS

- Previously undiagnosed lesions may be discovered by recognition of a murmur during routine physical examination. Common lesions include ASD, small VSD, PDA, PS, AS, and aortic coarctation. Adult patients with unrepaired ASD may present with atrial arrhythmias, often in the fourth decade of life.
- Patients with PDA and AS may present with dyspnea on exertion and fatigue.
- Patients with critical AS may present with syncope.
- Patients with critical PS may present with cyanosis on exertion, right-sided heart failure, or syncope.
- Patients with aortic coarctation are often diagnosed during evaluation for hypertension.

HYPERTROPHIC CARDIOMYOPATHY

- Hypertrophic cardiomyopathy, found in 1:500 of the general population, is a genetic disorder of sarcomeric proteins that results in varying degrees of left ventricular hypertrophy. It is a primary myopathy, not secondary to hypertension or aortic stenosis.
- Hypertrophic cardiomyopathy (HCM) includes both hypertrophic obstructive cardiomyopathy (HOCM) and idiopathic hypertrophic subaortic stenosis (IHSS), which are distinguished from each other by degree of outflow obstruction and/or asymmetric hypertrophy. The shared feature is diastolic dysfunction, with resultant impaired cardiac output on exertion.

- 50% of those with HCM have a family history. Young children may be asymptomatic, and the first clinical manifestation may be sudden death. Children and young adults may complain of dyspnea, angina, fatigue, or syncope, especially after strenuous exercise.
- Patients often have a harsh systolic murmur increasing after the first heart sound (“diamond-shaped”) best heard at the lower left sternal border or apex; this may be accompanied by an S4. The murmur may be more prominent with tachycardia or Valsalva maneuver; both of these reduce ventricular volume, mimicking the conditions of exercise. Conversely, the murmur may decrease with squatting, hand grip, or leg raise, which augment venous return and increase ventricular volume. Patients may also have a prominent apical impulse and rapidly rising carotid impulse on physical examination.
- ECG may show LVH and/or wide Q waves. Chest radiograph may be normal or reflect a mild-to-moderate increase in the size of the cardiac silhouette. Echocardiography is the mainstay of diagnosis.
- Treatment options span from medications such as β -blockers or calcium-channel blockers, to septal myectomy, to placement of an automated implantable cardioverter-defibrillator.
- Any child with high suspicion for HCM and a history consistent with arrhythmia or syncope warrants inpatient investigation, including urgent echocardiography and possibly electrophysiology studies. If the clinician has high suspicion of HCM in a currently asymptomatic child without high-risk historical features such as arrhythmia or syncope, nonemergent referral to a cardiologist is recommended. The child should avoid dehydration and refrain from strenuous activity until a definitive diagnosis is obtained.

EISENMENGER SYNDROME

- Patients with a large left-to-right shunt left unrepaired gradually develop pulmonary vascular disease due to the increased volume overload. When pulmonary hypertension becomes severe enough, the direction of shunting will reverse to right to left, and cyanosis ensues. Typically this occurs in adolescence to early adulthood. Patients may complain of decreased exercise tolerance, dyspnea on exertion, hemoptysis, palpitations due to atrial arrhythmias, and symptoms of hyperviscosity due to chronic polycythemia (vision disturbances, fatigue, headache, dizziness, paresthesias, and even cerebrovascular accident).
- Brain abscesses can occur with right-to-left passage of an infected embolus.
- On examination, the murmur may no longer be present with the disappearance of left-to-right shunting, and S2 is loud due to the pulmonary hypertension. CXR shows decreased vasculature (pruned pattern), and ECG shows RVH.
- Although no definitive therapy exists, other than heart–lung transplant, patients should avoid dehydration, heavy exertion, altitude, vasodilators, and pregnancy, which is associated with a high-mortality rate. Symptoms of hyperviscosity may be treated with phlebotomy and isovolemic replacement. Patients should be medically managed by a cardiologist to optimize cardiac function as long as possible.

CARE OF THE POSTCARDIAC SURGERY CONGENITAL HEART PATIENT

CATEGORIES OF REPAIR

- Patients with true complete repairs generally lead a normal life after repair: ASD, VSD, PDA, aortic coarctation, and TGA (switch procedure).
- Repairs of tetralogy of Fallot, AV canal, and valve obstructions typically result in anatomic repairs with residual lesions, and late complications may occur.
- Repairs requiring prosthetic materials such as pulmonary atresia, truncus arteriosus, and prosthetic valve replacements will require replacement of the prosthetic material due to growth of the child or degeneration of the material.
- Physiologic repairs improve the patient’s blood flow physiology but do not result in normal cardiac anatomy. These palliative repairs, which include the Fontan operation for lesions resulting in a functionally single ventricle, and the Mustard operation and Arterial Switch of Jatene for TGA, invariably produce late complications.
- The key concept for care of these children is a careful examination and consideration of complications such as arrhythmias, infective endocarditis (IE), and thromboembolism.

POSTOPERATIVE COMPLICATIONS

- Arrhythmias are the most common problem and may present with symptoms of palpitations, decreased appetite, emesis, and decreased exercise tolerance.
- Arrhythmias may be a result of the surgical repair, the underlying lesion (eg, Ebstein’s anomaly), or medical therapy (eg, digoxin toxicity).
- Supraventricular tachycardia (SVT) is the most common clinically significant arrhythmia; atrial fibrillation is another known complication.

- Endocarditis is a significant complication seen in congenital heart disease patients before and after surgical repair. The rate in patients with uncorrected congenital heart disease is 0.1% to 0.2% per patient-year; this decreases to 0.02% after correction for many lesions. Unrepaired complex congenital heart disease carries the highest risk, at 1.5% per patient-year.
- Antibiotic prophylaxis should be given to patients at high risk prior to invasive procedures likely to produce bacteremia, such as dental procedures (Tables 47-3, 47-4, 47-5).
- Other complications common in patients with congenital heart disease include poor growth, electrolyte disturbances due to medications, cerebral embolus in patients with right-to-left shunts, and increased susceptibility to respiratory illnesses.
- Cardiac patients may have a particularly complicated course with respiratory syncytial virus infections.

CARDIAC TRANSPLANT PATIENTS

- Patients who are status post-heart transplant are at risk for acute or chronic rejection, post-transplant lymphoproliferative disorder (PTLD), and infectious complications associated with their immunosuppressive or immunomodulating medication regimen.
- In contrast to other types of transplantation, acute and chronic cardiac rejection is not defined by the timing after the operation, but rather the clinical presentation. Acute rejection is defined as a distinct episode that prompts intensification of immunosuppressive therapy, either based on cardiac dysfunction or based on histologic diagnosis. Symptoms of acute rejection vary widely, and may include fever, myalgias, vomiting, and shock.

TABLE 47-3 Cardiac Conditions Associated with the Highest Risk of Adverse Outcome from Endocarditis

- Prosthetic cardiac valve or prosthetic material used for cardiac valve repair
- Previous infective endocarditis
- Congenital heart disease^a:
 - Unrepaired cyanotic CHD, including palliative shunts and conduits
 - Completely repaired congenital heart defect with prosthetic material or device, whether placed by surgery or by catheter intervention, during the first 6 mo after the procedure^b
 - Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device (which inhibit endothelialization)
- Cardiac transplantation recipients who develop cardiac valvulopathy

^aExcept for the conditions listed above, antibiotic prophylaxis is no longer recommended for any other form of CHD.

^bProphylaxis is reasonable because endothelialization of prosthetic material occurs within 6 mo after the procedure.

TABLE 47-4 Procedures for which Prophylaxis is Recommended

All dental procedures that involve manipulation of gingival tissue or the periapical region of teeth or perforation of the oral mucosa^a
Consider prophylaxis for incisional procedures on the respiratory tract, infected skin, or musculoskeletal tissue only for high-risk patients.

^aThe following procedures do not need prophylaxis: routine anesthetic injections through noninfected tissue, taking dental radiographs, placement of removable prosthodontic or orthodontic appliances, adjustment of orthodontic appliances, placement of orthodontic brackets, shedding of deciduous teeth, and bleeding from trauma to the lips or oral mucosa.

- Chronic rejection is an ongoing process that mostly involves development of atherosclerotic disease. Ischemic symptoms such as decreased exercise tolerance, fatigue, and chest pain may be present. Syncope or sudden death may result from an arrhythmia. As in acute rejection, the diagnosis is clinical.
- Post-transplant lymphoproliferative disorder (PTLD) is associated with the Epstein–Barr virus and occurs

TABLE 47-5 Regimens for Prophylaxis for Infective Endocarditis

SITUATION	AGENT	SINGLE DOSE 30–60 Min BEFORE PROCEDURE	
		ADULTS	CHILDREN
Oral	Amoxicillin	2 g po	50 mg/kg po
Unable to take oral medications	Cefazolin or Ceftriaxone	1 g IM or IV	50 mg/kg IM or IV
Allergic to penicillin or ampicillin—Oral	Cephalexin	2 g po	50 mg/kg po
	OR Clindamycin	600 mg po	20 mg/kg po
Allergic to penicillin or ampicillin—Unable to take oral medications	Azithromycin or Clarithromycin	500 mg po	15 mg/kg po
	Cefazolin or Ceftriaxone	1 g IM or IV	50 mg/kg IM or IV
	OR Clindamycin	600 mg IM or IV	20 mg/kg IM or IV

most often within a year of transplant, but may develop many years later. PTLD is a B-cell lymphoma resulting in masses throughout the body, most notably in the abdomen, chest, head and neck. Symptoms include a mononucleosis-like syndrome with fever, lymphadenopathy, and abdominal pain. The diagnosis is based on clinical suspicion from history and physical examination, with radiologic findings consistent with scattered lymphoid masses.

PREVENTION OF INFECTIVE ENDOCARDITIS

- See Tables 47-3, 47-4, 47-5.

OUTPATIENT REFERRAL

- Children with complications associated with congenital heart disease and those with a new diagnosis in the emergency department are invariably admitted for stabilization and/or investigation of the lesion.
- A currently asymptomatic child may have high-risk historical features, including symptoms associated with exertion, recurrent episodic symptoms, symptoms while recumbent, associated chest pain or palpitations, and family history of sudden death or childhood cardiac disease; children with one or more of these features should also be admitted in consultation with a pediatric cardiologist.
- An otherwise well child with no high-risk historical features and a normal ECG may present to the ED with signs and symptoms necessitating an expedited outpatient workup. Such symptoms include: otherwise well children with diaphoresis with eating, unexplained significant murmur, or exercise intolerance.
- Parent education is crucial in understanding both the importance of primary care and (if needed) subspecialty follow-up, as well as return precautions to the ED.

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QUESTIONS

1. A neonate is brought to the emergency department for “turning blue.” The parents’ state the infant was crying at the time. The infant was born full term without any complications. Regarding the cardiovascular system of the neonate, which of the following is TRUE?
 - A. At birth, the newborn’s pulmonary arterioles constrict and pulmonary vascular resistance increases.
 - B. The smooth muscle of the ductus arteriosus dilates in response to increased blood PO_2 ; it remains functionally open until approximately 15 days of life.
 - C. The foramen ovale closes by 3 years of age.
 - D. The young infant has a rate-dependent cardiac output, increased oxygen consumption, and a lower systolic reserve.
 - E. Young infants with congenital heart defects uniformly have a high pulmonary vascular resistance, which is not responsive to oxygen.
2. A full-term baby is born at home and brought in to the PED for evaluation. He is limp, has rapid breathing, and a faint bluish tinge to his lips. Regarding his resuscitation and possible etiologies of his presentation, which of the following is TRUE?
 - A. Lesions which are completely dependent on a patent ductus arteriosus for systemic or pulmonary blood flow present with acute onset circulatory failure and shock when the ductus closes, typically within the first week of life.
 - B. Ductal-dependent cardiac failure should be suspected only in infants of mothers with high-risk or complicated pregnancies.
 - C. PGE_1 is given to maintain the patency of the atrial septal defect.

- D. Infants started on a PGE₁ infusion should be monitored specifically for hyperpnea, tachycardia, and hypertension.
- E. Ventilatory and cardiovascular support is rarely needed when PGE₁ is given.
3. A two-month-old girl is brought in by parents for “not eating anything all day.” The parents only know that she was born with “a heart problem” and is taking medications for it. In evaluating this child for congestive heart failure (CHF), which of the following is TRUE?
- A. Causes other than congenital heart lesions include myocardial dysfunction such as cardiomyopathy and dysrhythmias
- B. Symptoms are always abrupt in onset and invariably involve cyanosis
- C. Jugular venous distension and peripheral edema are reliable indicators of CHF in children
- D. Fluid restriction is not necessary in children with CHF
- E. Continuous positive airway pressure (CPAP) is unsafe in children
4. A 2-year-old boy with a history of tetralogy of Fallot (TOF), uncorrected, presents to the ED after an episode of cyanosis and syncope. On arrival, he is crying and has slightly blue mucous membranes. Concerning his stabilization and treatment, which of the following is TRUE?
- A. A “tet spell” results in an alkalotic state
- B. The child with tetralogy of Fallot attempts to compensate by hyperventilating and squatting or drawing the knees to the chest
- C. As long as the child is crying, he is oxygenating well
- D. Morphine should never be given to children with potential respiratory issues such as seen in this case
- E. These children should be fluid-restricted, as the problem is congestive heart failure
5. A 13-year-old boy is sent to the ED from his primary physician’s office for work-up of a murmur found on routine physical examination. Regarding the presentation of congenital heart disease in older children, adolescents, and adults, which of the following is TRUE?
- A. Unapparent lesions are not detectable on routine physical examination
- B. Patients with patent ductus arteriosus (PDA) and aortic stenosis (AS) most commonly present with cardiac arrest
- C. Patients with critical aortic stenosis (AS) may present with plethoric facies
- D. Patients with critical pulmonic stenosis (PS) will present with pedal edema
- E. Patients with aortic coarctation are often diagnosed during evaluation for hypertension
6. A 9-month-old boy with a history of poor weight gain and possible developmental delay presents with vomiting, volume depletion, and an episode of color change. While keeping a wide differential, you mention to the parents that he should be admitted to the hospital and evaluated for possible congenital heart disease (among other things). They are very concerned and have many questions. Regarding congenital heart disease, which of the following statements is TRUE?
- A. Generally speaking, only patients with a corrected PDA can expect to lead a normal life after repair
- B. Repairs of tetralogy of Fallot, AV canal, and valve obstructions typically result in complete repair without residual defects; they are then re-categorized as “low-risk”
- C. Repairs requiring prosthetic materials are designed to unfold and “stretch” with the growth of the child
- D. Physiologic repairs improve the patient’s blood flow physiology but do not result in normal cardiac anatomy
- E. These children have the same risk of arrhythmia as the general population
7. A 6-month-old girl with a history of truncus arteriosus repair is brought in by her mother for a history of tactile fever, fussiness, and an episode of emesis. Which of the following statements involving her emergency department assessment is TRUE?
- A. Arrhythmias are common and may present with symptoms of palpitations, decreased appetite, emesis, and decreased exercise tolerance
- B. Arrhythmias are not seen out of the immediate post-operative period
- C. The most common arrhythmia is heart block
- D. Endocarditis is only a consideration with a documented fever without a source
- E. Antibiotic prophylaxis should be given to patients after simple laceration repair
8. A 6-year-old girl is sent from her dentist for a “full medical evaluation” before he will clean her teeth. They arrive to the ED without a chief complaint and ask to have a prescription filled for antibiotics; the child had an ASD repaired without complication at 6 months of age. In evaluating which children need antibiotic prophylaxis, which of the following is TRUE?
- A. Any child with a history of congenital heart disease requires antibiotic prophylaxis
- B. Any dental procedure requires antibiotic prophylaxis

- C. Not all congenital heart defects require prophylaxis
- D. If a child has had infective endocarditis previously, he is statistically unlikely to have a repeat episode
- E. Penicillin-allergic patients are limited to IV preparations of alternative medications

ANSWERS

1. D. The neonatal myocardium is inefficient in extracting oxygen at the cellular level; its baseline oxygen requirement is high and it is unable to increase its contractility in response to demand. When increased cardiac output is needed, the neonate responds with an increasing heart rate. "A" is incorrect, because the opposite is true: in the first hours of life, the newborn's pulmonary arterioles dilate and pulmonary vascular resistance begins to fall, resulting in increased pulmonary blood flow. Separation from the low-resistance placental circuit results in increased systemic blood pressure, which also reduces blood flow through the ductus arteriosus. "B" is incorrect, because the normal physiologic changes that result in the constriction of the ductus arteriosus occur begin immediately after birth; it is functionally closed by 15 hours of life. "C" is incorrect, because the foramen ovale closes by 3 months of age. Remembering the specific timing for each event is not important; rather, a general idea of the normal time course will aid the physician in understanding the basis of the pathophysiology of congenital heart disease. "E" is incorrect, because not all congenital heart lesions cause increased pulmonary vascular resistance, and because oxygen decreases pulmonary vascular resistance. Congenital lesions which result in high pulmonary vascular resistance respond well to oxygen.
2. A. Most infants with ductal-dependent decompensated shock present within the first week, but may present into the second week after birth. It is reasonable to keep this in the differential diagnosis for the first month after birth; it rarely presents beyond 4 weeks of age.
 "B" is incorrect, because congenital heart disease is multifactorial, and may not necessarily be diagnosed prenatally. Any neonate presenting in shock should be evaluated for congenital heart disease, among other more common etiologies, such as sepsis. "C" is incorrect, because PGE₁ is given to maintain the patency of the ductus arteriosus—not an atrial septal defect—which may help to shunt blood to the systemic circuit until a decision is made for surgical or palliative therapy. "D" and "E" are incorrect because the known, expected side effects of PGE₁ include *apnea, hypotension, and bradycardia*. Patients on PGE₁ therapy inevitably will need ventilator and circulatory support.
3. A. Children need not have an established diagnosis of congenital heart disease in order to present with CHF. Cardiomyopathies (congenital or acquired) and prolonged dysrhythmias also predispose infants to CHF. "B" is incorrect because symptoms of CHF are notoriously gradual in onset and may be subtle. Poor feeding, sweating while feeding, poor growth, and irritability are all consistent with CHF. Patients may present with increased respiratory effort, chronic cough or wheezing and increased frequency of respiratory infections.
 "C" describes the common presentation of right-sided heart failure in adults. Children more commonly present with hepatic congestion or hepatomegaly, as the relatively pliable liver becomes congested with venous blood. "D" is incorrect, because fluid and sodium restriction is the mainstay of therapy in adults and children, along with diuretics and ionotropes.
 "E" is incorrect because continuous positive airway pressure is commonly used in children for CHF; early and judicious use of CPAP may avert intubation in an exacerbation.
4. B. During a "tet spell," there is a clamping down of the pulmonary outflow tract, causing increased right-sided pressures, and therefore increased right-to-left shunting across the ventricular septal defect (VSD). The child attempts to compensate by hyperventilating (to improve oxygenation) and squatting or drawing the knees to the chest (to increase SVR/left heart pressure, which decreases right-to-left shunting of deoxygenated blood and therefore improves oxygenation). "A" is incorrect, because increased right-to-left shunting leads to hypoxemia, cyanosis, and acidosis. "C" is incorrect; although a crying child usually indicates an open airway and some evidence of cerebral perfusion and oxygenation, in this case it may be detrimental. Crying slightly decreases oxygen saturation, and the prolonged expiratory phase decreases venous return, which decreases preload, which in turn decreases flow to the pulmonary artery. Keeping the child calm can be key to breaking a "tet spell." "D" is incorrect because morphine is the first-line treatment for a "tet spell," along with a knee-to-chest position and oxygen. Its mechanism of action in these spells is not entirely understood, but is probably due to its sedating properties. "E" is incorrect, because on the contrary, a fluid bolus is indicated. Patients with TOF are preload dependent, and a 10 mL/kg bolus may help to increase blood flow

- to the pulmonary artery and improve oxygenation. This will further dilate the pulmonary artery, which decreases the right-sided pressures and thereby decreases the right-to-left shunt, breaking the spell.
5. E. Coarctation of the aorta varies in severity and patients may be asymptomatic for years prior to diagnosis. Hypertension may be noted on evaluation for symptoms such as headache, dizziness, palpitations, or epistaxis. Occasionally, there may be a complaint of claudication due to decreased perfusion of the lower extremities. “A” is incorrect because auscultation of a murmur on routine physical examination may uncover unapparent lesions such as ASD, small VSD, PDA, PS, AS, and aortic coarctation. “B” is incorrect because patients with PDA and AS most commonly present with dyspnea on exertion and fatigue, not cardiorespiratory arrest. “C” is incorrect because patients with critical AS typically present with syncope, not plethoric facies. “D” is incorrect because patients with critical PS may present with cyanosis on exertion, right-sided heart failure, or syncope, not pedal edema.
 6. D. Physiologic repairs are palliative or are an intermediate step to further repair; they invariably involve late complications. “A” is incorrect because there are many congenital lesions that can undergo an anatomic repair: ASD, VSD, PDA, aortic coarctation, and TGA (switch procedure). These children have an anatomic correction and often lead normal lives after surgery. “B” is incorrect because these patients are not “re-categorized” as low-risk. Although the repair is anatomic (and not physiologic or palliative), these lesions often result in residual defects. “C” is incorrect. Replacement of the prosthetic material—and therefore repeat surgery—is indicated when children outgrow previous valves or in cases of valvular degeneration.

“E” is incorrect, because children with repaired congenital heart disease are at higher risk for complications such as arrhythmias, infective endocarditis (IE), and thromboembolism.
 7. A. Arrhythmias are the most common post-heart surgery problem and may present with symptoms of palpitations, decreased appetite, emesis, and decreased exercise tolerance. “B” and “C” are incorrect, because the risk of arrhythmia after heart surgery does not end in the immediate post-operative period. The most common arrhythmia seen is SVT. “D” is incorrect, because endocarditis should always be considered in corrected or uncorrected congenital heart disease, especially without a known source. “E” Antibiotic prophylaxis guidelines do not include simple laceration repair.
 8. C. The guidelines for antibiotic prophylaxis for congenital heart disease patients have recently narrowed in scope. First, it is important to identify whether the patient is in a high-risk group; next, one can refer to a list of procedures where prophylaxis is recommended. High risk lesions include any residual defect or prosthetic material (Table 47-3). The girl in this scenario is completely repaired with regards to her ASD, and therefore does not require antibiotic prophylaxis. High-risk procedures (in patients at risk) include all invasive dental procedures, and procedures on the respiratory tract or infected tissue. “A” and “B” are incorrect because only high-risk patients (see list) undergoing certain procedures (see list) require antibiotic prophylaxis. “D” is incorrect, because a previous history of infective endocarditis puts the child at a higher risk for an adverse outcome from a repeat episode. “E” is incorrect, because many PO alternatives exist for antibiotic prophylaxis for congenital heart disease (Table 47-5).

48 CONGESTIVE HEART FAILURE

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INTRODUCTION

- Heart failure is a “clinical syndrome in which heart disease reduces cardiac output, increases venous pressures, and is accompanied by molecular abnormalities that cause progressive deterioration of the failing heart and premature myocardial cell death.”
- Cardiac output is determined by *preload*, or filling volume (increased in left-to-right shunts), *afterload*, or the resistance the ventricles face upon ejection of blood (important in outlet obstruction or systemic hypertension), *contractility* (altered in cardiomyopathy) and *rate* (too slow, resulting in inadequate output, or too fast, decreasing diastolic filling).

ETIOLOGIES OF CONGESTIVE HEART FAILURE (CHF)

- Congestive heart failure (CHF) has many etiologies:
 - Preload (volume overload): *left-to-right shunt*: VSD, PDA, AV fistula, atrioventricular canal defects, Epstein’s anomaly; anemia: iron deficiency, sickle cell, thalassemia; *iatrogenic*.

TABLE 48-1 Signs and Symptoms of CHF in Children

PULMONARY VENOUS CONGESTION (L → R SHUNT, PULMONARY EDEMA, POOR OXYGENATION)	SYSTEMIC VENOUS CONGESTION (INCREASED RIGHT-SIDED FILLING PRESSURE)	IMPAIRED CARDIAC OUTPUT (DECREASED CONTRACTILITY AND PERFUSION)
Tachypnea	Hepatomegaly	Decreased pulses
Wheezing (cardiac asthma)	Ascites	Delayed capillary refill (cool extremities)
Rales	Pleural effusion	Fatigue
Nasal flaring or grunting	Peripheral edema	Pallor
Retractions	Weight gain	Sweating
Cough or chest congestion	JVD (rare in children)	Poor weight gain (failure to thrive)
Poor feeding		Dizziness
Irritability		Altered consciousness
		Syncope

JVD, jugular venous distension.

- Afterload (increased SVR): *congenital*: coarctation of the aorta, aortic stenosis, pulmonary stenosis; *systemic hypertension*
- Contractility: *inflammatory*: infectious (myocarditis), cardiomyopathy; *rheumatic*: rheumatic fever, early Kawasaki, SLE; *toxin*: digoxin, Ca²⁺ channel/β-blocker, cocaine and other stimulants; *traumatic*: cardiac tamponade, myocardial contusion; *metabolic*: electrolyte abnormality, hypothyroidism; *other*: asphyxia, hypoglycemia, hypocalcemia, sepsis.

SIGNS AND SYMPTOMS

- Signs and symptoms of CHF vary with the age of the child (Table 48-1).
- When evaluating an *infant*, questions related to feeding and nutritional status are extremely important.
- In an *older child*, questions regarding exercise ability, fatigability, shortness of breath, weight gain, weight loss, abdominal pain, nausea, and anorexia are important when considering heart failure.

PHYSICAL EXAMINATION

- *Physical examination* should evaluate for resting tachycardia, tachypnea, hepatomegaly, ascites, edema, and diminished perfusion.
- *Cardiac examination* should focus on heart rate, presence of gallops, new murmurs, displaced point of maximal impulse, a right ventricular impulse, and perfusion deficits.

LABORATORY TESTING

- *Laboratory testing* includes arterial blood gas, complete blood cell count with hemoglobin concentration,

electrolytes, calcium, BUN, creatinine, lactic acid, liver function tests, and urinalysis.

- Newer laboratory tests assessing neurohormonal markers in heart failure have evolved.
 - The measurement of B-type natriuretic peptide (BNP) is useful in differentiating a pulmonary cause of dyspnea from a cardiac cause of dyspnea.
 - A BNP >100 pg/mL is considered abnormal; between 100 and 500, the test is inconclusive; and a level >500 pg/mL is indicative for heart failure in children.

ANCILLARY TESTING

- *Ancillary tests* include a chest radiograph, electrocardiogram (EKG), and echocardiogram.
 - *Chest radiograph* (CXR) typically demonstrates cardiomegaly with prominent pulmonary vascular markings of pulmonary edema. But, remember a normal CXR does not rule out heart failure
 - The EKG in addition to assessing chamber enlargement or hypertrophy is useful in picking up dysrhythmias or ST–T wave changes.
 - The quickest way to assess cardiac function in the emergency department (ED) is with *two-dimensional echocardiography* (ECHO). It is utilized to assess cardiac anatomy in congenital heart disease, but also in estimating gradients, shunting, and cardiac output.

MANAGEMENT

- Management of heart failure is multidimensional and requires a stepwise approach.
 - The goal is to reduce cardiac contractility, reduce afterload, improve oxygenation, and enhance nutrition.

- Medications to consider include diuretics, vasodilators, inotropes, and neurohumoral modulators.
- When the cause is known, correctable tasks must be undertaken. Examples include interventional techniques for obstructive lesions, exchange transfusion for profound anemia, or pericardiocentesis for cardiac tamponade.
- When the cause is unknown, empiric therapy is initiated based on the need to control rate, decrease preload, and improve afterload and/or contractility (Fig. 48-1).
- An algorithmic approach to CHF is presented in Fig. 48-2.
- Dosage guidelines with pharmacologic effects for the management of CHF are presented in Tables 48-2, 48-3, and 48-4.

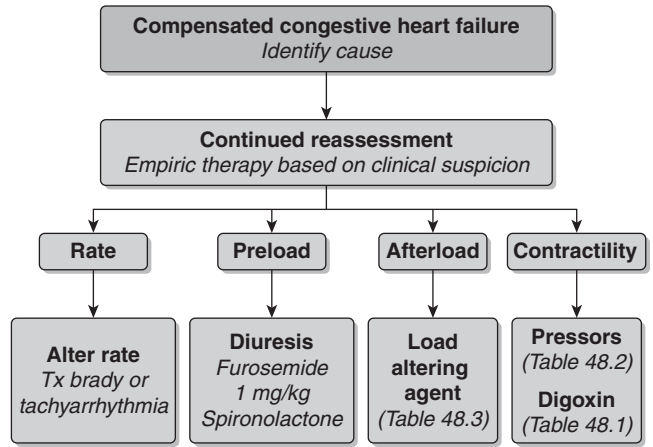


FIG. 48-1. Management of compensated (chronic) CHF in children.

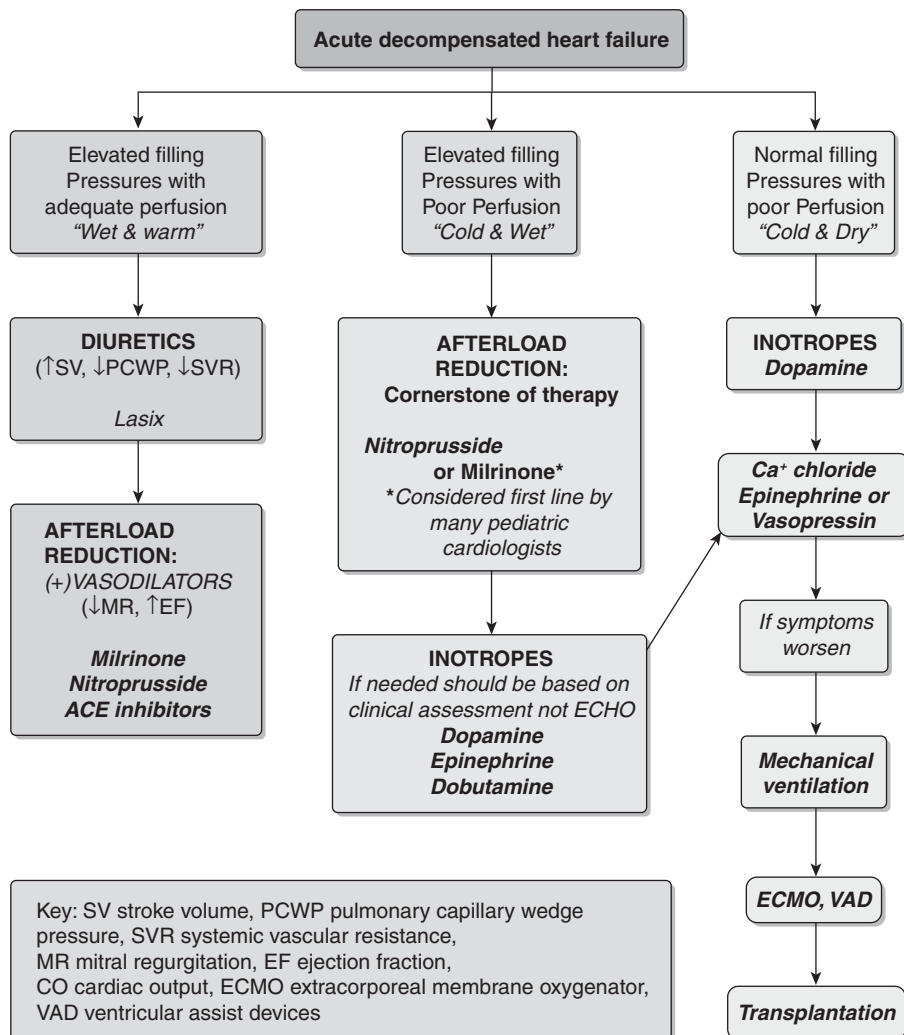


FIG. 48-2. Management of acute decompensated heart failure in children.

TABLE 48-2 Oral Dosing Guidelines for Digoxin^a

AGE AND WEIGHT	ACUTE DIGITALIZATION ($\mu\text{g}/\text{kg}$) ^b	MAINTENANCE
Premature infant	20	5 $\mu\text{g}/\text{kg}/\text{d}$
Full-term infant	30	4–5 $\mu\text{g}/\text{kg}$ q12h
2–24 mo	40–50	5–10 $\mu\text{g}/\text{kg}$ q12h
>24 mo	30–40	4–5 $\mu\text{g}/\text{kg}$ q12h

^aIV dose is 75% of po dose.

^bDaily dose = 1/2 given initially, then 1/4 given at 8 h, and 1/4 given at 16 h.

TABLE 48-4 Load-Altering Agents

DRUG	DOSE	COMMENTS
Nitroprusside	0.5–10 $\mu\text{g}/\text{kg}/\text{min}$ IV	Cyanide toxicity
Captopril	Infants: 0.1–0.5 mg/kg/d po q8–12h	Neutropenia, cough, proteinuria
Nitroglycerin	2–10 $\mu\text{g}/\text{kg}/\text{min}$ IV	Use not well established in children
Inamrinone	0.5–2 mg/kg, then 5–10 m/ kg/min IV	Hypotension, thrombocytopenia, hepatic dysfunction
Enalapril	0.1 mg/kg/d po div qd/bid, not to exceed 0.5 mg/kg/d	
Alprostadil (PGE1)	0.05–0.1 $\mu\text{g}/\text{kg}/\text{min}$ IV	For ductal-dependent lesions

TABLE 48-3 Inotropic Agents: Dosage and Pharmacologic Effects

DRUG	DOSE ($\mu\text{g}/\text{kg}/\text{min}$)	COMMENTS
Dopamine	1–5 6–20	Stimulates dopamine receptors in renal, cerebral, mesenteric, and pulmonary vasculature \uparrow HR, \uparrow contractility, and \uparrow afterload Side effects: tachycardia, dysrhythmias, increased myocardial oxygen consumption
Dobutamine	2–10	\uparrow HR, \uparrow contractility, and vasodilation Side effects: tachycardia, dysrhythmias Fallen out of favor
Epinephrine	0.05–1	\uparrow HR, \uparrow contractility, \uparrow afterload, and vasodilation Dose-dependent actions: tachycardia, proarrhythmia effects, increased myocardial oxygen consumption
Norepinephrine	0.1	\uparrow HR, \uparrow contractility, \uparrow afterload Side effects: bradycardia, arrhythmias
Inamrinone	5–10 Load: 1 mg/kg IV over 2–3 min	\uparrow HR, \uparrow contractility, \uparrow afterload Side effects: hypotension, tachycardia, myocardial ischemia
Milrinone	0.25–1 Load: 50 $\mu\text{g}/\text{kg}$ IV slowly over 15 min	\uparrow HR, \uparrow contractility, \uparrow afterload Side effects: hypotension, dysrhythmias, renal toxic in adults

SUMMARY

- CHF is a clinical syndrome and a directed history and physical examination can provide valuable clues to the presence and possible etiologies of heart failure.
- Abnormal vital signs such as unexplained tachycardia or tachypnea with normal temperature may suggest cardiac disease.
- Tachycardia of heart failure is often “monotonous” or incessant, and does not typically respond to treatment (ie, volume, antipyretics, pain medications, etc.)
- Tachypnea, failure to thrive, or diaphoresis with feeding, accompanied by an abnormal lung examination, tachycardia, hyperactive precordium, gallop, and hepatomegaly suggest CHF in an infant.
- New-onset heart failure may be less overtly symptomatic in older children. Malaise, decrease in the level of daily activity, and weight loss may be the only complaint. Symptoms of abdominal pain and nausea

and anorexia can be present, sometimes diverting attention from the real cause.

- New biomarker, BNP may help with the diagnosis of suspected CHF in some cases.
- Management is directed at the cause. Medications to consider include diuretics, vasodilators, Inotropes, and neurohumoral modulators.

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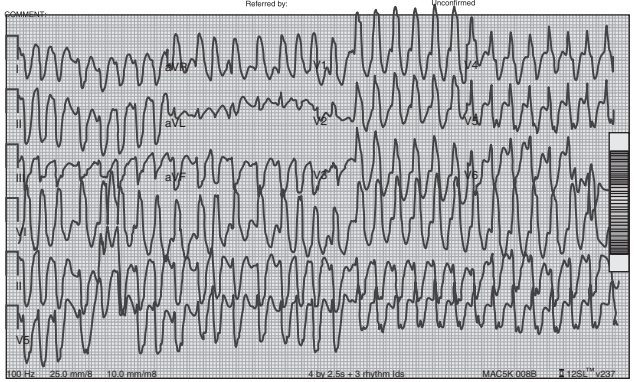
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QUESTIONS

1. A 2-month-old infant is brought to the ED by the mother who reports that the infant has been fussy for the past several days. He is taking less formula than previously and yesterday the mother noted him to be sweating profusely during feeding. He is afebrile with heart rate of 200 and respiratory rate of 60. What is the most likely cause of these complaints?
 - A. Myocarditis
 - B. Pericarditis
 - C. Endocarditis
 - D. CHF
 - E. Acute Rheumatic fever
2. On physical examination, the child described above has rales and a hyperactive precordium associated with a gallop. There is hepatomegaly but no pedal edema or neck vein distension. Chest x-ray shows pulmonary edema. Which of the following is true regarding the most appropriate management?
 - A. The child is kept at strict bed rest in the supine position.
 - B. Sedation is contraindicated since it will result in respiratory depression.
 - C. Oxygen can be given but concentrations are limited to <50%.
 - D. Digoxin is contraindicated due to its arrhythmogenicity.
 - E. Fluid overload is treated with furosemide, 1 mg/kg/dose IV.
3. A 4-year-old girl with known VSD and asthma presents to the ED with a 2-day history of fever, wheezing and tachypnea. Her respiratory rate is 40/min and pulse oximetry is 94%. She is home on enalapril, advair, and prn albuterol and according to her mother she has noted no response to the albuterol this morning. On arrival to the emergency room she is started on an asthma protocol, which includes three albuterol treatments every 20 minutes and PO steroids. You go to reassess her after one hour and note that she has progressive tachypnea with RR 70/min, pulse oximetry of 90% and persistent wheezing on examination. Which of the following is true regarding the most appropriate next step in her management?
 - A. The child should be placed on continuous albuterol at 20 mg/hr and reassess in another hour.
 - B. An IV should be placed and an additional dose of Solu-Medrol should be given
 - C. An IV should be placed and a dose of Furosemide 1 mg/kg/dose IV should be given.
 - D. A CXR should not be ordered because this is purely an asthma exacerbation.
 - E. Antibiotics should be started because the most likely reason for her deterioration is a concomitant pneumonia.
4. A 15-year-old boy with a 2-week history of vague abdominal pain and vomiting comes to the emergency room with increasing shortness of breath over the last 24 hours. He is placed on a monitor and the following rhythm is present. The first vital signs are HR 180, RR 40 with PO 88% RA. His physical examination is remarkable for rales bilaterally. What would be your treatment plan?
 
 - A. Place the patient on 40 % oxygen only.
 - B. Ask the respiratory therapist to begin an albuterol treatment for the shortness of breath.
 - C. Ask the nurse to place an IV and give a fluid bolus because of your concern for

- dehydration since he has been vomiting off and on for 2 weeks.
- D. Consider immediate synchronized cardioversion for the treatment of the above rhythm.
 - E. Order amiodarone at 10 mg/kg IVP and then reassess the rhythm.
5. The definitive test to perform in the ED for this patient to determine cardiac function would be
 - A. A repeat EKG
 - B. CXR
 - C. Chemistries with lactate
 - D. CBC
 - E. ECHO
 6. A 3-month-old presents to the ED in January with a four-day history of upper respiratory symptoms and cough. The patient is brought back to a room and placed on a monitor. The initial vital signs show a HR 180/min with RR 60/min with pulse oximetry of 94% on room air. The physical examination is remarkable for diffuse crackles and wheezing. Initial management includes placing the child on oxygen, deep suctioning and an L-epinephrine treatment but there is no change in the clinical status. A CXR is done and shows hyperinflation with prominent pulmonary vascular markings. What adjunct test may help with the additional management of this child?
 - A. ABG
 - B. EKG
 - C. BNP
 - D. Basic metabolic panel (BMP)
 - E. Calcium level
 7. 15-year-old white male with cardiomyopathy awaiting transplant presents to the ED with fatigue and weight gain over the last several days. Patient is known to be noncompliant with his medications. On arrival, patient is tachypneic with RR 40/minute with PO 92% RA. Physical examination is remarkable for pedal edema to the level of his knees and periorbital swelling with bilateral crackles on pulmonary examination with a liver edge at 4 cm below the right costal margin. The initial management for this patient would be?
 - A. Nasal cannula oxygen at 2 L
 - B. Normal saline bolus at 20 mL/kg
 - C. Furosemide (Lasix) at 1 mg/kg IV
 - D. Dopamine at 20 mcg/kg/min IV
 - E. Epinephrine at 1 mcg/kg/min
 8. 2-week-old white infant presents to the ED gray and mottled. Mother states the child has been feeding less over the last several days and has been more irritable. On arrival the child is noted to be cyanotic, tachypneic with weak thready peripheral pulses and a capillary refill of 5 seconds. The patient is placed on a cardiac monitor and noted to have a narrow complex rhythm with a rate of 300/min, RR 60/min with BP 50/30 and pulse oximetry 82% on room air. The initial management for this child would be
 - A. Immediate intubation with RSI drugs.
 - B. Defibrillation at 2 J/kg immediately.
 - C. Synchronized cardioversion at 1 J/kg.
 - D. Adenosine at 2 mg/kg IVP once an IV or IO was placed.
 - E. Digoxin at 20 mcg/kg IV.
 9. A 4-year-old Latin-American female presented to the ED with a 1-month history of weight loss and easy fatigability. On arrival to the ED she was noted to be pale and jaundiced in appearance with a resting heart rate of 180/minute with RR 30/min and PO 94% RA. Physical examination was remarkable for a gallop with a displaced PMI on her cardiac examination, fine crackles on lung examination and a liver edge at 5 cm below the right costal margin. A CXR was done which noted cardiomegaly with increased pulmonary markings consistent with CHF of unknown etiology. The child was admitted for an ECHO and medical management. The child was given a dose of Lasix at 1 mg/kg and placed on a dopamine drip at 10 mcg/kg/min yet over the next 24 hours her symptoms worsened. The dopamine was increased to 20 mcg/kg/min. An ECHO was done which was significant for an ejection fraction of 20%. Repeat vitals noted the child to have HR 190/min with BP 70/30. What would be your next agent in the care of this child?
 - A. Dobutamine
 - B. Epinephrine
 - C. Milrinone
 - D. Nitroprusside
 - E. Nestiritide
 10. After several days in the PICU the above patient was noted to worsen on maximum medical therapy and ventilatory support. The patient was noted to have an extremely irritable precordium and had to be resuscitated for runs of ventricular tachycardia and ventricular fibrillation. What would be your next step in this patient's care?
 - A. Muscle biopsy to determine the etiology of the cardiomyopathy
 - B. Ventricular assist device placement
 - C. ECMO
 - D. Transplantation
 - E. Cardiac catheterization

ANSWERS

1. D. Clinically, infants in CHF are irritable, feed poorly, and have poor weight gain. An acute weight gain may be due to edema. Diaphoresis during feeding is especially suggestive of CHF. Volume overload may present with respiratory symptoms. The lack of fever helps to differentiate from infection. Abnormal vital signs, such as unexplained tachycardia or tachypnea with normal temperature, may suggest cardiac disease.
2. E. Fluid overload is treated with furosemide. Upright positioning may be used as part of supportive care. Oxygen therapy need not be limited in the acute situation. Digoxin is contraindicated in CHF associated with myocarditis due to its arrhythmogenic effect on the irritable myocardium, but is indicated in other patients.
3. C. The most likely reason that she has not improved is secondary to her large VSD with left to right shunting resulting in worsening CHF. Additional albuterol will only drive her tachycardia but will not help to improve her tachypnea. Additional steroids again have no place in the treatment of CHF and the patient had already received a dose of steroids on arrival. A CXR would be extremely helpful which in this circumstance may help to confirm the presence of pulmonary edema. Antibiotics are not contraindicated and may be considered secondary to the fever but are not the most appropriate next step in her management.
4. D. The patient is in an unstable ventricular tachycardia and should immediately be given 0.5–1 J/kg of synchronized cardioversion to attempt to convert his rhythm. One should not limit the amount of oxygen he is given and he should be placed on a 100% nonrebreather mask. Albuterol has no play in the treatment of ventricular tachycardia. Even if one were to consider aberrant SVT, the addition of albuterol would only worsen the tachycardia and not treat the underlying rhythm. The patient is in CHF with a tachyarrhythmia. The dosing of Amiodarone is 5 mg/kg IV over 10 minutes to a max of 150 mg IV for the first dose.
5. E. The quickest way to assess cardiac function in the ED is with *two-dimensional echocardiography (ECHO)*. It is utilized to assess cardiac anatomy in congenital heart disease, but also in estimating gradients, shunting, and cardiac output. A repeat EKG is indicated to help to assess the additional care needed in this patient but would not help to determine the underlying cause. A CXR would help to confirm the pulmonary edema but remember a normal CXR does not rule out heart failure. Abnormalities on the chemistries may show hyponatremia and hypochloremia secondary to free water retention. An elevated creatinine due to poor renal perfusion and compromised renal function and an elevated lactic acid is present with significant tissue hypoxia. The CBC should be checked since severe anemia can precipitate high-output cardiac failure and accentuate heart failure but would not lead to a definitive diagnosis.
6. C. The measurement of BNP is useful in differentiating a pulmonary cause of dyspnea from a cardiac cause of dyspnea. A BNP >100 pg/mL is considered abnormal; between 100 and 500, the test is inconclusive; and a level >500 pg/mL is indicative for heart failure in children. An ABG would show a metabolic and respiratory acidemia due to pulmonary congestion and poor tissue perfusion but would not help with definitive management. An EKG may give information regarding atrial enlargement, ventricular hypertrophy, strain, and changes in ST segment or T-wave morphology. These findings are generally nonspecific. A BMP can show hyponatremia and hypochloremia secondary to free water retention and an elevated creatinine level due to poor renal perfusion and compromised renal function but are generally more apparent with chronic CHF. Finally a calcium level is critical in an infant when managing and treating the CHF but would not help with the preliminary diagnosis. An infant relies on the circulating calcium and, if low, will add to the difficulty in managing these infants.
7. C. The patient is in pulmonary edema and would benefit from a dose of Furosemide at 1 mg/kg IV. The patient should also be placed on supplemental oxygen and would need more than a nasal cannula. A normal saline bolus would be contraindicated in a patient in CHF and would only worsen the failure. Dopamine is a great inotropic agent if needed but one would not start at 20 mcg/kg/min. The goal of such a drug would be to improve contractility, rate, and increase afterload. Epinephrine increases heart rate, contractility, afterload, and vasodilation. Yet, epinephrine has dose dependent actions and if started at a higher dose may result in increased tachycardia, proarrhythmic effects, and increased myocardial oxygen consumption and would not be considered as the first line for treatment.
8. C. The patient is in CHF with unstable supraventricular tachycardia. Immediate management would be to synchronize cardiovert with 0.5–1 J/kg

and if unchanged to increase to 2 J/kg. This patient is not in Ventricular fibrillation or pulseless Ventricular tachycardia, which would require defibrillation instead of synchronized cardioversion. Immediate intubation with RSI without treating the underlying rhythm may result in death of the child and would not be the first line for airway management. When setting up for synchronized cardioversion, the child should be placed on 100% nonrebreather mask understanding the need for BVM if respiratory status worsens the intubation. Digoxin is useful in the stable, well-known patient in need of improved contractility by blocking the sodium–potassium pump and increasing intracellular calcium, making actin–myosin bridging more forceful. It also slows rate and relies diaphoresis through adrenergic withdrawal, but is contraindicated in CHF with myocarditis because of its arrhythmogenic effects on the irritable myocardium.

9. B. Epinephrine An excellent inotropic agent that will help the patient who is both tachycardic and hypotensive in CHF from a presumptive viral myocarditis. Epinephrine will play a key role in improving the blood pressure.

Contractility can be supported with intravenous mixed agents like dobutamine or milrinone. Milrinone, a phosphodiesterase inhibitor has gained popularity in many institutions as a first line agent. Milrinone increases cardiac muscle contractility, vascular smooth muscle relaxation, and cardiac output without increasing myocardial oxygen consumption or ventricular afterload. Milrinone is given as a loading dose of 50 mcg/kg slowly over 15 minutes. A key side effect being hypotension. Therefore, milrinone should not be used for this patient because the child is already hypotensive. As for dobutamine, it has fallen out of favor because of increased mortality in adult heart failure patients.

Nitroprusside is an afterload reduction agent. In the setting of low output with increased systemic resistance, afterload reduction may be helpful. The problem with this case is that the child has already hypotensive. Nitroprusside would not be indicated and would worsen the hypotension.

Finally, nesiritide (B-type natriuretic peptide) is one of the newest therapies in the treatment of decompensated CHF in adults. It possesses vasodilatory, natriuretic, diuretic, and neurohormonal effects. It has been shown to rapidly improve hemodynamics and induce diuresis in adult patients with moderate-to-severe CHF. This medication may only be con-

sidered after consultation with your cardiologist and is felt to be an alternative for decompensated heart failure in children.

10. C. ECMO Extracorporeal membrane oxygenation and ventricular assist devices are now being used to treat children with CHF who are not responding to medical management. The hope is to extend the child's life while awaiting cardiac transplantation or recovery from infection. ECMO would be what is needed for the above case; the child is dying on maximum therapy. The Ventricular assist device would only help in treating the arrhythmias. Transplantation would be the ultimate goal but requires time and availability of a heart. Muscle biopsy could help with determining the etiology but would not help support this patient at this juncture. Finally, cardiac catheterization would have been considered earlier if a structural lesion had been suspected which was not evidence by ECHO.

49 INFLAMMATORY AND INFECTIOUS HEART DISEASE

William T. Tsai

HIGH-YIELD FACTS

- Inflammatory diseases of the heart may affect the pericardium, myocardium, or endocardium.
- Pancarditis describes inflammation involving all layers of the heart. Such inflammatory cardiac disorders may be infectious, noninfectious, or rheumatologic and enter into the differential diagnosis in children presenting with complaints that range from chest pain, to acute gastrointestinal symptoms, to symptoms of cardiovascular collapse.
- Pericarditis presents with chest pain in the older child. Pleuritic or positional chest pain, fever, tachycardia, friction rub, and electrocardiographic changes may help narrow the differential.
- Myocarditis has protean manifestations with symptom complexes that range from sudden death to signs attributable to congestive heart failure and cardiogenic shock.
- Children with acute myocarditis should be admitted to a pediatric intensive care unit for careful monitoring and aggressive supportive management.
- Echocardiography should be performed in patients with suspected myocarditis.

TABLE 49-1 Etiology of Pericarditis

Infectious:	
Viral:	coxsackievirus, enterovirus, adenovirus, hepatitis B virus, human immunodeficiency virus, Epstein-Barr virus, cytomegalovirus
Bacterial:	<i>Streptococcus pneumoniae</i> , <i>Staphylococcus aureus</i> , <i>Haemophilus pneumoniae</i> , <i>Neisseria meningitidis</i>
Fungal:	histoplasmosis, coccidioidomycosis, Candida
Other:	Lyme disease, mycobacteria
Noninfectious	
Rheumatic fever	
Autoimmune:	juvenile rheumatoid arthritis, systemic lupus erythematosus, acute rheumatic fever
Uremia	
Radiation	
Hypersensitivity to drugs	
Postpericardiotomy syndrome	
Idiopathic	

- The at-risk patient with endocarditis presents with unexplained fever, myalgia, new murmur, and elevated acute-phase reactants.

PERICARDITIS

- Pericarditis usually follows a benign clinical course. Presenting symptoms include pleuritic chest pain, fever, dyspnea, or abdominal pain. Causes overlap with those of myocarditis (Table 49-1).
- Signs include a pericardial friction rub and tachycardia. In the presence of pericardial tamponade, distended jugular veins and hepatomegaly may become noticeable.
- As cardiac output decreases, delayed capillary refill, decreased urine output, and hypotension develop. Pulsus paradoxus, an exaggerated decrease in systolic blood pressure during inspiration, may be appreciated.
- Cardiomegaly occurs on chest radiography when moderate or large pericardial effusions are present. The electrocardiogram may be diagnostic with diffuse ST-T-wave changes. Echocardiography will rapidly demonstrate the presence, size, and location of a pericardial effusion and the presence of tamponade.
- Treatment is generally supportive and includes treatment with NSAIDs and cardiology consultation. Obtain urgent cardiology and critical care consultation in children with large effusions and hemodynamic instability.
- Children with pericardial effusion who exhibit signs of hemodynamic instability secondary to cardiac tamponade should have emergent pericardiocentesis.

MYOCARDITIS

- Acute myocarditis is a serious, but relatively uncommon diagnosis in the emergency department. Symptoms can progress from those of a nonspecific respiratory illness to those of cardiovascular collapse and death in a short period of time.
- Signs and symptoms may point to an obvious cardiac etiology, but subtler and misleading presentations require the clinician to have a high index of suspicion.
- In a recent survey of the incidence of pediatric cardiomyopathy in two regions of the United States, the incidence of myocarditis as a cause of dilated cardiomyopathy was approximately 0.2 per 100,000 children.

ETIOLOGY

- An etiologic agent is identified in less than one-third of the time. Viral etiologies predominate, however bacteria, rickettsia, fungi, and parasites are known agents (Table 49-1).

CLINICAL PRESENTATION

- Myocarditis is difficult to diagnose because signs and symptoms may mimic other very common disorders. Frequently, it is not until later in the clinical course that these symptoms are noted to be of cardiac origin.
- The clinical presentation can be divided into specific symptom complexes based on presentation (Table 49-2). In general, the pathophysiology of the symptom complexes follows the gradual onset of congestive heart failure to frank cardiogenic shock.
- Complaints include cough, wheeze, congestion, fever, or tachypnea. Bronchospasm responding poorly to conventional therapy may suggest early myocarditis.
- Red flags include the child who is tachypneic, but lack symptoms of wheezing or supporting evidence for the diagnosis of pneumonia.
- Other signs and symptoms include those associated with congestive heart failure, poor feeding, cyanosis, and grunting. Murmur, gallop rhythm,

TABLE 49-2 Myocarditis Presenting Symptom Complexes

Respiratory	Difficulty breathing, retractions, wheezing
Gastrointestinal	Vomiting, diarrhea, abdominal pain
Cardiac	Chest pain, palpitations, congestive heart failure
Hypoperfusion	Lethargy, syncope, shock

TABLE 49-3 Physical Findings

	<i>n</i> (%)
Respiratory distress, tachypnea	21 (68)
Tachycardia	18 (58)
Lethargy	12 (39)
Hepatomegaly	11 (36)
Abnormal heart sounds	10 (32)
Fever	9 (30)
Hypotension	7 (23)
Pallor	6 (19)
Peripheral edema, cyanosis	5 (16)
Cyanosis, hypoxia	3 (10)

rales, or organomegaly may confirm the diagnosis (Table 49-3).

- Myocarditis should be considered in any child who deteriorates despite aggressive treatment for bronchospasm or reactive airway disease.
- The onset of metabolic acidosis secondary to severe hypoperfusion (cardiogenic shock) accounts for some of the symptoms seen. Metabolic acidosis can cause tachypnea, retractions, and grunting.
- Severe acidosis may also affect mental status causing lethargy and coma.

DIAGNOSIS

- Diagnosis requires a high index of suspicion. Unfortunately, standard laboratory, radiographic, and electrocardiographic testing is nonspecific. Chest radiograph is positive in only 42% to 75% of cases (Fig. 49-1).
- Electrocardiographic changes include nonspecific ST-T wave and axis changes. Rarely, heart block or infarct patterns may emerge.
- All patients who may have myocarditis should receive echocardiography. Findings include increased end-diastolic chamber dimensions, reduced shortening fraction, atrioventricular valve regurgitation, and regional wall abnormalities (Fig. 49-2).

TREATMENT

- Children with acute myocarditis should be admitted to a pediatric intensive care unit (PICU) for continuous monitoring.
- Initial management includes treatment of congestive heart failure or cardiogenic shock. Invasive monitoring and inotropic support with dopamine, dobutamine, epinephrine, or milrinone may be necessary.
- An aggressive approach to dysrhythmias may prevent sudden death. Heart block is an indication for transvenous pacing.

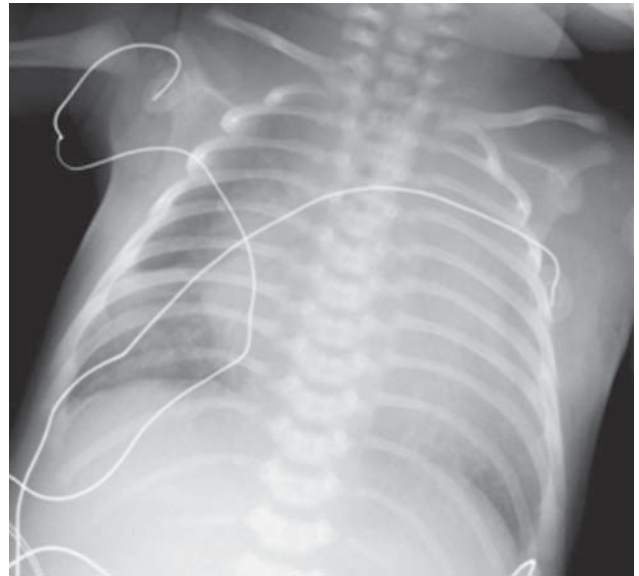


FIG. 49-1. Chest radiograph in an infant with myocarditis. Note cardiomegaly and increased vascular congestion consistent with pulmonary edema.

- The use of corticosteroids and other immunosuppressants is not well supported by current studies.
- The use of intravenous immunoglobulin remains controversial.
- Patients with fulminant myocarditis should receive aggressive mechanical support of the circulation (ECMO) because of the excellent long-term prognosis if these patients can survive the initial period of cardiogenic shock.
- Ultimately, heart transplantation may be necessary.

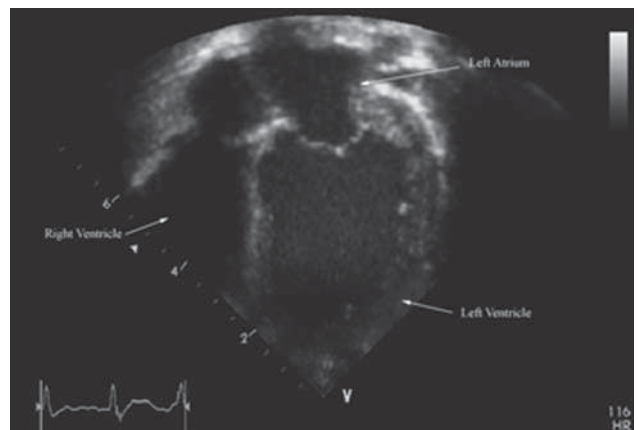


FIG. 49-2. Apical 4 chamber echocardiogram from infant in Figure 49-1, showing left ventricular dilation. Ejection fraction 28%.

ENDOCARDITIS

- Bacterial endocarditis occurs in children with congenital heart disease or central venous catheters or in adolescents who use intravenous drugs.
- Seeding can occur via dental caries, skin infections, and manipulation of the airway, gastrointestinal tract, or genitourinary tract.
- Staphylococcal and streptococcal species predominate, with HACEK organisms (*Hemophilus*, *Actinobacillus*, *Cardiobacterium*, *Eikenella*, and *Kingella*) and *Candida* as occasional offenders.
- Diagnosis is suspected in the at-risk patient in the presence of unexplained fever, weakness, myalgia, and arthralgia.
- A new murmur is present in fewer than 50% of cases. Other findings may include congestive heart failure secondary to valvular insufficiency, petechiae, or new neurologic findings. Adult cutaneous hallmarks such as Janeway lesions or Osler nodes are rare.
- Blood culture will identify the organism in 90% of cases. Other supportive data include elevated acute-phase reactants such as white blood cell count or erythrocyte sedimentation rate, anemia, hematuria, or embolic infiltrates.
- The echocardiogram has a 70% to 80% detection rate, with failure occurring in children who have complex congenital heart disease.
- Surgical intervention is indicated in cases of threatened or recurrent embolization, severe valve failure with heart failure or cardiogenic shock, recalcitrant arrhythmia secondary to the vegetation, or myocardial abscess.
- Because of the high mortality rate, prevention of endocarditis is important. The emergency physician plays an important role in endocarditis prophylaxis. American Heart Association guidelines for at-risk patients are summarized in Table 49-4.

ACUTE RHEUMATIC FEVER

- Valvular involvement characterizes the carditis of acute rheumatic fever. The acute phase begins 2 to 3 weeks after a group A streptococcal illness.
- The major Jones criteria include polymigratory arthritis, Sydenham's chorea, erythema marginatum, carditis, and subcutaneous nodules.
- The minor Jones criteria include fever, arthralgia, elevated acute-phase reactants, elevated or rising antistreptolysin (ASO) titer, leukocytosis, and EKG showing heart block.
- Mitral insufficiency is most common and is characterized by a holosystolic, high-pitched, blowing apical murmur radiating to the axilla. Regurgitant aortic

TABLE 49-4 Endocarditis Prophylaxis in Cardiac Conditions

Endocarditis Prophylaxis Recommended:

- Cardiac pacemakers and implanted defibrillators
- Complex cyanotic malformations
- Hypertrophic cardiomyopathy
- Mitral Valve Prolapse with regurgitation or thickened leaflets
- Most complex cardiac malformations
- Isolated secundum ASD
- Prosthetic valves
- Previous bacterial endocarditis
- Repaired secundum ASD, VSD, PDA, without residual, after 6 months
- Rheumatic or acquired valvular dysfunction
- Surgical systemic-pulmonary shunts

ASD, atrial septal defect; VSD, ventriculoseptal defect; PDA, patent ductus arteriosus.

Source: Adapted from Dajani AS, Taubert KA, Gerber MA, et al. Prevention of bacterial endocarditis: recommendations by the American Heart Association. *JAMA*. 1997;277:1794.

(http://www.heart.org/HEARTORG/Conditions/CongenitalHeartDefects/TheImpactofCongenitalHeartDefects/Infective-Endocarditis_UCM_307108_Article.jsp)

- murmurs are middiastolic, high-pitched, and blowing, located at the base, radiating into the neck.
- Other cardiac findings include tachycardia, gallop rhythm, pericardial rub, or congestive heart failure. The ECG may demonstrate PR prolongation, conduction delays, left ventricular hypertrophy, or dysrhythmia.
- Echocardiography is helpful in the follow-up of patients with rheumatic heart disease and may have a role in the evaluation of patients without murmur or with subclinical heart involvement.
- Treatment during the acute phase includes hospitalization and bed rest and cardiac rehabilitation follows. High-dose aspirin is started upon confirmation of the diagnosis.
- Penicillin or erythromycin is given to eradicate residual streptococci. Corticosteroids are controversial and may have a role in the treatment of carditis or chorea.
- Long-term follow-up of patients with acute rheumatic fever includes surveillance for recurrence, endocarditis prophylaxis, and treatment of chronic failure. Patients without early cardiac involvement are unlikely to develop delayed valvular disease.

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QUESTIONS

1. A teenage girl with a history of lupus presents to the emergency department with complaints of chest pain and difficulty breathing. The presence of which of the following is an indication for performing emergency pericardiocentesis?
 - A. Chest x-ray demonstrating severe cardiomegaly.
 - B. Electrocardiogram with severe, diffuse ST segment changes.
 - C. Hepatomegaly
 - D. Echocardiogram with pericardial effusion and bilateral pleural effusions.
 - E. Echocardiogram with a large pericardial effusion, tachycardia, and hypotension.
2. A 14-year-old boy presents to the emergency department with the complaint of positional chest pain. It hurts most when he is lying on his back and feels best when he is leaning forward. His electrocardiogram shows diffuse ST segment elevations. The presence of which of the below increases his likelihood of having pericarditis?
 - A. History of juvenile rheumatoid arthritis.
 - B. History of Noonan’s syndrome
 - C. History of phenylketonuria
 - D. History of DiGeorge syndrome
 - E. History of Williams’ syndrome
3. A 6-month-old infant presents to the emergency department with difficulty breathing. The parents report a low-grade fever, mild respiratory symptoms, and difficulty feeding. Which of the following signs and symptoms is concerning for a diagnosis of myocarditis?
 - A. History of URI exposure in the daycare setting.
 - B. Tachypnea and retractions with clear lung sounds.
 - C. Severe cough and multiple episodes of spitting up.
 - D. Foul urinary odor and dark-colored urine.
 - E. Green nasal discharge and swollen nasal mucosa.
4. A 1-year-old infant is transferred from another institution with the suspected diagnosis of myocarditis based on a chest x-ray that shows significant cardiomegaly. Which of the following signs or symptoms is most often seen in patients with myocarditis?
 - A. S4 gallop
 - B. Cyanosis
 - C. Tachypnea
 - D. Hypotension
 - E. Poor peripheral perfusion
5. A 5-year-old presents to the ED with fever and cough. Chest x-ray reveals cardiomegaly and bilateral pleural effusions. A diagnosis of myocarditis is suspected. Of the following etiologies below, which infectious agents are most often implicated in the diagnosis of myocarditis?
 - A. Viral
 - B. Bacterial
 - C. Protozoal
 - D. Spirochetal
 - E. Fungal
6. A 4-year-old presents to the emergency department with difficulty breathing. He has had cough and fever for approximately one week and parents have been giving him acetaminophen. They present today because of difficulty breathing. They describe significant retractions. On exam, he is indeed retracting but his breath sounds are clear with no wheezing. You suspect the diagnosis of myocarditis. Of the following, the best diagnostic test is
 - A. Electrocardiogram
 - B. Physical examination to detect the presence or absence of hepatomegaly.
 - C. Chest x-ray.
 - D. Echocardiography
 - E. Pulse oximetry

7. A 7-year-old girl is admitted to the PICU with the diagnosis of suspected myocarditis. Echocardiogram shows a dilated left ventricle with poor LV systolic function. Of the following therapies, which of the following is not well supported in the management of myocarditis?
 - A. Inotropes in setting of cardiogenic shock.
 - B. Transvenous pacing in heart block.
 - C. Diuretics in setting of congestive heart failure.
 - D. Extracorporeal membrane oxygenation (ECMO) in cases of refractory cardiogenic shock.
 - E. Steroids.
8. A 4-year-old boy with a history of complete atrioventricular canal repair presents with persistent fevers, Osler's nodes, and Janeway lesions. Echocardiogram reveals large vegetation on the mitral valve. Under which condition below should surgery be considered for removal of the vegetation?
 - A. Bacterial blood cultures are persistently positive for *Kingella* species.
 - B. Persistent diastolic murmur heard at left lower sternal border.
 - C. Difficulty breathing, bilateral pulmonary edema and severe mitral regurgitation.
 - D. Sedimentation rate greater than 60.
 - E. Severe allergy to penicillins and cephalosporins.
9. A 10-year-old girl presents to the emergency department after a fall while playing outdoors. She has sustained a through and through lip laceration extending from the philtrum to the oral mucosa. She will need this laceration repaired. Under which circumstance below will she need endocarditis prophylaxis?
 - A. She has a history of repaired atrial septal defect.
 - B. She has a pacemaker secondary to postoperative heart block.
 - C. She has a history of heart murmur.
 - D. She has a history of hypertrophic cardiomyopathy.
 - E. She has a history of Kawasaki disease without valve involvement.
10. A 16-year-old girl presents to the emergency department with polymigratory arthritis, an erythema marginatum rash, and subcutaneous nodules. She recalls having a sore throat 2 to 3 weeks ago but did not seek medical treatment. The diagnosis of acute rheumatic fever is considered. Which of the following is a mainstay of initial treatment?
 - A. High dose aspirin
 - B. Intravenous immunoglobulin

- C. High-dose ibuprofen
- D. Plasmapheresis
- E. Arthrocentesis

ANSWERS

1. E. In a patient with documented large effusion on echocardiogram and the presence of tachycardia and hypotension, consideration should be given toward the placement of an emergent pericardiocentesis catheter. The indications for emergent drainage of a documented pericardial effusion include signs of hemodynamic instability such as tachycardia, hypotension, poor perfusion, shock, and acidosis. The presence of cardiomegaly, EKG changes, hepatomegaly, and an echocardiogram with pericardial effusion and pleural effusion may indicate the presence of pericardial effusion, but are not indications for emergent drainage.
2. A. The presence of inflammatory autoimmune diseases such as juvenile rheumatoid arthritis, systemic lupus erythematosus, and acute rheumatic fever increases the likelihood of pericarditis. In addition, infectious etiologies such as viral, bacterial, and fungal infections, uremia, hypersensitivity to drugs, and postpericardiotomy states are associated with pericarditis and pericardial effusion. Noonan's syndrome, Williams' syndrome, DiGeorge syndrome and phenylketonuria are not associated with pericarditis.
3. B. While the diagnosis of myocarditis in the acute care setting may be difficult, there are a few red flag signs and symptoms that may be helpful. The presence of tachypnea and retractions in the setting of clear breath sounds suggests respiratory compensation for a metabolic acidosis. In the setting of myocarditis and cardiogenic shock, the metabolic acidosis may only manifest as difficulty breathing. In contrast, patients with bronchiolitis, reactive airways disease exacerbation, or pneumonia frequently have abnormal lung examinations. Another concerning scenario occurs during the treatment of wheezing. In children who deteriorate despite aggressive treatment for reactive airways disease, the clinician should consider the diagnosis of myocarditis. The other clinical scenarios listed are common emergency department presentations and are not helpful in identifying the child with myocarditis.
4. C. Tachypnea, along with tachycardia, is the most common signs seen in myocarditis. While these signs are sensitive, they are not specific for myocarditis.

S4 gallop, cyanosis, hypotension, and poor peripheral perfusion are infrequently seen in myocarditis but may develop as the condition worsens and signs of hemodynamic instability begin to occur.

5. A. Viruses are the most frequently identified etiologic agents in cases of myocarditis. Coxsackie viruses, enteroviruses, and adenoviruses are most frequently identified on culture, but can be detected by antigen detection tests such as PCR. In most cases however, the etiologic agent is never identified. Bacterial, protozoal, spirochetal, and fungal infection play a role in myocarditis, but are seen much less frequently than viruses.
6. D. Patients with suspected myocarditis should receive echocardiography. Echocardiography can reliably diagnose myocarditis by measuring chamber dimensions and myocardial function. The electrocardiogram shows nonspecific changes in the ST segment. Physical exam, chest x-ray, and pulse oximetry are not reliable in the diagnosis of myocarditis. While the chest x-ray may show cardiomegaly, cases of myocarditis may present with an initially normal chest x-ray.
7. E. The use of steroids remains controversial in the treatment of myocarditis. There has been no demonstrated improvement in a variety of measured endpoints in studies on adult patients with myocarditis. The data in pediatric patients is lacking. Inotropes, pacing, and diuretics are standard therapies in the acute management of myocarditis. The use of mechanical support of the circulation (ECMO) in the setting of fulminant myocarditis should be considered due to a relatively high rate of successful outcomes.
8. C. Indications for surgical treatment of bacterial endocarditis are: (1) Severe valvular failure with heart failure or shock, (2) threatened or recurrent embolization, (3) Recalcitrant arrhythmia due to vegetation, and (4) myocardial abscess.
9. D. Children with hypertrophic cardiomyopathy are at moderate risk for bacterial endocarditis. Children with cyanotic heart disease, prosthetic valves, cavopulmonary shunts, and a prior history of bacterial endocarditis are at high risk. Children with innocent murmurs repaired ASD without residual shunt, pacemakers, and Kawasaki disease without valve involvement do not require endocarditis prophylaxis.
10. A. High-dose aspirin is a mainstay treatment in acute rheumatic fever. The use of high-dose aspirin results in a rapid improvement in symptoms and attenuates the inflammatory response seen in this condition. Intravenous immunoglobulin, plasmapheresis, high-

dose ibuprofen, and arthrocentesis are not standard, initial treatments in this condition.

50 DYSRHYTHMIAS IN CHILDREN

Ghazala Q. Sharieff

INTRODUCTION

- Dysrhythmias in children are usually the result of cardiac lesions. Other causes include hypoxia, electrolyte imbalance, toxins, inflammatory disease, and cardioactive drugs, such as digoxin or over-the-counter cold remedies. A dysrhythmia associated with structural heart disease has a poorer prognosis than a one in a structurally normal heart.
- Evaluation of the child with idiopathic or unexplained dysrhythmia includes an electrocardiogram and an echocardiogram.
- Age is an important consideration in the child with dysrhythmia. Some ventricular dysrhythmias disappear with age; other conditions associated with an escape pacemaker, worsen with age. Age is also a factor in the clinical presentation. The infant may present with poor feeding, tachypnea, irritability, or signs of a low output state. The older child will have specific symptoms, such as syncope, chest pain, or palpitations. Active adolescents with syncope, palpitations, or exertional chest pain should be investigated promptly.
- The emergency management of dysrhythmias is dependent on rate, QRS width on a 12-lead ECG, and clinical stability, as determined by heart rate and blood pressure. Normal ranges for heart rate and blood pressure are listed in Tables 50-1 and 50-2.

SLOW RATES

SINUS BRADYCARDIA

- **Sinus bradycardia** can be a manifestation of serious underlying disease or a normal physiologic variant. Serious causes include hypoxia, hypothyroidism,

TABLE 50-1 Expected Heart Rates According to Age

AGE	RATE (MEAN)
0–3 mo	80–205 (140)
3 mo–2 y	75–190 (130)
2–10 y	60–140 (80)
>10 y	50–100 (75)

TABLE 50-2 Expected Systolic and Diastolic Blood Pressures According to Age

AGE	SYSTOLIC BP (mm Hg)	DIASTOLIC BP (mm Hg)
0 d	60–76	30–45
1–4 d	67–84	35–53
1 mo	73–94	36–56
3 mo	78–103	44–65
6 mo	82–105	46–68
1 y	67–104	20–60
2 y	70–106	25–65
7 y	79–115	38–78
15 y	93–131	45–85

increased intracranial pressure, or calcium channel blocker, beta-blocker, or digoxin toxicity.

- Treatment of the underlying condition will help to correct the rate. Since the most common cause of bradycardia is hypoxia, it is important to assess the ABC's first.
- If the patient remains bradycardic after oxygenation and ventilation are optimized, the drug of choice is epinephrine (1:10,000, 0.01 mg/kg IV/IO). Atropine (0.02 mg/kg, minimum dose of 0.1 mg) may be considered in cases of heart block or vagally mediated bradycardia. High dose epinephrine is no longer recommended unless a B-blocker overdose is suspected.

ATRIOVENTRICULAR BLOCKS

- **Complete atrioventricular (AV) block** may be congenital or acquired. Congenital block associated with structural abnormalities has a poorer prognosis than AV block associated with maternal collagen vascular disease. Rates of 50 to 80 bpm are typical in complete AV block and rates above 50 are rarely symptomatic. Complete AV block is suspected in utero in the setting of sustained fetal bradycardia, polyhydramnios, and congestive heart failure (CHF).
- Treatment of neonatal symptomatic bradycardia due to AV block includes control of CHF, atropine, or isoproterenol, and temporary transcutaneous, transthoracic, or umbilical transvenous pacing.
- **Acquired third degree block** is associated with myocarditis, endocarditis, rheumatic fever, cardiomyopathy, Lyme disease, or tumor. Postoperative blocks, which are less common today because of intraoperative mapping, may last for years or occur years after surgery. Unlike congenital third degree block, QRS complexes are usually wide.
- Treatment is similar, except patients with syncope must be paced immediately.

PACEMAKERS IN CHILDREN

- The indications for pediatric pacemakers include symptomatic bradycardia (most common), prolonged Q T syndrome, and cardioinhibitory syncope lasting longer than 10 seconds.
- Most permanent pediatric pacemakers are transvenous, with epicardial units being reserved for premature infants and those with right-to-left shunts. Choice of mode depends on disease. Most units can be programmed to sense, demand, or inhibit at the atrial or ventricular level and may also be programmed to sense motion or breathing.
- Syncope or palpitations in a child with a pacemaker suggests malfunction. Chest radiography may reveal wire fracture or lead displacement. Most malfunctions are not mechanical and require external reprogramming. Uncaptured paced beats outside the refractory period require investigation. If the problem is not easily resolved, the patient should be admitted.

FAST RATES

PAROXYSMAL SUPRAVENTRICULAR TACHYCARDIA

- The most common dysrhythmia in children is **paroxysmal supraventricular tachycardia (PSVT)**. PSVT is differentiated from sinus tachycardia by: abrupt onset, heart rate higher than 220 bpm in infants and 180 bpm in children, absence of normal P waves, and little to no beat to beat variation.
- In infants, symptoms include ill appearance, poor feeding, tachypnea, and irritability.
- Although it may be associated with fever, infection, drug exposure, or congenital heart disease, PSVT is usually caused by one of two mechanisms: younger children are more likely to have accessory pathway tachycardia. Adolescents may have AV nodal reentry.
- Accessory pathway tachycardia is usually orthodromic, with antegrade AV conduction and retrograde accessory pathway conduction. Conduction, during sinus rhythm, can be via the accessory pathway, resulting in a short PR interval and appearance of a delta wave. This characterizes the Wolff–Parkinson–White (WPW) syndrome. Some accessory pathways only conduct retrograde during bouts of PSVT and are termed “concealed” because they are not apparent on surface ECG.
- Lack of delta wave during sinus rhythm does not rule out concealed accessory tracts.

- Unstable PSVT is treated with synchronized cardioversion, 0.5 J/kg, increasing to 2 J/kg as needed. If unsuccessful, esophageal overdrive pacing may be necessary.
- Stable PSVT can be treated with vagal maneuvers by placing a bag of ice with some water over the nose and forehead for 15 to 20 seconds.
- If ice water fails, adenosine, 0.1 mg/kg, followed by 0.2 mg/kg, is recommended. Adenosine terminates nodal and accessory pathway tachycardia. Transient side effects include headache, flushing, chest pain, apnea, bronchospasm, and asystole.
- Digitalis is commonly used to prolong AV nodal conduction and refractoriness of fast and slow tracts (see Table 33-3). It may precipitate ventricular tachycardia (VT) and should be used under the supervision of a pediatric cardiologist. Digoxin may take hours to work and, if cardioversion is necessary, there is a risk of ventricular fibrillation.
- Procainamide is preferred in narrow complex tachycardia thought to be ventricular. A 15-mg/kg bolus is given over 20 to 30 minutes, watching for hypotension.
- Amiodarone can be used (5 mg/kg given over 20–60 minutes). However, it should not be given in conjunction with procainamide as the QT interval can be prolonged or severe hypotension may develop.
- Verapamil is not routinely used and is reserved for children >2 to 3 years of age. Hypotension, cardiovascular collapse, and death have occurred in infants. Older children with stable but recalcitrant PSVT may respond to IV verapamil, 0.1 mg/kg slowly. Calcium chloride, 10 mg/kg, and IV saline should be available to treat hypotension.
- Patients who are discharged are typically started on B-blocker therapy in consultation with a pediatric cardiologist.

Electro physiologic studies (EPS) and surgical ablation may be necessary.

ATRIAL FLUTTER AND FIBRILLATION

- **Atrial flutter and fibrillation** in children are rare. Children with congenital heart disease, rheumatic fever, or dilated cardiomyopathy are at highest risk. Patients with atrial flutter or fibrillation, in combination with an accessory pathway or hypertrophic cardiomyopathy, are at high risk for sudden death. Unstable patients should undergo cardioversion with 0.5 J/kg. Patients with long-standing atrial disease associated with a diseased sinus node are at a risk for bradycardia or asystole on termination. Pacing must be available.

PREMATURE VENTRICULAR CONTRACTIONS

- **Premature ventricular contractions (PVCs)** in the infant and young child are rare: unifocal PVCs begin appearing in healthy children during adolescence. The patient is usually asymptomatic and has a normal physical examination, chest x-ray, and ECG. Unusual morphology, such as multifocal PVCs, coupling, or the “R on T” phenomenon, are rarely cause for emergency intervention in the asymptomatic child with a normal Q T interval.
- PVCs that diminish during exercise or stress are benign and require no therapy. Patients with myocarditis, cardiomyopathy, congenital heart disease, those who are postoperative from cardiac surgery, or those who have syncope or exercise-induced PVCs, are at a greater risk and may require treatment. Lidocaine, procainamide, propranolol, or amiodarone may be useful using guidelines similar for ventricular tachycardia.

ACCELERATED IDIOVENTRICULAR RHYTHM

- **Accelerated idioventricular rhythm (AIVR)** is a benign pediatric dysrhythmia that has the appearance of ventricular tachycardia. It begins gradually with fusion beats and is a monomorphic, wide-complex rhythm that originates from an accelerated ventricular focus with rates that are rarely faster than 150 bpm. Patients with AIVR are stable. AIVR rarely responds to medication and can be a warning of a residual hemodynamic abnormality associated with corrected congenital heart disease.

VENTRICULAR TACHYCARDIA

- **Ventricular tachycardia (VT)** is rare in children. It is distinguished from PSVT by wide QRS complexes, more than 0.08 to 0.09 seconds, depending on age (complexes as narrow as 0.06 seconds have been noted in infantile VT). Wide complexes can also be seen in PSVT if conduction is antegrade through an accessory pathway. Rates averaging 250 bpm are rarely helpful in differentiating PSVT from VT. AV dissociation with P wave and QRS independence helps distinguish VT from PSVT.
- Idiopathic ventricular tachycardia can be seen in a child with a normal heart who is completely asymptomatic: it is usually not treated. Causes of VT include electrolyte disturbance, toxins, myocarditis, structural heart disease, tumor, cardiomyopathy, or long QT syndrome. Recurrent exercise-induced syncope is often due to VT and the initial work-up may be negative. EPS or biopsy may be necessary to guide treatment.

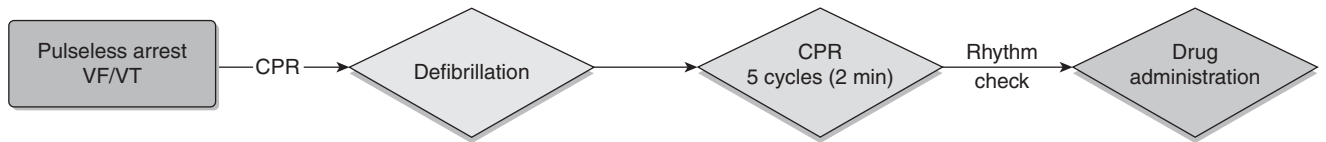


FIG. 50-1. Sequence of resuscitation, in pulseless arrest with ventricular fibrillation (VF) and ventricular tachycardia (VT).

- Regardless of etiology, unstable wide-complex tachycardia in patients with a pulse should be synchronously cardioverted with 0.5–1 J/kg. Any patient in cardiorespiratory arrest should be defibrillated.
- Amiodarone, (5 mg/kg over 20–60 minutes) can be given to stable patient. Procainamide may be useful for wide-complex tachycardia of uncertain origin because of its effect both above and below the AV node. An initial dose of 15 mg/kg over 30 to 45 minutes is followed by 20 to 80 $\mu\text{g}/\text{kg}/\text{min}$. Adenosine is safe and may be useful in the rare case of PSVT with aberrancy.
- It is important to note that stacked shocks are no longer recommended. A single shock is recommended followed by CPR largely because of the prolonged period of time to administer three shocks.
- CPR should not be interrupted until 5 cycles or 2 minutes for a pulse/rhythm check (Fig. 50-1) have been completed. More specifically, the treatment of each rhythm disturbance can be classified according to the tachycardia algorithm (Fig. 50-2). The presence or absence of a pulse determines which arm of the algorithm to initiate
 - Amiodarone (5 mg/kg, IV push) is recommended if defibrillation and epinephrine is not effective. It is important to note that amiodarone and procainamide should not be administered together as they can lead to severe hypotension and prolongation of the QT interval
 - Lidocaine is only recommended when amiodarone is unavailable.

VENTRICULAR FIBRILLATION

- Ventricular fibrillation is treated with defibrillation, at 2 J/kg and then 4 J/kg. Correction of precipitating factors, such as acidosis, hypoxia, or metabolic derangements, aids in conversion.

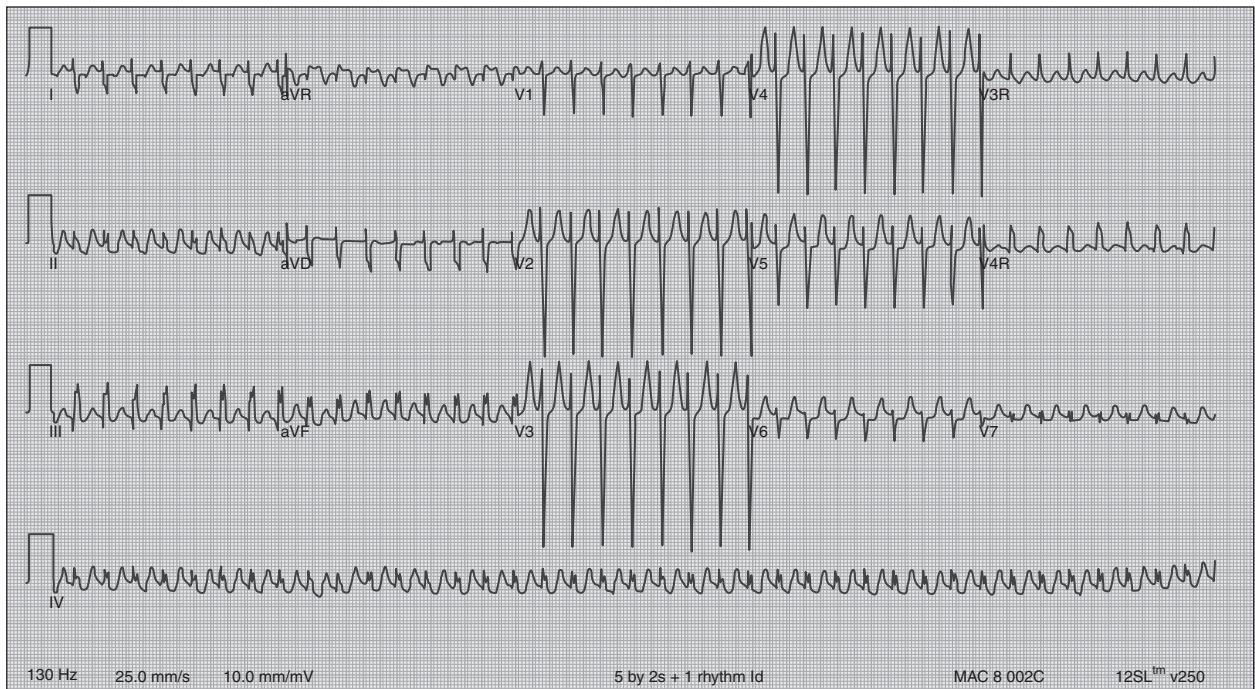


FIG. 50-2. SVT, with concomitant right ventricular hypertrophy. This 4-year-old male was postoperative from repair of congenital heart disease (Fontan repair). He was eventually converted to normal sinus rhythm after multiple doses of adenosine.

- Do not use high-dose epinephrine (1:1000 concentration via IV) as it is not recommended in any age group, and is actually associated with a worse outcome, especially in cases of asphyxia. The standard recommended dose is (0.01 mg/kg IV/IO) for all doses, which correlates to 0.1 mL/kg. Although the preferred routes of administration are intravenous or intraosseous, it may be given via the endotracheal tube when such access is unable to be obtained (0.1 mg/kg ETT). In exceptional cases, such as β -blocker overdose, high-dose epinephrine may be considered.
- Magnesium sulfate at a dose of 25 to 50 mg/kg (maximum 2 g) should be given for torsades de pointes.
- In the community, AEDs have been shown to increase survival rates. There has been sufficient evidence to show that AEDs can safely be used for those older than 1 year. In a sudden witnessed collapse, the AED should be used as soon as it becomes available. If the collapse is unwitnessed, CPR should be performed for 5 cycles or 2 minutes, prior to the use of the AED. Pediatric AED pads and energy levels should be used in those 1 to 8 years of age. If the pediatric dose is unavailable, the adult dose is a reasonable alternative.

OTHER CARDIAC CONDITIONS ASSOCIATED WITH DYSRHYTHMIAS

LONG QT SYNDROME

- Jervell and Lange-Nielsen first described the association of syncope, sudden death, deafness, and **long QT interval** in 1957. In 1963, Romano described the syndrome in normal-hearing patients. Congenital long QT syndrome (LQTS) is an inherited syndrome characterized by paroxysmal ventricular tachycardia and torsades de pointes. It can progress to ventricular fibrillation and sudden death. There may be a family history of syncope, sudden death, unusual seizures, drop attacks, or congenital deafness.
- Acquired QT prolongation associated with type IA antiarrhythmics, drugs, anorexia nervosa, bulimia, and electrolyte derangements can also predispose to dysrhythmia. A QTc longer than 0.44 seconds is a sign of delayed repolarization; more than 0.5 seconds is highly associated with sudden death. T wave alternans is also seen.
- Treatment includes β -blockers to help control sympathetic rushes and decrease the incidence of syncope, ganglionectomy, or pacing. Magnesium is used to treat torsades de pointes. Mortality from untreated congenital LQTS approaches 50%.

HYPERTROPHIC CARDIOMYOPATHY

- **Hypertrophic cardiomyopathy (HC)** is characterized by a hypertrophied, nondilated left ventricle. Symptoms include chest pain, dyspnea, syncope, CHF, or sudden death. Some patients are asymptomatic.
- Dysrhythmias include atrial fibrillation and ventricular tachyarrhythmia, the leading causes of sudden death. Atrial fibrillation associated hypertrophic cardiomyopathy puts a child at a high risk for 1:1 conduction, ventricular tachycardia, and sudden death.
- Outflow obstruction is rare. A late systolic murmur and paradoxical splitting of S2 may be present.
- Risk factors for sudden death include: presence in infancy, advanced symptoms at diagnosis, LV dysfunction, family history of sudden death, LV or septal hypertrophy on ECG is a poor prognostic sign.
- Echocardiogram is diagnostic.
- Therapy depends on the clinical manifestation: β -blockers are used for CHF but have no effect on rates of sudden death. Amiodarone can control atrial fibrillation but may cause sudden death; implantable defibrillators may be preferred. Surgical myectomy may be necessary for a significant outflow obstruction.

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QUESTIONS

- A 7-year-old boy collapses while playing baseball. Per bystanders, patient was stealing second base. Upon arrival of the paramedics, the patient has a heart rate of 180. Quick look paddles demonstrate an arrhythmia. What is the most common dysrhythmia in children?

 - Sinus bradycardia
 - Atrioventricular block
 - Atrial flutter
 - Paroxysmal supraventricular tachycardia
 - Premature ventricular contractions
- A 6-month old is brought by his parents for rapid breathing. The mother reports that since this morning the infant has had increased irritability and poor feeding. Upon arrival to the ED, the infant appears ill, tachypneic, and lethargic. The monitor shows PSVT. Which of the following is the most appropriate intervention for this infant?

 - Verapamil, 0.1 mg/kg slow IV push
 - Digitalis, 30 µg/kg IV slow IV for acute digitalization
 - Synchronized cardioversion, 0.5 J/kg, increasing to 2 J/kg as needed
 - Adenosine, 0.1 mg/kg IV push followed by 0.3 mg/kg, if needed
 - Vagal maneuvers, such as placing a bag of ice water over the nose and forehead
- A 17-year-old boy presents with syncope while playing football. He has no medical history and was in good health. He has no current complaints. His mother tells you that her brother was a victim of sudden death for unexplained reasons 20 years ago. His ECG is normal except for a QTc of 0.55 seconds. What is the prognosis?

 - The findings are a sign of delayed repolarization and unlikely to cause any significant problem.
 - This patient has congenital long QT syndrome (LQTS) and may experience recurrent syncope but is not at risk of significantly increased mortality
 - β-Blockers are contraindicated
 - Mortality of untreated congenital LQTS approaches 10%
 - QTc prolongation of more than 0.5 s is highly associated with sudden death
- A 10-year-old child presents after a syncopal episode. He reports progressive dyspnea on exertion. He is noted to have a late systolic murmur and paradoxical splitting of S2. On questioning, his uncle died a sudden death at age 25 years. Which of the following is correct regarding this patient?

 - Digoxin is the treatment of choice
 - It is not hereditary
 - The murmur may be increased by squatting or forceful hand gripping
 - An echocardiogram is the diagnostic study of choice
 - This patient may be safely discharged
- A 9-year-old boy is rescued from the nearby lake after a near drowning episode. Upon arrival of paramedics, patient is unconscious. CPR was started by bystanders. En route to the emergency department, the cardiac monitor shows sinus bradycardia. Which of the following is the most likely etiology for his bradycardia?

 - Hypoxia
 - Hyperthyroidism
 - Decreased intracranial pressure
 - Digoxin toxicity
 - Fever
- An 8-year-old with a history of congenital heart disease is sent from the pediatric clinic for evaluation of chest pain. You learn that the child recently had a pacemaker placed and was brought to the clinic by his parents “after fainting” at school. Which of the following is true regarding pacemakers in children?

 - Most are epicardial dual chamber units
 - The most common indication is symptomatic bradycardia
 - Syncope in a child with a pacemaker suggests overmedication
 - The mode is programmed to sense motion at the atrium
 - A CXR is not indicated in a child with a pacemaker malfunction
- A 4-month-old infant is brought to the emergency department for rapid breathing. The infant is breast fed and has been refusing breast feedings for the last 4 hours. The infant was a full term vaginal delivery with no complications and has been doing well at home. Upon arrival to the ED, the infant is febrile at 101.2 and an ECG reveals the absence of normal P waves, with little to no beat to beat variation. You suspect SVT. Supraventricular tachycardia in infants is defined as a heart rate greater than:

 - 180 bpm
 - 200 bpm
 - 190 bpm
 - 220 bpm
 - 150 bpm

ANSWERS

- D. The most common dysrhythmia in childhood is PSVT. PSVT is differentiated from sinus tachycardia by its abrupt onset, rates higher than 230 bpm, the

absence of normal P waves, or by little rate variation during stressful activities, such as phlebotomy. Symptoms include poor feeding, tachypnea, and irritability.

2. C. Unstable PSVT is treated with cardioversion. If unsuccessful, esophageal overdrive pacing may be necessary. Vagal maneuvers may be used to convert stable patients. Adenosine is effective but immediate recurrence rates approach 50%. Digitalis is best used in the well-known stable patient with AV nodal reentry. A pediatric cardiologist should be consulted prior to administration. Verapamil can cause hypotension, cardiovascular collapse, and death in infants and its use is not recommended for infants under 2 to 3 years of age.
3. E. QTc prolongation of more than 0.5 seconds is highly associated with sudden death. Mortality of untreated congenital LQTS approaches 50%. β -Blockers decrease the incidence of syncope and reduce mortality.
4. B. This patient has hypertrophic cardiomyopathy. The murmur decreases with squatting and forceful hand gripping. Echocardiography is the diagnostic study of choice. It is hereditary. Digoxin and nitrates should be avoided. Patients with syncope should be admitted.
5. A. Treatment of the underlying condition will help to correct the rate. The most common cause of bradycardia is hypoxia. Other causes of sinus bradycardia in young children include hypothyroidism, increased intracranial pressure, or calcium channel blocker, beta-blocker, or digoxin toxicity.
6. B. The most common indication for pediatric pacemakers is symptomatic bradycardia. Most permanent pediatric pacemakers are transvenous. Choice of mode depends on the disease and can be programmed to sense, demand, or inhibit at the atrial or ventricular level. A CXR may reveal wire fracture or lead displacement.
7. D. In infants, PSVT is defined as a heart rate greater than 220 beats per minute. In children, PSVT is defined as a heart rate greater than 180 BPM.

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The author wishes to thank William Toepper for these questions, which have been adapted.

51 PEDIATRIC HYPERTENSION

Emily C. MacNeill

HIGH-YIELD FACTS

- Hypertension is defined as blood pressures ≥ 90 th percentile for age and height matched normal values. Stage 2 hypertension (≥ 99 th percentile) requires urgent evaluation and treatment.
- The differential diagnosis of hypertension changes with the age of the patient.
- Evaluation of the hypertensive patient should focus on the cause of hypertension, and evaluate the patient for signs of end-organ damage.
- Initiation of oral antihypertensive agents should be done in conjunction with the physician who will be following up with the patients.
- Patients with a hypertensive emergency should be aggressively treated and admitted to a pediatric nephrologist or other hypertension specialist.

MEASURING BLOOD PRESSURE

- Blood pressures must be measured in the seated and relaxed pediatric patient
- The cuff bladder width should be 40% of the circumference of the width of the arm at mid point between the acromion and the olecranon process; length should reach 80% to 100% of the circumference of the arm.
- Automated oscillometric devices can be accurate at all age ranges; abnormal readings must be repeated by manual sphygmomanometer.
- If abnormally high, measurements should be obtained from both upper and at least one lower extremity.

DEFINING HYPERTENSION

- Hypertension is a systolic (sBP) or diastolic pressure (dBp) ≥ 95 th percentile of age and height matched normal values; pre-hypertension is between the 90th and 95th percentile (Fig. 51-1).
- Stage 1 hypertension is a systolic BP or diastolic BP between the 95th and 99th percentile. Patients can be discharged with close follow-up.
- Stage 2 hypertension systolic BP or diastolic BP ≥ 99 th percentile. Requires urgent evaluation and treatment.
- Hypertensive urgency is present when there is an elevated blood pressure with *potential* to cause end-organ damage *without* evidence of damage.

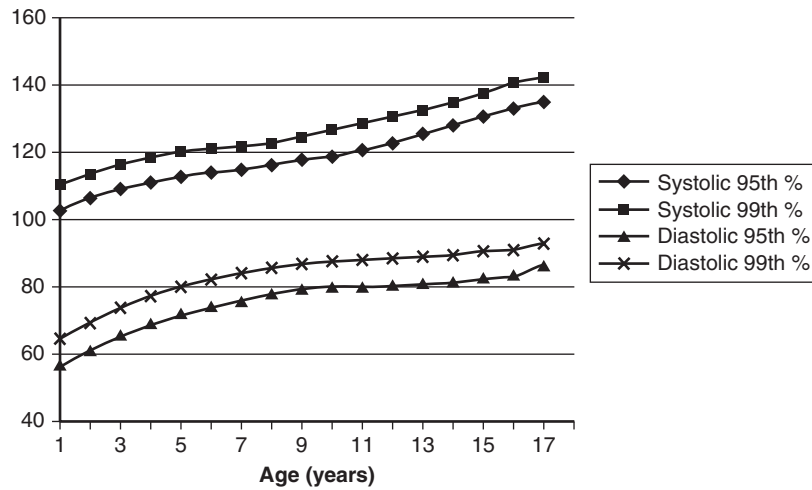


FIG. 51-1. Hypertensive values according to age. From the Fourth Report on the Diagnosis, Evaluation, and Treatment of High Blood Pressure in Children and Adolescents from 2004. Data points are taken from males at the 50th percentile for height. Females and smaller children will have slightly lower blood pressure thresholds.

- Hypertensive emergency is present when a patient has an elevated blood pressure with end-organ damage.
- Secondary hypertension is common in children; these children often present with stage 2 hypertension and signs of end-organ damage (encephalopathy, congestive heart failure, etc).
- There is an increase in the number of patients with essential hypertension, especially with positive family histories and elevated body mass index.

DIFFERENTIAL DIAGNOSIS

- The differential diagnosis for the etiology of secondary hypertension in the pediatric patient is broad (Table 51-1) and age dependent (Table 51-2).

ASSESSMENT

HISTORY

- Ascertain history of hypertension and medication use for this problem; sudden withdrawal of medication can lead to a pathologic increase in blood pressures.
- Obtain medication history of both prescription and recreational drugs, especially oral contraceptives, steroids, cocaine, and amphetamines.
- Ask about birth history, especially umbilical artery catheterization and chronic lung disease.

TABLE 51-1 Etiology of Secondary Hypertension in Children

Renal	Endocrine
Glomerulonephritis	Hypercortisol states
Obstructive uropathy	Adrenal dysfunction
Hemolytic uremic syndrome	Hyperparathyroidism
Renal tumors	Hyperthyroidism
Pyelonephritis	Toxicologic
Acute renal failure	Oral contraceptives
Congenital malformation	Steroids (corticosteroids and anabolic)
Renovascular	Stimulants (amphetamines, ephedrine)
Cardiovascular	Heavy metal exposures
Coarctation of aorta	Cyclosporine
Aortic insufficiency	
Vasculitis	
Neurologic	
Trauma	
Increased intracranial pressure	
Guillain–Barre syndrome	

TABLE 51-2 Common Etiology of Hypertension in Children According to Age

AGE	ETIOLOGY
Infancy	Congenital renal disease, aortic coarctation, renal artery/vein thrombosis
Early childhood	Renal parenchymal disease, aortic coarctation, renal vessel disease
School age	Renal parenchymal disease, essential hypertension, renal vessel disease
Adolescent	Essential hypertension, renal parenchymal disease

- Inquire about symptoms of renal disease, specifically gross hematuria, edema, generalized fatigue, and recent infections.
- In the obese patient, inquire about sleep disturbance, as sleep-disordered breathing is associated with hypertension.
- Inquire about symptoms of elevated blood pressures: chest pain, exertional dispend, palpitations, headaches, visual disturbances, and in more severe cases, altered mental status and convulsions.

PHYSICAL EXAMINATION

- Blood pressure measurements should be performed in both upper extremities and at least in one lower extremity. Leg pressures should measure at least 10 to 20 mm Hg higher than arm pressures.
- Tachycardia can point toward an endocrine etiology, whereas bradycardia can signify increased intracranial pressure and impending herniation.

- The most important part of the physical examination is the neurologic examination (including fundoscopy) as altered mental status can be a cause or a result of elevated pressures.
- Cardiovascular system exam: check pulses in all four extremities, evaluate for murmurs and gallops.
- Look for signs of heart failure (pulmonary edema or hepatomegaly), renal disease (peripheral or periorbital edema), endocrine pathology (striae, flushing, acne, and hirsutism) and urogenital abnormalities (ambiguous genitalia).

TESTING

- Refer to Fig. 51–2.
- An electrocardiogram, CBC, basic metabolic panel, urinalysis and urine culture should be done on every patient with blood pressures > 95th percentile prior to deciding disposition.

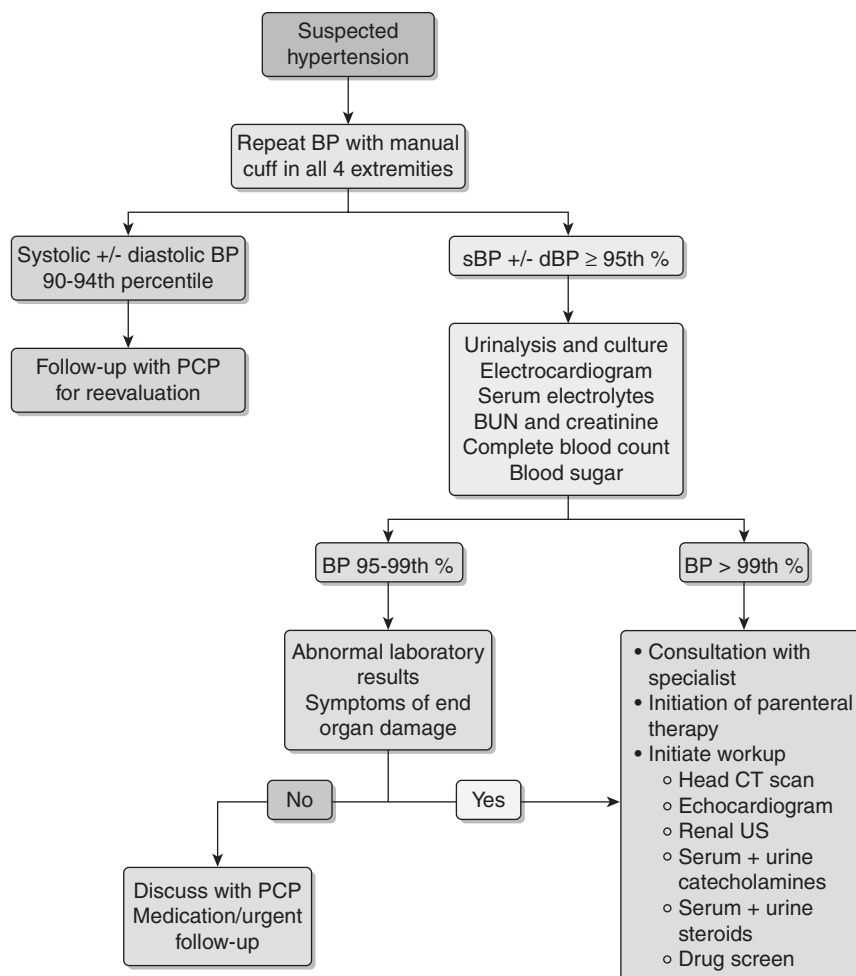


FIG. 51-2. Algorithm for the assessment of pediatric hypertension.

TABLE 51-3 Medications Useful in Hypertensive Emergency

ROUTE	DRUG	DOSE	ONSET OF ACTION	COMMENTS
Continuous IV drip	Nitroprusside	0.3–8 µg/kg/min	Seconds	May cause increased ICP Contraindicated with asthma; avoid use with congenital heart disease/heart failure
	Labetolol	5–3.0 mg/kg/h ^a	2–10 min	
IV bolus	Esmolol	50–300 µg/kg/min ^b	Seconds	Avoid with asthma
	Nicardipine	0.5–3.0 µg/kg/min	2–5 min	Caution with increased ICP
	Hydralazine	0.1–0.5 mg/kg/dose (max. 20 mg/dose)	5–30 min	May cause reflex tachycardia
	Enalapril	5–10 µg/kg/dose	15–60 min	Do not use with bilateral renal artery stenosis

^aAn initial bolus of 0.2–1 mg/kg can be given followed by 0.25–1.5 mg/kg/h drip. Labetolol can also be given at 0.2–1 mg/kg in single doses (with a maximum of 20 mg/dose).

^bA loading dose of 100–500 µg/kg is used to start this medication.

- If a child is to be admitted, start the work-up by obtaining the following tests: renin levels, plasma and urine steroids, plasma and urine catecholamines.
- Drug screening, heavy metal levels, head CT and echocardiogram should be ordered if appropriate and guided by the history and physical examination.

TREATMENT

GOALS

- Refer to Table 51-3 for doses and side effects for each parenteral anti-hypertensive agent and Table 51-4 for enteral agents.
- Parenteral medications should be reserved for patients with hypertensive emergencies with a goal of slow reduction of pressure so as not to underperfuse vital organs.
- Decrease blood pressure by 25% over the first 8 hours; then titrate medications to normalize blood pressure over the next 26 to 48 hours.

- Children with stage 2 hypertension who have no evidence of end-organ damage can be started on oral antihypertensive agents.

INDIVIDUAL AGENTS

SODIUM NITROPRUSSIDE

- Mechanism of action: dilates arterial and venous side of the circulation.
- Pharmacokinetics: Quick onset of action and short half-life.
- Dosing: start at 0.3 to 0.5 µg/kg/min with a maximum of 8 µg/kg/min.
- Side effects: increase in intracranial pressure
- Complications: nitroprusside is metabolized into cyanide, which is metabolized into thiocyanate by the liver and then excreted by the kidneys. Prolonged use, and/or higher doses in small children can lead to cyanide poisoning. Thiocyanate levels need to be followed in prolonged administration and the drug should be discontinued after approximately 48 hours.

TABLE 51-4 Oral Agents for the Management of Pediatric Hypertension

AGENT	INITIAL DOSE (mg/kg/dose)	MAXIMUM DOSE PER DAY, (mg/kg)/ MAX mg	FREQUENCY
ACE Inhibitors			
Captopril	0.3–0.5	6	tid
Lisinopril	0.07	0.6/40	qid
Angiotensin-receptor blocker	0.7	1.4/100	qid
Losartan			
Diuretics			
Furosemide	0.5	6	qid–bid
Hydrochlorothiazide	1	3/50	qid
Spirolactone	1	3.3/100	qid–bid
Calcium channel blocker			
Nifedipine	0.25–0.5/d	3/120	qid–bid
β-blocker			
Atenolol	0.5–1	2/100	qid–bid
Propranolol	1–2	4/640	bid–tid

Because of its metabolic pathway, Use with caution in children with renal insufficiency or liver impairment.

α OR β BLOCKADE

- *Labetolol* (both an α - and a β -blocker)
 - Mechanism of Action: peripheral vessel smooth muscle relaxation
 - Pharmacokinetics: half-life of 3 to 5 hours; overshooting the desired blood pressure decrease is not easily reversed.
 - Contraindications: children with asthma or chronic lung disease and children with congenital heart disease due to negative inotropy.
- *Esmolol* (selective β -blocker)
 - More cardioselective; some recommend its use in children with congenital heart disease
- *Prazosin and clonidine* (selective α blocker)
 - Can be used in the setting of asthma or chronic lung disease.

CALCIUM CHANNEL BLOCKADE

- Nicardipine
 - Administered intravenously and can be titrated carefully with rapid onset of action and short half-life.
- Nifedipine
 - Oral agent available in liquid-filled capsules and is not used often in acute settings. The capsule can be opened and administered sublingually as a last resort for children with hypertensive emergency and no IV access.
 - Difficult to dose and can cause excessive drop in blood pressure.
 - Short half-life with potential for rebound hypertension.
 - Better used for long-term management of hypertension in its extended release form.

ACE INHIBITORS

- Mechanism of action: inhibits angiotensin-converting enzyme (ACE) that converts renin to angiotensin; results in vascular relaxation and decrease in blood pressure.
- Oral forms are both efficacious and safe in pediatric populations.
- Intravenous version of enalapril (enaloprilat) can theoretically be used for pediatric hypertensive emergency (most effective with children who have high renin states).

HYDRALAZINE

- Mechanism of Action: pure arteriolar dilatation.
- No more effective than sodium nitroprusside, with a less predictable effect on blood pressure, and can also cause significant tachycardia.

DIURETICS

- Mechanism of action: decreasing intra- and extravascular volumes.
- The body becomes accustomed to this effect, leading to medication resistance.
- Commonly used agents: loop diuretics, thiazides, and potassium-sparing diuretics, and all are safe to use initially.
- Side effects include electrolyte imbalances that should be evaluated in patients who take them for prolonged periods of time. Both loop diuretics and thiazides can cause hypokalemia and hyperlipidemia; loop diuretics also cause hypercalcemia and thiazides can cause calciuria.

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QUESTIONS

1. A 10-year-old obese male presents to the emergency department with a chief complaint of “ankle sprain.” As you review the chart you see that his blood pressure is 125/85. The next most appropriate step is
 - A. Address his ankle sprain and have him follow-up for his blood pressure
 - B. Check electrocardiogram, blood and urine studies

- C. Start the patient on hydrochlorothiazide
D. Check manual blood pressure and check in both upper and one lower extremity
E. Perform a fundoscopic exam
2. A 15-year-old girl is brought to the emergency department for evaluation of vomiting and diarrhea for 3 days. Her triage blood pressure is 130/90. Her other vital signs are normal. In addition, her physical examination is normal. Which of the following substances can cause hypertension in pediatric patients?
A. Baclofen
B. Marijuana
C. Oral contraceptive pills
D. NSAIDs
E. Fluoroquinolones
3. A 7-year-old boy with a history of pharyngitis is brought to the Emergency department by his father for vomiting and decreased urinary output. He is currently on no medications. His blood pressure is 145/99. He is unable to provide any urine. Which of the following is an optimal treatment for a child with new diagnosis of hypertensive emergency?
A. Hydralazine
B. Esmolol
C. Nitroprusside
D. Furosemide
E. Enalaprilat
4. A 5-year-old girl presents with a chief complaint of “blood in urine.” Her blood pressure is 120/80. She is otherwise asymptomatic. Her ECG is normal and her creatinine is 0.5. This is an example of:
A. Primary hypertension
B. Hypertensive urgency
C. Hypertensive emergency
D. Stage 1 hypertension
E. Normotensive
5. A 12-year-old obese boy is referred to the ED for an elevated blood pressure. The patient was being evaluated in the clinic for ear pain when found to have a blood pressure of 140/85. The patient has no complaints. Which of the following is true regarding the measurement of pediatric blood pressures
A. The cuff bladder width should be 40% of the circumference of the proximal third of the distance between the acromion and olecranon process
B. The cuff bladder length should be 60% of the circumference of the midpoint of the arm between the acromion and olecranon process
C. Automated oscillometric devices are accurate even in the abnormal ranges
D. All abnormal pressures need to be repeated in a supine and standing position
E. All abnormal pressures need to be repeated and confirmed in all four extremities with a manual cuff
6. A 14-year-old obese male patient with known primary hypertension presents to the ED with headaches. He is well appearing and describes the headaches as intermittent. His blood pressure (confirmed by manual measurement) is 170/95. He takes lisinopril for his blood pressure. What is the most likely cause of his increased blood pressure?
A. Pain from headaches
B. Subarachnoid hemorrhage
C. Insufficient dosing due to weight gain and growth
D. Medical non-compliance
E. Suboptimal dosing due to his body becoming accustomed to the lisinopril
7. Which of the following patients can be sent home after evaluation for *hypertension* in the emergency department?
A. 1-year-old girl born at 30 weeks gestation with a history of an umbilical artery catheter who presents with upper respiratory infection symptoms and has a blood pressure of 120/60
B. 14-year-old obese female who presents with exertional dyspnea and has a blood pressure of 160/70
C. 6-year-old male who presents with a supracondylar fracture and has an initial blood pressure of 120/60; repeat after analgesics and splinting is 90/40.
D. 12-year-old who presents with abdominal pain and has a blood pressure of 150/100
E. 10-year-old who presents with contact dermatitis who is found to have a blood pressure of 120/80
8. Which of the following is a correct association between blood pressure medication and potential complication?
A. Esmolol: bronchoconstriction
B. Nicardipine: oral formulation only
C. Hydralazine: bradycardia
D. Nitroprusside: increased intracranial pressure
E. Labetolol: short half-life

ANSWERS

1. D. While practitioners accustomed to taking care of adult patients might glance over the blood pressure, this level lies in the 95th to 99th percentile for age. Merely having the patient follow-up is not the most appropriate step. The patient will likely require an initial work-up for that pressure: EKG, CBC, BMP, and urine studies. The first step, however,

is to ascertain that this elevated reading is correct. Treating pain in this case might be important as blood pressures rise in the setting of pain. The elevated reading must be confirmed by a good manual pressure. While this is being done, pressures can be obtained in the other extremities.

2. C. The progesterone component of birth control pills is known to cause hypertension although usually it yields a mild increase in diastolic pressure. Nonsteroidal anti-inflammatory medications, though excreted through the kidneys, have not been shown to cause hypertension. Baclofen, a drug used to treat hypertonicity in patients with cerebral palsy does not cause hypertension unless a patient experiences sudden withdrawal from the drug. There is some controversy over the effect of marijuana on blood pressure, but it has not been shown to cause hypertension. Fluoroquinolones are not indicated for use in children but not because of any effects on blood pressure.
3. C. Although all of the above agents are effective at acutely decreasing blood pressure, nitroprusside is usually the drug of choice due to its easy titratability and few contraindications. The only real downside of nitroprusside is that it can't be used long term due to toxic metabolites. The other drugs have clinical situations where they may work better (ie, esmolol in the setting of congenital heart disease); however, nitroprusside is effective in these patients as well and allows for more careful titration for safe reduction of blood pressure. Enalaprilat is more effective in children with high renin states and may not be effective for all pediatric hypertensive emergencies. Furosemide can be helpful in high volume states but can also lead to electrolyte abnormalities and is not as titratable. Hydralazine is frequently used; however it can lead to reflex tachycardia and does not have as predictable of a dose response.
4. B. This child is hypertensive for her age. In fact, her pressure is above the 99th percentile for age implying that she has stage 2 hypertension. Hypertensive emergency means that she has evidence of end-organ damage. With no ventricular hypertrophy or "strain" on the ECG, she has no evidence thus far of heart damage. She has no evidence of renal damage with a normal creatinine and she is otherwise asymptomatic. She does have hematuria which likely means that she may have developed glomerulonephritis which is causing her high blood pressure.
5. C. The blood pressure cuff selected to measure blood pressure in the pediatric patient should have a width of 40% and a length of 80–100% of the circumference of the upper arm midway between the acromion and the olecranon process. The automated blood pressure devices are usually quite accurate, even outside of the normal ranges. That being said, all abnormal readings should be confirmed with manual sphyngomanometry. Blood pressures do not need to be measured in the supine and standing positions; this is useful for diagnosing orthostatic hypotension. Abnormal blood pressures should be repeated in both upper extremities and one lower extremity as this rapidly evaluates for clinically significant aortic coarctation.
6. D. One of the common issues in management of chronic disease in adolescents is medical non-compliance. Fear of being different, denial, and rebellious behavior play some role in this but non-compliance (a problem with people of all ages) is also due to lack of side-effects and thus lack of perceived illness when medication is missed. A well-appearing adolescent is unlikely to be hypertensive because of pain, although this can quickly be ruled out by treating the headache with acetaminophen and evaluating the blood pressure after the pain has resolved. Subarachnoid hemorrhage in a well-appearing adolescent with intermittent headaches is incredible unlikely. Although younger children need constant dose adjustments to keep their medicine therapeutic during periods of growth, this is less likely than non-compliance to be the cause of his hypertension. The body can develop tachyphylaxis to certain medications and decrease their effectiveness. This is a problem with diuretic medications but is not as much with ACE-Inhibitors.
7. E. Only the patient in the last scenario has a blood pressure in the 95th to 99th percentile. These patients can undergo a brief evaluation in the emergency department and can follow up with their primary care provider or a nephrologist. Despite knowing that the hypertension in the 1 year old with a history of prematurity likely has hypertension due to her umbilical artery catheter, her blood pressure is much too high to send home with follow-up. The same is true for the 14 year old who likely has secondary hypertension. The 6 year old likely had an elevated blood pressure due to pain as is evidenced by the resolution of the hypertension with analgesics. The 12 year old with abdominal pain has a pressure above the 99th percentile and needs further evaluation.
8. D. Nitroprusside causes increased intracranial pressure and should be used with caution in patients with hypertensive encephalopathy or head injury. Esmolol is a selective β -blocker and has fewer effects on the airways. Thus it is safe to use this drug in children with asthma. Nifedipine, like nitroprusside, is available in a titratable drip. It is nifedipine that is only

available in an oral formulation. Hydralazine is known for causing a reflex tachycardia. Labetolol has a long half-life which is why it is not often used in a continuous infusion; it is too easy to “overshoot” the desired blood pressure.

52 THROMBOEMBOLIC DISEASE

Lee S. Benjamin

HIGH-YIELD FACTS

THROMBOEMBOLIC DISEASE

- Deep vein thrombosis (DVT) is the most common cause of thromboembolism (TE) in children (Figure 52-1). Pulmonary embolism (PE) represents only 8% of pediatric venous thrombotic events (VTE) (Figure 52-2).
- Arterial thromboembolism (ATE) is the most rare thromboembolic event, found mostly in neonates and children with cardiac disorders, likely due to the

use of umbilical artery catheters, cardiac catheters, ECMO circuits, and valvular disease.

RISK FACTORS FOR THROMBOEMBOLISM

- Risk factors for developing VTE assume two primary forms: inherited and acquired.
- Inherited thrombophilias such as protein C and S deficiencies, antithrombin deficiency, and the presence of lupus anticoagulant are considered high-risk states, with factor V Leiden disease, prothrombin mutation, elevated factor VIII, hyperhomocysteinemia, elevated lipoprotein (a), dysfibrinogenemia, and hypo/dysplasminogenemia considered slightly lower in risk.
- Healthy children with a single thrombophilic trait rarely present with TE, but the risk increases with multiple thrombophilic traits or with the addition of acquired risk factors.
- The most consistent acquired risk factor for development of VTE is central venous catheter placement.
- Among medical conditions, one of the most concerning risk factors for TE is cancer. Both acute leukemia and sarcoma carry high risk of VTE.

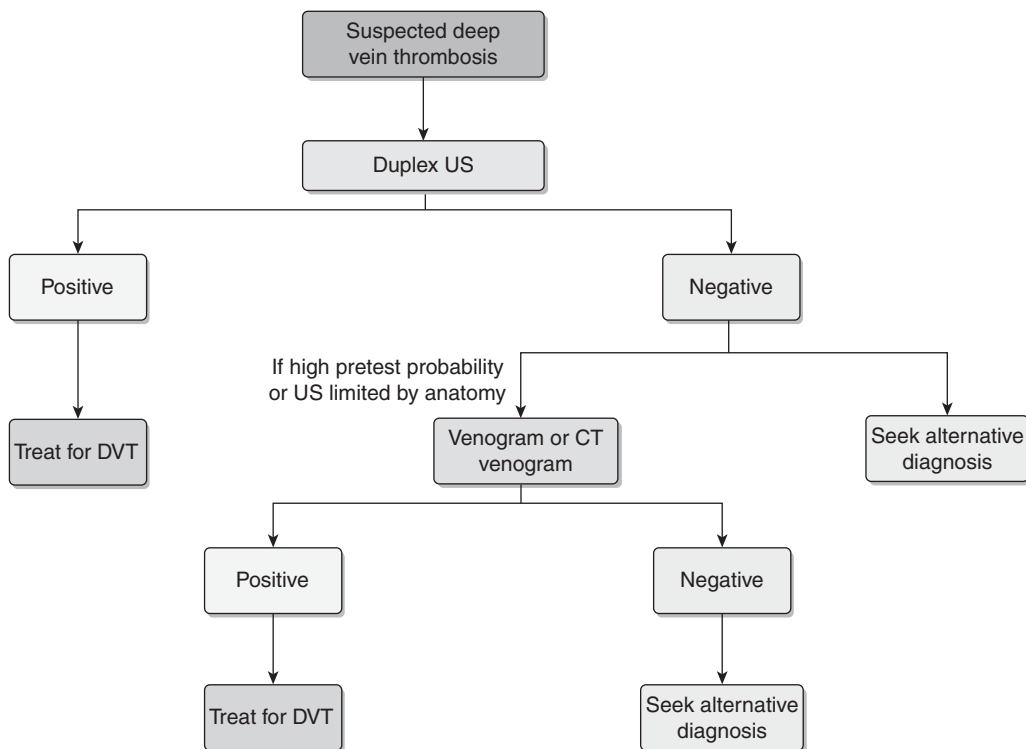


FIG. 52-1. Diagnostic algorithm for deep vein thrombosis. DVT, deep vein thrombosis; US, ultrasound; CT, computed tomography.

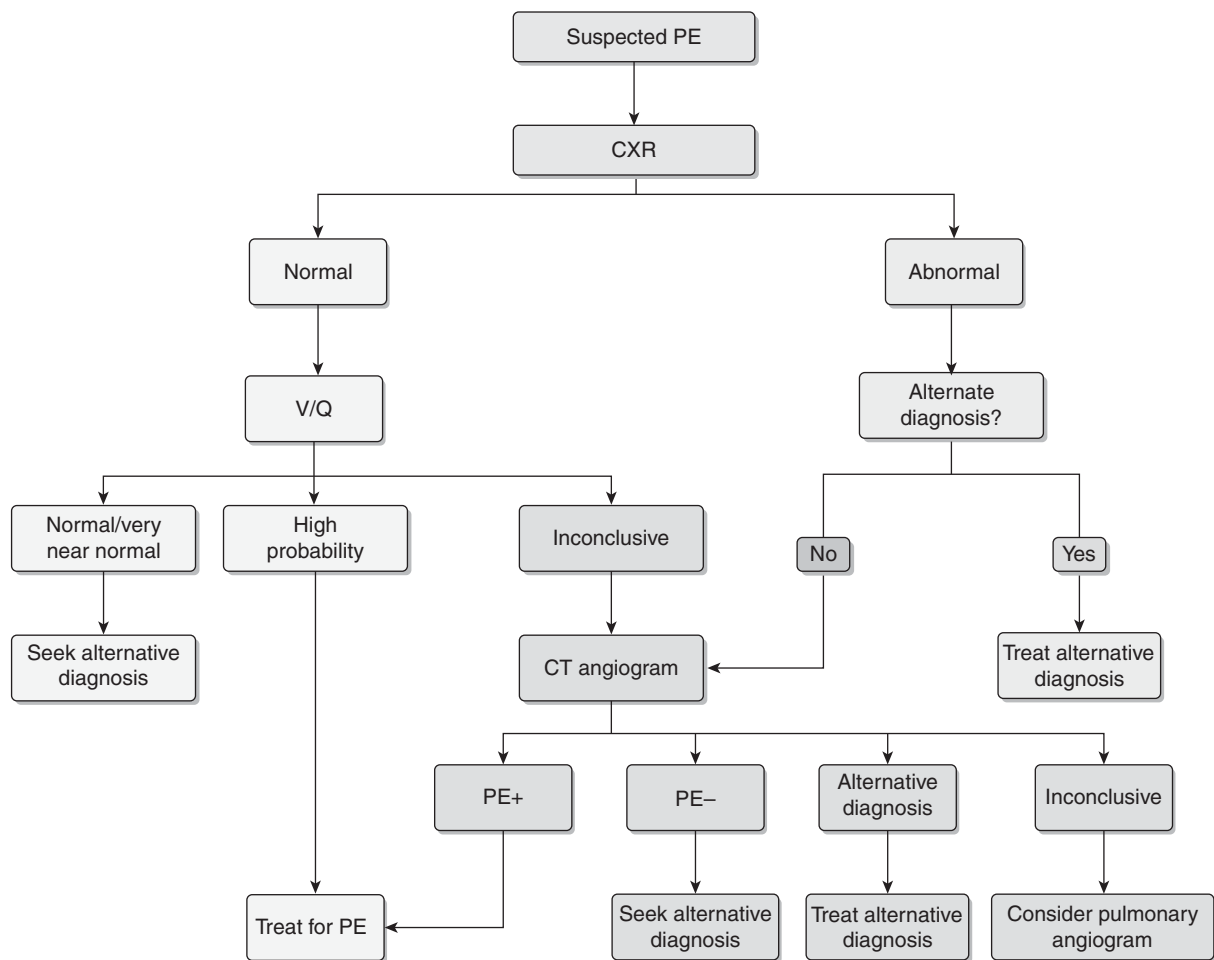


FIG. 52-2. Diagnostic algorithm for pulmonary embolism. PE, pulmonary embolism; CXR, chest radiograph; V/Q, ventilation-perfusion scan; CT, computed tomography.

- Trauma has anecdotally been considered a risk factor for TE. However, when specifically studied, VTE appears quite rare in pediatric trauma. Incidence rate for all children younger than 17 years admitted to a trauma center after injury was only 0.06%.

ARTERIAL THROMBOEMBOLISM

- Previously healthy children with no underlying risk factors rarely present with arterial thromboembolism (ATE) although 22% of pediatric patients developing ischemic stroke have no identifiable underlying risk factors.
- Among critically ill children, the vast majority of ATE is catheter related either secondary to peripheral catheter use or cardiac catheterization.
- Noncatheter-related ATE occur in patients with underlying hematologic risk factors similar to those

correlated with VTE, yet also include organ transplantation and vasculitides such as Kawasaki disease and Takayasu arteritis.

- Complications of ATE include stroke, limb loss, and dysfunction of the involved distal organs.

VENOUS THROMBOEMBOLISM

CLINICAL PRESENTATION

- DVT presents as pain, swelling, and erythema of the involved extremity. The differential diagnosis includes musculoskeletal injury, tumor, infection, arteriovenous malformation, and cystic lesions in the lower extremity.
- In contrast to adults, DVTs in children commonly occur in the upper venous system correlating strongly with the common locations of central venous catheters.

- The clinical presentation of PE varies widely. Even when well studied in adults, a significant proportion of patients lack the characteristic clinical findings of pleuritic chest pain, shortness of breath, tachycardia, tachypnea, hypoxia, and signs or symptoms of DVT.

LABORATORY TESTING

- Baseline CBC, BMP, PT, PTT, ABG, and type and screen should be considered for a patient with suspected VTE requiring treatment.
- Serum D-dimer testing has not been validated in children and up to 40% of pediatric patients with proven VTE have negative assays.

IMAGING

- Ultrasound (US) is the most commonly accepted initial imaging modality for DVT.
- Ultrasound sensitivity drops in upper extremity DVT due to poor visualization of the vasculature due to skeletal structures. Venography, CT venogram, MR venogram, and echocardiogram, should be considered if US is unable to visualize the potential thrombosis.
- Use of both perfusion lung scans (V/Q) and computed tomography (CT) imaging may lead to the diagnosis of PE. No studies document the efficacy of these tests in children; however, V/Q scans with perfusion defects should be assumed to be PE as most pediatric patients lack chronic pulmonary diseases.
- If a chest radiograph is void of significant disease, and the patient is able to comply, V/Q could be the imaging study of choice for PE. If the study is nondiagnostic, further imaging with CT should be obtained.
- Pulmonary angiography, the previous standard diagnostic tool, remains an option as well.
- US remains the standard imaging modality for evaluating renal vein, inferior vena cava, and right atrium thrombosis.

TREATMENT

- The mainstay of treatment in thromboembolic disease is anticoagulation to prevent further clot formation.
- Anticoagulation is achieved acutely with unfractionated heparin (UH) or low-molecular weight heparin (LMWH), followed by long-term anticoagulation with either LMWH or warfarin.
- Heparin dosing in children is not well studied; however, infants appear to require higher doses per unit body weight than do older children due to

physiologically low levels of antithrombin. Older children who have acquired antithrombin deficiency usually require higher doses of heparin as well.

- Thrombolytics are effective in the pediatric population, but not without risk at higher doses. Tissue plasminogen activator (tPA) can be given continuously for the treatment of arterial thrombosis, extensive DVT, or massive PE. The coadministration of heparin is necessary when using tPA, as tPA does not inhibit clot propagation or alter hypercoagulability.

COMPLICATIONS

- Mortality directly attributable to VTE has been reported as 2% to 9%.
- Recurrent thrombosis occurs in 8% to 18.5%.
- Postphlebotic syndrome develops in 12% to 21%.
- The complication of heparin-induced thrombocytopenia (HIT) is reported primarily in children undergoing anticoagulation with UH, yet there are case reports of children developing HIT while using LMWH as well.

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QUESTIONS

- A previously healthy 16-year-old female with dysmenorrhea presents to your emergency department complaining of shortness of breath and pleuritic chest pain. Which of the following statements best describes this patient's potential risk factors for venous thromboembolism?
 - Cancer
 - Central line
 - Familial thrombophilia
 - Oral contraceptive use
 - Trauma
- A 12-year-old patient under therapy for osteosarcoma presents to the emergency department complaining of a swollen and painful leg. On examination, he is well appearing, afebrile, and vital signs are appropriate for age. The left calf and lower thigh is erythematous and tender. Which of the following statements describes the most appropriate initial evaluation for suspected deep vein thrombosis in this patient?
 - CT venogram
 - Impedance plethmography
 - MR venogram
 - Ultrasound
 - Venogram
- A 4 year old with leukemia presents from the oncology clinic to rule out DVT. Which of the following statements is most correct regarding utility of lab testing in this patient?
 - D-dimer testing is recommended to rule out hypercoaguable state in the pediatric population
 - D-dimer testing is not recommended to rule out hypercoaguable state in the pediatric population
 - Factor Xa testing is recommended to rule out hypercoaguable state in the pediatric population
 - Fibrinogen testing is recommended to rule out hypercoaguable state in the pediatric population
 - PT and PTT testing is recommended to rule out hypercoaguable state
- A 7 year old is transferred by EMS to your emergency department after a motor vehicle collision. He has a tibia fracture and pulmonary contusion. Which of the following statements is most correct regarding this patient?
 - Anticoagulation should be initiated upon admission
 - Immobilization in admitted pediatric trauma patients leads to high risk of deep vein thrombosis
 - Pulmonary embolism is the most common thromboembolic event in the pediatric population
 - Tibia fractures carry high risk for venous thromboembolism
 - Venous thromboembolism is uncommon in pediatric trauma
- An 8-year-old female presents with a complaint of weakness of the left upper and lower extremity. On examination she has complete left sided hemiparesis. Which of the following is true regarding stroke in children?
 - Arterial stroke usually occurs in children with known risk factors
 - Catheter use is not considered a risk factor for stroke in children
 - Immobilization is a risk factor for stroke
 - Venous thromboembolism is a risk factor for stroke
 - Trauma is a risk factor for stroke in children
- A 16-year-old postpartum female presents to your emergency department complaining of sudden onset chest pain and difficulty breathing. Which of the following studies is the most appropriate imaging study to identify a pulmonary embolism in your patient?
 - Angiogram
 - CT angiogram
 - MR angiogram
 - Ultrasound of the lower extremity
 - Ultrasound of the thorax
- A 12-year-old patient with sickle cell disease has an indwelling line in the left upper extremity. He presents with swelling of the extremity for three days. Which of the following is true regarding the diagnosis of upper extremity DVT?
 - Echocardiogram of the chest is the most appropriate initial test
 - MR venogram of the extremity may assist in diagnosis
 - Ultrasound is the gold standard of diagnosis
 - Venogram of the extremity is the most appropriate initial test
 - VQ scan of the chest may assist in diagnosis
- A 10 year old is diagnosed with a deep vein thrombosis. Which of the following is the most appropriate initial treatment option?
 - Clopidogrel
 - Dalteparin
 - Heparin
 - Tissue plasminogen activator
 - Warfarin
- A 12 year old is sent to your emergency department for calf swelling, erythema, and tenderness,

concerning for DVT. Which of the following are considered mimics of lower extremity DVT?

- A. Arterial thromboembolism
- B. Baker cyst
- C. Endocarditis
- D. Erythema nodosum
- E. Henoch–Schönlein purpura

ANSWERS

1. D. Although cancer, central line placement and familial thrombophilia are all risk factors for developing venous thromboembolism, there is no history in this patient consistent with these risk factors. However she does have dysmenorrhea, commonly managed medically with oral contraceptive use, also a known risk factor for venous thromboembolism. Trauma is rarely considered a risk factor in the pediatric population, and although this 16 year old is approaching adult physiology, there is no mention of trauma preceding her symptoms.
2. D. All of the other studies have utility in making the diagnosis of deep vein thrombosis, and may be indicated if ultrasound is negative. However due to technical limitations and availability, ultrasound remains the standard.
3. B. D-dimer has not been well studied in children, and has reported to be negative in 40% of patients with documented deep vein thrombosis. Therefore it should not be used in the pediatric population to rule out venous thromboembolism. Factor Xa assesses for low molecular weight heparin efficacy in anticoagulation, and is not used as a diagnostic tool PT and PTT testing are recommended in a patient if anticoagulation is to be started, but cannot rule out hypercoagulable state. Abnormalities of fibrinogen may cause either bleeding or thrombosis, but routine testing is not recommended in evaluating for venous thromboembolism.
4. E. Contrary to adult patients, pediatric trauma patients rarely develop venous thromboembolism, especially if young. Specific characteristics that carry higher risk in children include spinal cord injury, major vascular injury, older age, central line placement, and operative interventions. Of all venous thromboembolic events in all pediatric patients, deep vein thrombosis is the most common.
5. A. The majority of strokes in children occur in patient with known underlying disease or recent catheterization. Although they share common risk factors, venous thromboembolism does not predispose to arterial disease. Neither immobilization nor trauma is considered high risk factors for stroke.
6. B. The initial studies of choice in evaluating for pulmonary embolism include either a ventilation perfusion scan or a CT angiogram of the pulmonary vasculature. The latter has the advantage of identifying other thoracic pathology, yet may expose the patient to higher radiation doses. Pulmonary angiogram has been considered the gold standard of diagnosis, yet carries technical challenges and a substantial complication rate. MRI is not sensitive in evaluating for pulmonary embolism. Ultrasound of the chest has utility limited to superior and inferior vena cava evaluation. Ultrasound of the lower extremity is the test of choice to evaluate for lower extremity deep vein thrombosis.
7. B. The initial test of choice to evaluate for upper extremity venous thrombosis is ultrasound. However the sensitivity of the test drops drastically if the venous system is obstructed by the skeletal structures of the chest wall. MR venogram, CT venogram, or venogram may all evaluate the venous system if ultrasound is unsuccessful or if there is high index of suspicion. Echocardiogram evaluates the heart and great vessels, and VQ is used to evaluate for pulmonary embolism.
8. C. Heparin, either unfractionated or low molecular weight, may be used as initial options for anticoagulation. Dalteparin is a similar medication used infrequently in the USA. Warfarin is utilized for long-term anticoagulation. Tissue plasminogen activator is a thrombolytic used to treat thrombosis in select cases only. Clopidogrel is an antiplatelet medication used in the emergency department most commonly in adults with acute coronary syndromes.
9. B. Baker's cyst is a popliteal cyst commonly found in adult patients with osteoarthritis, and may present with popliteal swelling and tenderness. Other mimickers include popliteal artery aneurysms, tumor, infection, and musculoskeletal injury. Arterial occlusion presents with signs of ischemia and pain. Endocarditis can present with findings of the hands and feet, but not with diffuse swelling of the calf. Erythema nodosum is a nodular, tender rash noted over the anterior surface of the lower extremity, not involving the calf. Henoch-Schönlein purpura is a vasculitis that commonly presents with a petechial or purpuric rash isolated to the lower extremities, however not significant swelling or tenderness.

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53 SYNCOPE

Susan Fuchs

INTRODUCTION

Syncope refers to a sudden and transient loss of consciousness and postural tone. Although in the pediatric age group it accounts for less than 1% of emergency department visits, 15% to 50% of children will have experienced a syncopal episode by age 18. Unlike the adult population, in which syncope often results from malignant cardiac arrhythmias, in the pediatric population it is more often secondary to neurally mediated causes.

PATHOPHYSIOLOGY

- The pathophysiology of syncope varies with etiology (Table 53-1), but it always results from momentarily inadequate delivery of oxygen and glucose to the brain. Syncope can result from inadequate cardiac output, which can be secondary to obstruction of blood flow, or to an arrhythmia. It can also result from inappropriate autonomic compensation for the normal fall in blood pressure that occurs on rising from a sitting or a supine position. Respiratory disturbances, especially hyperventilation that results in hypocapnia and cerebral vasoconstriction, can also cause syncope.

HISTORY

- The first component in the evaluation of a patient with syncope is to determine that momentary loss of consciousness actually occurred. A prodrome

of light-headedness, nausea, dizziness or vision changes, a sudden change in posture, emotional excitement, respiratory difficulty, palpitations, exercise, and any history of trauma are essential information. Past history of syncope, history of medication or drug ingestion, any history of congenital heart disease, any family history of heart disease, or family history of sudden death in children or young adults are obtained.

- An important consideration in any patient with a history of loss of consciousness is the possibility that the patient may have suffered a seizure. In contrast to syncope, seizures are usually accompanied by some form of muscle twitching or convulsions, and are usually followed by a postictal phase, during which the patient has confusion, disorientation, or other mental status changes, usually lasting more than 5 minutes. Convulsions are unusual during syncopal episodes except during very severe events, or with reflex anoxic seizures, and patients generally have normal mental status upon recovery from the episode.

PHYSICAL EXAMINATION

- Particular attention is paid to vital signs, especially to pulse and orthostatic blood pressure. A positive test is a decrease in systolic blood pressure by 20 mm Hg or an increase in heart rate (20 beats per minute), on going from lying to sitting, or sitting to standing.
- The patient's mental status is carefully evaluated, and a full neurologic examination is performed.
- For all patients, a careful cardiac examination is indicated. Diminished pulses should prompt blood pressure measurements in all extremities to check for coarctation of the aorta.

TABLE 53-1 Causes of Syncope

Neurocardiogenic/vasodepressor/vasovagal
Orthostatic hypotension
Environmental triggers
Sympathetic withdrawal
Situational syncope
Reflex syncope (pallid breath-holding spells)
Cardiac mediated syncope
Arrhythmias
Supraventricular tachycardias
Atrial flutter
Wolfe–Parkinson–White syndrome
Ventricular tachycardia
Ventricular fibrillation
Conduction disturbances
Atrioventricular block
Prolonged QTc
Short QTc
Brugada syndrome
Sick sinus syndrome
Arrhythmogenic right ventricular dysplasia (ARVD)
Obstructive lesions
Aortic stenosis
Pulmonic stenosis
Idiopathic hypertrophic subaortic stenosis
Hypertrophic cardiomyopathy
Mitral stenosis
Coarctation of the aorta
Tetralogy of Fallot
Anomalous origin of the left coronary artery
Tumors
Other
Myocarditis
Pericarditis
Cardiac tamponade
Cardiomyopathy
Pulmonary hypertension
Noncardiac
Metabolic
Hypoglycemia
Hypocalcemia
Hypomagnesemia
Toxic
Seizures
Psychogenic
Hyperventilation
Hysteria

Data from Chaves-Carbello E. Syncope and paroxysmal disorders other than epilepsy. In: Swaiman KF, Ashwal S, Ferriero DM, eds. *Pediatric Neurology: Principles and Practice*. 4th ed. Philadelphia, PA: Mosby/Elsevier; 2006:1209–1223.

Data from Coleman B, Salerno JC. Causes of syncope in children and adolescents. In: Rose BD, ed. *UpToDate*. Waltham, MA: UpToDate; 2007.

Data from Strieper MG. Distinguishing benign syncope from life-threatening cardiac causes of syncope. *Semin Pediatr Neurol*. 2005;12:32–38.

LABORATORY STUDIES

- The selection of laboratory studies of use in the evaluation of the syncope patient is largely guided by the history and physical examination (Fig. 53-1).

- For all patients with a history of syncope, a 12-lead electrocardiogram (ECG) is indicated. Special attention is paid to determination of the corrected QT interval (QTc), since prolonged QT syndrome, as well as a short QTc, is a cause of syncope in children. If abnormalities are seen, or if a cardiac abnormality is strongly suspected, further evaluation will include a 24-hour ambulatory (Holter) monitor or continuous loop event monitoring and cardiology consultation.

SPECIFIC ETIOLOGIES OF SYNCOPE

NEUROCARDIOGENIC (AUTONOMIC, VASODEPRESSOR, VASOVAGAL) SYNCOPE

- The most common syncope in children is neurocardiogenic (vasodepressor or vasovagal) syncope. There is a sudden brief loss of consciousness because of vasodilatation and decreased peripheral resistance, resulting in decreased arterial pressure, hypotension, bradycardia, and then decreased cerebral blood flow (Bezold–Jarisch reflex). Orthostatic hypotension may be a result of volume depletion, anemia, or drugs, but it can also be because of a paradoxical response to the vasodepressor reaction.
- A tilt-table test can be performed by a cardiologist to diagnose true neurocardiogenic syncope. A positive head upright tilt-table test response, consisting of an initial increase in heart rate followed by bradycardia and syncope, may warrant drug therapy if frequent episodes occur.
- Environmental factors, such as prolonged standing, heat, fatigue, crowding, or hunger, can trigger syncope. Emotional stress or a recent illness can also play a role. Patients may have symptoms beforehand such as blurred vision, dizziness, nausea, or pallor. This is what is commonly referred to as a “simple faint.” Placing the person in a supine position with the head down usually results in improvement although the patient may still complain of dizziness.
- The term situational syncope can be used for those patients who have syncope triggered by specific events. The common denominator is that these actions are accompanied by a Valsalva-like maneuver. This includes coughing, micturition, hairgrooming, diving, weight lifting, and sneezing. Another form is carotid sinus syncope, which occurs with head rotation or pressure on the carotid sinus. This can occur with shaving or tight collars.
- Breath-holding spells are another example of reflex syncope. The age of onset of breath-holding spells

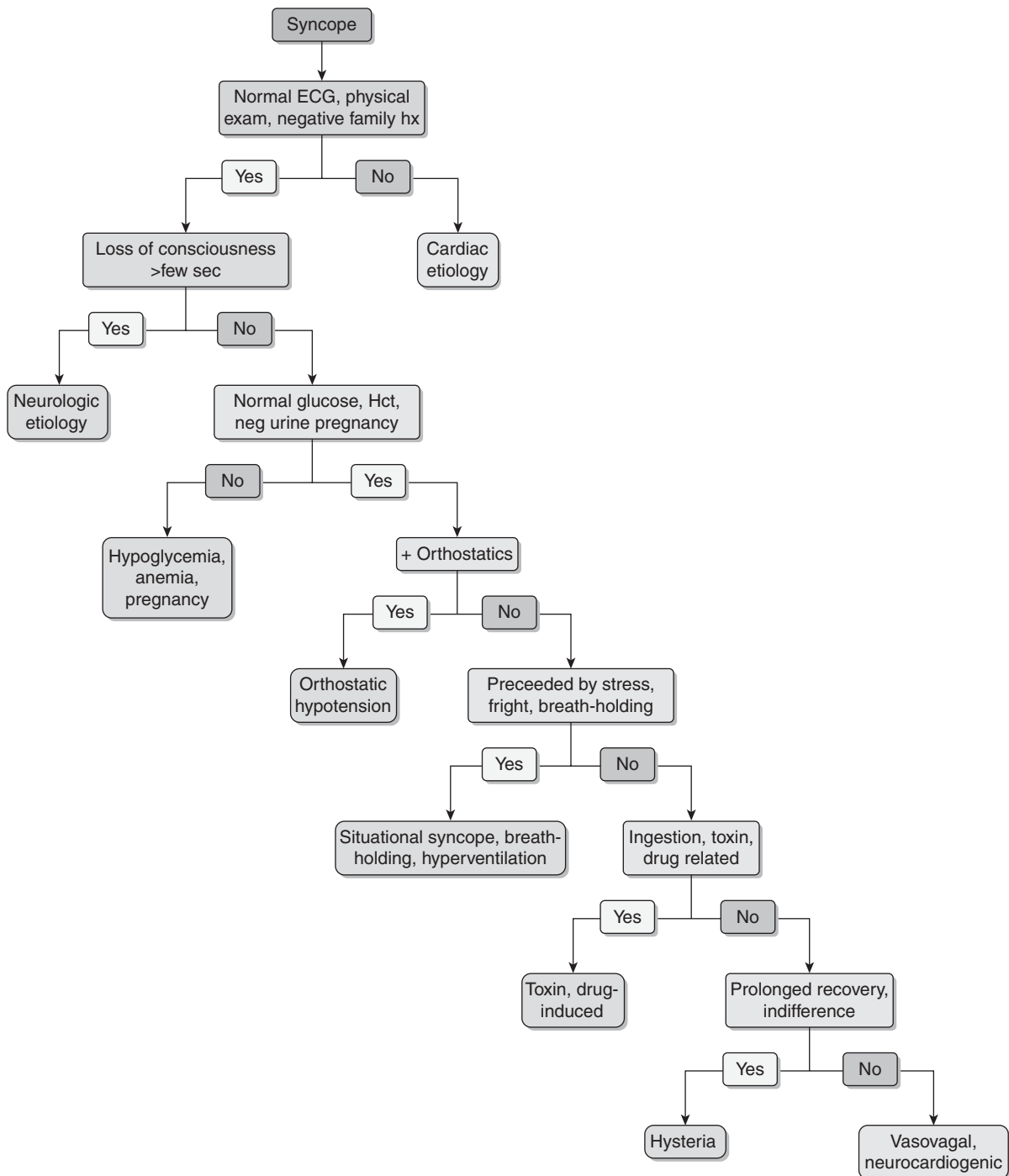


FIG. 53-1. Diagnostic approach to the patient with syncope.

is 6 to 18 months. The two types of breath-holding spells are classified based on the color change:

- The pallid breath-holding spell is a form of reflex syncope. Pallid breath-holding spells are usually provoked by some mild antecedent trauma (usually to the head), pain, or fright. The child may gasp and

cry, then become quiet, lose postural tone and consciousness, and become pale. The child may have clonic movements and incontinence in more severe episodes. The child regains consciousness in less than 1 minute, but may be tired after the episode for several hours.

- A cyanotic breath-holding spell is often precipitated by anger or frustration. The child cries, becomes quiet, and holds the breath in expiration. This apnea is associated with cyanosis and there may be a loss of consciousness, limpness, or opisthotonic posturing, with recovery usually within 1 minute.

CARDIAC SYNCOPE

- Cardiac syncope can be due arrhythmia, obstruction, cyanosis, and other cardiac etiologies.
- Arrhythmias that result in a heart rate that is too fast or too slow can cause a decrease in cardiac output and lead to decreased cerebral perfusion.
- Conduction abnormalities such as AV block, sick sinus syndrome (may occur after cardiac surgery), long QT syndrome, congenital short QT, or Brugada syndrome may be present.

LONG QT SYNDROME

- Long QT syndrome is a disorder of myocardial repolarization that can lead to polymorphic ventricular tachycardia (torsades de pointes) (Fig. 53-2). There are congenital forms: Romano–Ward syndrome (autosomal dominant) and Jervell and Lange–Nielsen

syndrome (autosomal recessive). The latter form is associated with sensorineural deafness. There are also acquired forms usually the result of the metabolic disorders such as hypokalemia and hypomagnesemia, some medications (quinidine, procainamide), or a combination of medications (erythromycin or ketoconazole and terfenadine).

- The definition of corrected QTc changes with the patient's age and sex. (QTc is >450 ms in males and >460 in females). To calculate the QTc, use Bazett's formula $QTc = QT/\sqrt{RR}$.² After evaluation by a cardiologist, treatments include β -blockers, implantable defibrillators, and avoidance of medication that prolong the QT interval.

CONGENITAL SHORT QT SYNDROME

- This syndrome has led to sudden cardiac death, syncope, and atrial fibrillation. The QTc is ≤ 0.30 ms.

BRUGADA SYNDROME

- Patients with this inherited autosomal dominant disorder have a characteristic ECG pattern: ST segment elevation (≥ 2 mm) in leads V_1 – V_3 . There is an increased risk of sudden cardiac death resulting from polymorphic ventricular tachyarrhythmias. Although it is more

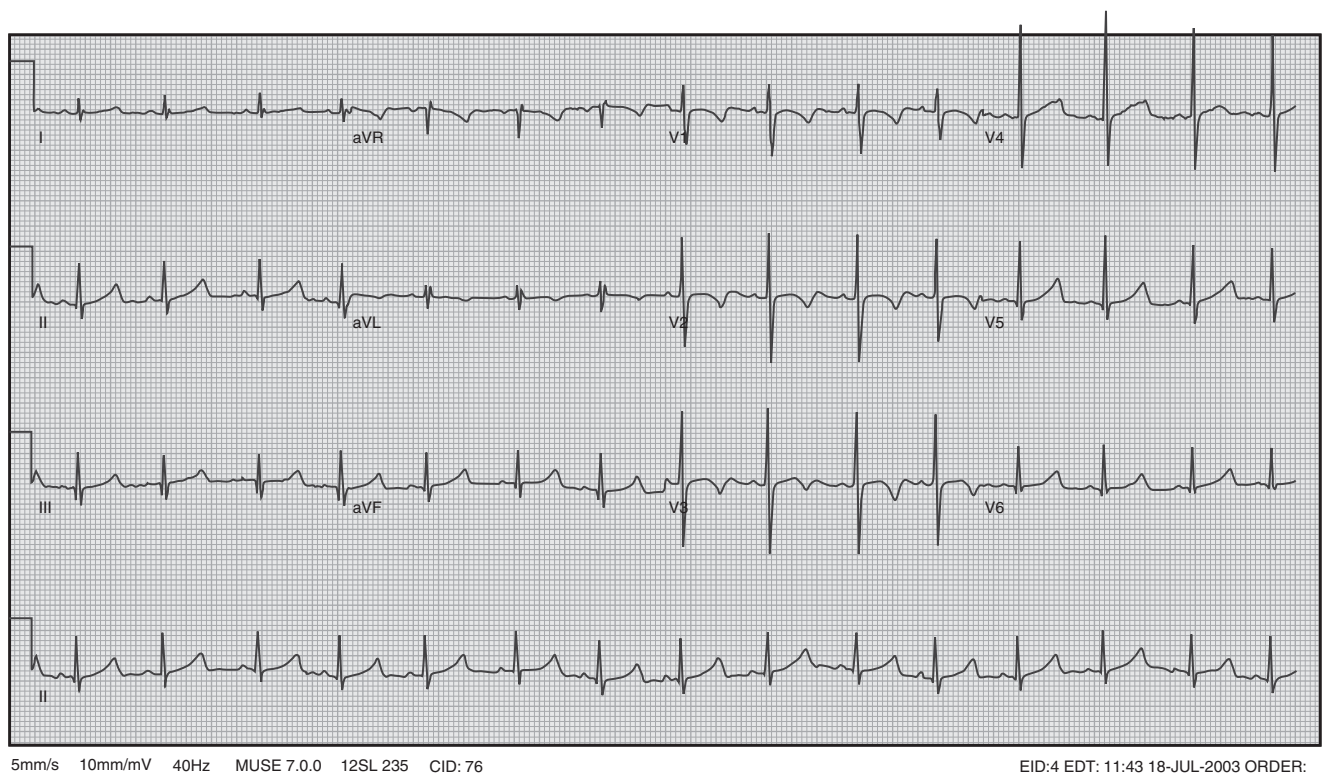


FIG. 53-2. Long QT syndrome.

likely to occur in young males, with a reported mean age of sudden death at 40 years, children have been detected with this disorder during family screening; there was no male predominance, and febrile illness was the most important precipitating event.

ARRHYTHMOGENIC RIGHT VENTRICULAR DYSPLASIA/CARDIOMYOPATHY

- Arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD) is characterized by ventricular tachycardia and abnormalities of the right ventricle caused by myocyte replacement by fibrosis or adipose tissue. Although sudden cardiac death may be the presenting feature (mean age of 30 years), PVCs, syncope, and ventricular tachycardia with left bundle branch block may be forewarning findings. Diagnosis is based on ECG findings (epsilon waves after the QRS) as well as structural and functional criteria.
- Obstructive lesions can impair cardiac output and cerebral blood flow, leading to syncope. These include congenital lesions such as aortic stenosis, pulmonic stenosis, idiopathic hypertrophic subaortic stenosis (IHSS), hypertrophic cardiomyopathy, mitral stenosis, coarctation of the aorta, tetralogy of Fallot, anomalous origin of the left coronary artery, and transposition of the great vessels. Children who have undergone surgical correction of tetralogy, transposition, and aortic stenosis are at a greater risk of arrhythmias.
- The presence of chest pain and syncope with exercise, as well as a murmur on physical examination, can suggest left ventricular outflow obstruction because of IHSS or aortic stenosis.
- Cyanosis with or without syncope can result from increased resistance to pulmonary blood flow, causing an increase in the right-to-left shunting of blood. These spells can occur in children with tetralogy of Fallot, tricuspid atresia, and Eisenmenger's syndrome.

HYPERTROPHIC CARDIOMYOPATHY

- This autosomal dominant disorder is characterized by asymmetric hypertrophy of the left ventricle, without dilatation. It is a common cause of sudden death associated with exercise in young patients. Syncope is a major risk factor for sudden death. The mechanisms that cause syncope and sudden death include bradyarrhythmias, ventricular arrhythmias, severe outflow tract obstruction, and decreased blood pressure in response to exercise. Implantable defibrillators have been used to treat this disorder with some success.
- Acquired lesions include cardiac tumors and conditions secondary to myocarditis, pericarditis, cardiac tamponade, cardiomyopathy, and pulmonary hypertension. Primary pulmonary hypertension can cause dyspnea

on exertion but can also cause syncope from inadequate cardiac output. Evaluation includes a 12-lead ECG, chest radiograph, and prompt cardiology referral.

NONCARDIAC SYNCOPE

- Seizures should be considered whenever there is a loss of consciousness, especially if accompanied by increased muscle tone or tonic-clonic movements. If syncope occurs while the child is in a recumbent position, a seizure is a likely diagnosis. The diagnostic workup should proceed based on the most likely etiology and type of seizure.
- Hypoglycemia is the main metabolic disorder that can cause syncope. Prior to a loss of consciousness there is often a period of confusion and weakness. Hypocalcemia and hypomagnesemia can also cause syncope, but this is secondary to the arrhythmias generated by these disorders.
- Psychologic causes of syncope include hyperventilation and hysteria. Hyperventilation results in hypocapnia, which causes cerebral vasoconstriction and decreased cerebral blood flow. The patient may complain of shortness of breath, chest tightness, and numb fingers before syncope ensues.
- Hysterical syncope occurs when the patient mimics a loss of consciousness and falls to the ground without injury. No abnormalities of heart rate, blood pressure, or skin color are detected, and clues regarding surrounding events may point to the correct diagnosis, such as prolonged recovery after the event, and indifference to syncope.
- Drug-induced syncope can be caused by prescription drugs, over-the-counter medications, or drugs of abuse. Inhalant use can result in ventricular tachycardia and death. Antihypertensive agents, phenothiazines, calcium channel blockers, nitrates, and barbiturates can block the increased blood pressure response, and β -blockers and digitalis will block the tachycardia needed to respond to decreased systemic vascular resistance prior to syncope. Some of the newer antihistamines can cause prolonged QT and even torsades de pointes, if given with macrolides or ketoconazole.

TREATMENT

- Although most children will not require specific therapy, treatment should be based on the etiology and frequency of syncopal episodes.
- If a cardiac etiology is suspected, depending on the urgency of the situation, further studies could be performed on an inpatient or outpatient basis, but activity restrictions may be needed while awaiting evaluation.

- Treatment of a child with orthostatic syncope with β -blockers or mineralocorticoids should await formal head upright tilt-table testing. If a neurologic etiology is suspected, treatment with anticonvulsive medications should be based on neurologic consultation.
- If the emergency department evaluation and workup is negative, with the likely etiology being neurocardiogenic, or a simple faint, reassurance may be all that is needed. Avoidance of specific triggers or environmental factors can prevent most attacks. Those with a prodromal phase can be taught to sit or lie down before the loss of tone and consciousness occurs.

DISPOSITION

- Most patients with syncope can be discharged from the emergency department with appropriate follow-up.
- Patients with a cardiac origin for syncope require urgent evaluation. Patients with arrhythmias precipitated by drugs require inpatient monitoring, for at least the half-life of the offending agent.
- Patients with cyanosis with syncope, those with abnormal neurologic examinations, and those with orthostatic syncope who do not resolve with fluids and have no other etiology should also be admitted.

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QUESTIONS

1. A 12-year-old female presents to the ED after collapsing at church. She felt hot, was nauseous, and developed blurred vision before she passed out. She was caught by her older brother and did not hit her head. She was out for 30 seconds, and felt better after lying down. In the ED her vital signs and physical exam are normal. Family history is negative for sudden death or cardiac problems. Which of the following is the most likely etiology of her syncopal episode?
 - A. Hyperventilation
 - B. Vasovagal
 - C. Situational
 - D. Arrhythmia
 - E. Hysteria
2. A 14-year-old female collapses while playing basketball in the park. She had a brief loss of consciousness and denies hitting her head. This has happened before and she ignored it. The patient says an uncle died of a heart attack at age 24. Which of the following is the most important test to perform in the ED?
 - A. Blood glucose
 - B. Pregnancy test
 - C. ECG
 - D. Chest radiograph
 - E. Echocardiogram
3. An 18-month-old boy is brought to the ED after “passing out.” Mom describes the child playing happily when she called him for dinner. He said no, and kept playing. Mom then took his toys away and he began to cry. His face turned blue and he fell to the ground. In the ED he is acting normally and his vital signs are normal. Pulse oximetry on room air is 100%. This is an example of which of the following?
 - A. Cyanotic breath-holding spell
 - B. Pallid breath-holding spell
 - C. Situational syncope
 - D. Arrhythmia
 - E. “Simple faint”
4. A 15-year-old male with known long QT syndrome, on terfenadine for allergies presents to the ED with a persistent cough and fever. On exam he has crackles in his left lower lobe, confirmed on x-ray. You suspect Mycoplasma, and feel he is stable for home therapy. Which of the following antibiotics are most appropriate for this patient?
 - A. Azithromycin
 - B. Clarithromycin
 - C. Penicillin
 - D. Amoxicillin/clavulanate
 - E. Doxycycline

5. An 8-year-old male comes to the ED after passing out at school. He ate lunch and felt fine, but in class felt his heart racing. He became sweaty and nauseous, then slumped over at his desk. On EMS arrival his heart rate was 210, and he was awake. In the ED his heart rate is 120 and regular, and the ECG is normal. The most appropriate study for this patient is which of the following?
- Blood glucose
 - Chest radiograph
 - Serum electrolytes
 - Echocardiogram
 - Holter monitor

ANSWERS

1. B. Vasovagal. The most common syncope in children is neurocardiogenic (vasodepressor or vasovagal) syncope. Environmental factors, such as prolonged standing as in church, heat, fatigue, crowding, or hunger, can trigger syncope. Patients may have symptoms beforehand such as blurred vision, dizziness, nausea, or pallor. Situational syncope can be used for those patients that have syncope triggered by specific events. The common denominator is that these actions are accompanied by a Valsalva-like maneuver. This includes coughing, micturition, hairgrooming, diving, weight lifting, and sneezing. Psychologic causes of syncope include hyperventilation and hysteria. Cardiac syncope can be due to an arrhythmia; however the patient can often tell that their heart is beating too fast.
2. C. ECG. This patient has a family history of a “heart attack” in a young adult, so sudden death is a concern. An ECG to check for long QT syndrome, short QT syndrome or Brugada syndrome should be performed. While blood sugar and pregnancy test are important, they may not be diagnostic. A chest radiograph and echocardiogram may be helpful, but may not reveal the diagnosis.
3. A. Cyanotic breath-holding spell. Breath-holding spells are an example of reflex syncope. The age of onset of breath-holding spells is 6 to 18 months. The two types of breath-holding spells are classified based on the color change. A cyanotic breath-holding spell is often precipitated by anger or frustration. The child cries, becomes quiet, and holds the breath in expiration. This apnea is associated with cyanosis and there may be a loss of consciousness, limpness, or opisthotonic posturing, with recovery usually within 1 minute. The pallid breath-holding spell is a form of reflex syncope. Pallid breath-holding spells are usually provoked by some mild antecedent trauma (usually to the head), pain, or fright. The child may gasp and cry, then become quiet, lose postural tone and consciousness, and become pale. The child may have clonic movements and incontinence in more severe episodes. The child regains consciousness in less than 1 minute, but may be tired after the episode for several hours. Situational syncope can be used for those patients who have syncope triggered by specific events. The common denominator is that these actions are accompanied by a Valsalva-like maneuver. This includes coughing, micturition, hairgrooming, diving, weight lifting, and sneezing. An arrhythmia is unlikely in this setting, especially with the antecedent history and cyanosis. Environmental factors, such as prolonged standing, heat, fatigue, crowding, or hunger, can trigger syncope. Emotional stress or a recent illness can also play a role. Patients may have symptoms beforehand such as blurred vision, dizziness, nausea, or pallor. This is what is commonly referred to as a “simple faint.”
4. E. Doxycycline. Doxycycline is the appropriate treatment. A patient with long QT syndrome already on terfenadine should not be prescribed a macrolide, such as azithromycin or clarithromycin. Penicillin and amoxicillin/clavulanate are not effective against *Mycoplasma pneumoniae*. A second line option is ciprofloxacin.
5. E. Holter monitor. Since this patient’s heart was racing fast and found to be 210 by EMS, an arrhythmia is likely. Even if it is not seen in the ED on an ECG, a holter monitor may catch a recurrence. Blood glucose would be helpful if the patient had skipped lunch. A chest radiograph and echocardiogram may be helpful, but may not be as diagnostic. Electrolytes are likely to be normal as well.

54 ATAXIA

Susan Fuchs

INTRODUCTION

- Ataxia is a disorder of intentional movement, characterized by impaired balance and coordination. It can variably affect the trunk or extremities.
- Ataxia can be congenital or acquired. Acquired ataxia is often classified as acute, episodic/intermittent, or chronic.

PATHOPHYSIOLOGY

- Ataxia can result from a variety of lesions, including damage to the peripheral nerves, spinal cord, cerebellum, and cerebral hemispheres. Lesions of the

cerebellum can be further categorized as affecting the hemispheres, which results in ipsilateral limb hypotonia, tremor, and dysmetria, but spare speech. With walking, these patients tend to veer to the side of the lesion. Lesions of the midline vermis cause truncal ataxia or titubation, dysarthria, and gait abnormalities. Damage to the spinal cord can cause ataxia when the patient stands with the eyes closed, which is referred to as a positive Romberg's sign. Patients with cerebellar ataxia have findings whether their eyes are open or closed.

- Metabolic and systemic disorders can also cause ataxia. One of the most common etiologies is drug intoxication, especially with alcohol, benzodiazepines, or phenytoin.

EVALUATION

True ataxia must be distinguished from problems with similar neurologic manifestations.

- Vestibular disorders can cause vertigo, a sensation of abnormal movement or spinning that can cause a severe gait disturbance, nausea, and vomiting. Vertigo is often accompanied by nystagmus.
- Myopathies can be confused with ataxia, as can peripheral neuropathies. On physical examination, myopathies are characterized by muscle weakness, whereas peripheral neuropathies are accompanied by decreased reflexes.
- Chorea is a disorder characterized by involuntary movements and incoordination. It is distinguished from ataxia in that it occurs at rest, whereas ataxia is manifested during intentional movement.

PHYSICAL EXAMINATION

- Findings of cerebellar dysfunction include nystagmus, staggering, wide-based gait, titubation, and speech abnormalities. Specific neurologic tests include the following:
 - Finger to nose with eyes closed (to look for intention tremor).
 - Finger to nose to (examiner's) finger; dysmetria is poor coordination of voluntary movements, and results in under- or overshooting the target (tests cerebellar integrity when limb strength and sensation is intact).
 - Heel-to-shin maneuvers (test cerebellar integrity when limb strength and sensation are intact).
 - Rapid alternating hand movements—difficulty is termed dysdiadochokinesia (tests cerebellar function).
 - Heel and toe walking (with hemispheric lesions there is a tendency to veer in one direction)

- Tandem gait (with hemispheric lesions there is a tendency to veer in one direction).
- Walking in a circle (with hemispheric lesions there is a tendency to veer in one direction).
- A sensory examination for light touch and pinprick, position, and vibration sense should be performed because lower extremity sensory impairment can cause ataxia.
- For diagnostic purposes, it is useful to categorize ataxia as acute, intermittent, or chronic. Chronic ataxia is further categorized as progressive or nonprogressive (Table 54-1).

TABLE 54-1 Causes of Ataxia

Acute
Postinfectious/immune mediated
Acute cerebellar
Polymyoclonus/opsoelonus
Guillain-Barré/Miller-Fisher syndromes
Tick paralysis
Acute demyelinating encephalomyelitis (ADEM)
Posttraumatic
Hematoma
Mass
Vertebrobasilar dissection
Infection
Meningitis
Encephalitis
Polyneuritis
Posterior fossa tumors
Intoxications
Alcohol
Anticonvulsants
Benzodiazepines
Cyclic antidepressants
Sedative-hypnotics
Inhalants
Carbon monoxide
Lead
Intermittent
Migraine & migraine variants
Seizures
Multiple sclerosis
Metabolic/Inborn errors
Urea cycle defects
Amino acid disorders
Organic acid disorders
Recurrent genetic ataxias
Chronic
Progressive
Tumor
Abscess
Hydrocephalus
Degenerative
Nonprogressive
Cerebral palsy
Sequelae of
Head trauma
Lead poisoning
Cerebellar malformations: Dandy-Walker cysts, Agensis, Hypoplasia

ACUTE ATAXIA

- Acute ataxia generally has an onset of <24 hours.
- Drug toxicity and infections are the most common etiologies. Specific toxins to assess include anticonvulsants, benzodiazepines, tricyclic antidepressants, sedative/hypnotics, and alcohol.
- Acute metabolic processes, such as hypoglycemia, are also implicated although they are usually accompanied by multiple systemic manifestations. Any ataxic patient in whom an acute infectious process is considered requires a lumbar puncture.
- Immunizations have been postulated as inciting an autoimmune phenomenon, which can result in acute cerebellar ataxia, acute demyelinating encephalomyelitis (ADEM), or brain stem encephalitis.

ACUTE CEREBELLAR ATAXIA

- A common cause of ataxia in children younger than 5 years is acute cerebellar ataxia.
- The onset of ataxia is insidious and predominantly affects the gait although dysmetria, nystagmus, and dysarthria can occur. The affected child is afebrile, and has a normal level of consciousness.
- Acute cerebellar ataxia is thought to be a postinfectious phenomenon and often occurs 2 weeks after a viral illness. Ataxia has been reported after infection with varicella, influenza, mumps, Epstein–Barr virus, echovirus 6, coxsackie B virus, and other viruses.
- The gait abnormalities come on abruptly, and are maximum at onset, with the trunk affected more than the extremities.
- It is a self-limiting illness with an excellent prognosis. Although therapies such as steroids and intravenous immune globulin have been used, there is really no specific therapy.

MYOCLONIC ENCEPHALOPATHY OF INFANCY (OPSOCLONUS/MYOCLONUS)

- Myoclonic encephalopathy of infancy (polymyoclonus/opsoclonus) is another cause of acute ataxia.
- This syndrome occurs in children from 6 months to 3 years of age, in association with occult neuroblastoma, viral disease, aseptic meningitis (especially mumps), and unknown or miscellaneous causes.
- It is differentiated from acute cerebellar ataxia by its association with opsoclonus (rapid, chaotic conjugate eye movements), which occur in association with

polymyoclonus (severe myoclonic jerks of the limbs and trunk or head).

- Diagnostic tests that should be initiated in the emergency department (ED) include chest and abdominal CT, urine vanillylmandelic acid (VMA), and lumbar puncture to exclude aseptic meningitis

ACUTE POSTINFECTIOUS DEMYELINATING ENCEPHALOMYELITIS

- This is a multifocal immune-mediated encephalopathy characterized by ataxia, alteration in level of consciousness, and neurological deficits. Seizures, cranial neuropathies, hemiparesis, and transverse myelitis are common. The patient may have fever, headache, and meningismus. It develops during the recovery from a viral illness, or after vaccination. Repeated episodes of this should prompt a workup for multiple sclerosis.

TUMORS

- Up to 60% of childhood brain tumors occur in the brain stem or cerebellum. Those that arise in the posterior fossa often have a slow onset of symptoms including ataxia, headache, and vomiting. However, hydrocephalus from obstruction or a bleed into the tumor can cause abrupt symptoms and decompensation.

GUILLAIN–BARRÉ SYNDROME (GBS)/MILLER–FISHER SYNDROME

- This is an acute demyelinating polyradiculoneuropathy that occurs after an infection or immunization.
- Although it predominantly affects the motor nerves resulting in weakness, 15% of patients develop a loss of sensory input to the cerebellum resulting in a sensory ataxia. The ataxia and weakness is progressive over several days.
- The Miller–Fisher syndrome is a form of GBS that results in ataxia, areflexia, and ophthalmoplegia. It occurs 5 to 10 days after an infectious illness, especially *Campylobacter gastroenteritis*.
- Diagnostic tests include lumbar puncture looking for cytoalbuminologic dissociation and anti-GQ1b antibody for the Miller–Fisher variant.
- Patients suspected of these syndromes should be admitted to the hospital for further testing and monitoring as disease progression is variable. Electromyography is helpful for diagnosis.

INTERMITTENT/EPISODIC ATAXIA

- In children, the most common cause of intermittent ataxia is a migraine headache that involves the basilar artery. Besides ataxia, associated symptoms include blurred vision, visual field deficits, vertigo, and headache. In a child experiencing the first basilar migraine, it is essential to exclude an acute infectious process, toxic ingestion, or mass lesion.
- Partial complex seizures can also cause intermittent ataxia but are often associated with alteration of consciousness and possibly characteristic motor manifestations.
- Rarely, inborn errors of metabolism result in intermittent ataxia.

CHRONIC PROGRESSIVE ATAXIA

- Chronic progressive ataxia has an insidious onset and progresses slowly over weeks to months.
- The combination of ataxia, headache, irritability, and vomiting in a child younger than 9 years is characteristic of a cerebellar tumor, most commonly a medulloblastoma. Cerebellar astrocytomas are located in the cerebellar hemispheres and cause ipsilateral limb ataxia, headache, and double vision. They occur most commonly in school-age children. Brain stem gliomas present with ataxia and are often accompanied by cranial nerve palsies or spasticity. In some cases, posterior fossa tumors have a relatively acute presentation.

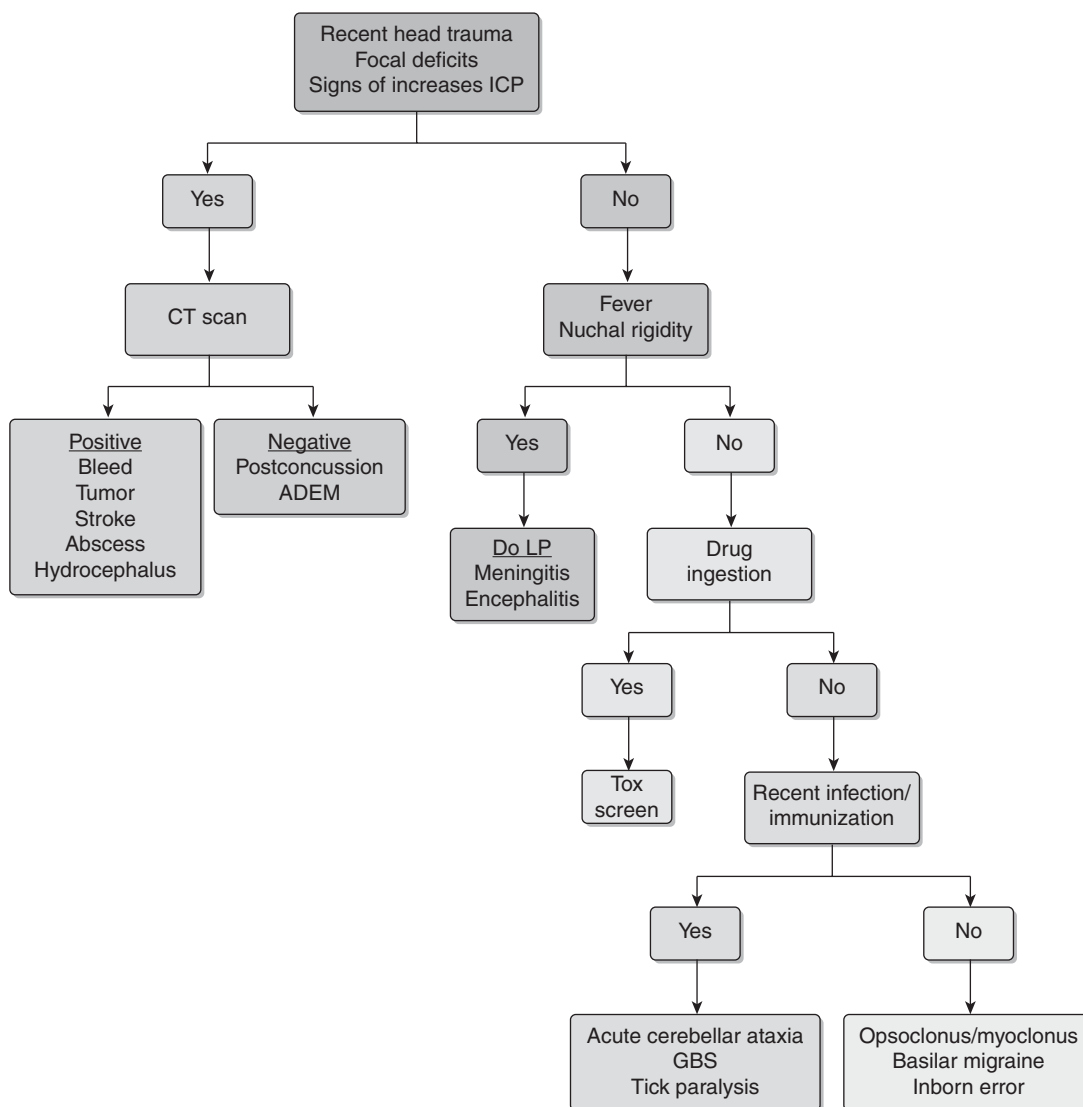


FIG. 54-1. ED evaluation of acute ataxia.

- Hydrocephalus, whether congenital or acquired, can cause ataxia. It is often accompanied by headache and vomiting and, when the patient presents late in the course of illness, can be associated with critically increased intracranial pressure.
- Neurodegenerative diseases are a group of inherited disorders that can cause spinocerebellar degeneration and progressive ataxia.
- Other hereditary causes of ataxia include spinocerebellar ataxias, of which there are 28 types. Friedrich's ataxia is the most common autosomal recessive ataxia, which usually manifests before 10 years of age. It is characterized by ataxia, nystagmus, kyphoscoliosis, cardiomyopathy, and distal muscle wasting. Patients with ataxia-telangiectasia present with ataxia before the onset of the ocular and facial telangiectasias.
- Patients with progressive ataxia require an aggressive evaluation in the ED. All patients are examined for signs of increased intracranial pressure, which in some cases can be severe enough to result in the threat of uncal herniation. A CT scan of the brain is indicated in any patient with signs of a mass lesion or hydrocephalus.

CHRONIC NONPROGRESSIVE ATAXIA

- Chronic nonprogressive ataxia may be a sequela of head trauma, meningitis, or lead poisoning. It can also result from congenital malformations, such as cerebellar agenesis or hypoplasia or the Chiari type I malformation (herniation of the cerebellar tonsils into the foramen magnum).
- The ED evaluation of chronic nonprogressive ataxia consists of ensuring, by a careful history and physical examination, that the problem is indeed stable. Patients with an unknown diagnosis may benefit from CT or magnetic resonance imaging of the brain (see Fig. 54-1).

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QUESTIONS

1. A 3-year-old male is brought to the ED due to a wobbly gait. The child has been well, with no recent illnesses, fever, or vomiting. When he woke up this morning he almost fell out of bed. Mom describes him as “acting like he is drunk.” On exam he is awake, but his gait is ataxic. The medications Mom knows are at home include: multivitamins, amoxicillin, risperidone, loratadine, and alprazolam. Of these medications, which is the most likely cause of these symptoms?
 - A. Multivitamins
 - B. Amoxicillin
 - C. Risperidone
 - D. Loratadine
 - E. Alprazolam
2. A 4-year-old male had hand-foot-and mouth disease 2 weeks ago. This has resolved, but this morning the child had an unsteady gait. In the ED, the child has a normal exam except for ataxia upon walking, and mild truncal ataxia when sitting. Of the following, which is the most likely etiology of his ataxia?
 - A. Drug ingestion
 - B. Acute cerebellar ataxia
 - C. Brain tumor
 - D. Guillain-Barre syndrome
 - E. Brain abscess
3. Parent bring their 8-month-old infant to the ED due to funny eye movements. The child has been well, has had no fever, no vomiting, has not been irritable, is up to date on shots, and they deny trauma. On exam the infant has chaotic eye movements, and jerks of the head and arms. Which of the following are of most concern as the etiology?
 - A. Meningitis
 - B. Seizure
 - C. Child maltreatment
 - D. Opsoclonus/myoclonus
 - E. Drug ingestion

4. For the above case, which is the most appropriate test to perform?
 - A. Head CT
 - B. Lumbar puncture
 - C. Urine toxicology screen
 - D. Serum glucose
 - E. Chest and abdominal CT
5. A 12-year-old female comes to the ED with progressive ataxia. On exam you find symmetrical lower extremity weakness, but intact sensation. Deep tendon reflexes are absent in the lower extremities. Her upper extremities have good strength and reflexes. Her gait is ataxic. There is no history of trauma or back pain. The most appropriate diagnostic test is which of the following?
 - A. Head CT scan
 - B. CT scan of the back
 - C. Lumbar puncture
 - D. Serum electrolytes
 - E. Urine toxicology screen
6. A 10-year-old female with a history of headaches presents to the ED with vertigo, blurred vision, and ataxia. She has no fever or stiff neck. There have been no recent illnesses. Mom has a history of migraine headaches. On exam she has no focal findings and no papilledema. The most likely etiology of these symptoms is which of the following?
 - A. Seizure
 - B. Drug ingestion
 - C. Brain tumor
 - D. Basilar migraine
 - E. Meningitis
7. A 7-year-old male comes to the ED with a history of a headache for 2 weeks. He developed vomiting 3 days ago, and he seems to have trouble keeping his balance. The vomiting tends to be in the morning. On exam there are no focal findings, but he needs to hold onto the wall to walk. Which of the following is the most important tests to perform to diagnose this child's problem?
 - A. Head CT scan
 - B. Lumbar puncture
 - C. CT scan of the back
 - D. EEG
 - E. Urine toxicology screen

ANSWERS

1. E. Alprazolam. Alprazolam (Xanax) is a benzodiazepine, so can cause ataxia. Drug toxicity and infections are the most common etiologies of ataxia. Specific toxins to assess include anticonvulsants, benzodiazepines, tricyclic antidepressants, sedative/

hypnotics, and alcohol. Side effects of risperidone include abdominal pain, dizziness, and headache. The other medications do not cause ataxia.

2. B. Acute cerebellar ataxia. A common cause of ataxia in children younger than 5 years is acute cerebellar ataxia. The onset of ataxia is insidious and predominantly affects the gait. The affected child is afebrile, and has a normal level of consciousness. Acute cerebellar ataxia is thought to be a postinfectious phenomenon and often occurs 2 weeks after a viral illness. Ataxia has been reported after infection with varicella, influenza, mumps, Epstein–Barr virus, echovirus 6, coxsackie B virus, and other viruses. The gait abnormalities come on abruptly and are the maximum at onset, with the trunk affected more than the extremities.

The sudden onset can also make drug ingestion a possibility, but is less likely with the antecedent illness. A brain tumor is unlikely due to the sudden onset. A brain abscess is unlikely without a history of cardiac problems, or an illness such as sinusitis or periorbital abscess. The presentation does not fit that of GBS.

3. C. Opsoclonus/myoclonus
4. E Chest and abdominal CT scan. Myoclonic encephalopathy of infancy (polymyoclonus/opsoclonus) is another cause of acute ataxia. This syndrome occurs in children from 6 months to 3 years of age, in association with occult neuroblastoma, viral disease, aseptic meningitis (especially mumps), and unknown or miscellaneous causes. It is differentiated from acute cerebellar ataxia by its association with opsoclonus (rapid, chaotic conjugate eye movements), which occur in association with polymyoclonus (severe myoclonic jerks of the limbs and trunk or head). Diagnostic tests that should be initiated in the emergency department (ED) include chest and abdominal CT, urine vanillylmandelic acid (VMA), and lumbar puncture to exclude aseptic meningitis.

Although meningitis is a possibility, the infant has not had a fever, vomiting or irritability. The type of seizure that can occur with random jerks in an infant is infantile spasms or West Syndrome. Its onset is before 1 year of age and consists of muscles spasm of the trunk, neck, and extremities that increase in frequency. There are no specific eye movements associated with it. The diagnosis is made by the classic EEG finding of hypersarrhythmia. Although child maltreatment and drug ingestion should be considered, they are less likely with this presentation.

5. C. Lumbar puncture. This presentation is typical of Guillain–Barre Syndrome, which is acute

demyelinating polyradiculoneuropathy that occurs after an infection or immunization. It predominantly affects the motor nerves resulting in symmetrical ascending weakness. The ataxia and weakness are progressive over several days. Diagnostic tests include lumbar puncture looking for cytoalbuminologic dissociation. With no history of trauma or back pain, a CT scan of the back is unlikely to be diagnostic. With no headache or vomiting, a head CT scan is not needed. Serum electrolytes may be useful as a baseline, but urine toxicology screen is not needed.

6. D. Basilar migraine. In children, the most common cause of intermittent ataxia is a migraine headache that involves the basilar artery. Besides ataxia, associated symptoms include blurred vision, visual field deficits, vertigo, and headache. In a child experiencing the first basilar migraine, it is essential to exclude an acute infectious process, toxic ingestion, or mass lesion. Since this patient has had headaches in the past, these are less likely but should be excluded. Partial complex seizures can also cause intermittent ataxia but are often associated with alteration of consciousness and possibly characteristic motor manifestations.
7. A. Head CT scan. The progressive nature of this child's worrisome symptoms: ataxia, headache, irritability, and vomiting in a child younger than 9 years is characteristic of a cerebellar tumor, most commonly a medulloblastoma. Although MRI may show small tumors better than a CT scan, in the ED a CT scan is often easier to obtain. A CT of his back is unlikely to determine the etiology. The patient has no fever to suggest meningitis. In fact, an LP is ill-advised in this patient. The progressive nature of the symptoms makes drug ingestion unlikely; therefore, a urine toxicology screen is not needed. There is also no evidence of seizures to make an EEG necessary.

55 WEAKNESS

Susan Fuchs

INTRODUCTION

- The term “weakness” can refer to a general phenomenon that affects all or most of the body or may refer to a specific area, such as an extremity. The primary focus in this chapter is on weakness arising from neuromuscular disorders.

PATHOPHYSIOLOGY

- Paresis implies a complete or partial weakness.
- Paraparesis is weakness of the lower half of the body.
- Quadraparesis is weakness involving all limbs.
- Hemiparesis is weakness of one side.
- Quadriplegia is paralysis of all limbs; it usually results from a spinal cord lesion.
- Hemiplegia, involving one side of the body, generally results from a lesion in the brain.
- Abnormalities of the neuromuscular system are further classified as arising from an upper or lower motor neuron unit:
 - The upper motor neuron unit arises in the motor strip of cerebral cortex, traverses the corticospinal tract, and ends in the spinal cord adjacent to the anterior horn cell. Upper motor neuron diseases involving the cerebral cortex or spinal cord usually present with asymmetrical weakness that is contralateral to the lesion, and are associated with hyperreflexia, increased muscle tone, and the absence of atrophy or fasciculations.
 - The lower motor neuron unit includes the anterior horn cells, peripheral nerve, neuromuscular junction, and muscle fibers. Lower motor neuron diseases present with symmetrical weakness that can be isolated to specific muscle groups, and are associated with decreased muscle tone and depressed reflexes. Depending on whether the disorder is acute or chronic, atrophy, and fasciculations may be present (Table 55-1).
- Involvement of bulbar muscles is manifested by cranial nerve findings, facial muscle weakness, and chewing or swallowing difficulties. Bulbar involvement can occur in both upper and lower motor neuron disorders.
- Neuropathies are disorders of nerves, and tend to produce distal muscle weakness, hypesthesias or paresthesias, and decreased reflexes, especially early in the disease. The progression of weakness and sensory loss is in a stocking and glove distribution.

TABLE 55-1 Upper Versus Lower Motor Neuron Findings

Upper motor neuron lesions
1. Asymmetric weakness
2. Increased muscle tone
3. No atrophy/fasciculations
4. Hyperreflexia/clonus
Lower motor neuron lesions
1. Symmetrical weakness
2. Decreased muscle tone
3. Atrophy/fasciculations
4. Diminished reflexes

- Myopathies are disorders of muscle, and can be inflammatory, infectious, congenital, or metabolic. Inflammatory myopathies usually involve proximal muscles and are often associated with muscle pain or tenderness. Reflexes become decreased late in the disease. Congenital myopathies tend to involve specific muscle groups, and can present at birth with hypotonia and weakness, or in older children with a more insidious progression.

DIAGNOSIS

HISTORY

- It is vital to distinguish between acute and chronic disorders. The location of the initial weakness is elicited, and weakness is established as focal or general. Focal weakness is further characterized as predominantly proximal or distal. The rate of progression of symptoms is characterized as acute, which implies minutes to hours; subacute, meaning hours to days; and slowly progressive, which involves a prolonged period. Acute onset or rapid progression implies spinal cord compression or a vascular event involving the spinal cord or brain. Subacute progression can be due to infection, inflammation, toxin, or tumor. Slowly progressive symptoms imply a chronic or congenital disorder. The loss of developmental milestones implies a degenerative disorder.

PHYSICAL EXAMINATION

- The physical examination begins with observation of mental status, posture, gait, and the ability of the child to get on the examining table or to sit unaided.
- The vital signs are assessed with particular attention to respiratory rate and effort since many neuromuscular disorders are associated with a risk of respiratory failure. Blood pressure and pulse are carefully monitored, since some neuromuscular disorders, such as Guillain-Barré syndrome, are associated with autonomic instability.
- The patient's general appearance and facial expression is noted, with attention given to general muscular development and the presence of kyphosis, scoliosis, or lordosis. Lack of facial expression, snarl, or slack jaw suggests myasthenia gravis.
- Gross inspection of the muscles is performed, noting the presence of wasting, fasciculation, or hypertrophy.
- The neurologic examination includes an evaluation of pupillary size and reactivity and the remainder of

TABLE 55-2 Evaluating Muscle Strength

0: Total lack of contraction
1: Muscle twitch/trace contraction
2: Movement/weak contraction without gravity
3: Movement/weak contraction against gravity
4: Movement against some but not full resistance
5: Normal motor strength against full resistance

the cranial nerves. One way to test facial strength is to have children blow out their cheeks and resist compression.

- For patients old enough to cooperate, motor strength in the extremities is evaluated and rated on a scale of 0 to 5 (Table 55-2). For infants who cannot cooperate with the examination, it is possible to perform a general assessment of muscle tone and integrity by holding the baby under arms and placing the feet on the bed. Infants with normal tone will not slide through the examiner's hands and will actively kick both legs against the resistance of the bed. In an outstretched prone position, infants supported on the trunk should hold their head up, flex the limbs, and keep their back straight.
- Deep tendon reflexes at the knees, ankles, elbows, and wrists are elicited (Table 55-3).
- Hyperreflexia or sustained clonus indicate an upper motor neuron lesion, whereas absent or decreased reflexes imply a problem in a lower motor distribution.
- Sensory evaluation includes touch, pain, position, vibration, and temperature. Touch and pain are evaluated by assessing soft versus sharp stimulation and two-point discrimination. Position sense is assessed by asking the child to indicate the direction in which an examiner moves a finger or toe. Temperature sensation can be assessed by the use of a cold stethoscope or by touching the child with cold or warm water. Vibration can be tested using a tuning fork on both thumbs (interphalangeal joint) and big toes.
- An abnormality of touch and position on one side and pain and temperature on the other suggests a cord lesion.
- The unilateral loss of all sensations suggests a brain lesion.
- A stocking and glove distribution of sensory loss suggests a peripheral neuropathy.

TABLE 55-3 Evaluating Deep Tendon Reflexes

0: Absent
1: Reduced
2: Normal
3: Increased
4: Clonus

LABORATORY EVALUATION

- The laboratory evaluation is based on the provisional diagnosis. Generally, complete blood counts and serum electrolytes are indicated.
- Electromyography and nerve conduction studies are indicated if lower motor neuron disease is suspected, and to distinguish neurogenic from myopathic etiologies but they are not ED studies.

SPECIFIC CAUSES OF WEAKNESS

GUILLAIN–BARRÉ SYNDROME

- Also known as acute inflammatory demyelinating polyradiculoneuropathy (AIDP), Guillain–Barré syndrome (GBS) occurs in both children and adults. It is more common in the adult patient population. The pathogenesis is unknown, but it is thought to result from an immune response to an antecedent viral infection that triggers demyelination of nerve roots and peripheral nerves. *Campylobacter* infection is the most common preceding illness, occurring in 30% of cases.
- The syndrome often starts with paresthesias in the toes and fingers, and nonspecific muscular pain, most often in the thighs. The pain is followed by weakness, which is most often symmetric and distal, and results in trouble walking. Weakness progresses upward and, in some cases, results in total paralysis within 24 hours. Cranial nerve involvement is common, with bilateral facial weakness. Deep tendon reflexes are usually absent, but plantar responses remain down going.
- Autonomic involvement can produce labile changes in blood pressure and bowel and bladder incontinence.
- Laboratory findings are generally not helpful, although spinal fluid analysis may reveal high protein (>45 mg/dL) and usually has fewer than 10 white blood cells.
- Electrophysiologic studies demonstrate motor conduction block, and slowed nerve conduction velocities.

The treatment for Guillain–Barré syndrome is supportive care. Patients with vital capacity $\frac{1}{2}$ normal for age usually require ventilatory support. Steroids and other immunosuppressive agents are of no value. Intravenous immunoglobulin (IVIG) has been shown to shorten the course, as have limited studies on plasmapheresis. The American Academy of Neurology states that IVIG or plasmapheresis are treatment options for children with severe GBS.

- There are variants of GBS/AIDP in children:
 - Miller–Fisher syndrome is characterized by ophthalmoplegia, hyporeflexia, and ataxia.
 - Acute motor and sensory axonal neuropathy (AMSAN) and acute motor axonal neuropathy

occur mainly in China, and are associated with a preceding *Campylobacter* infection.

The latter two forms consist of acute ascending weakness, hyporeflexia, and elevated CSF protein, with AMSAN having a more prolonged course and limited recovery.

TRANSVERSE MYELITIS

- Transverse myelitis is a syndrome characterized by acute dysfunction at a level of the spinal cord. The onset can be insidious, but is usually over 24 to 48 hours. Patients may initially complain of paresthesias and weakness of the lower extremities. Those with the rapid form often complain of back pain. Progressive weakness, paraplegia, and urinary retention usually result, and a sensory level may develop, most commonly in the thoracic area.
- Flaccid paralysis and decreased reflexes are characteristic early in the process but are later followed by increased muscle tone.
- For a patient with signs of a rapidly advancing spinal cord lesion, it is imperative to exclude a treatable mass lesion that could be compressing the cord, such as an epidural abscess or hemorrhage. This is usually done by MRI, which shows a gadolinium signal abnormality over the spinal cord, and may show swelling on weighted T2 images.
- Most patients with transverse myelitis recover some function. Corticosteroids may benefit some patients.

TICK PARALYSIS

- Tick paralysis is caused by a neurotoxin from the Rocky Mountain wood tick (*Dermacentor andersoni*) or the Eastern dog tick (*Dermacentor variabilis*). The tick produces a neurotoxin that prevents liberation of acetylcholine at neuromuscular junctions. Several days after the tick attaches, the patient begins to experience paresthesias, fatigue, and weakness, which progresses to ataxia and difficulty walking. Deep tendon reflexes are absent.
- If the tick is not removed, flaccid paralysis and death can result. Removal of the tick is generally curative within a few hours.

BOTULISM

- Infection with *Clostridium botulinum* can produce three neurologic diseases. Ultimately, symptoms result from a toxin generated from spores of the bacteria that inhibits calcium-dependent release of acetylcholine at the prejunction of terminal nerve fibers.

- Food-borne botulism results from ingestion of food containing the toxin. Diarrhea and vomiting are followed by neurologic symptoms, often secondary to cranial nerve dysfunction. Blurred vision, dysarthria, and diplopia can occur and can be followed by weakness of the extremities. Mucous membranes of the mouth and pharynx may be dry. Deep tendon reflexes may be weak or absent. Antitoxin may be effective in food-borne botulism.
- Wound botulism results from infection of a contaminated wound, 4 to 14 days after the wound is infected. Clinically, it is usually indistinguishable from food-borne botulism. Treatment includes wound debridement and antibiotic therapy. Antitoxin may also be useful.
- Infantile botulism is caused by colonization of the intestinal tract by spores of *C botulinum*, which releases toxin that is systemically absorbed.
 - It has been related to the ingestion of contaminated honey, but many cases are linked to nearby construction projects, as soil harbors the spores. The affected age group is 6 weeks to 9 months. A prominent manifestation is constipation.
 - The infant develops a descending paralysis, with ptosis, difficulty in sucking and swallowing, and reduced facial expression and can become hypotonic. Symmetrical paralysis can develop with occasional respiratory involvement.
 - Diagnosis is by isolating the toxin in the infant's stool.
 - Electromyography is also useful.
 - The management of infant botulism is supportive and may require mechanical ventilation.
 - Treatment with antitoxin and antibiotics does not seem to be of benefit.

MYASTHENIA GRAVIS

- Myasthenia gravis comprises a group of autoimmune diseases characterized by easy fatigability. It is associated with antiacetylcholine receptor antibodies resulting in decreased transmission of nerve impulses across the neuromuscular junction. It is the most common disorder of the myoneural junction in children. The two basic categories of myasthenia gravis in the pediatric population are as follows:
 - Neonatal transient myasthenia.
 - Juvenile myasthenia gravis.
- Neonatal transient myasthenia gravis occurs in infants born to mothers with the disease, and is caused by maternal antiacetylcholine receptor antibodies that cross the placenta. It affects 10% to 15% of infants whose mothers have myasthenia gravis. The infant

may have a weak cry or suck, ptosis, generalized weakness, hypotonia, and respiratory distress. It presents in the first few hours of life, but may be delayed up to 3 days. Treatment is with neostigmine or pyridostigmine. The disease usually improves in 4 to 6 weeks, but may last for months.

- Juvenile myasthenia gravis is similar to that seen in adults. It commonly has its onset at around 10 years of age, and is more common in females. Ptosis, ophthalmoplegia, and weakness of other facial muscles are commonly present. This results in difficulty in chewing, dysarthria, and dysphagia. Bulbar weakness develops in 75% of patients. Symmetrical limb weakness is usually present, and affects the proximal muscles more than the distal muscles. The disease not only tends to become worse throughout the day, but can also worsen with stress or exertion. Both remissions and exacerbations are common.
- To diagnose myasthenia gravis, the edrophonium (tensilon) test is helpful, but electrophysiologic testing and the presence of serum antiacetylcholine receptor antibodies are more specific.
- The primary treatment is with anticholinesterase agents.
- In refractory or severe cases, immunosuppressive agents, corticosteroids, IVIG, plasmapheresis, or thymectomy may be necessary.
- Erythromycin, tetracycline, aminoglycoside antibiotics, anesthetic drugs, neuromuscular blockers, and muscle relaxants therapy can exacerbate symptoms and should be avoided.

MYASTHENIC CRISIS

- Myasthenic crisis results in profound weakness, difficulty in swallowing secretions, and respiratory insufficiency.
- It is often precipitated by infection, surgery, or decreasing immunosuppressive drugs. It can also be exacerbated by the use of antibiotic therapy, especially aminoglycosides as well as antiarrhythmics, and ophthalmologic medications.
- If a myasthenic crisis is suspected, the patient is admitted to an intensive care unit where respiratory status can be monitored, and elective intubation can occur if needed.

CHOLINERGIC CRISIS

- An overdose of anticholinesterase medications, which can provoke a cholinergic crisis. Unfortunately, the symptoms of cholinergic excess are similar to those of a myasthenic crisis. In both, increasing weakness

is the predominant finding. Patients suffering a cholinergic crisis may also have associated vomiting, diarrhea, and hypersalivation.

- In patients with obvious severe cholinergic excess, atropine or glycopyrrolate may be useful in drying airway secretions. However, in most cases, it will be difficult to distinguish between a cholinergic crisis and an exacerbation of myasthenia, and hospital admission and close observation are indicated.

BELL'S PALSY

- Bell's palsy is a condition that results in unilateral facial weakness. In severe cases, there can be total paralysis of the facial muscles. It is thought to result from swelling and edema of cranial nerve VII, the facial nerve, as it traverses the facial canal within the temporal bone.
- In most cases, Bell's palsy is idiopathic. Certain conditions are associated with unilateral facial weakness, including viral infections, otitis media, Lyme disease, and temporal bone trauma.
- Symptoms may begin with ear pain, which is followed by the development of facial weakness, characterized by a drooping mouth and inability to close the eye on the affected side. In some cases, lacrimation, and taste are impaired. Inability to close the mouth can make eating and drinking difficult.
- Laboratory studies are not necessary in uncomplicated cases.
- The prognosis of Bell's palsy is generally good, with recovery usually beginning in 2 to 4 weeks, but may take 6 to 14 weeks to resolve fully.
- Steroid therapy may be beneficial if started early in the course of illness, and is given for 1 week. Treatment includes lubricating solutions for the eye on the affected side to maintain moisture of the cornea. Patients with inability to close the eye may require patching. In young children, ophthalmologic consultation may be advisable.

MYOPATHIES

- Myopathies are diseases that affect skeletal muscle. Many myopathies are congenital.

MUSCULAR DYSTROPHIES

- Muscular dystrophies are disorders associated with progressive degeneration of muscle. The most common is Duchenne muscular dystrophy, usually an X-linked recessive disorder resulting in the absence of the protein dystrophin.

- Clinical manifestations usually become apparent before age 4, when patients begin to develop weakness of the hip girdle and shoulder muscles. Patients may have difficulty standing and characteristically rise from all fours by placing their hands on the thighs and pushing up (Gower's sign). There is hypertrophy of the calf muscles. The disease is characterized by a progressive loss of muscle strength.
- Lordosis and kyphoscoliosis develop as the disease progresses. Pulmonary involvement due to weakness of the diaphragmatic and intercostal muscles results in impaired lung function. Cardiac insufficiency and cardiomyopathy of varying degrees occur in the majority of children.
- The use of daily prednisone is the only treatment. It has been shown to improve ambulation, reduce contractures, and preserve respiratory function.

PERIODIC PARALYSIS

- Periodic paralysis is an example of metabolic myopathies that result in muscle weakness. The disorders are usually inherited, but can also be acquired. The inherited forms are autosomal dominant. There are several varieties, which include the channelopathies affecting the potassium, sodium, and calcium channels in skeletal muscle.
- Hypokalemic periodic paralysis is actually one of five channelopathies that is due to a disorder in the skeletal muscle sodium, calcium, or potassium channel gene, or is associated with thyroid disease, or cardiac dysrhythmias (Anderson syndrome).
 - Most episodes of hypokalemic periodic paralysis have their onset during the first or second decade of life.
 - They are often precipitated by excitement, cold, rest after exercise, or ingestion of high-carbohydrate meals, but can also be associated with hyperthyroidism, excessive insulin, renal tubular acidosis, or laxative abuse.
 - Paralysis usually begins proximally and spreads distally. The patient may be areflexic. The episode can last for 6 to 12 hours.
 - Serum potassium during an attack is usually decreased compared with a baseline value. Treatment with oral potassium during an attack may be helpful; intravenous potassium may be needed for severe attacks.
 - Long-term therapy with a low-sodium and low-carbohydrate diet, avoidance of exposure to cold, and potassium supplement can be beneficial. Acetazolamide taken daily may reduce the number of attacks.

- The three forms of hyperkalemic periodic paralysis are also autosomal dominant conditions associated with intermittent attacks beginning in the first decade of life.
 - Attacks can be provoked by cold exposure, periods of rest following heavy exertion, and oral potassium loads.
 - Weakness can develop rapidly, and lasts a shorter period of time than that of hypokalemic periodic paralysis. Myalgia develops at the outset, and is followed by proximal then distal muscle weakness. Some patients develop myotonia during attacks.
 - The serum potassium is elevated above baseline values, although the degree of hyperkalemia varies.
 - Treatment with oral glucose may speed recovery. Most attacks respond to treatment with glucose and insulin. In severe cases, intravenous calcium gluconate is necessary. In such cases, nebulized albuterol is also helpful.
 - Preventive treatment includes avoidance of exposure to cold and avoidance of fasting. Eating frequent high-carbohydrate meals may be helpful, as is treatment with hydrochlorothiazide or acetazolamide.

VIRAL MYOSITIS

- Viral myositis is a common cause of weakness in children. It may follow influenza or other viral illnesses. Fever and other constitutional symptoms are accompanied by myalgias. Affected muscles are tender to touch, and may be boggy. The creatine kinase is often elevated. The urine should be examined for myoglobinuria, as this may indicate rhabdomyolysis, a serious complication. Most cases are self-limited, and are treated with bed rest, hydration, and NSAIDs or acetaminophen.

TRICHINOSIS

- This is caused by ingestion of inadequately cooked meat (usually pork) containing the parasitic nematode *Trichinella spiralis*. While most infections are asymptomatic, invasion of the muscles results in an acute systemic infection characterized by fever, headache, generalized myalgias, abdominal pain, and weakness 2 to 12 days after ingestion of the meat. Myalgias and weakness are more profound in the third week of infection. Other complications include myocarditis and CNS infection.
- Diagnosis is by serum antibody levels, which peak 3 to 4 weeks after infection. Many patients also

have elevated creatine kinase and eosinophilic leukocytosis.

- Treatment includes thiabendazole and prednisone, to prevent a Herxheimer-like reaction after degeneration of the larvae.

PYOMYOSITIS

- This is an abscess or multifocal abscesses within the muscle. While it is more likely to occur in an immunocompromised host, it has been occurring more frequently with the emergence of methicillin resistant *Staphylococcus aureus* (MRSA). Other bacterial causes include streptococci, *Escherichia coli*, *Yersinia*, and *Legionella*.
- The child presents with fever, muscle pain, and tenderness. The abscess can be seen on ultrasound, CT scan, or MRI.
- Treatment involves appropriate intravenous antibiotics.

POLIOMYELITIS

- The virus attacks the anterior horn cells, resulting in asymmetrical weakness. The child may develop nuchal rigidity, muscle tightness, and fever. In infants younger than 1 year, spasm of the back muscles is also prominent. Bulbar involvement results in respiratory compromise, circulatory and autonomic instability, and mandates ventilatory support.
- The cerebrospinal fluid has pleocytosis with mononuclear cells, and a normal or slightly elevated protein.

WEST NILE VIRUS

- This RNA virus has rapidly become endemic in the United States, due to its vector of the *Culex* mosquito.
- Symptoms include fever, headache, nausea, vomiting, and meningoencephalitis. Weakness may develop, and may be asymmetric or involve the face.
- Diagnosis is by demonstrating a rising IgM titer, or by reverse transcriptase PCR.

DISPOSITION

- The disposition of a patient with weakness depends on the degree of disability and the nature of the underlying problem. In any patient in whom the development of respiratory compromise is a possibility, hospital admission and close observation is recommended.

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QUESTIONS

1. A 6-year-old female presents to the ED with weakness of her right leg for 3 days. She denies any trauma. She is noted to have decreased muscle tone and decreased reflexes in the right leg, especially her foot and calf. Her left leg is normal. Which of the following do these findings indicate?
 - A. Upper motor neuron lesion
 - B. Lower motor neuron lesion
 - C. Neuropathy
 - D. Myopathy
 - E. Guillain–Barre syndrome
2. A 6-month-old male presents with constipation. On examination you note ptosis and a flat facial expression. The most likely diagnosis is
 - A. Food-borne botulism
 - B. Infantile botulism
 - C. Myasthenia gravis
 - D. Muscular dystrophy
 - E. Wound botulism
3. A 15-year-old female presents to the ED with trouble walking. She was well until 1 week ago when she had diarrhea for 3 days. After it resolved, she developed muscle pain in her thighs. The pain has gotten better, but she was unable to get out of bed or walk without assistance today. On examination, both lower extremity reflexes are absent. There is no evidence of muscle atrophy. Which of the following are true about this disorder?
 - A. LP reveals low protein
 - B. Bowel and bladder incontinence is uncommon
 - C. Thought to be an immune response
 - D. EMG shows increased nerve conduction
 - E. Is associated with Shigella infection
4. An 8-year-old boy presents to the ED with back pain. On examination there is bilateral weakness of the lower extremities, and a sensory level at T-10. The patient denies trauma. The most important diagnostic test is which of the following?
 - A. Spine x-ray
 - B. CT of head
 - C. MRI of spine
 - D. EMG
 - E. Lumbar puncture
5. An 11-year-old female presents with slurred speech and trouble swallowing. On examination you note ptosis and weak facial muscles. She also has some limb weakness, more proximal than distal. In the ED a helpful diagnostic test would be
 - A. CT of head
 - B. Lumbar puncture
 - C. Tensilon test
 - D. Antistreptolysin O titers
 - E. Tick removal
6. An 8-year-old female presents with an uneven smile for 1 day. She denies trauma, or travel. She has no ear, jaw or tooth pain. She has unilateral facial weakness. The rest of her exam is normal. Appropriate treatment includes which of the following?
 - A. IV steroids
 - B. PO steroids
 - C. Cycloplegic drops

- D. Treatment for Lyme disease
E. Plasmapheresis
7. A 15-year-old male presents to the ED complaining of thigh pain that developed while snowboarding. He then had trouble going from a sitting to standing position. On examination you note proximal muscle weakness. The most appropriate test at this time to diagnose his problem is
- Serum glucose
 - Serum Na
 - Myoglobin
 - Serum K
 - CPK
8. A 4-year-old male presents with a red, swollen thigh. At the center is a pus-filled pimple. There is no fluctuance, but there is induration underneath. The patient has decreased range of motion of the right hip. He is otherwise a healthy male. Mom says he has had MRSA in the past. The most important treatment is
- Incision and drainage of the pimple
 - X-ray of hip
 - CBC
 - X-ray of femur
 - Ultrasound of thigh

ANSWERS

1. C. Neuropathies are disorders of nerves, and tend to produce distal muscle weakness, hypesthesias, or paresthesias, and decreased reflexes, especially early in the disease. The progression of weakness and sensory loss is in a stocking and glove distribution. Inflammatory myopathies usually involve proximal muscles and are often associated with muscle pain or tenderness. Reflexes become decreased late in the disease.
- Upper motor neuron diseases involving the cerebral cortex or spinal cord usually present with asymmetrical weakness that is contralateral to the lesion, and are associated with hyperreflexia, increased muscle tone, and the absence of atrophy or fasciculations.
- Lower motor neuron diseases present with symmetrical weakness that can be isolated to specific muscle groups, and are associated with decreased muscle tone and depressed reflexes. Guillain–Barre syndrome often starts with paresthesias in the toes and fingers, and nonspecific muscular pain, most often in the thighs. The pain is followed by weakness, which is most often symmetric and distal, and results in trouble walking.
2. B. Infantile botulism is caused by colonization of the intestinal tract by spores of *C botulinum*, which releases toxin that is systemically absorbed. It has been related to the ingestion of contaminated honey, but many cases are linked to nearby construction projects, as soil harbors the spores. The affected age group is 6 weeks to 9 months. A prominent manifestation is constipation. The infant develops a descending paralysis, with ptosis, difficulty in sucking, and swallowing, and reduced facial expression and can become hypotonic. Symmetrical paralysis can develop with occasional respiratory involvement. Food-borne botulism results from ingestion of food containing the toxin. Diarrhea and vomiting are followed by neurologic symptoms, often secondary to cranial nerve dysfunction. Blurred vision, dysarthria, and diplopia can occur and can be followed by weakness of the extremities. Mucous membranes of the mouth and pharynx may be dry. Deep tendon reflexes may be weak or absent. Wound botulism results from infection of a contaminated wound, 4 to 14 days after the wound is infected. Clinically, it is usually indistinguishable from food-borne botulism. Neonatal myasthenia presents in the first few hours of life, but may be delayed up to 3 days. Clinical manifestations of muscular dystrophy usually become apparent before age 4, when patients begin to develop weakness of the hip girdle and shoulder muscles. Patients may have difficulty standing and characteristically rise from all fours by placing their hands on the thighs and pushing up (Gower’s sign). There is hypertrophy of the calf muscles. The disease is characterized by a progressive loss of muscle strength.
3. C. Guillain–Barré syndrome (GBS) occurs in both children and adults. It is more common in the adult patient population. The pathogenesis is unknown, but it is thought to result from an immune response to an antecedent viral infection that triggers demyelination of nerve roots and peripheral nerves. Campylobacter infection is the most common preceding illness, occurring in 30% of cases. Autonomic involvement can produce labile changes in blood pressure and bowel and bladder incontinence. Laboratory findings are generally not helpful, although spinal fluid analysis may reveal a high protein (>45 mg/dL) and usually has fewer than 10 white blood cells. Electrophysiologic studies demonstrate motor conduction block, and slowed nerve conduction velocities.
4. C. Transverse myelitis is a syndrome characterized by acute dysfunction at a level of the spinal cord. The onset can be insidious, but is usually over 24 to 48 hours. Patients may initially complain of paresthesias and weakness of the lower extremities.

Those with the rapid form often complain of back pain. Progressive weakness, paraplegia, and urinary retention usually result, and a sensory level may develop, most commonly in the thoracic area. Flaccid paralysis and decreased reflexes are characteristic early in the process but are later followed by increased muscle tone. For a patient with signs of a rapidly advancing spinal cord lesion, it is imperative to exclude a treatable mass lesion that could be compressing the cord, such as an epidural abscess or hemorrhage. This is usually done by MRI, which shows a gadolinium signal abnormality over the spinal cord, and may show swelling on weighted T2 images. A spinal xray is unlikely to show the spinal abnormality. A CT scan of the head will not show the spinal pathology. A lumbar puncture is not indicated, especially with signs of cord compression. An EMG cannot usually be performed in the ED.

5. C. Juvenile myasthenia gravis commonly has its onset at around 10 years of age, and is more common in females. Ptosis, ophthalmoplegia, and weakness of other facial muscles are commonly present. This results in difficulty in chewing, dysarthria, and dysphagia. Bulbar weakness develops in 75% of patients. Symmetrical limb weakness is usually present, and affects the proximal muscles more than the distal muscles. To diagnose myasthenia gravis, the edrophonium (tensilon) test is helpful, but electrophysiologic testing and the presence of serum antiacetylcholine receptor antibodies are more specific. A CT of the head, lumbar puncture, antistreptolysin O titer would not be diagnostic.
6. B. Bell's palsy is a condition that results in unilateral facial weakness. In severe cases, there can be total paralysis of the facial muscles. It is thought to result from swelling and edema of cranial nerve VII, the facial nerve, as it traverses the facial canal within the temporal bone. In most cases, Bell's palsy is idiopathic. Certain conditions are associated with unilateral facial weakness, including viral infections, otitis media, Lyme disease, and temporal bone trauma. Symptoms may begin with ear pain, which is followed by the development of facial weakness, characterized by a drooping mouth and inability to close the eye on the affected side. In some cases, lacrimation and taste are impaired. Inability to close the mouth can make eating and drinking difficult. Oral steroid therapy may be beneficial if started early in the course of illness, and is given for 1 week. Treatment includes lubricating solutions for the eye on the affected side to maintain moisture of the cornea. (not cycloplegics) Patients with inability to close the eye may require patching. Plasmapheresis is not used to treat Bell's Palsy.
7. D. Hypokalemic periodic paralysis is often precipitated by excitement, cold, rest after exercise, or ingestion of high-carbohydrate meals, but can also be associated with hyperthyroidism, excessive insulin, renal tubular acidosis, or laxative abuse. Paralysis usually begins proximally and spreads distally. The patient may be areflexic. The episode can last for 6 to 12 hours. Hyperkalemic periodic paralysis attacks can be provoked by cold exposure, periods of rest following heavy exertion, and oral potassium loads. Weakness can develop rapidly, and lasts a shorter period of time than that of hypokalemic periodic paralysis. Myalgia develops at the outset, and is followed by proximal then distal muscle weakness. Some patients develop myotonia during attacks. The serum potassium is elevated above baseline values, although the degree of hyperkalemia varies. Although there are channelopathies that involve sodium and calcium channels, ones involving potassium are more common. Although there has been exercise, a myoglobin and CPK are unlikely to determine the cause of this patients' weakness.
8. E. This presentation describes pyomyositis, an abscess or multifocal abscesses within the muscle. While it is more likely to occur in an immunocompromised host, it has been occurring more frequently with the emergence of MRSA. The child presents with fever, muscle pain, and tenderness. The abscess can be seen on ultrasound, CT scan, or MRI. An incision and drainage of the pimple may result in a positive culture, but will not diagnose the underlying abscess in the muscle. A CBC will likely be elevated, but not diagnostic. An xray of the hip is helpful if septic arthritis is a possibility. An x-ray of the femur mat shows some swelling of the muscle tissue, but an ultrasound is much more diagnostic.

56 HEADACHE

Susan Fuchs

INTRODUCTION

- As many as 82% of children experience headache by the age of 15 years. Headaches are sometimes the manifestation of life-threatening illness.
- Headaches can be classified as primary or secondary. Primary headaches include migraine, tension-type,

and cluster headaches. Secondary headaches have identifiable etiologies based on history and physical examination. These include headaches attributed to head or neck trauma, infection, a vascular disorder such as a stroke, a nonvascular intracranial disorder such as elevated intracranial pressure or a neoplasm, or a toxic substance such as carbon monoxide.

- Facial pain, sinus, and dental problems can also cause headaches. Some patients with psychiatric disorders will complain of headache.

EVALUATION

- The evaluation of a child for a headache includes information on the headache history. Based on this information, the headache can be classified as acute (sudden, first), acute and recurrent (episodic), chronic and progressive, or chronic and nonprogressive. One way to help determine headache etiology is demonstrated in Table 56-1.
- The following information should be obtained: age at onset, frequency and duration, time of onset, location, quality of pain, associated symptoms such as nausea, vomiting, photophobia, warning signs or aura, precipitating factors, relieving factors, recent trauma, change in school or home environment, response to treatment at home, and family history of migraines.
- The physical evaluation includes general appearance, blood pressure and temperature, height, weight, and head circumference. The eyes are assessed for extraocular nerve palsies or nystagmus. A fundoscopic examination evaluates the possibility of papilledema. Examination of the head assesses the temporomandibular joints and sinuses. The neck is auscultated for bruits that would indicate an arteriovenous malformation, and assessed for the presence of meningismus or rigidity. The skin is examined for café-au-lait spots, neurofibromas, and ash leaf spots. The neurologic examination includes strength testing, deep tendon reflexes, Romberg test, gait testing, cerebellar tests, Brudzinkis, and Kernig's sign.

- The main concern for a physician is whether intracranial pathology exists. One study identified seven predictors for space-occupying lesions. These included sleep-related headache, absence of family history of migraine, vomiting, absence of visual symptoms, headache less than 6 months, confusion, and an abnormal neurologic examination.
- Brain tumor headaches in children are associated with neurologic findings, including papilledema, ataxia, and weakness in 85% of cases within 8 weeks of onset, and in virtually all cases by 24 weeks. Although many brain tumors in children have a midline cerebellar origin, signs and symptoms will still occur. Look for nonlateralizing signs such as vomiting, head tilt, double vision or, a change in personality or gait.
- Other life-threatening causes of headache include bacterial meningitis, orbital or cerebral abscess, viral encephalitis, hydrocephalus, intracranial hemorrhage, hypertensive encephalopathy, and carbon monoxide poisoning.

LABORATORY STUDIES

- Laboratory studies are based upon the suspected etiology of the headache. If the blood pressure is elevated, electrolytes, BUN, creatinine, and urinalysis are appropriate. For a child with a fever, CBC, blood cultures, and cerebrospinal fluid (CSF) studies are appropriate.

NEUROIMAGING

- Neuroimaging should be performed for a child with signs of increased intracranial pressure, focal symptoms, an abnormal neurologic examination, skin lesions suggestive of a neurocutaneous syndrome, recent head trauma, and a progressive neurologic disorder. While a computed tomography (CT) scan is usually adequate to see a space-occupying lesion, bleed, hydrocephalus, and abscess, a magnetic resonance imaging (MRI) may be needed to demonstrate sellar

TABLE 56-1 Headache Etiology

ACUTE, LOCALIZED	ACUTE, GENERALIZED	ACUTE, RECURRENT	CHRONIC, PROGRESSIVE	CHRONIC, NONPROGRESSIVE
Sinusitis, otitis, viral infection: flu	Systemic infection: flu, meningitis	Migraine	Idiopathic intracranial hypertension	Tension-type
Posttrauma	Hypertension		Space occupying lesion (tumor, abscess, hemorrhage, hydrocephalus)	Psychiatric (depression, school phobia)
Dental abscess, TMJ	Hemorrhage			
First migraine	Exertional, first migraine			

lesions, some small posterior fossa lesions, white matter abnormalities, and congenital anomalies.

- The American Academy of Neurology and the Child Neurology Society developed a practice parameter for the evaluation of children and adolescents with recurrent headaches. They stated that routine neuroimaging is not recommended in children with recurrent headaches and a normal neurologic examination. Neuroimaging should be considered in the child with headache who has an abnormal neurologic examination consisting of focal findings, signs of increased intracranial pressure, altered level of consciousness, or coexistent seizures or both.
 - Neuroimaging should be considered in children with historical features to suggest recent onset of severe headache, change in the type of headache, or features that suggest neurologic dysfunction.

LUMBAR PUNCTURE

PRIMARY HEADACHES

- The characteristics of primary headaches are described in Table 56-2.

MIGRAINE HEADACHES

- Migraine headaches tend to be unilateral, throbbing, or pulsating, last 1 to 48 hours, are often associated with nausea and vomiting, and are relieved with sleep. There is a positive family history in most cases.
- The classification of migraines includes migraine with aura (classic), migraine without aura (common), and childhood periodic syndromes that are commonly precursors of migraine, including abdominal migraines, benign paroxysmal vertigo of childhood, and cyclic vomiting.
- The IHS classification of pediatric migraine without aura, which accounts for 60% to 85% of cases, includes the following:
 - A. At least five attacks fulfilling criteria B to D.
 - B. Headache attacks lasting 1 to 72 hours.

- C. Headache has at least two of the following: either bilateral or unilateral location (frontal/temporal, but not occipital), pulsating quality, moderate-to-severe intensity aggravated by routine physical activities such as walking.
 - D. At least one of the following during the headache: nausea and/or vomiting, photophobia, and phonophobia (may be inferred from behavior).
- Pediatric migraine with aura occurs in 15% to 30% of patients and the diagnostic criteria includes the following:
 - A. At least two attacks fulfilling criteria B to D.
 - B. An aura consisting of at least one of the following: (1) fully reversible visual symptoms such as flickering lights or spots, (2) fully reversible sensory symptoms such as pins and needles or numbness, or (3) fully reversible dysphasic speech disturbances. Motor weakness is not a criterion.
 - C. At least two of the following: (1) homonymous visual symptoms or unilateral sensory symptoms, (2) at least one aura symptom develops gradually over ≥ 5 minutes or different aura symptoms occur in succession ≥ 5 minutes, (3) each symptom lasts ≥ 5 minutes and ≤ 60 minutes.
 - D. Symptoms are not attributable to another disorder.
 - Benign paroxysmal vertigo is a migraine precursor. This occurs in children aged 2 to 6 years and consists of sudden, brief episodes when the child cannot stand upright without support. There is no loss of consciousness, but nystagmus is often seen. The episode lasts for several minutes, and then the child recovers completely.
 - Cyclic vomiting syndrome is recurrent episodes of severe vomiting that last for a few hours to days, separated by symptom-free periods. Treatment includes IV fluids with glucose, antiemetic medication, and sometimes sedation.
 - Abdominal migraine is recurrent episodes of abdominal pain lasting 1 to 72 hours, usually vague, and midline or periumbilical in location. It may be accompanied by nausea, vomiting, anorexia, or pallor.

TABLE 56-2 Primary Headache Characteristics

SYMPTOM	MIGRAINE	TENSION	CLUSTER
Location	Unilateral 60%–70%, bilateral 30%	Bilateral	Always unilateral, begins around eye, temple
Characteristics	Gradual onset, crescendo, pulsating, mod-severe	Pressure, tightness, waxes, and wanes	Begins quickly, crescendo, deep, continuous, excruciating pain
Patient appearance	Prefers rest in dark, quiet room	May remain active, or may rest	Remains active
Duration	4–72 h	Variable	30 min–3 h
Associated symptoms	Nausea, vomiting, photophobia, phonophobia, \pm aura	None	Ipsilateral eye lacrimation, redness, rhinorrhea, sweating, Horner syndrome

TREATMENT

- Reassurance and patient/parental education are the first steps. Patients should keep a headache diary in order to identify any precipitating factors.
- Certain foods such as those that contain tyramine (aged cheese), sodium nitrite (hot dogs and smoked meats), or monosodium glutamate (Chinese food) can precipitate migraines, as can caffeine-containing beverages, chocolate, red wine, and certain drugs (such as oral contraceptives, antihypertensive medications, cimetidine, and H₂ blockers).
- ED treatment of migraines consists of providing analgesia and treating associated symptoms. Analgesics such as acetaminophen or ibuprofen are often effective. If the child is unable to tolerate oral NSAIDs, intravenous ketorolac can be used.

Sumatriptan is a selective 5-HT agonist that can be given subcutaneously (6 mg) or as a nasal spray for children older than 12 years. There are no data regarding oral use of sumatriptan, and other triptans are not approved for children.

- For children older than 10 years who present to the ED with a migraine headache, dihydroergotamine mesylate (DHE) 0.25 to 1.0 mg over 3 minutes intravenously may be beneficial, especially if given with metoclopramide.
- For nausea, antiemetics such as ondansetron, promethazine, metoclopramide, or prochlorpromazine have been used.
- Preventive treatments include cyproheptadine, β -blockers, antidepressants, anticonvulsants, and calcium channel blockers, but conclusive data are lacking regarding their use in children and adolescents.
- Methods such as relaxation therapy and biofeedback may be beneficial.
- Those children who have frequent migraines and those with migraines that are unresponsive to abortive measures should be placed on prophylactic medications after consultation with a neurologist.

TENSION-TYPE HEADACHES

- Tension-type headaches or muscle contraction or stress headaches tend to be chronic and nonprogressive in nature, with the pain described as band-like, bilateral, or generalized. There is no accompanying aura, and nausea is rare.
- The headache can last for 30 minutes to days and can be accompanied by photophobia or phonophobia but is not aggravated by physical activity.
- The diagnosis of tension-type headache is usually based upon clinical criteria, but the differential

diagnosis includes infection, increased intracranial pressure, Chiari I malformation, and chronic sinus infection. The neurologic examination of a child with tension-type headaches is normal.

- Tension-type headaches are generally managed with emotional support, and mild analgesics, such as acetaminophen and ibuprofen. Reassuring the family that the problem is not organic and advising the patient to avoid precipitating factors, such as stress, is an important part of therapy.
- Behavioral techniques, such as biofeedback, and relaxation exercises may be useful.
- In cases of frequent or chronic headaches, treatment with an antidepressant such as amitriptyline may be of benefit.

CLUSTER HEADACHES

- Cluster headaches are uncommon in children younger than 10 years of age. The headache is unilateral, occurs in the frontal or periorbital region, and is often described as ice-pick like.
- The headache lasts 15 minutes to 3 hours, and is associated with ipsilateral lacrimation, redness of the eye, and ipsilateral nasal congestion; the cheek may become flushed and warm. The patient may develop Horner syndrome—miosis, ptosis, and, facial anhidrosis—on the side of the headache. The headache tends to occur at the same time each day during a cluster. Patients are unable to lie down or rest because of the pain.
- Abortive treatment consists of 100% oxygen given for 20 minutes; sumatriptan and octreotide have been used in adults.

SECONDARY HEADACHES

BRAIN TUMORS AND HYDROCEPHALUS

- The presence of a headache that is made worse by lying down, or that comes on with coughing, sneezing, or straining at stool and then disappears, suggests increased intracranial pressure.
- Headaches associated with disorders related to increased intracranial pressure are of a progressive nature. Papilledema is often found on funduscopic examination.
- A complete neurologic examination may disclose other abnormalities, such as ataxia with a cerebellar tumor, or cranial nerve findings with hydrocephalus.
- Differentiation of these disorders is by CT scan or MRI with appropriate consultation if hydrocephalus or a tumor is found.

IDIOPATHIC INTRACRANIAL HYPERTENSION (PSEUDOTUMOR CEREBRI)

- Idiopathic intracranial hypertension or pseudotumor cerebri is a condition associated with increased intracranial pressure in the absence of a mass lesion or other obvious etiology. It results from impaired reabsorption of CSF.
- While many cases are idiopathic, it is associated with obesity, pregnancy, high doses of vitamin A, birth control pills, retinoic acid, tetracycline, infections including mastoiditis and otitis media, endocrinopathies, systemic lupus erythematosus, and steroid withdrawal. Patients may have papilledema on examination.
- Lumbar puncture will reveal an opening pressure >25 cm H₂O, with normal protein, glucose, and cell count.
- Therapy includes treating the underlying cause, serial lumbar punctures to relieve acute symptoms, and acetazolamide 25 mg/kg to reduce the formation of CSF.

HYPERTENSIVE ENCEPHALOPATHY

- Severe elevation in blood pressure can cause headache and if untreated can result in the development of encephalopathy and seizures.
- This should be suspected in a patient with a severe headache whose diastolic blood pressure is greater than the 95th percentile for age.
- In young children, the development of hypertension is often secondary to an acute illness, such as fulminant glomerulonephritis.
- Acute therapy in the ED is based on complete physical examination findings, degree of hypertension, and past history of hypertension. Patients with encephalopathy and seizures require rapid reduction of blood pressure with an agent such as nitroprusside. The patient is admitted for blood pressure control and complete evaluation.

ACUTE HEMORRHAGE

- The child presenting with a severe headache of sudden onset, with or without neck or back pain, may have suffered an intracranial hemorrhage.
- Spontaneous intracranial hemorrhage usually results from either a ruptured aneurysm or arteriovenous malformation. It can also occur in association with coagulopathies.

MENINGITIS/ENCEPHALITIS/BRAIN ABSCESS

- The association of headache with a fever and stiff neck implies an infectious etiology, such as bacterial meningitis, encephalitis, or brain abscess.
- A brain abscess can result from orbital cellulitis, extension from sinusitis, or in children with congenital heart disease caused by right-to-left intracardiac shunt.
- Viral encephalitis can present with mild neck stiffness along with a change in behavior, fever, and headache. Specific etiologies to consider include herpes simplex and West Nile virus.
- If there are no focal findings or signs of increased intracranial pressure on physical examination, a lumbar puncture is performed and will provide the diagnosis. CSF is sent for culture, cell count, protein, and glucose, with bacterial and viral cultures.
- If there are focal neurologic abnormalities or signs of increased intracranial pressure, a CT scan or MRI of the brain with and without contrast is performed prior to a lumbar puncture to avoid the potential for herniation.
- Hospital admission is required for children with these problems; the neurologic examination, serum electrolytes, and fluid status need to be monitored closely.
- Antibiotics are required for bacterial meningitis and brain abscess; acyclovir is the treatment for herpes simplex virus infection.

OTHER ETIOLOGIES OF HEADACHE

- Included in this group are problems that originate outside the calvarium but that can result in headache, either directly or through referred pain. They include sinusitis, otitis media, dental caries or abscess, pharyngitis, temporomandibular joint abnormalities, postconcussion or posttraumatic syndrome, and ophthalmologic problems, such as refractive errors or astigmatism.
- It can also include systemic infections, such as influenza, or strep throat. Toxic exposures, especially to carbon monoxide, can also cause headache.

PSYCHIATRIC/PSYCHOGENIC HEADACHES

- Psychiatric-related or psychogenic headaches tend to be chronic and nonprogressive and are characterized by vague complaints and nonspecific symptoms. They may result from stress, adjustment reactions, conversion reactions, depression, and malingering. In the ED, they are a diagnosis of exclusion.

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QUESTIONS

1. A 6-year-old is brought to the emergency department by concerned parents. The child has been complaining of a headache fairly consistently and recently his mother states the “child does not seem the same”. Which of the following neurologic findings are predictors of a space-occupying lesion in a child with a headache?
 - A. Family hx of migraine
 - B. Blurred vision
 - C. Headache >6 months

- D. Vomiting
 - E. Early morning headache
2. A 12-year-old female presents to the ED with a history of a frontal headache. She has had this headache over a one-month period, and they usually resolve with NSAIDs. Today the headache lasted longer than usual. Her neuro exam and blood pressure are normal. The most appropriate evaluation would be
 - A. Lumbar puncture
 - B. Head CT while in the ED
 - C. Outpatient head CT
 - D. Reassurance
 - E. BP of all four extremities
 3. An 8-year-old male presents to the ED with vomiting. Mom states that when he begins to vomit, he cannot stop, and gets IV fluids, and is usually admitted. The child denies headache, fever, and diarrhea. There are no specific precipitants. His abdominal exam reveals diffuse tenderness, but is not distended, and bowel sounds are present. His neurologic exam is normal. Appropriate treatment for this child in addition to IV fluids includes
 - A. CT abdomen
 - B. Ondansetron
 - C. CT head
 - D. Ketorolac
 - E. Sumatriptan
 4. An 18-year-old female has a history of migraines. She states that they are triggered when she eats hot dogs. Besides meats with sodium nitrite, other foods she should avoid include
 - A. Goat cheese
 - B. Mexican food
 - C. American cheese
 - D. White wine
 - E. Chinese food
 5. A 14-year-old male comes to the ED with a right sided headache. He describes the pain as unilateral and boring. On exam, you note that he has redness and tearing of his right eye, as well as the pupil on the right is smaller than that on the left. There is slight ptosis on the right. The patient says he has never had a headache like this before. The most appropriate therapy is
 - A. CT head
 - B. Sumatriptan
 - C. 100% oxygen
 - D. IV ketorolac
 - E. IV diazepam
 6. A 13-year-old obese female presents to the ED with a headache. She has had this headache off and on for 2 weeks. She has a history of asthma and was

admitted to the hospital, 3 weeks ago, and was discharged on oral steroids for 7 days. On physical exam, she has no focal neurologic findings, her blood pressure is normal, no fever, and her neck is supple. Which of the following are likely findings on lumbar puncture?

- A. WBC of 1000 cells
- B. Protein of 80
- C. Glucose of 10
- D. Opening pressure of 35 cm H₂O
- E. WBC of 100 cells

ANSWERS

1. D. One study identified seven predictors for space-occupying lesions. These included sleep-related headache, absence of family history of migraine, vomiting, absence of visual symptoms, headache less than 6 months, confusion, and an abnormal neurologic examination.

2. D. At this point, since the child has no focal findings, no vomiting, and the only change in headache character is it lasted longer than normal, reassurance is the best treatment. Neuroimaging should be performed for a child with signs of increased intracranial pressure, focal symptoms, an abnormal neurologic examination, skin lesions suggestive of a neurocutaneous syndrome, recent head trauma, and a progressive neurologic disorder

Based on this a CT in the ED is not necessary. An outpatient CT could be arranged based on the AAN practice parameter. The American Academy of Neurology and the Child Neurology Society developed a practice parameter for the evaluation of children and adolescents with recurrent headaches. They stated that routine neuroimaging is not recommended in children with recurrent headaches and a normal neurologic examination. Neuroimaging should be considered in the child with headache who has an abnormal neurologic examination consisting of focal findings, signs of increased intracranial pressure, altered level of consciousness, or coexistent seizures or both. Neuroimaging should be considered in children with historical features to suggest recent onset of severe headache, change in the type of headache, or features that suggest neurologic dysfunction. The child has no findings that suggest pseudotumor or meningitis that would prompt a lumbar puncture. This child does not have elevated blood pressure, so coarctation is less likely.

3. B. Cyclic vomiting syndrome is recurrent episodes of severe vomiting that last for a few hours to days, separated by symptom-free periods. Treatment

includes IV fluids with glucose, antiemetic medication, and sometimes sedation. A head or abdominal CT are not necessary. Even though these are a migraine variant, sumatriptan is not the medication of choice. A NSAID will not improve this patient's condition either.

4. E. Certain foods such as those that contain tyramine (aged cheese), sodium nitrite (hot dogs, smoked meats), or monosodium glutamate (Chinese food) can precipitate migraines, as can caffeine-containing beverages, chocolate, red wine, and certain drugs (such as oral contraceptives, antihypertensive medications, cimetidine, and H₂ blockers).

Although many restaurants specifically say they do not use monosodium glutamate (MSG), this food additive is a well-known trigger for migraine headaches. Goat cheese is usually fresh and without preservatives. Mexican food does not use MSG, white wine does not contain the caffeine contained in red wine, and American cheese, while processed, is not aged.

5. C. This patient has a classic cluster headache. The headache is unilateral, occurs in the frontal or periorbital region, and is often described as ice-pick like. The headache lasts 15 minutes to 3 hours, and is associated with ipsilateral lacrimation, redness of the eye, and ipsilateral nasal congestion; the cheek may become flushed and warm. The patient may develop Horner syndrome—miosis, ptosis, and, facial anhidrosis—on the side of the headache. The headache tends to occur at the same time each day during a cluster. Patients are unable to lie down or rest because of the pain. Abortive treatment consists of 100% oxygen given for 20 minutes. Although sumatriptan and octreotide have been used in adults, they have not been studied in children. IV ketorolac is unlikely to help. IV diazepam may make the patient sleepy, but will not abort the headache. Because the findings are classic, a head CT is not needed.

6. D. Idiopathic intracranial hypertension or pseudotumor cerebri is a condition associated with increased intracranial pressure in the absence of a mass lesion or other obvious etiology. It results from impaired reabsorption of CSF. While many cases are idiopathic, it is associated with obesity, pregnancy, high doses of vitamin A, birth control pills, retinoic acid, tetracycline, infections including mastoiditis and otitis media, endocrinopathies, systemic lupus erythematosus, and steroid withdrawal. Patients may have papilledema on examination. Lumbar puncture will reveal an opening pressure >25 cm H₂O, with normal protein, glucose, and cell count.

57 HYDROCEPHALUS

Susan Fuchs

INTRODUCTION

- Hydrocephalus refers to the excess accumulation of CSF. This can occur because of obstruction of CSF flow, reduced absorption, or excess production. Most (70%) of CSF is produced by the choroid plexus and absorbed by the arachnoid villi and granulations CSF is absorbed through arachnoid villi covering the brain and leptomeninges, across the ependymal lining of the ventricles, and the spinal subarachnoid space.
- Most cases of hydrocephalus result from congenital or acquired obstructions to the flow of CSF from the brain to the spinal canal. Congenital malformations include the Arnold–Chiari malformation, which is elongation and downward displacement of the brain stem into the fourth ventricle, and the Dandy–Walker malformation, which is a posterior fossa cyst that causes obstruction at the outlet of the fourth ventricle at the foramen of Luschka and Magendie.
- Intrauterine infections such as toxoplasmosis, rubella, cytomegalovirus, herpes, and syphilis can lead to hydrocephalus through inflammation of the ependymal lining of the ventricular system and lead to CSF flow obstruction. Intraventricular hemorrhage in preterm newborns can also lead to hydrocephalus.
- Beyond the neonatal period, the most common causes of acquired hydrocephalus are mass lesions, which include tumors, cysts, and abscesses. Other acquired causes of hydrocephalus are meningitis, encephalitis, posthemorrhagic adhesions, and vascular malformations.

CLINICAL PRESENTATION

- The clinical presentation of hydrocephalus depends on the age of the patient and the rate at which it develops.
- Infants with hydrocephalus are often diagnosed on routine examination by finding head circumference disproportionately large for age or splitting of the cranial sutures.
- An infant may initially experience irritability, poor feeding, or other behavioral changes. When intracranial pressure becomes severely elevated, the infant develops vomiting and lethargy, which can signal impending herniation. In addition to split sutures, the physical examination may reveal a bulging anterior fontanel and engorged scalp veins. Dysfunction of cranial nerve III may result in loss of upward

gaze, or the “sundown or setting-sun” sign. Bobble-head doll movements may also occur, especially with aqueductal stenosis or a third ventricle cyst.

- Older children with hydrocephalus will usually complain of headache, which is often progressive in nature, worse in the morning, awakens the patient from sleep, and is exacerbated by lying down or straining. Gait disturbances can occur, especially ataxia, which is characteristic of children with posterior fossa tumors.
- Older children develop vomiting as intracranial pressure begins to become severely elevated. Papilledema is a late finding in children and is rarely found in infants, but it implies a severe increase in intracranial pressure.

MANAGEMENT

- The primary goal of management of the child with hydrocephalus is the assessment and control of elevated intracranial pressure.
- Cushing’s triad of hypertension, bradycardia, and abnormal respiratory patterns is a late sign of elevated intracranial pressure.
- Specific signs of herniation depend on the part of the brain involved. In uncal herniation, there is compression of the third cranial nerve with dilation of the ipsilateral pupil and contralateral hemiparesis. Herniation of the cerebellar tonsils through the foramen magnum is preceded by headache and stiff neck and characterized by fixed, dilated pupils. The loss of leg function on one side suggests herniation under the falx. Central herniation occurs when both cerebral hemispheres compress the midbrain and results in decreased level of consciousness, constricted pupils, and Cheyne–Stokes respirations.
- Patients who are lethargic on presentation, those with a Glasgow Coma Scale <8, or those who deteriorate in the emergency department are intubated following rapid sequence induction. Prior to intubation, ventilation with a bag-valve-mask device to attain a P_{CO_2} of 35 torr may provide sufficient cerebral vasoconstriction to reduce intracranial pressure enough to avert herniation. If there are signs of herniation such as unequal pupils, fixed and dilated pupils, or posturing, mild hyperventilation can be helpful on a short-term basis.
- Patients who do not respond with an improved mental status to intubation and ventilation may benefit from diuretic therapy with mannitol (0.25–1 g/kg) or furosemide (1 mg/kg). It is appropriate to elevate the head of the bed 15° to 30°.
- After the patient is stabilized, a computed tomography scan or magnetic resonance image of the brain

is performed to define the lesion and plan definitive treatment.

- Those children with hydrocephalus due to an obstruction in CSF flow, require insertion of a catheter from the ventricle to a distal site by a neurosurgeon.
- If the cause of the increased intracranial pressure is due to trauma resulting in bleeding or cerebral edema, therapy usually includes placement of an intracranial pressure monitoring device by a neurosurgeon. In dire circumstances, a percutaneous ventricular tap may be performed.

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QUESTIONS

1. A 4-month-old female with known mild hydrocephalus is brought to the ED for vomiting and increased irritability. The parents state she is flown by a neurosurgeon at a children's hospital 40 miles away. On examination you note that her eyes constantly look down. The parents say this is new. Her vital signs are HR 120 RR 30, BP 90/50. The most appropriate management plan for this infant is
 - A. Head CT and admit to your hospital
 - B. Arrange transfer to the children's hospital
 - C. Intubate then transfer to the children's hospital
 - D. Perform MRI then transfer
 - E. Have parents drive patient to children's hospital
2. A 12-year-old male with a right-sided ventriculoperitoneal shunt presents to the ED with lethargy, and vomiting. On exam he has left pupil is 6 mm and nonreactive, his right pupil is 4 mm and reactive. His HR is 40, RR 10, BP 160/90. The most important treatment at this time is
 - A. Mannitol
 - B. Furosemide

- C. Begin bag mask ventilation and prepare to intubate via RSI
- D. Intubate without medications
- E. Begin bag mask ventilation, and get Pco₂ to 20

ANSWERS

1. B. Since this infant's vital signs are stable, even though she is "sun-setting", the most appropriate management is to transfer this child to the hospital where she is being followed by a neurosurgeon. Based on their knowledge of this patient and her new symptoms, they may need to insert a ventriculo-peritoneal shunt. The specific method of transfer (advanced life support ambulance or specialized pediatric critical care unit) can be discussed with the receiving hospital based on their capabilities. Admitting her to your hospital, will not fix her problems unless a neurosurgeon at your hospital will treat her. An MRI is not necessary and will delay transfer. She does not need to be intubated at this time, as her vital signs are stable. The parents should not drive this infant in their car to the hospital.
2. C. This patient is showing signs of herniation, probably due to a VP shunt malfunction. His heart rate is low, BP elevated, and respirations slow (Cushing's triad). The best treatment is to begin bag mask ventilation, as this patient may not be ventilating effectively, and is at risk for hypoxia. While ventilations are being performed, preparation for intubation using rapid sequence induction medications should be initiated. Intubating this patient without medication will likely increase his increased intracranial pressure. Ventilating him to a Pco₂ of 20 is too much. He can be ventilated to a Pco₂ of 35, watching for changes in his papillary findings. Mannitol, and furosemide can both reduce the amount of CSF, but this patient needs his ABCs taken care of first.

58 CEREBRAL PALSY

Susan Fuchs

INTRODUCTION

- Cerebral palsy (CP) is a nonprogressive motor disorder reported to occur in 1.2 to 2.5 per 1000 children. It is usually evident within the first 3 to 4 years of life. The injury that results in CP can occur during the antepartum, peripartum, or postnatal period.

Prematurity remains the most important risk factor, but there is also a higher risk of CP with multiple births, even those not born prematurely.

- The task force on Neonatal Encephalopathy has developed four essential criteria for asphyxial CP: evidence of metabolic acidosis, early onset of severe or moderate neonatal encephalopathy in infants older than 34 weeks gestation, CP of the spastic quadriplegia or dyskinetic type, and exclusion of other identifiable etiologies. They have also developed a list of peripartum events that may be related to the development of CP, but are not specifically asphyxial in nature: a sentinel hypoxic event occurring immediately before or after delivery, a sudden and sustained fetal bradycardia or absence of fetal heart rate variability, Apgar score of 0 to 5 at 5 minutes, onset of multisystem involvement within 72 hours, and early imaging studies showing evidence of an acute nonfocal cerebral abnormality.
- Other etiologies of CP include congenital abnormalities, brain malformations, stroke in the perinatal period, intracranial hemorrhage, intrauterine infection, premature birth, genetic abnormalities, metabolic abnormalities, and kernicterus.
- Although the disorder is not in itself an emergency department diagnosis, children with CP have associated problems that often result in emergency department visits. The emergency department physician must realize that each child with CP has different abilities and problems and that each family has different parent–child relationships and coping mechanisms.

CLINICAL PRESENTATION

- There are several forms of CP, with the classification systems based on the extremities involved, tone, and the ability to perform normal activity. The major disorder is of muscle tone, but there can also be neurologic disorders such as seizures, vision disturbances, and impaired intelligence.
 - Spastic CP includes several variants: spastic quadriplegia, spastic diplegia, and spastic hemiplegia. Spastic CP is the most common variant, with 70% to 80% of children with CP in one of these groups.
 - Spastic quadriplegia is characterized by a generalized increase in muscle tone, deep tendon reflexes, and rigidity of the limbs on both flexion and extension. Although the lower extremities are generally more severely affected, in severe forms the child is stiff and assumes a posture of decerebrate rigidity. Many children have pseudobulbar

involvement, resulting in swallowing difficulties and recurrent aspiration. Intellectual impairment is severe, and half have a tonic–clonic seizure disorder.

- Spastic diplegia is characterized by bilateral spasticity, with greater involvement of the lower extremities than the upper. It is the most common form of CP diagnosed in preterm infants. Early in life, when rigidity predominates, the legs are held in extension and in a scissored pattern because of adductor spasm. As spasticity progresses, flexion of the hips and knees develops, ultimately leading to contractures. In those less severely affected, dorsiflexion of the feet with increased ankle tone results in toe-walking. Other manifestations include convergent strabismus, delayed speech development, and seizure disorders. Intellectual impairment parallels the motor deficit.
- Spastic hemiplegia (hemiparesis) is a unilateral paresis that usually affects the upper extremity more than the lower. Some degree of spasticity and flexion contracture usually results. One of the initial symptoms is fisting, which is an exaggerated palmar grasp reflex. The gait may be circumductive, with swinging of the affected leg like an arc. The extent of functional impairment varies, with fine movements of the hand affected most. Sensory impairment, growth disturbance, and involuntary movements of the affected limbs can occur. In addition, facial weakness, visual disturbances, and seizures can occur.
- Another classification of CP is extrapyramidal or dyskinetic, which accounts for 10% to 15% of cases. Dyskinesia is difficulty performing voluntary movements.
- Athetoid CP involves slow, continuous writhing motions and usually involves the distal limbs.
- Ballismus is characterized by violent, jerky motions of the arms and legs.
- Choreic CP movements, which are rapid and irregular muscle contractions, result from disorganized tone and can involve the limbs, face, or trunk. Those with tremulous CP have involuntary, rhythmic contractions of opposing muscles.
- Dystonic CP results in repetitive twisting movements of the limbs and trunk, although the head and neck can also be involved.

COMPLICATIONS

- The most common problem in CP patients presenting to the emergency department is breakthrough seizures.

- Respiratory difficulties also commonly present to the emergency department. Chronic aspiration can result in reactive airway disease and, for some patients, chronic hypoxia, and hypercarbia. Acute pneumonia is common after aspiration and is often difficult to diagnose in patients whose baseline chest radiographs are abnormal. Poor coughing mechanisms contribute to pulmonary pathology. In CP patients with evidence of pneumonia, aggressive antibiotic therapy is indicated. Admission to the hospital may be necessary if the child is unable to take oral antibiotics or needs chest physiotherapy or supplemental oxygen.
- Many children with CP have significant feeding difficulties that require placement of a gastrostomy tube or button, with or without a fundoplication, (often used when aspiration is a risk). In some patients, a gastrojejunal feeding tube is inserted. Feeding tube malfunction is common.
- Many patients with CP who are significantly impaired are not toilet trained and are vulnerable to urinary tract infections and perineal skin breakdown that can result in infection.
- In addition to a chest radiograph to rule out pneumonia, all febrile CP patients require a urinalysis and urine culture.

is 39.0°C, and you hear rales in the right base. A chest radiograph confirms pneumonia. The most likely etiology of his problems is

- Aspiration pneumonia
 - Mic-Key (Gtube) malfunction
 - Acute reactive airway disease
 - Seizure disorder
 - Foreign body aspiration
2. A 4-year-old with CP presents to the ED with vomiting at midnight. The child has a GJ tube for all feeds due to chronic aspiration, and also has seizures, which have increased with the vomiting. An abdominal x-ray reveals that the GJ tube tip is out of position; it is in the stomach. Interventional radiology will not replace the tube at this time. The most appropriate treatment at this time is
 - Try to reposition the GJ tube yourself.
 - Remove the GJ tube and place a G tube for now, and discharge home.
 - Insert and IV and admit the patient to the hospital.
 - Discharge and have Mom return with the patient for a new GJ tube in the morning.
 - Transfer to another hospital.

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QUESTIONS

1. A 7-year-old male with CP presents to the ED with breathing difficulties. The child is non-verbal, and has developmental delay. He has a Mic-key (G tube) for feeds, and has been vomiting. In the ED his temp

ANSWERS

1. A. Children with CP commonly present to the ED with respiratory difficulties. Chronic aspiration can result in reactive airway disease and, for some patients, chronic hypoxia, and hypercarbia. Acute pneumonia is common after aspiration and is often difficult to diagnose in patients whose baseline chest radiographs are abnormal. Poor coughing mechanisms contribute to pulmonary pathology. The Mic-Key (G tube) malfunction may allow food to be aspirated, but is not the direct cause of respiratory problems. A seizure can result in aspiration of mucus, but there is no history to support this. Foreign body aspiration is less likely.
2. C. This patient requires either a GJ tube or an IV for seizure medications. A GJ tube must be placed under fluoroscopic guidance to ensure that it is in the jejunum. Because this patient has chronic aspiration, placing a G tube will just lead to aspiration problems. Having the patient go home and return in the morning is not appropriate, as she needs her seizure medication. Transferring this patient to another hospital is also inappropriate, unless you are sure the GJ tube will be replaced right away.

59 CEREBROVASCULAR SYNDROMES

Susan Fuchs

INTRODUCTION

- In U.S. children, hemorrhagic strokes are more common than ischemic strokes.
- Ischemic strokes can be categorized as arterial ischemic strokes (AIS) and cerebral sinovenous thrombosis (CSVT). In the pediatric population, arterial ischemic stroke usually results from a thromboembolism. Occlusion of venous sinuses or cerebral veins can result in cerebral sinovenous thrombosis. The peak age of AIS is neonatal and childhood, whereas the peak age of CSVT is neonatal and adolescence.
- The signs of AIS vary with age and the area of the brain affected by ischemia; they include focal neurologic findings such as hemiparesis, cranial nerve palsies, visual field deficits, and aphasia. Seizures may or may not occur. CSVT may present with diffuse neurologic signs and seizures, but symptoms vary with age and etiology. Infants present with seizures, jitteriness, or lethargy, while older children have headaches, vomiting, possible seizures, fever, and focal neurologic deficits.
- The arterial circulation to the brain is via the anterior carotids and the posterior vertebral and basilar arteries, which link via communicating arteries to form the circle of Willis. Cerebral arteries can thrombose due to damage to the arterial wall, emboli, or prothrombotic conditions. Infarction occurs when loss of blood supply to cerebral tissue results in ischemia, hypoxia, and depletion of energy and carbohydrate stores. The extent of neuronal damage depends on the severity and length of time of ischemia, the availability of collateral circulation, and the metabolic needs of the brain.
- In children, risk factors associated with AIS include cardiac disease, coagulation and hematologic disorders, infection, vasculitis, cancer, metabolic disorders, moyamoya, sickle cell anemia, and perinatal complications.
- Sinovenous thrombosis can occur due to thrombophlebitis, hemoconcentration, or coagulation abnormalities.
- Risk factors associated with CSVT are prothrombotic disorders, dehydration, systemic infection, otitis media, mastoiditis, sinusitis, hematologic disorders, drugs, cardiac disease, cancer, and perinatal complications.
- The underlying diseases that cause AIS and CSVT are listed in Tables 59-1 and 59-2, respectively.

- Hemorrhagic strokes involve the rupture of cerebral blood vessels with leakage of blood into the brain parenchyma, subarachnoid space, or ventricular system. The location of the hemorrhage defines the two major types of stroke as intracerebral/intraparenchymal or subarachnoid, and determines the pathophysiology, risk factors, and clinical findings.
- Intracerebral hemorrhage occurs when arteries or veins rupture into intracerebral areas or brain parenchyma. In children, the greatest risk factor is head trauma, followed by aneurysms and vascular malformations.
- Subarachnoid hemorrhage results from rupture of an aneurysm (usually proximal arteries at the circle of Willis) or an arteriovenous malformation (AVM). Risk factors include disorders associated with vascular malformations, aneurysms, sickle cell disease, and hypertension. However, hemorrhagic strokes are occasionally associated with systemic diseases and coagulopathies. These conditions are summarized in Table 59-3.

DIAGNOSIS

HISTORY

- AIS usually have a rapid onset, so there may be little in the history to warn of the impending event. Patients often suffer sudden seizures, or focal neurologic findings, especially hemiplegia. A history of recurrent headaches, transient ischemic attacks, or focal seizures may be obtained, but these do not provide a specific diagnosis, and often confuse the issue. An AIS involving a large vessel may present with loss of consciousness and multiple focal neurologic deficits.
- An older child with a CSVT may present with slowly progressive signs, such as fever, vomiting, or headache. A young infant may have dilated scalp veins, eyelid swelling, and a large anterior fontanel.
- An older child with a hemorrhagic stroke may have a history of severe headache or, especially in the case of subarachnoid hemorrhage, neck pain. A large bleed will usually result in a sudden alteration in consciousness and perhaps seizures, but a small bleed may result in subtle focal neurologic signs, including cranial nerve palsies.
- A history of cardiac disorders, especially complex congenital heart disease, prosthetic heart valve, or recent cardiac surgery should raise suspicion of an embolic phenomenon.
- Inherited coagulation disorders such as deficiency of protein C, protein S, antithrombin III, and plasminogen, or the presence of factor V Leiden,

TABLE 59-1 Predisposing Condition for Ischemic Stroke

Cardiac	Antithrombin III deficiency
Congenital heart disease (s/p Fontan, ECMO)	Plasminogen deficiency
Rheumatic heart disease	Pregnancy
Bacterial endocarditis	Congenital prothrombotic disorders
Arrhythmias	Factor V Leiden gene defect
Cardiomyopathy	Prothrombin gene G20 210A mutation
Prosthetic heart valves	Methyltetrahydrofolate reductase
Ventriculoseptal defect/atrial septal defect	Antithrombin III deficiency
Patent foramen ovale	Trauma
Infection	Head injury
Meningitis	Neck injury
Encephalitis	Intraoral trauma
Vasculopathy	Child abuse
Moyamoya disease	Drugs
Postvaricella angiopathy	Cocaine
Postradiation vasculopathy	Oral contraceptives
Transient cerebral arteriopathy	Antineoplastic agents (e.g., L-asparaginase)
Systemic disorders	Steroids
Systemic lupus erythematosus	LSD
Polyarteritis nodosa	Amphetamines
Nephrotic syndrome	Metabolic disorders
Inflammatory bowel disease	Hyperhomocystinuria
Takayasu's arteritis	Mitochondrial encephalomyopathy (MELAS)
Dermatomyositis	Hyperlipidemia
Rheumatoid arthritis	Cerebral autosomal dominant arteriopathy with subcortical
Diabetes mellitus	infarct and leukoencephalopathy (CADASIL)
Hematologic disorders	Neurocutaneous syndromes
Sickle cell disease	Neurofibromatosis
Leukemia	Sturge–Weber syndrome
Iron deficiency anemia	Tuberous sclerosis
Polycythemia	Hereditary disorders
Thrombocytosis	Ehlers–Danlos syndrome
Thrombotic thrombocytopenic purpura	Fabry's disease
Idiopathic thrombocytopenic purpura	Vasospastic disorders
Acquired prothrombotic states	Migraine
Lupus anticoagulant	
Anticardiolipin antibodies	
Protein S deficiency	
Protein C deficiency	

Data from DeVeber Ga. Cerebrovascular disease. In: Swaiman KF, Ashwal S, Ferriero DM, eds. *Pediatric Neurology: Principles and Practice*. 4th ed. Philadelphia, PA: Mosby/Elsevier; 2006:1759–1801.

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anticardiolipin antibody, or lupus anticoagulant can all lead to thromboembolism.

- A history of sickle cell disease is extremely important to elicit, because 25% of patients will develop cerebrovascular problems.
- Systemic lupus erythematosus and other forms of vasculitis such as polyarteritis nodosa, mixed connective tissue disease, or Takayasu's arteritis have all been associated with arterial ischemic and sinovenous thrombosis.
- Metabolic disorders, such as homozygous homocystinuria (hyperhomocysteinemia), which have a thrombotic effect, can cause arterial and venous throm-

bosis. Fabry's disease (deficiency of α -galactosidase A) can lead to lacunar infarcts, and hyperlipidemia has also been associated with childhood strokes.

- The MELAS syndrome (mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episodes) is characterized by episodes of nausea, vomiting, headaches, seizures, and hemiparesis, which initially resolve, but ultimately lead to persistent deficits and cortical blindness.
- Neurocutaneous disorders such as neurofibromatosis, Sturge–Weber syndrome, and tuberous sclerosis are all associated with both ischemic and hemorrhagic strokes.

TABLE 59-2 Risk Factors for CSVT

Septic
Otitis media
Mastoiditis
Sinusitis
Indwelling central lines
Prothrombotic
Protein S deficiency
Protein C deficiency
Antiphospholipid antibodies
Antithrombin III deficiency
Factor V Leiden gene defect
Prothrombin G20 210A mutation
Hyperhomocysteinemia
Dehydration
Systemic infection
Systemic lupus erythematosus
Nephrotic syndrome
Hematologic disorders
Polycythemia
Malignancy
L-asparaginase treatment
Cardiac disease
Oral contraceptives
Head injury
Pregnancy

Data from DeVeber Ga. Cerebrovascular disease. In: Swaiman KF, Ashwal S, Ferriero DM, eds. *Pediatric Neurology: Principles and Practice*. 4th ed. Philadelphia, PA: Mosby/Elsevier; 2006:1759–1801.

Data from Ferro JM, Canhao P. Etiology, clinical features, and diagnosis of cerebral venous thrombosis. In: Rose BD, ed. *UpToDate*. Waltham, MA: UpToDate; 2008.

Data from Goldenberg NA, Bernard TJ. Venous thromboembolism in children. *Pediatr Clin North Am*. 2008;55:305–322.

- Adolescents in particular are questioned regarding illicit drug ingestion, particularly cocaine. Additional questions are directed toward detecting one of the underlying etiologies noted in Tables 59-1, 59-2, and 59-3.

PHYSICAL EXAMINATION

- Stabilization of the patient is the first priority, since seizures may occur in younger children at the time of or shortly after the stroke. Complete vital signs include temperature and blood pressure. If trauma is suspected, the head and neck are immobilized. A thorough examination includes auscultation over the head, eyes, and carotid arteries listening for bruits, as well as a careful auscultation of the heart for murmurs, clicks suggestive of valvular disease, arrhythmias, or indications of prior cardiac surgery. The eyes are examined for extraocular movements, pupillary responses, and the visual fields. The eyes will look toward the lesion if the cerebral hemisphere is involved, but away with brain stem involvement. The skin is examined for petechiae, café au lait spots, neurofibromas, or telangiectasias.

TABLE 59-3 Conditions Predisposing to Hemorrhagic Stroke

Vascular malformations
Aneurysms
Arteriovenous malformations
Cavernous malformation
Coagulation defects
Hemophilia
Disseminated intravascular coagulation
Idiopathic thrombocytopenia purpura
Vitamin K deficiency
Anticoagulation treatment
Leukemia
Aplastic anemia
Factor deficiencies
Systemic disorders
Hypertension
Sickle cell disease
Drug
Amphetamines
Phenylpropanolamine
Cocaine
Head trauma
Child abuse
Brain tumors
Infection
Herpes simplex
Varicella
Congenital syndromes
Ehlers–Danlos Syndrome
Neurofibromatosis
Tuberous sclerosis

With permission from DeVeber Ga. Cerebrovascular disease. In: Swaiman KF, Ashwal S, Ferriero DM, eds. *Pediatric Neurology: Principles and Practice*. 4th ed. Philadelphia, PA: Mosby/Elsevier; 2006:1759–1801. Copyright Elsevier 1997.

- Neurologic assessment includes determination of degree of weakness, cranial nerve dysfunction, and the side and extent to which the extremities are involved. If the facial muscles and tongue are involved, there is dysarthria, but involvement of the basal ganglia, thalamus, or cerebral hemispheres can result in aphasia. It may be difficult to assess sensory impairment due to aphasia.

DIAGNOSTIC EVALUATION

- Baseline laboratory studies include a complete blood count with differential and platelet count and coagulation studies. If sickle cell disease is a possibility, a sickle cell preparation and hemoglobin electrophoresis are performed. Further coagulation studies are indicated if hemophilia or other coagulopathies, such as protein S or C or antithrombin III deficiencies, are suspected.
- Additional studies should include electrolytes, BUN, creatinine, glucose, sedimentation rate, and CRP, as

well as a urinalysis looking for red cells or protein, and a urine pregnancy test in females.

- Studies evaluating a hypercoagulable/prothrombotic state include antinuclear antibodies, protein S, protein C, factor V Leiden mutation, prothrombin 20210A, antithrombin III activity, homocysteine concentration, and anticardiolipin levels.
 - If a fever is present, blood culture, urine culture, and CSF studies are indicated. Blood and CSF viral titers of varicella zoster, herpes zoster, EBV enterovirus, and parvovirus may be helpful.
- If a metabolic disorder is suspected, blood lactate, pyruvate, carnitine, and serum amino acids are ordered, and urine is sent for organic acids.
- An electrocardiogram and an echocardiogram should be performed on all children in whom underlying heart disease is suspected.
- Magnetic resonance imaging (MRI) is more sensitive than a computed tomography (CT) scan in detecting small infarcts, infarcts of the brain stem and cerebellum, and infarcts that become hemorrhagic. A CT scan will show a tumor or abscess, and may show loss of gray/white differentiation and the dense triangle sign (hyperdense thrombus in posterior part of superior sagittal sinus). However, a CT scan may not detect a small acute hemorrhage. CT scan is also normal in up to 30% of CSVT, so MRI is the preferred study in these cases.
- Magnetic resonance angiography (MRA) correlates well with angiography, and can be done at the time of the MRI to visualize the flow through the cerebral arteries. MRI can also be used with magnetic resonance venography (MRV) to diagnose CSVT. The visualization of a thrombus and the absence of a flow-related signal provide the diagnosis
- The gold standard to visualize intracranial and extracranial vessels is cerebral angiography.
- For patients in whom a hemorrhagic stroke is suspected, in whom the CT scan is negative, and who have no signs of increased intracranial pressure, a lumbar puncture is indicated. Particularly in a small subarachnoid hemorrhage, the CT scan may not reveal blood. The CSF is evaluated for the presence of red blood cells, which, in the absence of a traumatic lumbar puncture, indicates hemorrhage, especially if the blood does not clear during CSF collection. In some cases, the CSF may appear xanthochromic, which is also consistent with hemorrhage.

TREATMENT

- The key function of the ED is stabilization of the patient's respiratory and cardiovascular status, especially the blood pressure. In the event of an ischemic

infarct, a precipitous decline in blood pressure is avoided, since it can worsen cerebral ischemia. If hypotension is present, careful fluid resuscitation and inotropic support may be needed.

- If there are signs of impending herniation, mannitol (0.25 to 1 g/kg intravenously over 20 minutes) and controlled ventilation may be required. Serum glucose should be monitored closely because hypoglycemia can worsen the effect of the stroke, and hyperglycemia can increase the infarct size. Maintenance of normal body temperature is also important, since hyperthermia can worsen ischemic brain damage. The patient should be kept with the bed at 0° to 15° of head elevation for AIS.
- Specific therapy is directed at the etiology of the stroke, such as correction of clotting abnormalities, antibiotics for infections, antiepileptic medication for seizures, and surgery for evacuation of a hematoma.
- In patients with sickle cell disease, hydration and exchange transfusion to reduce hemoglobin S to <30% is indicated (see Chapter 2).
- Depending on the etiology of the stroke in children, there is some use for antithrombotic agents, but dosing and efficacy still need to be determined for some therapies.

DISPOSITION

- Children who have suffered strokes are admitted to an intensive care setting for close monitoring of blood pressure, fluid status, temperature, glucose, neurologic function, and antithrombotic therapy.

OUTCOME/DISABILITY

- For children with AIS, a middle cerebral artery territory stroke volume >10% of intracranial volume and initial presentation with altered level of consciousness are predictors of poor outcome. The risk of recurrence ranges from 6% to 40%; the presence of vasculopathy appears to be an important risk factor.
- For CSVT, outcome depends on etiology, but mortality ranges from 4% to 20%, with a recurrence rate of 17%.
- For children with sickle cell disease, effective prevention or recurrent stroke includes chronic transfusion therapy, and hydroxyurea.

SPECIFIC CONDITIONS

MOYAMOYA

- Moyamoya is characterized by an abnormal network of small collateral vessels, which develop due to

idiopathic stenosis and occlusion of large cerebral arteries involving the circle of Willis. The “puff of smoke” appearance on angiogram is due to the collateral vessels. The etiology is unknown, but there is a relatively high incidence in Asian and Japanese populations.

- In children, the presentation is more often with ischemic events such as TIA or stroke, rather than a hemorrhagic stroke and seizures, which occur more in adults. The prognosis in children is worse in children <3 years of age compared to patients between 3 and 6 years; both age groups have a worse prognosis than adults.
- Moyamoya syndrome includes the typical angiographic pattern, but is secondary to an etiology such as trisomy 21, sickle cell disease, and cranial radiation.

POSTVARICELLA ANGIOPATHY AND TRANSIENT CEREBRAL ARTERIOPATHY

- In children with AIS, varicella-associated angiopathy accounts for one-third of cases. The mechanism is thought to be a transient acute vasculitis. Characteristic findings include basal ganglia infarction and intracranial narrowing of the distal internal carotid or proximal anterior, middle, or posterior cerebral arteries.
- Transient cerebral angiopathy is characterized by unilateral focal or segmental stenosis of the distal carotid arteries and vessels in the proximal circle of Willis vessels. These lesions may resolve or stabilize within 6 months. This results in infarction in the internal capsule or basal ganglia. The average age of onset is 5 years, and etiology is thought to be viral, and when preceded by varicella within 12 months is termed postvaricella angiopathy.

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QUESTIONS

1. A 3-month-old male infant presents via EMS after having a 30-second seizure at home. The child had stopped seizing on EMS arrival, but has been very lethargic since. Blood glucose was 100. The infant had been born at 30 weeks and spend 4 weeks in the NICU on a ventilator, then additional time there for feeding issues and to gain weight. He was discharged 2 weeks ago. He has been afebrile, but not taking formula well for the past 2 days. There are no focal findings on exam, and a head CT shows no bleed or hydrocephalus. The next most appropriate step is
 - A. Lumbar puncture
 - B. MRA/MRV
 - C. Head CT with contrast
 - D. Metabolic work up
 - E. Acyclovir
2. A 6-year-old male with sickle cell disease presents to the ED with a headache and new onset of left sided weakness. He is able to talk to you, and has no fever, or neck pain. Appropriate diagnostic studies include which of the following?
 - A. MRI
 - B. EEG
 - C. CT
 - D. ECG
 - E. Lumbar puncture

ANSWERS

1. B. CSVT may present with diffuse neurologic signs and seizures, but symptoms vary with age and etiology. Infants present with seizures, jitteriness, or lethargy. Because this child likely had an umbilical artery catheter in place for a long time while on the ventilator, thrombosis is likely, especially now that he is not drinking and could be dehydrated. MRI is more sensitive than a CT scan in detecting small

infarcts, infarcts of the brain stem and cerebellum. A CT scan will show a tumor or abscess, and may show loss of gray/white differentiation and the dense triangle sign (hyperdense thrombus in posterior part of superior sagittal sinus). However, a CT scan may not detect a small acute hemorrhage. CT scan is also normal in up to 30% of CSVT, so MRI is the preferred study in these cases. MRA correlates well with angiography, and can be done at the time of the MRI to visualize the flow through the cerebral arteries. MRI can also be used with MRV to diagnose CSVT. The visualization of a thrombus and the absence of a flow-related signal provide the diagnosis. A CT with contrast will not provide any additional information, and a metabolic work-up may be helpful to ensure that the infant does not have a prothrombotic state such as hyperhomocysteinemia. Acyclovir would be appropriate if he has herpes meningitis/encephalitis, but the history does not support this.

2. A. Since this child has sickle cell disease, he is at risk of having a stroke. The signs of AIS vary with age and the area of the brain affected by ischemia; they include focal neurologic findings such as hemiparesis, cranial nerve palsies, visual field deficits, and aphasia. Seizures may or may not occur. MRI is more sensitive than a CT scan in detecting small infarcts, infarcts of the brain stem and cerebellum, and infarcts that become hemorrhagic. A CT scan will show a tumor or abscess, and may show loss of gray/white differentiation and the dense triangle sign (hyperdense thrombus in posterior part of superior sagittal sinus). However, a CT scan may not detect a small acute hemorrhage. CT scan is also normal in up to 30% of CSVT, so MRI is the preferred study in these cases. A lumbar puncture may be appropriate after the MRI, based on the findings. An EEG is not needed, as the child had no seizure. An ECG will not help in the diagnosis.

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Section 8

INFECTIOUS EMERGENCIES

60 INFLUENZA

Karen C. Hayani
Arthur L. Frank

INTRODUCTION

- Influenza is made up of single-stranded RNA surrounded by an envelope, which contains proteins M1 and M2, as well as glycoproteins, 80% of which are hemagglutinin (HA) and 20% neuraminidase (NA).
 - The HA and NA glycoproteins determine the strain of the virus.
 - Minor changes in these glycoproteins are called antigenic drift (occurs yearly).
 - Major changes in these glycoproteins are called antigenic shift (cause worldwide pandemics).
 - Antigenic drift and shift are important in the spread of influenza because new strains are able to evade the host immune system (Table 60-1 and Fig. 60-1).

EPIDEMIOLOGY AND TRANSMISSION

- Influenza is found worldwide.
- In temperate areas, increased incidence usually in winter.
- Usually spreads by inhalation of infected droplets or aerosols (from cough or sneeze).
- Also spreads by direct contact with infected animals or fomites.
- Viral shedding lasts about 7 days.
 - Shedding can be prolonged in young children and immunocompromised patients.
- In recent years, influenza A strains with the H1N1 and H3N2 serotypes have been present, with either type being predominant during any 1 year.

- When there is coinfection with human and animal influenza viruses, reassortment between the viruses can occur, resulting in new serotypes with new HA or NA glycoproteins (and when populations lack immunity, worldwide pandemics can result).

PATHOPHYSIOLOGY

- When virus is inhaled, it causes damage to the respiratory epithelium (potential portal of entry for secondary bacterial infections).
- Infection can spread to lower airways by inhalation into alveoli or by contiguous spread.
- Host defenses include production of interleukin-6 and interferon, which result in many of the systemic clinical symptoms of influenza.

CLINICAL PRESENTATION

- Incubation period is 2 to 4 days and varies with viral strain and host factors.
- Onset of symptoms is usually acute.
 - Symptoms include fever, chills, headache, sore throat, dry cough, myalgias, otitis media, and conjunctivitis.
 - Upper respiratory tract symptoms are most common.
- Croup, bronchitis, and pneumonia can also develop.
 - Pneumonia can be either due to the influenza virus or secondary bacterial infection.
- In older children, acute calf pain and refusal to walk may indicate myositis (mostly following influenza B rather than influenza A.)
- Young children may present with fever alone (“rule out sepsis”) or may have GI symptoms such as vomiting, abdominal pain and diarrhea.
- Infants may present with apnea.

TABLE 60-1 Terms Associated with Influenza

TERM	COMMENTARY
Influenza	Orthomyxoviridae family of viruses Single-stranded, enveloped, RNA virus (negative sense) Segmented genome (7 or 8 segments)
Major types	A, B, and C
Serotypes	Determined by hemagglutinin (HA) and neuraminidase (NA) surface glycoproteins
Strains (strain variants)	Identified by World Health Organization (WHO) naming system Major type (A, B, or C) Geographic area of origin Isolate number Year of isolation HA and NA serotype (if influenza A) Examples: Influenza A/New Caledonia/20/99 (H1N1) Influenza A/Texas/1/77 (H3N2) Influenza B/Hong Kong/20/2003
Hemagglutinin (HA)	Surface glycoprotein Important for attachment to the sialic acid receptor on the host epithelial cell Neutralizing response of host is mostly determined by ability to recognize HA Influenza A has 16 different HA (designated as H1 to H16)
Neuraminidase (NA)	Surface glycoprotein Cleaves host cell membrane sialic acid, enabling release of virions Prevents viral aggregation, thus promoting infectivity and free release of virions Target of host immune response Influenza A has 9 different NA (designated as N1–N9) NA inhibitors include oseltamivir and zanamivir
Matrix protein 1 (M1)	Protein located on the inner surface on the viral envelope Provides stability to virus
Matrix protein 2 (M2)	Protein located within the envelope of influenza A only Forms an ion channel Facilitates uncoating of the virus M2 inhibitors include amantadine and rimantadine (active against influenza A only)
Antigenic drift	Minor changes in HA and NA Due to random, frequent genetic mutations Occurs in both influenza A and influenza B Leads to seasonal, yearly epidemics Drift is anticipated in yearly vaccine formulation Example: drift within H1N1 or within H3N2
Antigenic shift	Major changes in HA and NA Due to rare genetic events: when an animal strain mutates and is able to infect human hosts, or when a human strain takes on the genes from an animal strain (reassortment) Occurs only in influenza A Leads to large, worldwide pandemics, many years apart Pandemics occur because human populations lack immunity Example: shift from H1N1 to H2N2, and then to H3N2, etc.
Epidemic	Occurs annually Due to antigenic drift Seasonal (November to March in temperate climates) Caused by one or two predominant serotypes
Pandemic	Occurs rarely, only at infrequent intervals (many years) Due to antigenic shift

- Rarely, influenza can cause myocarditis, seizures, encephalopathy, encephalitis, transverse myelitis or Guillain–Barré syndrome.
 - In Japan, a severe, acute necrotizing encephalopathy due to influenza has been reported in young children (associated with seizures and coma).
- Reye syndrome can develop if aspirin is used during influenza.
- Deaths usually occur due to complications, including secondary bacterial infections like community-acquired–methicillin-resistant *Staphylococcus aureus* (CA-MRSA) pneumonia.

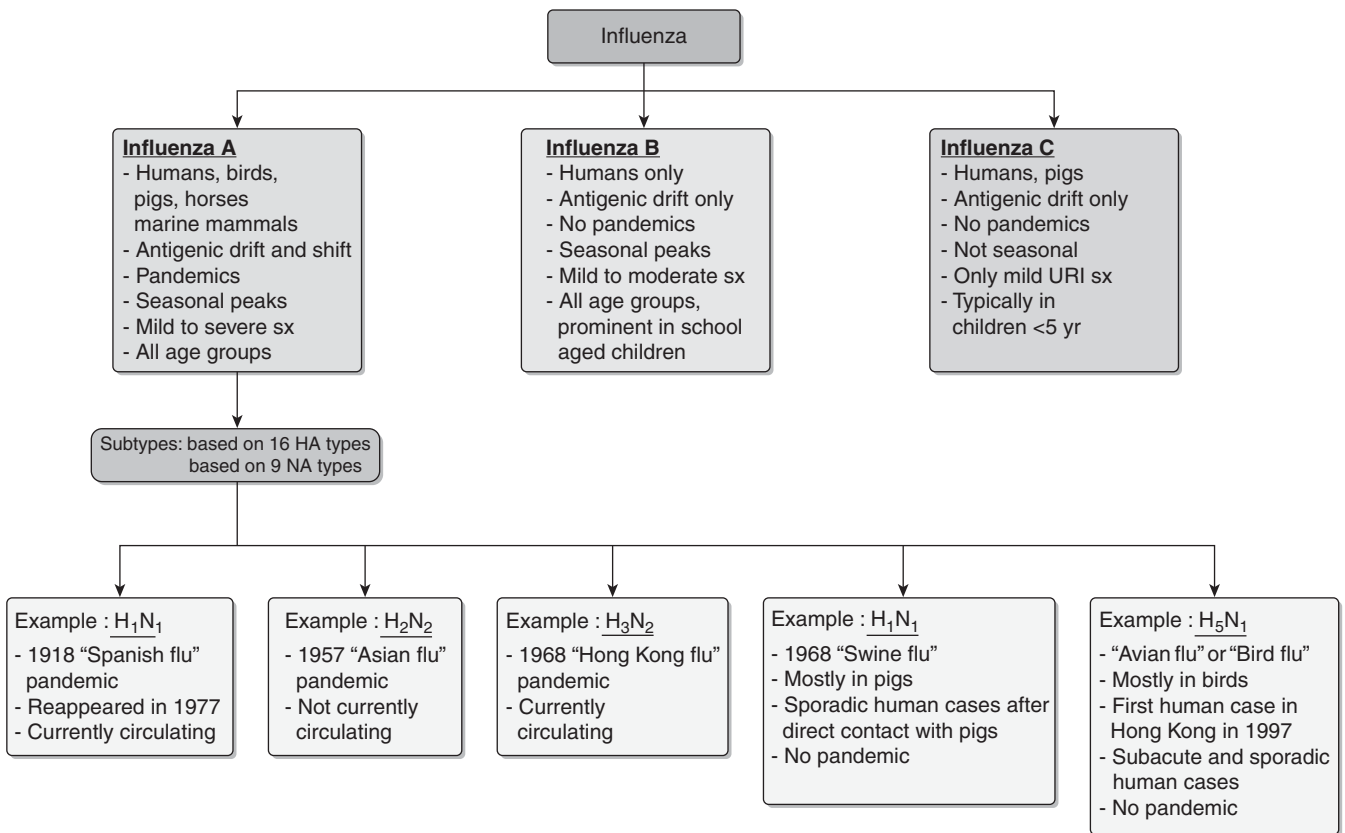


FIG. 60-1. Classification of influenza viruses.

DIFFERENTIAL DIAGNOSIS AND CLINICAL MANAGEMENT

- Clinical presentation of influenza overlaps with other respiratory viruses, especially respiratory syncytial virus and human metapneumovirus.

- Table 60-2 outlines some features of various respiratory viruses that are a part of the differential diagnosis of influenza.
- Figure 60-2 summarizes an approach to the management of infants and children who may present to the clinic or emergency department with influenza.

TABLE 60-2 Respiratory Viruses to Consider in the Differential Diagnosis of Influenza (with Accompanying Clinical Tips)

Influenza A—"Common flu" (example: H1N1 or H3N2)	Respiratory syncytial virus (RSV)
Wide variety of symptoms with potentially severe systemic disease	Seasonal overlap with influenza (winter in temperate climates)
Influenza A—"Avian flu" (example: H5N1)	Large number of hospital admissions in infants with bronchiolitis
Usually bird exposure history, but risk of future pandemic with human-human spread	Premature infants may have received preventative monthly monoclonal anti-RSV antibody
Influenza A—"Swine flu"	Transmission especially by contaminated fomites
Specific pig exposure history	Human Metapneumovirus (hMPV)
Influenza B	Clinically and epidemiologically similar to RSV, but smaller number of cases
School-age children predominant	Adenovirus
Rarely can cause myositis	Wide range of clinical severity (common cold to fatal disseminated infection)
Influenza C	Acute pneumonia can be the initial clinical presentation
Rare and clinically insignificant	Coronaviruses (Co-V)
Parainfluenza 1, 2, and 3	Severe acute respiratory syndrome (SARS-CoV)
Season is different than influenza (spring and fall in temperate climates)	Severe life-threatening disease is likely
Non-specific febrile illness with hospital admission in young infants (para 3)	History of travel to an area where SARS is occurring
Croup (para 1 and 2)	Non-SARS Co-V
	Generally mild upper respiratory infections
	Bocaviruses
	Recently discovered by molecular testing
	Full clinical spectrum unclear, but likely mild respiratory disease

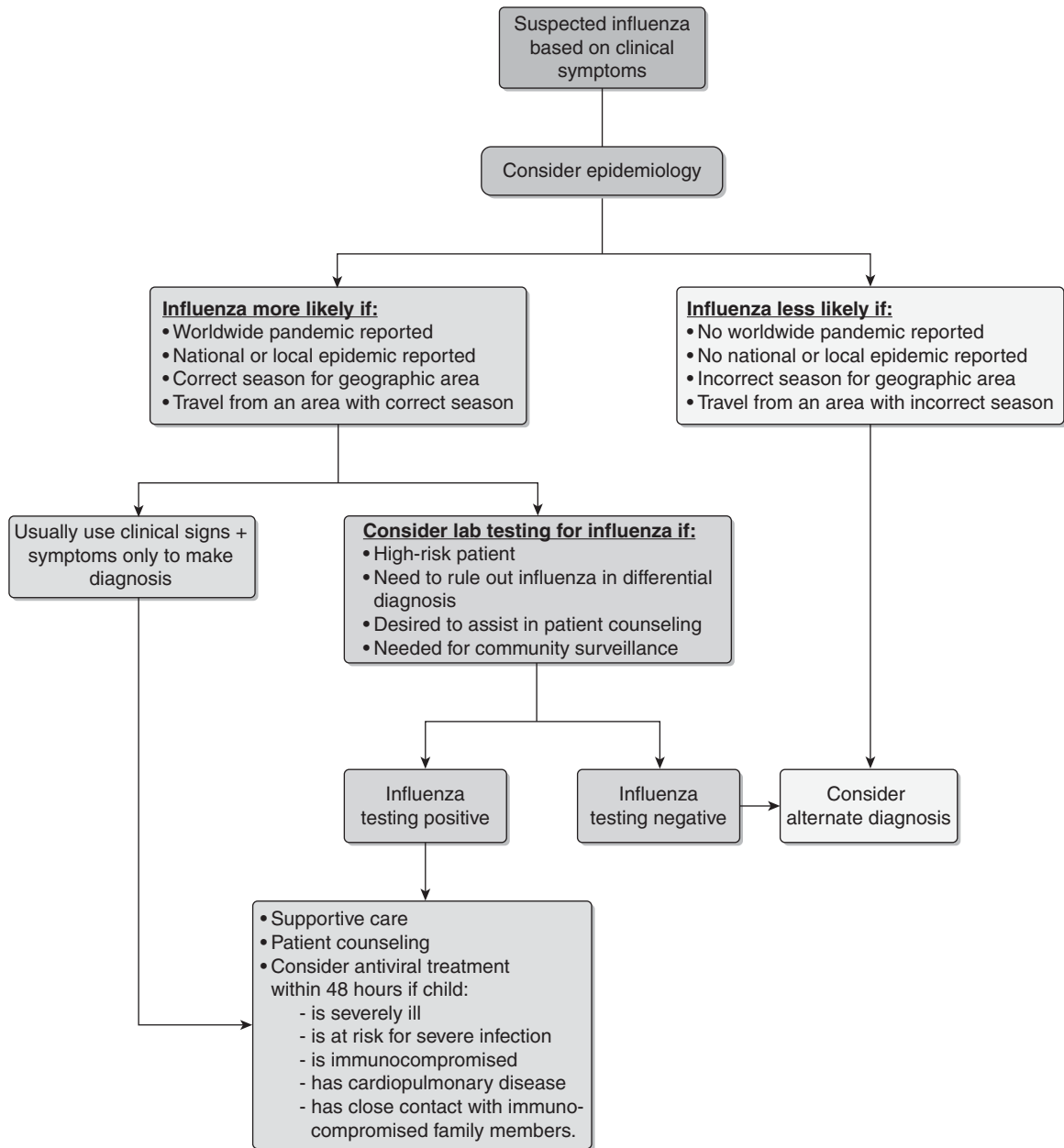


FIG. 60-2. Management of suspected influenza in children.

LABORATORY AND RADIOGRAPHIC FINDINGS

- Most cases of influenza have only mild symptoms and supportive care alone is appropriate.
- Specific laboratory diagnosis may be helpful when patients are infants, have underlying diseases or have severe symptoms.
 - Table 60-3 outlines the features of the laboratory tests most commonly available.

- Influenza PCR, done on nasopharyngeal specimens, is the most sensitive and results are obtained more quickly than by viral culture.
- Routine laboratory tests, such as CBC, are often not needed but may be helpful in patients with lower respiratory tract symptoms.
 - WBC counts of $<5000/\text{mm}^3$ are often seen with influenza and other viruses and elevated WBC counts with predominantly polymorphonuclear cells suggest secondary bacterial infection.

- Serology is not useful acutely.
- Chest radiographs are not routinely indicated.

TREATMENT

- Most influenza infections require only supportive care.
- Aspirin (salicylates) should NEVER be given to children with respiratory viral infections, because of the risk of Reye syndrome.
- Decongestants are not recommended, especially in children younger than 2 years of age.
- Antiviral medications approved for use in children are outlined in Table 60-4.
 - Antiviral medications should not be given routinely.
 - Antiviral medications can be considered in children who are at risk for severe or complicated infection (for example, immunocompromised children or children with underlying cardiopulmonary disease,

TABLE 60-3 Laboratory Testing in Influenza Virus Infection

Polymerase chain reaction (PCR) testing for influenza A and B:
Done on nasopharyngeal specimens
Often part of a PCR panel that may include parainfluenza, respiratory syncycial virus (RSV), and human metapneumovirus (hMPV)
High sensitivity and specificity
Rapid results
Direct fluorescent antigen (DFA) and indirect fluorescent antigen (IFA) testing
Done on nasopharyngeal specimens
Rapid results
Viral culture:
Done on nasopharyngeal specimens
Viral isolation within 2–6 d
Complete blood count (CBC) with differential:
Usually not helpful, nonspecific
If done for other reasons, it may show mild WBC elevation with predominance of lymphocytes
Serology
Usually not helpful in acute infection:
Can be helpful in making the diagnosis retrospectively, but only if acute and convalescent sera (obtained 1–2 wk apart) are available

TABLE 60-4 Antiviral Medication used for Influenza Prophylaxis and Treatment^a

GENERIC NAME	BRAND NAME (IN THE UNITED STATES)	CLASS OF DRUG	ACTIVE AGAINST INFLUENZA TYPES	ROUTE OF ADMINISTRATION	APPROVED FOR TREATMENT	APPROVED FOR PROPHYLAXIS	SIDE EFFECTS OR PRECAUTIONS
Amantidine	Symmetrel	Adamantane (inhibits M2 protein)	A only	Oral	≥1 y of age but not currently recommended in the United States due to resistance	≥1 y of age but not currently recommended in the United States due to resistance	Hypotension, Edema Dizziness, Headache Confusion Livedo reticularis Nausea, Vomiting Urinary retention
Rimantidine	Flumadine	Adamantane (inhibits M2 protein)	A only	Oral	≥13 y of age but not currently recommended in the United States due to resistance	≥1 y of age but not currently recommended in the United States due to resistance	Hypotension Dizziness, Headache Confusion CNS effects less than with amantidine Nausea, Vomiting Urinary retention
Oseltamivir	Tamiflu	Neuraminidase (NA) inhibitor	A and B	Oral	≥1 y of age	≥1 y of age	Unstable angina Arrhythmia Dizziness, Headache Rash Worsening of diabetes Nausea, Vomiting Anemia Hepatitis Conjunctivitis Neuropsychiatric effects ^b
Zanamivir	Relenza	Neuraminidase (NA) inhibitor	A and B	Inhaled	≥7 y of age	>5 y of age	Bronchospasm Allergic reactions

^aRibavirin has been studied for use in influenza infections for many years, but despite positive results initially, there has been no consistent documentation of significant clinical benefit in practice. Ribavirin is currently not recommended for either prophylaxis or treatment of the influenza viruses.

^bAs of November 2007, the precautions for oseltamivir include neuropsychiatric effects. This is based on rare postmarketing reports (mostly from Japan) of delirium and abnormal behavior leading to injury and, in some cases, resulting in fatal outcomes. These events were reported primarily in pediatric patients and often had an abrupt onset and rapid resolution.

TABLE 60-5 Influenza Vaccines^a

VACCINE	BRAND NAMES	ROUTE GIVEN	APPROVED FOR AGE GROUPS	DOSE (ML)	NUMBER OF DOSES	CONTRAINDICATIONS
Trivalent inactivated influenza vaccine (TIV)	Fluzone	IM	6–35 mo	0.25 mL	1 or 2 ^b	Children <6 mo of age Persons with history of hypersensitivity including anaphylactic reaction to chicken or egg proteins or other TIV vaccine components
		IM	≥36 mo	0.5 mL	1 or 2 ^b	
	Fluvirin	IM	≥4 y	0.5 mL	1 or 2 ^b	Persons with moderate or severe acute illness
	Fluarix	IM	≥18 y	0.5 mL	1	
	FluLaval	IM	≥18 y	0.5 mL	1	
Live attenuated influenza vaccine (LAIV)	Flumist	Nasal spray	2–49 y of age if healthy and nonpregnant	0.2 mL (spray 1/2 dose into each nostril)	1 or 2 ^c	Persons <2 y of age or >50 y of age Persons considered to be at high risk for severe influenza (including those with asthma) Persons with history of hypersensitivity including anaphylactic reaction to egg proteins or other LAIV vaccine components Children or adolescents receiving aspirin Known or suspected immunodeficiency History of Guillain–Barré syndrome Pregnant women

^aWith permission from www.cdc.gov/flu. 2007–2008 Influenza Prevention and Control Recommendations. Recommendations of the Advisory Committee on Immunization Practices (ACIP).

^bFor children aged 6 mo to 8 y who are receiving TIV for the first time, two doses administered at least 1 mo apart are recommended. For those who received only one dose in their first year of vaccination, two doses should be given the following year.

^cFor children aged 5 to 8 y who are receiving LAIV for the first time, two doses administered at least 6 wk apart are recommended. For those who received only one dose in their first year of vaccination, two doses should be given the following year.

healthy children with severe symptoms or children with special environmental circumstances, like close contact with immunocompromised family members.

- Adamantanes (amantidine and rimantidine) and neuaminidase inhibitors (oseltamavir and zanamavir) can shorten the duration of symptoms if started within 24 to 48 hours of symptoms
- Prevention of influenza complications with antivirals is less well documented.
- Level of antiviral resistance in a community is important in determining which medication to prescribe (can get info from the CDC or local health department).
- Oseltamivir has become the drug of choice; however, as of February 2008, 8% of U.S. influenza A (H1N1) isolates were oseltamivir-resistant.
- In cases of life-threatening influenza illness, use of two antiviral medications (one from each class of drug) should be considered.
- If secondary bacterial pneumonia is suspected, physicians should be aware of the prevalence of CA-MRSA in their communities when choosing empiric antibiotics.

PREVENTION: IMMUNIZATION AND PROPHYLAXIS

- Influenza vaccines are newly formulated every year.
 - Each year, the trivalent inactivated (TIV) and the live attenuated (LAIV) influenza vaccines are based on the same strains (usually two influenza A strains and one influenza B strain).
 - Usually, one of the strains is new and two are strains that were in the formulation of the vaccine from the previous year.
- Features of the two types of influenza vaccines (TIV and LAIV) are outlined in Table 60-5.
- Prevention of influenza can also include prophylaxis with antiviral medications in certain circumstances.
 - Medications approved for prophylaxis in various age groups are outlined in Table 60-4
- Helpful Web sites for up-to-date information on influenza are listed in Table 60-6.

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TABLE 60-6 Helpful Web Sites

Centers for Disease Control and Prevention (CDC)

including the Advisory Committee on Immunization Practices (ACIP)—www.cdc.gov

American Academy of Pediatrics (AAP) including the Red Book Committee (Committee on Infectious Diseases)—www.aap.org

World Health Organization (WHO)—www.who.org

Infectious Diseases Society of America (IDSA)—www.idsociety.org

Pediatric Infectious Diseases Society (PIDS)—www.pids.org

American Academy of Emergency Medicine (AAEM)—www.aaem.org

National Network for Immunization Information (NNII)—www.immunizationinfo.org

These Web sites are regularly updated and can provide:

handouts for patients and parents (easily downloaded)
 educational materials (posters, pamphlets) for health care professionals and staff
 recommendations for immunizations and antiviral medications
 recent trends in influenza epidemiology, antiviral drug resistance patterns, etc.
 upcoming meetings, lectures, and seminars for health care professionals
 contact phone numbers and e-mail addresses

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QUESTIONS

- Influenza is the cause of yearly, seasonal epidemics. This is due to minor changes in the HA and NA antigens and is referred to by which of the following?
 - Strain typing
 - Antigenic drift
 - Antigenic shift
 - Serotype variation
 - Serotype introduction
- Large worldwide influenza pandemics occurring at irregular intervals are due to major changes in the HA and NA antigens. These rare genetic events enable an animal influenza strain to infect human hosts, or allow human strains to take on the genes from an animal strain. The process by which this takes place is called which of the following?
 - Gene splicing
 - Conjugation
 - Antigenic mutation
 - Viral assortment
 - Gene reassortment
- A 6-year-old male presents to the ED with fever, chills, generalized myalgias, cough, and is diagnosed with having influenza. What is the duration of viral shedding in this child with influenza?
 - 24 hours before clinical symptoms, until fever resolves.
 - 24 hours before clinical symptoms, lasting 1 to 2 weeks.
 - 24 hours before clinical symptoms, lasting 4 to 6 weeks.
 - 48 hours before clinical symptoms, lasting 1 to 2 weeks.
 - 48 hours before clinical symptoms, lasting 4 to 6 weeks.
- An 8-year-old, generally healthy girl, living on a farm in the Midwest, is brought to a local emergency room in November with a history of 3 days of fever, dry cough and myalgias. She is hemodynamically stable, her respiratory rate is 22/minute, and her oxygen saturation is 96%. Her physical examination is normal except for a few coarse rhonchi in the lungs bilaterally. The most appropriate diagnostic approach at this time would most likely be which of the following?
 - CBC with differential
 - Chest X-ray
 - Nasal wash specimen for respiratory viral PCR panel, which includes influenza.
 - CBC and nasal wash specimen for respiratory viral PCR panel, which includes influenza.
 - Chest x-ray and nasal wash specimen for respiratory viral PCR panel, which includes influenza.
- Many children with influenza infection develop fever and mild symptoms, but require only supportive care. Which statement is most correct for treatment of influenza in a 2 year old?
 - Fever can be treated with acetaminophen or ibuprofen.
 - Fever can be treated with baby aspirin.
 - Fever from influenza does not respond to treatment.
 - Fever is always an indication for antiviral treatment.
 - Fever is a poor prognostic indicator.
- A 12-year-old female develops fever, acute calf pain, and refuses to walk (suggestive of myositis). This patient is most likely to have which of the following as the etiology of her symptoms?
 - Influenza A
 - Influenza B
 - Influenza C
 - “Bird flu”
 - Human metapneumovirus
- Most influenza infections are self-limited, and require only supportive care; however, antiviral medications can be considered in some cases. These include children with severe disease, children who are at risk of complications (due to an underlying medical condition), or children in special epidemiologic circumstances (such as those living with immunocompromised family members). If oseltamivir is given, how is it usually administered?
 - Given by directly observed therapy by health care or health department personnel.
 - Given twice a day for a minimum of 1 week.
 - Given 3 times a day for 10 days.
 - Started within the first 5 days of symptoms and given for 10 days.
 - Started within the first 24–48 hours of symptoms and given for 5 days
- Which of the following statements is true regarding antiviral therapy?
 - Amantidine, Rimantidine and Oseltamivir are all given orally and Zanamivir is given by inhalation.
 - Amantidine and Rimantidine and Zanamivir are given orally, and Oseltamivir is given by inhalation.
 - Amantidine and Rimantidine are given orally, and Oseltamivir and Zanamivir are given by inhalation.

- D. Amantidine, Rimantidine, Oseltamivir and Zanamivir are all given orally.
- E. Amantidine, Rimantidine, Oseltamivir and Zanamivir are all active against both influenza A and influenza B.
9. Influenza vaccination is an important public health measure to help prevent influenza disease. The TIV and LAIV influenza vaccines are both newly formulated every year in the summer, depending on predictions of the most likely strains to circulate the following winter. Which of the following statements is true?
- A. The TIV and LAIV influenza vaccines usually contain different strains.
- B. The TIV and LAIV influenza vaccines usually contain 4 to 6 different strains.
- C. The TIV and LAIV influenza vaccines both include two influenza A strains and one influenza B strain.
- D. The TIV and LAIV influenza vaccines are both given by injection.
- E. The TIV and LAIV influenza vaccines can be given to patients 6 months of age or older.
10. 5-year-old and 7-year-old brothers present with influenza-like symptoms to the ED. Another important aspect of influenza prevention involves infection control. In addition to good hand washing, what type of control measures should generally be used for these patients with proven or suspected influenza infection?
- A. Standard precautions and droplet precautions.
- B. Standard precautions and contact precautions.
- C. Standard precautions and reverse isolation.
- D. Standard precautions and prophylaxis with antiretroviral medications for all close contacts of patients.
- E. Standard precautions only.
- strains are simultaneously in the same host, and it allows one influenza strain to acquire RNA from the other influenza strain, thereby conferring new properties.
3. B. Influenza is transmitted by inhalation of infected droplets and aerosols (such as from a cough or sneeze), or by direct contact from fomites. The incubation period is generally 1–4 days, and the viral shedding starts 24 hours before clinical symptoms and lasts about 1–2 weeks.
4. C. This 8-year-old, who does not have any underlying chronic medical conditions, is suspected as having a typical, uncomplicated case of influenza. She does not need a CBC or a chest x-ray, however, PCR testing would be the most appropriate. The PCR for influenza, is often included in a respiratory viral PCR panel, and is the most sensitive and specific to identify the major type of influenza. Generally PCR tests identify the major type of influenza (ie, influenza A or influenza B) rather than identifying the specific strain. During the 2009 H1N1 influenza epidemic, however, a PCR was developed (and was commercially available) which was strain specific.
5. A. The fever that accompanies influenza can be treated with either acetaminophen or ibuprofen or both (alternate dosing). Aspirin should NEVER be given to a child with influenza or any other viral infection, because of the risk of developing Reye syndrome. If a child is already receiving aspirin (for other medical conditions such as Kawasaki syndrome), the aspirin should be stopped. Fever may persist for many days; it is neither an indication for antiviral medications nor a poor prognostic sign.
6. B. Influenza B is the most likely cause of myositis. If it occurs, it is often in school age children, and can be confused with discitis.
7. E. Antiviral medications for influenza should be started within the first 24–48 hours of symptoms and are generally given for 5 days. Although influenza epidemiology is closely followed by public health officials, there is no need for directly observed therapy.
8. A. Amantidine, Rimantidine, and Oseltamivir are all given orally and Zanamivir is given by inhalation. Amantidine and Rimantidine belong to the adamantane class of drug, and they inhibit the M2 protein of influenza A. They are active only against influenza A. Oseltamivir and Zanamivir are both NA inhibitors, and are active against both influenza A and influenza B

ANSWERS

1. B. Antigenic drift is due to random, frequent genetic mutations and occurs in both influenza A and influenza B. Antigenic drift is anticipated when the yearly influenza vaccines are formulated. In contrast, antigenic shift is due to major changes in HA and NA antigens, and occurs only in influenza A, and leads to large, worldwide epidemics.
2. E. The major changes in HA and NA that are seen in influenza pandemics (antigen shift) can occur because strains of the virus sometimes undergo gene reassortment. This occurs when two influenza

9. C. The TIV and LAIV influenza vaccines are based on the same strains. These include the predicted prevalent strains for any given year, and have recently included two influenza A strains and one influenza B strain.
10. A. In addition to good hand washing, standard precautions and droplet precautions are the recommended infection control measures. Under local or special conditions, additional measures can be added. Antiviral prophylaxis is not usually needed, but is recommended in certain high-risk patients, or depending on special circumstances.

61 MENINGITIS

Steven Lelyveld
Gary R. Strange

PATHOPHYSIOLOGY

- Pathogens enter the subarachnoid space by hematogenous spread, by mechanical disruption, as in a fracture of the base of the skull, or by direct extension from an infection in an adjacent structure.
- Once the blood–brain barrier is breached, natural defense mechanisms are unable to stop the multiplication of organisms.
- Cerebral edema causes increased intracranial pressure, decreased cerebral blood flow, regional hypoxia, focal ischemia, and ultimately death.

PRESENTATION

- The “classic” signs and symptoms of meningitis are headache, photophobia, stiff neck, change in mental status, bulging fontanelle, nausea, and vomiting.
- The younger the child, the less obvious the signs and symptoms until late in the course of the disease. Young infants present with poor feeding, irritability, inconsolability, or listlessness.
- Nuchal rigidity occurs in <15% of children under 18 months with meningitis.
- The Brudzinski sign occurs when the irritated meninges are stretched with neck flexion causing the hips and knees to flex involuntarily (Fig. 61-1).
- The Kernig sign is when the hip is flexed to 90° and the examiner is unable to passively extend the leg fully to the 180° position (Fig. 61-2).
- Meningitis is either insidious (90%) or fulminant (10%).

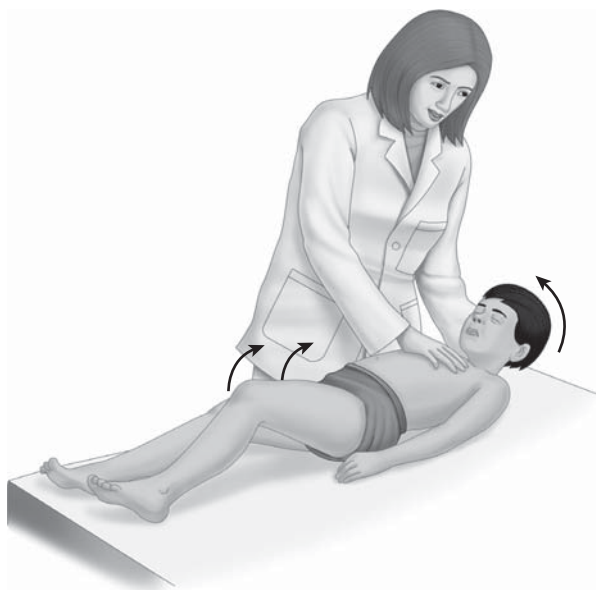


FIG. 61-1. Brudzinski sign. With the patient supine and the examiner's hand on the patient's chest, passive neck flexion (arrow at right) results in flexion at the hips (arrows at left), which is often asymmetric. The sign is present with meningeal irritation and inflammation such as that seen in meningitis, but also in subarachnoid hemorrhage.

- Insidious meningitis (frequently caused by *Pneumococcus*) can present to a physician as a non-specific illness up to two weeks before the diagnosis, with a median of 36 to 72 hours. Many children receive partial antibiotic treatment that may complicate the diagnostic process. However, prior antibiotic may be associated with reduction in meningitis-related sequelae.

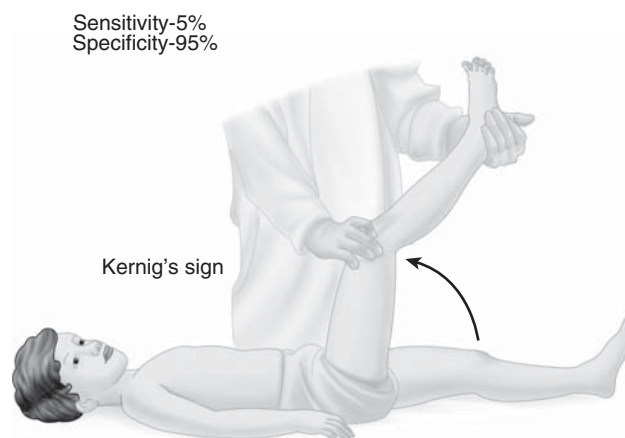


FIG. 61-2. Kernig sign. With the patient supine, the hip is flexed to 90° and the examiner attempts to passively extend the leg fully. Inability to passively extend the leg to its full extent is a positive Kernig sign.

- The more fulminant the course, the worse the prognosis. Concomitant bacteremia rapidly progresses to petechiae, purpura fulminans, and cardiovascular collapse.
- The management of any of the bacterial meningitides may be complicated by hemorrhage into the adrenal cortex, the Friderichsen–Waterhouse syndrome.

DIFFERENTIAL DIAGNOSIS

- Early meningitis may be confused with gastroenteritis, upper respiratory infection, pneumonia, otitis media, or minor viral syndromes.
- When alteration of mental status occurs, the diagnoses of encephalitis, subarachnoid or subdural hemorrhage with or without direct trauma or abuse, cerebral abscess, Reye's syndrome, toxic ingestions, seizure disorders, diabetic ketoacidosis, and hypothyroidism may be considered. A young child with intussusception may present with vomiting, altered mental status, and cardiovascular collapse. Many of these children are evaluated for meningitis before their diagnosis is clear.
- Presence of fever helps to refine the differential diagnosis.

MANAGEMENT OF UNSTABLE PATIENTS

- The diagnostic test of choice for meningitis is the lumbar puncture with cerebrospinal fluid (CSF) analysis.
- In the unstable child, lumbar puncture should be withheld until after stabilization and antibiotic administration. Although the early administration of antibiotics may prevent recovery of the organism on culture of CSF, there is no definite correlation between early treatment and clinical outcome. A blood culture prior to administration of antibiotics may help identify a specific pathogen.
- The ABCs of resuscitation take precedence over diagnostic procedures.
- The unstable patient with meningitis may have respiratory compromise, shock, increased intracranial pressure, seizures, or hypoglycemia.
 - Ensure an open airway and adequate ventilation, with supplemental oxygen administered with the bag-valve-mask technique, followed by endotracheal intubation.
 - Treat shock with rapid intravenous or intraosseous infusion of crystalloid solution in 20 mL/kg aliquots. Continually reassess to avoid fluid overload that can lead to worsening of cerebral edema. Fluid restriction is not routinely indicated.
 - Treat signs of increased intracranial pressure by elevating the head at 30 degrees and keep the PaCO₂

TABLE 61-1 Normal Cerebrospinal Fluid (CSF) Values

PARAMETER	PRETERM INFANT	TERM INFANT	CHILD
Cell count	9 (0–25) WBC/mm ³	8 (0–22) WBC/mm ³	0–7
Differential	57% PMNs	61% PMNs	0% PMNs
Glucose	24–63 mg/dL (mean 52)	34–119 mg/dL (mean 52)	40–80 mg/dL
CSF/blood glucose ratio	55%–105%	44%–128%	50%
Protein	65–150 mg/dL (mean 115)	20–170 mg/dL (mean 90)	5–40 mg/dL

between 30 and 35 mm Hg with controlled ventilation. Patients who do not respond may benefit from diuretic therapy with mannitol (0.25–1 g/kg) or furosemide (1 mg/kg).

- Seizures are controlled with rapid-acting benzodiazepines followed by phenytoin (see Chapter 7).
- If the blood glucose is <40 mg/dL, administer glucose (250 to 500 mg/kg) as 10% dextrose for neonates, 25% dextrose for infants younger than 2 years, and 50% dextrose for older infants and children.

MANAGEMENT OF STABLE PATIENTS

- In stable patients with manifestations suggestive of meningitis, phlebotomy for diagnostic studies is followed promptly by lumbar puncture. The threshold for clinical suspicion of meningitis should be particularly low for
 - Neonates
 - Immunocompromised children
 - Children who have been in close contact with known cases of meningitis
 - Children with documented bacteremia
- The initial workup includes a complete blood count (CBC), electrolytes, glucose, renal functions, and blood culture. Tests for bacterial capsular antigens are not often helpful in the acute situation.
- The expected laboratory parameters for CSF analysis are age related (Table 61-1).
- A low white blood cell count, with predominantly mononuclear cells, and normal glucose and protein are more consistent with a viral etiology.
- High protein, low sugar, and elevated polymorphonuclear leukocytes suggest bacterial etiology.

ANTIBIOTIC TREATMENT

- Antibiotic treatment is directed by the specific organisms, or by the predominant organisms found for the age group of the patient.

- Newborns are generally treated with an initial dose of ampicillin, 100 mg/kg, and an aminoglycoside, such as gentamicin, 2.5 mg/kg. A cephalosporin active against gram-negative bacilli, such as cefotaxime, 50 mg/kg, may be substituted for the aminoglycoside.
- Infants and children are generally treated with a cephalosporin (ceftriaxone 100 mg/kg/dose qd or cefotaxime 50 mg/kg/dose tid). The combination of ampicillin, 100 mg/kg/dose qid, and chloramphenicol, 25 mg/kg/dose bid, is an option.
- If the organism is known to be *Streptococcus pneumoniae* or if gram-positive cocci are seen on gram stain of the CSF, penicillin and cephalosporin resistance is possible. Vancomycin, 15 mg/kg/dose bid, is the only antibiotic to which all strains of pneumococci are currently susceptible, and is added to the cephalosporin for comprehensive therapy.

CORTICOSTEROID TREATMENT

- When given prior to the antibiotic, the anti-inflammatory effect of dexamethasone (0.15 mg/kg intravenously) decreases intracranial pressure, cerebral edema, and CSF lactate concentrations.
- Dexamethasone significantly decreases hearing loss and other neurologic sequelae in meningitis caused by *Haemophilus influenzae* type B.
- Dexamethasone therapy may improve the outcome of meningitis caused by other organisms. Some authorities argue that steroids may strengthen the blood–brain barrier and limit the penetration of intravenously administered antibiotics into the CSF. There is no clear consensus on the empiric use of steroids for meningitis when the bacterial agent is unknown.

SEQUELAE

- The vast majority of children with aseptic meningitis have a self-limited illness without subsequent problems.
- In spite of treatment, the mortality of *H influenzae* type B meningitis is 5% to 10%, and for *S pneumoniae* meningitis is 20% to 40%.
- Up to 20% of survivors will have long-term sequelae, including mild learning defects, sensorineural hearing loss, afebrile seizures, and multiple neurologic defects, retardation and blindness.
- Other neurologic defects, such as hemiparesis, ataxia, cranial nerve palsy, and abnormal extensor reflexes, may be present initially but resolve in a few months.

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QUESTIONS

1. A parent presents to the ED with a 12-month-old having a first seizure that lasted several minutes. The vital signs are $T = 40.1^{\circ}\text{C}$, $P = 178$, $R = 46$, $\text{BP} = 68/40$, $\text{Pox} = 98\%$. Upon inspection, the child has stiff limbs, marbled skin with delayed capillary refill, and eyes deviated to the left. In the management of this patient, the most important initial step would be which of the following?
 - A. Give ceftriaxone 100 mg/kg IM.
 - B. Establish an IV and give 20 cc/kg of normal saline.
 - C. Place a nasal cannula and give $\text{O}_2 @ 5 \text{ L/min}$.
 - D. Rapidly assess the glucose and infuse 0.25–0.5 gm/kg if glucose $<40 \text{ mg/dL}$.
 - E. Give a rapidly acting benzodiazepine to stop the seizure.
2. A 3-year-old girl presents with a history of fever to 39°C , cough, frontal headache, sore throat, and decreased appetite. Upon examination, the Kernig and Brudzinski signs are negative, the throat has no exudates, the lungs are clear, and the rest of the examination is unremarkable. Your most important action is to
 - A. Reassure the parents that this is just a virus and no follow-up is necessary.
 - B. Administer an antipyretic and base your next action on the temperature in one hour.
 - C. Order a monospot test and treat with penicillin if monospot test is positive
 - D. Do a CBC, blood culture, urinalysis, and give IM ceftriaxone at 100 mg/kg.

- E. Give antipyretics, fluids and arrange for a second contact with this family in 24–48 hours.
3. An 18-month-old male infant is seen in your ED with a low-grade fever and is diagnosed with acute otitis media. A first dose of amoxicillin is given and the patient is discharged on antibiotics with instructions to follow up with the pediatrician in 48–72 hours. One week later the child presents lethargic to the triage desk. The following vital signs are obtained: temperature 37.7°C, respiratory rate 30, heart rate 140, blood pressure 98/55, Pulse oximetry 98%. He is responsive to tactile stimuli, pupils are mid-position and equal, discs are flat, Kernig and Brudzinski signs are negative, and there are no other findings on physical examination. The best course of action to take is
- Intubate, establish a large bore intravenous line and give 20 cc/kg of normal saline.
 - Insert a nasogastric tube and send the aspirate for toxicology.
 - Administer 100 mg/kg of ceftriaxone IV and arrange a CT scan of the head.
 - Draw blood culture, CBC, metabolic panel, liver functions, obtain a CT scan, and perform a lumbar puncture.
 - Question the parents independently about possible intentional or unintentional trauma.
4. A 5-year-old presents to your ED in August complaining of a headache, vomiting, and photophobia. She has gotten progressively worse over three days. Her vital signs are normal except for a temperature of 38.6°C. Her Kernig and Brudzinski signs are positive. Her discs are flat. You suspect meningitis. After obtaining a CT to rule out increased intracranial pressure, you perform a lumbar puncture. The CSF most likely will be
- Xanthochromic with 0 WBC/mm³ and 2,000 RBC/mm³, normal protein and a negative gram stain for bacteria.
 - Clear with 20 WBC/mm³ (90% mononuclear, 10% polymorphonuclear cells), normal glucose 50 mg/dL, normal protein and a negative gram stain for bacteria.
 - Cloudy with 1250 WBC/mm³ (5% mononuclear, 95% polymorphonuclear cells), low glucose, high protein and a gram stain with gram-negative intracellular diplococci.
 - Cloudy with 100 WBC/mm³ (50% mononuclear, 50% polymorphonuclear cells), low glucose, high protein and a gram stain with gram-positive cocci in chains.
 - Cloudy with 50 WBC/mm³ (95% mononuclear, 5% eosinophils), low glucose, high protein and a gram stain negative for bacteria.

ANSWERS

- B. It is reasonable to consider meningitis or other serious bacterial infection in any 12-month-old with a high fever and seizure, and give appropriate broad-spectrum antibiotics (a). Seizing infants can rapidly deplete their glycogen stores and lower metabolic substrates to levels that will not sustain cell life. Assessing and replacing glucose (d) is a vital part of this infant's management. Similarly stopping the seizure with a benzodiazepine (e) followed by phenytoin if it does not stop spontaneously is necessary for a healthy brain. However, the most important action is to address the ABC's of resuscitation first. This child is maintaining normal oxygen saturation. The marbled skin (*cutis marmorata*), rapid heart rate, and low blood pressure indicate this infant is in shock. It is essential to establish a large bore IV or, failing that, to establish an intraosseous line and give normal saline at 20 cc/kg rapidly. After the vital signs stabilize, then reassess oxygenation, glucose, and seizure activity. The antibiotics can be given before lumbar puncture and blood culture, as diagnostic tests should be deferred until stability is assured.
- E. While a simple, self-limited viral infection is the most common final diagnosis (a) in this child, the clinician does not have enough information to simply reassure the parents of this child. Looking for markers of bacteremia (d) are appropriate if the child has a temperature over 40°C, is under 36 months of age, and does not have a source. This is not the case here. This child has a history compatible with early pharyngitis. Group A beta strep cannot be excluded on purely clinical grounds, so testing for it (c) would be appropriate. A monospot test is used to confirm a diagnosis of mononucleosis, which is not treated with antibiotics. There is strong evidence that response to antipyretics (b) cannot be used as a marker for serious bacterial disease. Finally, many cases of insidious onset meningitis present with a nonspecific symptom complex within two weeks prior to the diagnosis, with an average time to presentation of 36–72 hours. Therefore, the most appropriate action is to ensure a follow-up appointment (e), no matter what other actions are taken.
- D. Addressing the ABCs of resuscitation is always the first thing to consider (a). However, this child has normal vital signs and oxygenation, so there is time for other interventions. This child has altered mental status. At this age, the differential includes toxic ingestion (b), head trauma/child abuse (e), metabolic derangement, including diabetic ketoacidosis (d) and pre-treated or partially treated meningitis or other bacterial

infection. The child is clinically stable. One does not need to treat first and investigate the source later (c), as timing of treatment within the single ED visit has not correlated with outcome. Pretreated meningitis can present without a fever, as can other infections and metabolic derangements. Therefore, the best action is (d), looking for infection or clues to investigate toxins and other causes of altered mental status.

4. B. It is possible that this child has a process mimicking meningitis, such as upper lobe pneumonia or a subarachnoid hemorrhage but less likely. Xanthochromia is the yellow discoloration of CSF indicating the presence of bilirubin and is used to differentiate in vivo subarachnoid hemorrhage from a traumatic lumbar puncture. With the introduction of the Hib and pneumococcal vaccines, *Neisseria meningitidis* has become the leading cause of bacterial meningitis in this age group followed by *S pneumoniae*. It is possible for both of these organisms, after they invade the brain, to multiply so rapidly that they overwhelm host defenses and can present with sheets of bacteria without a large inflammatory response. TB meningitis (e) is increasing in incidence as resistance to current therapy expands. However, it has a more insidious course than described. The majority of all cases of meningitis are aseptic. Over 80% are seasonal enteroviruses, predominantly echovirus, and coxsackievirus. There is a peak in the summer months. The low number of cells found may be predominantly polymorphonuclear early in the illness, but rapidly shift to mononuclear cells. Glucose and protein are normal, and the gram stain is negative. The best answer, therefore, is (b).

62 TOXIC SHOCK SYNDROME

Eiman Abdulrahman
Shabnam Jain

ETIOLOGY AND PATHOGENESIS

- Toxic shock syndrome toxin-1 (TSST-1), a superantigen, causes activation of blood vessel muscle cells leading to vasodilation and hypotension, activation of skin cells leading to rash, activation of gut cells causing diarrhea, and activation of muscle cells causing pain and cramps.
- Massive vasodilatation and rapid movement of serum proteins and fluid from the intravascular to the extravascular space results in oliguria, hypotension,

edema, and low central venous pressure. The multi-system collapse seen in TSS may be either a reflection of the rapid onset of shock versus direct effects of toxin(s) on the parenchymal cells of the involved organs.

- Majority of TSS cases are caused by coagulase-positive *Staphylococcus aureus*, although recently, coagulase-negative strains have been isolated, often developing from a site of colonization rather than infection. STSS is used to designate TSS caused by invasive group A streptococci (GAS), which produce the streptococcal enterotoxin.

EPIDEMIOLOGY

- According to CDC, only half of the cases of TSS are associated with tampon use; nonmenstrual cases are associated with postpartum or cutaneous/subcutaneous *Staphylococcus aureus* infections with predisposing factors such as burns, abrasions, abscesses, and nasal packing. TSS is more common in children with burns of relatively low body surface area.
- Overall mortality rate is 5% and is lower in children compared to adults. Recurrence rates are about 30%; secondary cases are milder and occur within 3 months of the original episode.
- STSS occurs most commonly following varicella in previously healthy children or during the use of non-steroidal antiinflammatory agents.

CLINICAL MANIFESTATIONS

- Sudden onset of fever over 38.9°C (with chills), vomiting, diarrhea, myalgia, dizziness, and diffuse rash. Additional symptoms include headache, arthralgia, sore throat, abdominal pain, stiff neck or tender, edematous external genitalia between third and fifth day of menses in menstruating females.
- Physical examination shows an acutely ill-appearing patient with hypotension, hyperemia of the conjunctiva and vagina, scant purulent cervical discharge, and bilateral adnexal tenderness in menstrual TSS. In the acute state, the patient may be agitated, disoriented, or obtunded. On the fifth to tenth hospital day, a generalized, diffuse, blanching, pruritic maculopapular rash develops in about 25% of patients, fades within 3 days of its appearance, and is followed by a generalized desquamation of the skin involving palms, soles, toes, and fingers.
- There is no diagnostic laboratory test for TSS. Therefore, diagnosis depends on the presence of typical signs and symptoms along with the development

of the typical desquamation of palms, soles, toes, and fingers.

DIFFERENTIAL DIAGNOSIS

- Kawasaki disease is clinically similar but lacks many of the features of TSS, including diffuse myalgia, vomiting, abdominal pain, diarrhea, azotemia, thrombocytopenia, and shock. Additionally, Kawasaki disease occurs typically in children younger than 5 years.
- Streptococcal scarlet fever is rare after the age of 10 and its rash is a nontender erythematous sandpaper-like rash as opposed to tender erythroderma seen commonly with TSS.
- Sepsis syndrome results from multiplying bacteria without development of superantigens; both sepsis and TSS may present with hypotension and initial management is always the same. However, the appearance of a rash and the laboratory abnormalities will distinguish between the two diseases.
- Other differentials include Rocky Mountain spotted fever, leptospirosis, meningococemia, Stevens–Johnson syndrome, and staphylococcal scalded skin syndrome, all of which may resemble TSS.

MANAGEMENT

- Important initial therapy is aggressive management of hypovolemic shock with large volumes of crystalloids or fresh frozen plasma; pressors may be added if fluids alone are not sufficient. Early identification and removal of material such as nasal packing or retained tampons is critical to remove the source of toxin production.
- Adjunctive therapy with intravenous immunoglobulins or fresh frozen plasma to neutralize toxins by binding to superantigens stops activation of cells and prevents further damage. Corticosteroids have not been shown conclusively to affect outcome.
- Initial parenteral antibiotic coverage with an antistaphylococcal agent and a protein synthesis-inhibiting antimicrobial, such as clindamycin, to cover for both GAS and *S aureus* infection should be instituted because of the similarity in the clinical appearance between TSS and STSS. Additionally, clindamycin may help suppress toxin production.

RECURRENCES

- Antibiotic use is important for eradication of the organism and reducing recurrence rate; more than

half of the patients not treated with a β -lactamase-resistant antibiotic have recurrences.

- Most recurrent episodes occur by the second month following the initial episode and have a milder course, and occur on the same day of menses as the prior attack.

THE FUTURE

- Toxoid vaccines against superantigens and monoclonal antibody against TSST-1 are being studied. It remains unexplained why some people develop TSS while others do not.

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QUESTIONS

1. A 17-year-old-female with a psychiatric disorder is brought to the ED from a group home with a 2-day history of fever, nausea, vomiting, and diffuse erythroderma. Vitals: T 39.5°C, HR 138, RR

- 16, BP 90/42. The most appropriate initial management for this patient is which of the following?
- Start broad-antibiotic coverage with ceftriaxone and nafcillin.
 - Give NS bolus and gram-negative coverage.
 - Start dopamine at 5mcg/kg/min.
 - Check for retained tampons or foreign body, give NS bolus and IV clindamycin.
 - IV fluid with D5 ½ NS + 20 KCL @ 125ml/hr.
2. The CDC case definition for diagnosis of TSS include all except which of the following?
- Vomiting or Diarrhea
 - Mucous membrane hyperemia
 - Leukocytosis
 - Hypotension
 - Rash
3. A 14-year-old female was admitted and treated for TSS. She was discharged and later presents to her pediatrician with skin changes. Which of the following cutaneous findings develop weeks after hospitalization in a patient with TSS?
- Vesicular eruption
 - Petechiae coalesce into purpuric lesions
 - Erythroderma
 - Desquamation especially pronounced in palms and soles
 - Sandpaper-like rash
4. A 16-year-old male presents with fever, rash, myalgias, vomiting, hypotension and has a retained nasal packing placed for epistaxis 3 days previously. The preferred initial antibiotic of choice in this patient would be which of the following?
- Vancomycin and meropenem
 - Vancomycin and clindamycin
 - Nafcillin and clindamycin
 - Nafcillin and Ceftriaxone
 - Unasyn
5. Which of the following statements most accurately describes TSS?
- Annual incidence of TSS has decreased but menstrual TSS still accounts for a high percentage of TSS cases.
 - More common in children with large body surface area burns compared to minor burns.
 - Children have higher incidence of morbidity and mortality compared to adults.
 - Recurrence rate for TSS is about 30%.
 - Overall mortality rate is around 25% in children.
6. A 12-year-old male presents with a classic presentation for TSS. The family asks why their son developed this condition. Which of the following best explains the pathophysiology of TSS?
- It is primarily a result of high bacterial load causing shock.
 - Most commonly caused by group A streptococcus.
 - The pathogenesis is thought to be related to TSST-1.
 - The pathophysiology includes massive vasoconstriction.
 - The late complication of TSS mainly involves the respiratory system.
7. Which of the following are predisposing factors in patients presenting with TSS?
- Immunocompromised patients.
 - Males with frequent minor nosebleeds.
 - Toddlers who attend daycare
 - Children with minor burns
 - Patients with history of Kawasaki disease.
8. A 5-year-old female presents several days after a small surface area burn with fever, nausea, vomiting, abdominal pain, a generalized pruritic maculopapular rash, and hypotension. Which of the following tests can help confirm the diagnosis of TSS in this patient?
- Azotemia and abnormal urinary sediments.
 - Elevated liver enzymes and bilirubin
 - Abnormal clotting factors and thrombocytopenia
 - No specific laboratory test can make the diagnosis
 - Leucocytosis and lymphopenia
9. Which of the following case scenarios is a classic presentation of staphylococcal TSS?
- 3-year-old boy with fever, conjunctival hyperemia, and erythema of mucous membranes with desquamation.
 - 3-year-old boy with fever and hypotension who responds to treatment with fluids and antibiotics.
 - 3-year-old boy with fever, hypotension, and rash that involves palms and soles
 - 3-year-old with fever, hypotension, and purpuric rash mostly in the extremities
 - 3-year-old with a two day old small body surface area burn who suddenly presents with hypotension.
10. A 14-year-old female presents with fever, abdominal pain, and hypotension several months after being treated for TSS. Which statement is accurate about the recurrences of TSS?
- Patients treated with antibiotics have similar recurrences to those who do not receive antibiotics.
 - Most recurrences occur one year following the initial episode of TSS.

- C. The majority of patients have an increased severity in illness in the recurrent episode
- D. The secondary cases are milder with an overall mortality rate of 5%
- E. Patients with TSS have a significant antibody response so recurrence rate is very low.

ANSWERS

1. D. The history and physical examination in this case strongly suggests TSS. Removing toxin producing retained tampons or other packings and adequate resuscitation with NS bolus with antibiotic coverage with clindamycin are all-important initial steps in this case. *S aureus* is the most likely pathogen. Starting broad-spectrum antibiotic coverage is important if case presentation is unclear but in this case it would be important to cover for *S aureus* and nafcillin is no longer recommended because of high resistance. Gram-negative coverage without coverage of gram positives to cover *S aureus* is inadequate. Pressors are usually used after the patient has been given adequate fluid boluses.
2. C. All the choices make up the signs and symptoms of the CDC criteria of TSS except leukocytosis.
3. D. Between the fifth and tenth hospital day, a generalized pruritic maculopapular rash develops in 25% of patients with TSS. The skin rash is diffuse and blanches; it fades within 3 days of appearance and is followed by a fine generalized desquamation of the skin, with peeling over palms, soles, toes, and fingers. Sandpaper rash is more characteristic of streptococcus and petechiae/purpuric rash are seen in other conditions such as Henoch-Schonlein purpura.
4. B. The most common pathogen for TSS is *S aureus* and less commonly *Streptococcus pyogenes*. Initial antimicrobial therapy should include an antistaphylococcal agent and a protein synthesis-inhibiting antimicrobial agent such as clindamycin. Clindamycin is more effective in treating *S pyogenes*. Given the growing high resistance to β -lactamases with the emergence of methicillin-resistant *S aureus* (MRSA) in many communities in the United States, nafcillin is not recommended for initial coverage. Therefore, the best choice is vancomycin and clindamycin.
5. D. Recurrent TSS is milder and occurs in about 30% of cases. According to annual incidence reports from the CDC, the rates of menstrual TSS have decreased. It is children with relatively low body surface area burns that, for unknown reasons, tend to present with TSS. Overall mortality is around 5% in children, which is lower than the morbidity and mortality reported in adults.
6. C. Most cases of TSS have been directly associated with *S aureus*. The pathogenesis is thought to be related to production of a toxin, currently referred to as TSST-1. The most impressive aspect of the pathophysiology of TSS is the massive vasodilation and rapid movement of serum proteins and fluid from the intravascular to the extravascular space. Abnormal laboratory values in these patients reflect the multisystem involvement of TSS.
7. D. The Centers for Disease Control and Prevention have reported a decrease in the annual incidence of TSS, presumably from the increased awareness of risk associated with tampon use. TSS is seen in menstruating women but cases are also reported in nonmenstruating women, children, and men. Nonmenstrual cases occur in a variety of clinical settings, but are chiefly associated with postpartum or cutaneous/subcutaneous *S aureus* infections. Predisposing factors include burns, abrasions, abscesses, and nasal packing. The differential diagnosis may include Kawasaki disease, staphylococcal scarlet fever, streptococcal scarlet fever, and septic shock. Immunocompromised patients, day-care attendants, or patients with Kawasaki disease have not been described as having higher incidence of TSS.
8. D. All the abnormal values in the answer choices may be seen and reflect the multisystem involvement in TSS; however, no specific laboratory test can make the diagnosis. Diagnosis of TSS requires clinical suspicion and CDC case definition criteria must be used.
9. E. The cases listed are all in the differential diagnosis when considering TSS. The clinical scenario in choice "A" has characteristics typically seen in Kawasaki disease. The case scenario in choice "B" is nonspecific but consistent with sepsis syndrome and is not classic for TSS. Rocky Mountain spotted fever comes to mind in clinical scenario "C" given the characteristic distribution in the palms and soles. Purpuric rash is most concerning for meningococemia in choice "D". The correct answer in choice "E" is a toddler with a low body surface area burn who has sudden clinical deterioration likely secondary to staphylococcal infection and toxin production.
10. D. More than half of patients not treated with a β -lactamase-resistant antibiotic have recurrences. Most recurrent episodes occur by the second

month following the initial episode on the same day of menses as the prior attack. In the majority of patients, the initial episode is the most severe. Studies have shown that patients with TSS do not develop significant antibody response to TSS-1 so the recurrence rate is around 30%. The secondary cases are milder with an overall mortality of 5% making choice “D” the correct choice.

63 KAWASAKI DISEASE

*Anthony Cooley
Shabnam Jain*

ETIOLOGY AND PATHOGENESIS

- Kawasaki disease (KD), or mucocutaneous lymph node syndrome, is an acute, self-limited, multisystem vasculitis of unknown etiology.
- Given its clinical presentation and epidemiology, an infectious agent or immune response to an infectious agent could be argued.
- The principal pathologic feature of this syndrome is an acute, nonspecific vasculitis that affects the microvessels (arterioles, venules, and capillaries). Nearly every organ system is involved.
- The peak incidence is in children 18 to 24 months of age, with 80% to 85% of cases occurring in children younger than 5 years.
- It is most common in children of Asian and Pacific Island descent.

CLINICAL FINDINGS TO MAKE THE DIAGNOSIS

- Without pathognomonic laboratory or clinical findings, the diagnosis is established using clinical criteria. Classically, KD is diagnosed by the presence of fever for 5 days and four of five other physical findings (Table 63-1):
 - Conjunctival injection without discharge.
 - Lip and pharyngeal erythema without exudate.
 - Cervical adenopathy.
 - Hand and foot swelling and erythema.
 - Skin rash.
- All symptoms do not need to be present simultaneously to make the diagnosis. When at least four criteria are present, most experts will make the diagnosis of KD on the fourth day of fever.

TABLE 63-1 Diagnostic Criteria for Kawasaki Syndrome

Fever persisting for ≥ 5 d
<i>And at least four of the following findings:</i>
Bilateral, painless bulbar conjunctival injection without exudate
Mucous membrane changes of the oropharynx including injected, dry, cracked lips, and oral mucosa; pharyngeal injection; “strawberry tongue”
Peripheral extremity changes including erythema and edema of hands and feet in the acute phase; periungual and generalized desquamation in the convalescent phase
Polymorphous truncal exanthema usually erythematous but may be pustular
Acute, nonpurulent cervical lymphadenopathy usually >1.5 cm
Findings cannot be explained by some other known disease process

- Up to 20% of children with coronary artery, aneurysms fail to meet the classical definition of KD. Therefore, incomplete KD should be considered in children with 5 days of fever and at least two of the classic criteria. Then, a stepwise approach considering acute-phase reactants (C-reactive protein or erythrocyte sedimentation rate), supplemental laboratory criteria and echocardiogram is taken to make the diagnosis (Fig. 63-1) Infants younger than 6 months with fever lasting for more than 7 days should be included in this algorithm, even without any physical findings.

PHASES OF KD

ACUTE OR FEBRILE PHASE

- The acute phase lasts 7 to 15 days and is the period when most diagnostic clinical features occur. Fever defines this phase.
- All physical findings are a consequence of the vasculitis.
- Bulbar conjunctivitis is bilateral, nonexudative, and spares the limbus.
- Mucocutaneous changes may include bright red erythema of the lips with cracking, a strawberry tongue and erythema of pharynx without exudate.
- Cervical adenopathy may not be prominent but nodes should be at least 1.5 cm in diameter.
- Rashes are red, polymorphous and develop in most children.
- Any part of the hands and feet may be edematous. The palms and soles may be erythematous.
- Involvement of almost any system can occur.
 - Urethritis is common, affecting 70% of patients. A sterile pyuria is characterized by white blood cells on microscopy but absence of leukocyte esterase on urinalysis.

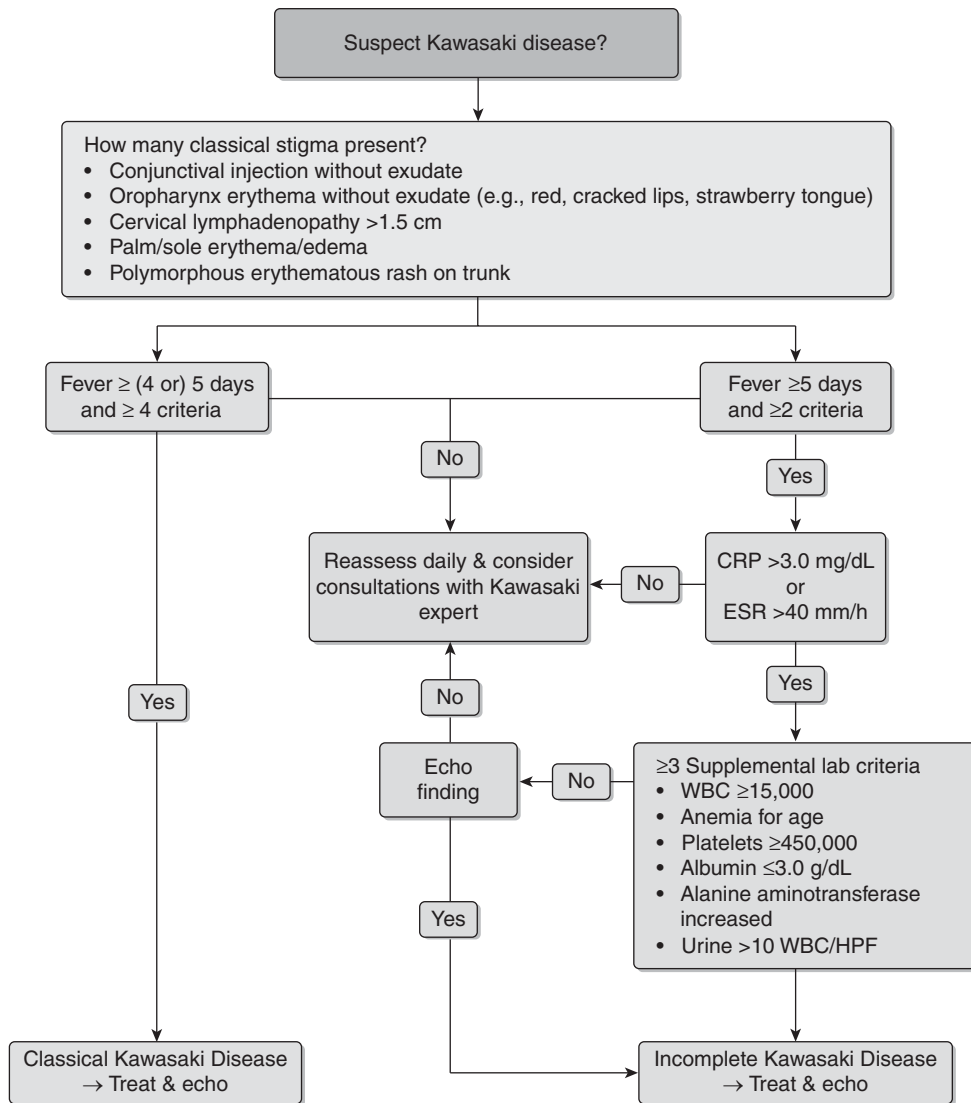


FIG 63-1. Algorithm for diagnosing Kawasaki disease.

- Gallbladder hydrops (acute acalculous distention of gallbladder) occurs in 15% of patients. A tender right upper quadrant mass may be palpated. Serum bilirubin will be elevated and the diagnosis can be confirmed by ultrasonography.
- Children may have abdominal pain, vomiting, or diarrhea because of gut vasculitis.
- Uveitis occurs in 25% to 50% of patients.
- Between 10% and 20% develop arthralgia or arthritis.

SUBACUTE PHASE

- The subacute phase lasts for approximately 2 to 4 weeks. It starts with the resolution of fever and

rise in platelet count; it ends with the resolution of thrombocytosis.

- The subacute phase is dominated by desquamation that may begin before the resolution of fever. It is noted first in the periungual region, with peeling underneath the fingernails and toenails. It may also be prominent in the diaper area.
- Platelet counts can range from 500,000 to 3,000,000/ mm^3 during this phase.

RECOVERY OR CONVALESCENT PHASE

- The recovery phase may last months to years. Some coronary artery disease is first recognized during this phase. Resolution of coronary artery disease hopefully occurs during this phase.

ANCILLARY DATA

- Laboratory findings are nonspecific in KD.
- The complete blood count often shows an elevated white blood cell count with a left shift. A mild hemolytic anemia may be present.
- Elevated platelet counts occur in the subacute phase but are usually normal in the acute phase.
- Acute-phase reactants (CRP, ESR) are markedly elevated.
- Urinalysis demonstrates moderate pyuria from urethritis.
- Bilirubinuria may occur as an early sign of hydrops of the gall bladder.
- Chest radiographs may show evidence of pulmonary infiltrates or cardiomegaly.
- The electrocardiogram may show dysrhythmias, prolonged PR or QT intervals (QTc) and nonspecific ST-T wave changes.
- Two-dimensional echocardiography may demonstrate coronary artery dilation or aneurysms, pericardial effusion or decreased contractility.

DIFFERENTIAL DIAGNOSIS

- The differential diagnosis is extensive because of the nonspecific nature of the clinical features (Table 63-2).
- Positive screening tests for infection may identify a concomitant infection, carrier-state, or viral shedding. This does not always rule out KD.

TABLE 63-2 Differential Diagnosis For Kawasaki Syndrome

Viral Illnesses	
	Rubeola (measles)
	Rubella
	Epstein-Barr virus infection
	Adenovirus infection
	Enterovirus infection
Bacterial Infections	
	Toxic shock syndrome
	Scarlet fever
Rickettsial Diseases	
	Rocky Mountain spotted fever
	Leptospirosis
Rheumatologic Disease	
	Juvenile rheumatoid arthritis
	Systemic lupus erythematosus
	Acute rheumatic fever
Drug/Toxin Reactions	
	Serum sickness
	Stevens-Johnson syndrome
	Mercury hypersensitivity (acrodynia)

CARDIOVASCULAR COMPLICATIONS

- The most serious manifestation of KD is cardiac involvement. KD has surpassed rheumatic fever as the leading cause of acquired heart disease in American children.
- Patients with KD have a 20% to 25% risk of developing coronary aneurysms without treatment. With treatment, this risk drops to 5%. Coronary aneurysms develop during the acute or subacute phase.
- With coronary aneurysms, sudden cardiac death or myocardial infarction may occur. Mortality peaks 15 to 45 days after the first day of fever.
- Coronary artery aneurysms typically occur after the first week but before the fourth week of illness. It is uncommon for them to develop after the sixth week.
- Echocardiography is the most sensitive technique for delineating coronary aneurysms. It should be performed as soon as the diagnosis is made, but treatment should not be delayed waiting for echocardiography. Myocarditis is universal and patients may have decreased left ventricular function
- Late death may occur from coronary occlusive disease, rupture of an aneurysm several years after onset or small blood vessel disease of the heart.

MANAGEMENT

- All patients diagnosed with KD should be hospitalized immediately for administration of intravenous gamma globulin (IVGG), aspirin therapy, and cardiac evaluation (echocardiography).
- IVGG has been shown to decrease the incidence of coronary artery aneurysm from 25% to 5%, when given during the acute phase of disease. The dose of IVGG is 2 g/kg infused over 8 to 12 hours as a single dose. IVGG given before the eighth day of fever reduces the risk of cardiac sequelae. However, IVGG should be given to any child who presents later with KD with continuing fever or aneurysm with persistent signs of inflammation.
- Aspirin has two roles in the treatment of KD. High-dose aspirin (100 mg/kg/d divided into four doses) is used for its anti-inflammatory effect. Duration of high-dose treatment varies from 48 hours after defervescence to the 14th day of the illness, depending on the center. This is followed by low-dose aspirin (3–5 mg/kg/d as a single-day dose) for its antiplatelet effects; this is continued for at least 6 to 8 weeks and until resolution of coronary abnormalities.
- Ten percent of children will have refractory disease, with persistence of fever 36 hours after completion of IVGG. The majority will respond to a second dose

of IVGG. For those refractory to this second dose of IVGG, there is limited data about how best to treat them. A third dose of IVGG, high-dose corticosteroids and tumor necrosis factor inhibitors have been used by experts in some centers.

PROGNOSIS

- The prognosis for patients receiving treatment within the first 8 to 10 days of the illness is excellent.
- Of the 5% with coronary aneurysms after treatment, 1% persists as giant aneurysms and the remainder regress. Even in patients with regression, there is debate about cardiac risk later in life. Patients may be at greater risk for atherosclerosis and some patients may have poorly compliant fibrotic vessel walls. All patients with a history of KD should be followed by a cardiologist at regular intervals.
- The overall mortality rate of Kawasaki syndrome in American children is between 1 and 2 per 1000. It is higher in infants younger than 1 year.

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QUESTIONS

1. A 5-year-old is brought to the ED by her parents with a history of fever, conjunctival injection, hand

swelling, and rash. You are suspicious of Kawasaki's disease. What is the minimum duration of fever required to make the diagnosis of Kawasaki disease?

- A. 2 days
 - B. 3 days
 - C. 5 days
 - D. 7 days
 - E. 10 days
2. Which of the following may be seen with Kawasaki disease?
 - A. Conjunctivitis
 - B. Erythematous pharynx with exudate
 - C. Swollen red hands
 - D. A and C
 - E. A, B, and C
 3. A 3-year-old male is brought in to the ED with a history of prolonged fever lasting over 6 days. The parents are concerned about the persistence of fever. Your differential includes Kawasaki disease. Which of the following may be seen with Kawasaki disease?
 - A. Neck stiffness
 - B. Abdominal pain
 - C. Joint pain
 - D. Difficulty breathing
 - E. All of the above
 4. Which of the following conditions can mimic Kawasaki disease?
 - A. Acute gastroenteritis
 - B. Scarlet fever
 - C. Meningitis
 - D. Lymphadenitis
 - E. All of the above
 5. A 4-year-old girl presents with a history of fever for 5 days, rash, and swelling to her hands. Which of the following laboratory results may be seen with Kawasaki disease?
 - A. Low hemoglobin
 - B. Elevated white blood cell count
 - C. Nitrates on urinalysis
 - D. A and B
 - E. A, B, and C
 6. Which of the following echocardiogram and electrocardiogram findings may be seen with Kawasaki disease?
 - A. Coronary aneurysms
 - B. Prolonged PR interval
 - C. Poor contractility
 - D. Pericardial effusion
 - E. All of the above

7. Which of the following statements about treatment of KD is true?
- Patient may be discharged home from emergency department after dose of IVGG
 - Do not give IVGG until echocardiogram confirms presence of aneurysms
 - Do not give IVGG if echocardiogram is normal
 - Aspirin dose should never exceed that of single baby aspirin (81 mg) per day
 - None of the above
8. A 6-year-old girl is diagnosed with probable KD in the ED. On echocardiogram she has evidence of a coronary aneurysm. The parents are very concerned and ask about the prognosis for her recovery. Which of the following is true about the prognosis of Kawasaki disease?
- IVGG reduces risk of coronary aneurysm from 25% to 5%, if given within 8 to 10 days of onset of fever.
 - Most cardiac deaths occur between 15 and 45 days from onset of fever.
 - Of children who have aneurysms in spite of IVGG treatment, 80% regress eventually.
 - Children who have regression of aneurysms may be at risk for cardiac disease than the general population.
 - All of the above
8. E. All are true. The prognosis for patients receiving treatment within the first 8 to 10 days of the illness is excellent. Of the 5% with coronary aneurysms after treatment, 1% persists as giant aneurysms and the remainder will regress.

64 THE PEDIATRIC HIV PATIENT IN THE ED

John F. Marcinak

INTRODUCTION

- Since human immunodeficiency virus (HIV) infection was first identified in 1981, the epidemic of HIV in the pediatric age group has affected a disproportionate number of infants born to mothers with HIV infection, and many infants with perinatal HIV infection have now reached adolescence.
- Up to 25% of perinatally infected children develop severe immunosuppression in the first year of life, but the majority of children have a slower progression to AIDS with a mean time period of 6 to 9 years.
- Use of highly active antiretroviral therapy (HAART) in children and adolescents has improved survival, decreased hospitalization rates, and transformed HIV infection from a uniformly fatal to a chronic disease.
- Not all children treated with HAART have been able to achieve or sustain viral suppression over time, and these children have a poorer prognosis and are at risk for complications of AIDS compared to children who achieve complete viral suppression.
- The natural history of HIV infection in adolescents with horizontally acquired HIV infection is similar to that of adults in that without treatment with antiretroviral (ARV) therapy, a 13-year-old adolescent would be expected to develop AIDS 10 to 12 years after primary infection with HIV.

EPIDEMIOLOGY

- In the United States, HIV infection affects a disproportionate number of non-Hispanic Black and Hispanic children and adolescents, and non-Hispanic Black and Hispanic women are also overrepresented in the HIV epidemic.
- There has been a significant decline in mother-to-child transmission (MTCT) of HIV since 1994, which has been attributed to U.S. Public Health Service Guidelines regarding use of zidovudine (ZDV) to reduce MTCT of

ANSWERS

1. C. A minimum of 5 days of fever is required, although some experts will diagnose KD if fever present for 4 days and at least four stigma present.
2. D. Exudative pharyngitis is not seen with KD and suggests infection.
3. E. KD can affect every organ in the body causing meningeal inflammation (neck pain), gastrointestinal complaints, joint involvement, and trouble breathing from either lung or cardiac involvement.
4. E. Careful history, physical examination, and interpretation of studies should distinguish diseases that mimic Kawasaki disease.
5. D. Laboratory findings are nonspecific in Kawasaki disease. The complete blood count often shows an elevated white blood cell count with a left shift. A mild hemolytic anemia may be present. Nitrates are not present with the sterile pyuria seen in Kawasaki disease.
6. E. All are possible.
7. E. All patients should be admitted and IVGG and aspirin started prior to and regardless of echocardiogram results.

TABLE 64-1 Distribution of HIV Infection and AIDS Cases Reported Among 13- to 19-Year Old Adolescents (by Sex) in 2005

SEX	HIV INFECTION (%) ^a	AIDS (%) ^b
Female	36	43
Male	64	57

^aData from 33 states with confidential name-based HIV infection reporting since 2001.

^bReported in 2005—United States and dependent areas.

Source: US Centers for Disease Control.

HIV (1994) and universal HIV counseling and voluntary HIV testing of pregnant women (1995).

- Compared to adolescents with newly acquired HIV infection, adolescents with perinatal HIV infection are more likely to have received HAART, thus prolonging life in this group.
- Newly acquired HIV in adolescents is still a significant public health problem with half of new HIV infections in the United States occurring in youths aged 13 to 24 years with 75% of infected youths in racial or ethnic minority groups.
- The epidemic of HIV in pediatrics has changed from a predominance of infection in infants with perinatal HIV infection to adolescents between 13 and 24 years of age infected through sexual transmission, either heterosexual or men who have sex with men.
- In 2005, adolescent females aged 13 to 19 years accounted for over one-third of the cases of HIV infection reported to the Centers for Disease Control and Prevention (CDC) (Table 64-1).
- Approximately 25% of sexually transmitted infections (STIs) reported in the United States are among adolescents.
- Female adolescents have unique risk factors of biological vulnerability for HIV acquisition, including the cervix during puberty being composed of single-layer columnar epithelial cells, STIs often being asymptomatic in women and transmission of HIV from male to female being more efficient than from female to male.

DIAGNOSIS

- The diagnosis of HIV infection in the pediatric and adolescent age groups depends upon a number of factors, the most important of which include age, clinical presentation, and the diagnostic test employed.
- HIV antibody testing by enzyme immunoassay (EIA) should not be used for diagnosis of HIV in infants younger than 18 months or for diagnosis of acute HIV infection in an adolescent patient.

- In September 2006 the CDC recommended routine HIV testing for all patients 13 to 64 years of age in health care settings, including the emergency department (ED), with consent for testing included in the consent for care, with testing performed unless the patient declines (referred to as “opt-out” screening).
- There are currently six rapid HIV tests approved by the U.S. Food and Drug Administration using blood (either whole blood, serum, or plasma) or oral fluid (one only), and four are easy to perform with negligible chance for error and therefore CLIA (Clinical Laboratory Improvements Amendments) waived.
- Rapid HIV tests have shown a very high sensitivity and specificity (both >99%) for diagnosing HIV infection, and the use of rapid HIV testing has been shown to be feasible and acceptable for patients as part of routine health care services in the ED setting.
- There is currently no information available regarding the acceptance of opt-out HIV testing of adolescents in an ED setting.
- The viral load, as measured by quantitative RNA PCR, is the test with the shortest time from infection to development of a positive test (Table 64-2), and testing for both HIV RNA and HIV antibody should be performed when acute HIV infection is being considered.

CLINICAL PRESENTATIONS

- Important principles that guide ED management of the child or adolescent known to have HIV infection include understanding of appropriate infection-control procedures, noting the patient’s CD4 count/percentage, and consultation with the patient’s primary HIV care provider.
- In pediatric patients <13 years of age, a CD4 percentage $\geq 25\%$ indicates no immune suppression, while a CD4 percentage of 15 to 24 indicates moderate immune suppression and <15% represents severe immune suppression.

TABLE 64-2 Time of HIV Infection for Various Diagnostic HIV-1 Markers

MARKER	TIME AT WHICH HIV CAN FIRST BE DETECTED
Antibody ^a	22 d
p 24 antigen	16 d
RNA by PCR	9 d

^aThird-generation, IgM sensitive EIA (enzyme immunoassay).

Data from Fiebig EW, Wright DJ, Rawal BD, et al. Dynamics of HIV viremia and antibody seroconversion in plasma donors: implications for diagnosis and staging of primary HIV infection. *AIDS*. 2003;17:1871.

Data from Branson BM. State of the art for diagnosis of HIV infection. *Clin Infect Dis*. 2007;45:S221.

- An HIV infected infant with the diagnosis of Pneumocystis pneumonia (PCP) caused by *Pneumocystis jirovecii* is most likely to present at 3 to 6 months of age with diffuse interstitial infiltrates on chest x-ray and respiratory failure.
- Common serious bacterial infections such as pneumonia, recurrent otitis media and sinusitis, meningitis, and sepsis can be first manifestations of disease in children with HIV infection.
- An adolescent with acute HIV infection will likely present with fever, pharyngitis, oral ulcers, lymphadenopathy, gastrointestinal symptoms and a maculopapular rash (seen in more than 50% of cases reported in most series), but a papulovesicular rash can also be seen (Fig. 64-1).
- More unusual manifestations of acute HIV infection include clinical hepatitis, acute pneumonitis, aplastic anemia, vasculitis, rhabdomyolysis, Guillain–Barré syndrome, and facial nerve palsy.
- The most common laboratory abnormalities found in acute HIV infection include leukopenia, lymphopenia, and thrombocytopenia.



FIG 64-1. Primary HIV infection. A maculopapular rash is seen in more than half of persons with symptomatic acute HIV infection. This less typical papular/vesicular rash was present in a patient with primary HIV infection. (Courtesy of Gregory K. Robbins, MD, MPH.)

COMPLICATIONS IN HIV-INFECTED PATIENTS

- In a large multicenter cohort study of HIV infected children receiving HAART, the four most common first-time infections were bacterial pneumonia, herpes zoster, oral candidiasis, and dermatophyte infections. One-half of the children had a CD4 percentage of at least 25% at the time of first infection.
- Infections most likely to be seen in the pediatric or adolescent HIV patient with a CD4 percentage <25 include herpes zoster and oral candidiasis, while development of opportunistic illness and infection is strongly associated with a CD4 percentage <15.
- Children and adolescents with HIV infection can have significant oral and dental lesions including dental caries, gingivitis, aphthous ulcers, herpes stomatitis, oral candidiasis, and tongue lesions, such as depapillated tongue and median rhomboid glossitis. Oral lesions are most commonly seen in children with severe immunosuppression (CD4 percentage <15).
- Bilateral cervical lymph node, as well as parotid gland, enlargement can also occur and is more common with severe immunosuppression, and those children with generalized cervical, axillary, and inguinal lymphadenopathy are more likely to have uncontrolled viral replication with elevated viral loads as measured by RNA PCR.
- An HIV-infected child or adolescent with asymmetric lymphadenopathy that has persisted for weeks in spite of antibiotic therapy should raise consideration of a malignancy such as lymphoma (most commonly non-Hodgkin lymphoma) or Hodgkin disease.
- Psychiatric hospitalizations among HIV-infected children and adolescents younger than 15 years are significantly higher than for the same age group in the general pediatric population, with most common reasons for hospitalization including depression, behavioral disorders, and suicidal ideation/attempts.
- ARV therapy in children and adolescents can be associated with a number of adverse effects including development of drug resistance, toxicity and long-term complications.
- Important distinct adverse drug effects include mitochondrial dysfunction (including lactic acidosis and hepatic toxicity), metabolic abnormalities (such as fat maldistribution, body habitus changes, hyperlipidemia, hyperglycemia, and insulin resistance, osteopenia, osteoporosis, and osteonecrosis), hematologic complications, and allergic reactions.
- An important life-threatening complication of antiretroviral therapy seen in the ED is lactic acidosis.
- The complete list of the ARV medications the pediatric or adolescent patient is receiving is important to determine since significant adverse events, including life-threatening and fatal cases of lactic acidosis, are associated with specific individual or groups of ARV medications (Table 64-3).

TABLE 64-3 Distinct Adverse Drug Events With Individual or Classes of Antiretroviral (ARV) Therapy

ADVERSE EVENT	CLASS OF ARV	ASSOCIATED INDIVIDUAL ARVS
Lactic acidosis	NRTIs	Didanosine, Stavudine, Zidovudine Lamivudine
Hepatic toxicity	All NRTIs, NNRTIs, PIs Fusion inhibitor	Common to all the classes Enfuvirtide
Hyperglycemia	PIs	Common to the entire class
Hematologic (anemia and neutropenia)	NRTIs	Zidovudine, Didanosine, Stavudine
Skin rash (maculopapular)	NNRTIs PIs	Common to the entire class Amprenavir, Fosamprenavir, Atazanavir, Darunavir, Tipranavir
Hypersensitivity reaction	NRTIs	Abacavir
	NRTIs	Abacavir Nevirapine
	Fusion inhibitor	Enfuvirtide
Stevens-Johnson syndrome	NNRTIs	Nevirapine, Efavirenz, Delaviridine

NRTIs, nucleoside analogue reverse transcriptase inhibitors; NNRTIs, nonnucleoside analogue reverse transcriptase inhibitors; PIs, protease inhibitors.

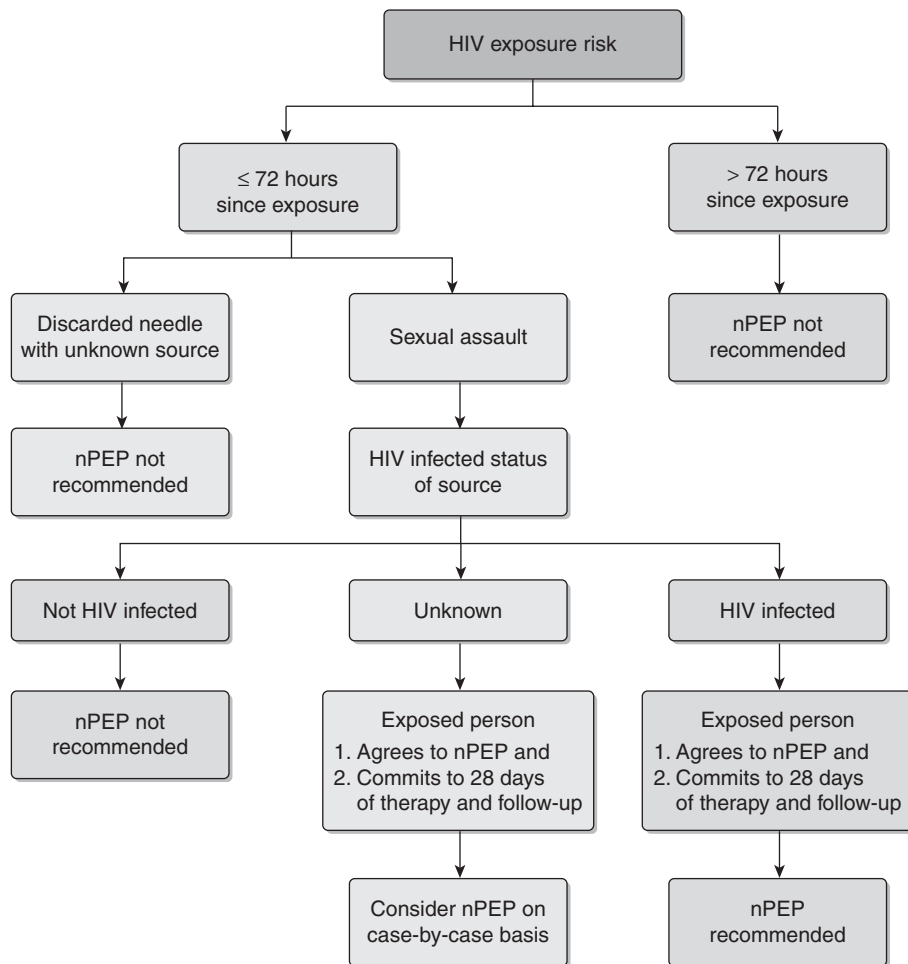


FIG 64-2. Evaluation and treatment of children and adolescents for possible nonoccupational exposure to HIV from a discarded needle or sexual assault.

POSTEXPOSURE PROPHYLAXIS

- The issue of postexposure prophylaxis for children and adolescents for nonoccupational exposure to HIV is likely to be encountered in the ED setting with the two most likely clinical settings involving a needle-stick injury or sexual assault.
- Guidelines have been established to aid in the decision-making process for the appropriate use of antiretroviral medications for nonoccupational postexposure prophylaxis (nPEP) in the pediatric and adolescent patient.
- There have been no confirmed reports of HIV transmission from percutaneous injury by a needle found in the community, while victims of sexual assault are at much higher risk for HIV transmission.
- Use of nPEP should be considered only if the exposure has occurred within 72 hours, the exposed patient voluntarily accepts nPEP and agrees to 28 days of ARV medications for prophylaxis and appropriate follow-up (Fig. 64-2).
- The preferred ARV medications recommended by the CDC for nPEP include a three drug HAART regimen that is either nucleoside reverse transcriptase inhibitor (NRTI)- or protease inhibitor (PI)-based.
- Completion of a 28-day course of ARV medications and close follow-up may be quite challenging for adolescents who are victims of sexual assault.
- In adolescent victims of sexual assault in whom nPEP is recommended or considered, a 3- to 5-day starter pack of ARV medications should be offered if nPEP is accepted by the adolescent along with close follow-up with a pediatric and adolescent HIV provider.

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QUESTIONS

1. A term newborn infant is born to a 25-year-old female with HIV infection. Compared to infants born to HIV infected pregnant women in the 1980s, the infant now is less likely to acquire perinatal infection for which of the following reasons?
 - A. Antiretroviral prophylaxis in the pregnant woman.
 - B. Antiretroviral prophylaxis in the newborn infant.
 - C. Antiretroviral prophylaxis in both the pregnant woman and newborn infant.
 - D. HIV is now less virulent resulting in lower risk of perinatal transmission.
 - E. There is now less HIV drug resistance.
2. A 15-year-old adolescent female diagnosed with *Chlamydia trachomatis* infection is also tested for HIV and is found to be HIV positive. The most likely method of transmission of HIV in this case would be which of the following in this patient?
 - A. Heterosexual contact
 - B. Homosexual contact
 - C. Intravenous drug use
 - D. Blood transfusion
 - E. Needle stick from an unknown source
3. You suspect infectious mononucleosis in a 17-year-old adolescent female but the monospot test is negative. Your next consideration is acute HIV infection and the diagnostic test most likely to confirm that diagnosis is which of the following for this patient?
 - A. HIV enzyme immunoassay
 - B. HIV RNA PCR
 - C. HIV DNA PCR
 - D. HIV culture
 - E. Lymphocyte subsets

4. You are considering introducing rapid HIV testing in the ED of the hospital. An important characteristic of all the currently licensed rapid HIV tests is which of the following?
 - A. Lower specificity compared to the HIV enzyme immunoassay.
 - B. Specimens tested can include whole blood and oral fluid.
 - C. Quality control required by a hospital clinical laboratory.
 - D. An average time to obtain results of 12 hours.
 - E. Similar sensitivity compared to the HIV enzyme immunoassay.
5. A 6-year-old child with perinatal HIV infection presents to the ED with high fever, difficulty breathing, and generalized lymphadenopathy. A CD4 cell count percentage that is indicative of moderate immunosuppression would be which of the following?
 - A. 8%
 - B. 14%
 - C. 22%
 - D. 25%
 - E. 30%
6. A 16-year-old adolescent male who has a recent history of infection with *Neisseria gonorrhoeae* presents to the ED with fever, pharyngitis, cervical lymphadenopathy, and a maculopapular rash. You suspect infectious mononucleosis caused by Epstein–Barr virus, but a monospot test for heterophil antibody is negative. The most likely diagnosis in this clinical setting is which of the following?
 - A. Cytomegalovirus infection
 - B. Acute Hepatitis A
 - C. Adenovirus
 - D. Acute HIV
 - E. Human herpes virus type 6
7. An 8-year-old child with HIV infection presents to the ED with a history of fever for 10 days. You determine after speaking with the child's pediatric HIV provider that the most recent CD4 percentage is 11%. The infection most likely to be associated with a CD4 percentage <15 includes which of the following?
 - A. Herpes Zoster
 - B. *Escherichia coli* urinary tract infection
 - C. Tinea capitis
 - D. Sinusitis
 - E. *Staphylococcus aureus* cellulitis
8. A 13-year-old adolescent female with perinatal HIV infection on HAART including one PI and 2 NRTIs presents to the ED with weakness, vomiting, diarrhea, and difficulty breathing. The adolescent's HIV infection is well controlled with an undetectable HIV viral load measured by HIV RNA PCR, and an absolute CD4 count of 600/ μ L. The most serious complication to consider in this patient is which of the following conditions?
 - A. Viral gastroenteritis
 - B. Gastrointestinal adverse drug reaction from PIs
 - C. Cryptosporidium
 - D. Salmonellosis
 - E. Lactic acidosis
9. A 14-year-old adolescent female presents to the ED after sexual assault from an unknown assailant. You are considering whether or not to consider HIV prophylaxis in this setting. The maximum time period after the assault in which non-occupational postexposure prophylaxis for HIV should be considered is which of the following?
 - A. 24 hours
 - B. 48 hours
 - C. 72 hours
 - D. 96 hours
 - E. 120 hours
10. You decide to recommend HIV postexposure prophylaxis to the adolescent who is a victim of sexual assault. An important initial step in increasing the success of the ARV prophylaxis to prevent HIV infection is to do which of the following?
 - A. Provide a 3–5 day starter pack of the ARV medications.
 - B. Write a prescription for the ARV medications to be filled at a local pharmacy.
 - C. Prescribe one medication for ARV prophylaxis.
 - D. Schedule follow-up within 7 days at the ED.
 - E. Prescribe a 14-day course of 3 ARV medications.

ANSWERS

1. C. Antepartum ARV prophylaxis with HAART starting anytime after 14 weeks gestation, intravenous ZDV during labor and delivery and followed by oral ZDV to the newborn infant as soon as possible after birth has resulted in a decrease in perinatal HIV transmission to less than 2%. Administration of ZDV alone to infants born to pregnant women identified during labor and delivery within 48 hours of birth has also been shown to decrease transmission but is not nearly as effective. HIV drug resistance has increased in frequency since the first clinical trials conducted in the 1990s demonstrated the effectiveness of ARV prophylaxis for the prevention of MTCT.

2. A. Acquisition of HIV during adolescence occurs primarily as a result of sexual exposure with little contribution secondary to intravenous drug use. In adolescent females, heterosexual transmission predominates while adolescent young men who have sex with men are at high risk for acquiring HIV. Needlestick injuries from an unknown source found in the community have not been confirmed as a method of HIV transmission. The risk of HIV transmission via a blood transfusion is very low with the estimated per-risk of contamination at 1 in 675,000.
3. B. With acute HIV infection, antibody as measured by HIV EIA can be negative because even with the new generation HIV EIAs that also detect IgM antibody, the time period from infection to seroconversion is 3–4 weeks while HIV RNA can be detected as early as 9 days after onset of infection. HIV culture is not available routinely in clinical laboratories and results take up to 28 days. HIV DNA PCR is a qualitative test and is the preferred test to diagnose HIV in infants.
4. E. Rapid HIV tests are single-use EIAs that can provide results in as short a time as 15–20 minutes. The licensed tests that have received a Clinical Laboratory Improvements Amendment (CLIA) waiver can use whole blood, but only one CLIA-waived HIV test is approved for use with an oral fluid sample. Rapid HIV tests have similar high sensitivity and specificity when compared to HIV EIAs. Therefore, a negative result for a rapid HIV test is conclusive and usually requires no follow-up.
5. C. Moderate immunosuppression is defined as a CD4 percentage of 15–24 while with severe immunosuppression the CD4 percentage is less than 15. Like adults, the normal absolute CD4 T-lymphocyte count is $\geq 500/\mu\text{L}$ in children 6 years and older. Also for children 6 years and older, a CD4 T-lymphocyte count of $200/\mu\text{L}$ or less is indicative of severe immunosuppression. In children under the age of 5 years, the CD4 percentage is preferred for monitoring the immune status because the absolute CD4 T-lymphocyte count varies more with age among individual children in this age group.
6. D. An adolescent with an infectious mononucleosis like-syndrome should be evaluated for acute HIV infection, especially with a history of a STI. The other diagnoses listed can also present as an infectious mononucleosis-like syndrome that is indistinguishable from Epstein–Barr virus. Cytomegalovirus infection is usually inapparent, and human herpes virus type 6 usually occurs in children between 6 months and 3 years of age. Adenovirus infection will often have conjunctivitis, and respiratory tract symptoms predominate. An adolescent with symptomatic Hepatitis A infection will likely have low-grade fever, nausea, and vomiting, with or without jaundice.
7. A. In perinatally HIV infected children, herpes zoster continues to occur, and a CD4 percentage < 15 is a significant risk factor. Infections seen commonly in healthy children, such as otitis media, sinusitis, and tinea capitis, also are frequently seen in children with perinatal HIV infection. HIV infected children also have an increased incidence of urinary tract infections. *S aureus* is the most common skin infection that occurs in HIV infected children.
8. E. Lactic acidosis is a severe and life threatening complication of ARV therapy that is important to recognize. Decompensated lactic acidosis has been reported in the pediatric patient, and the predominant presenting symptoms are gastrointestinal and non-specific. The ARV medication should be held when the diagnosis is being considered. Mitochondrial toxicity related to NRTI administration is the cause of this adverse event. The most common adverse effects of PIs are nausea, vomiting, and diarrhea that may lead to discontinuation, but these effects are not life-threatening. Cryptosporidium can occur in the HIV infected pediatric patient. Cryptosporidium causes severe and protracted diarrhea with abdominal pain and anorexia in HIV-infected children with advanced HIV infection. Salmonella can cause disseminated infection in children with HIV infection, resulting in bacteremia, pneumonia, osteomyelitis or meningitis. A low CD4 count and advanced HIV disease increases the risk of Salmonella infection.
9. C. The U.S. Department of Health and Human Services (DHHS) has provided recommendations for use of antiretroviral drugs to prevent HIV infection after unanticipated nonoccupational exposure to HIV. Evidence from both animal studies and human observational studies have demonstrated that nPEP administered within 48–72 hours might reduce the risk of acquiring HIV after mucosal or other non-occupational exposure. The sooner nPEP is administered after exposure, the more likely for transmission to be interrupted. A caveat is included that a clinician could consider nPEP for exposures >72 hours if the exposure confers a serious risk for HIV transmission, and the diminished potential

benefit of nPEP outweighs the risks for adverse events.

10. A. Providing a 3–5 day starter pack of ARV medication is important and is recommended by the U.S. DHHS. All ARV medications are not routinely available on an immediate basis at many pharmacies. A 3-drug HAART regimen is preferred based on the observation that the maximal viral suppression as has been demonstrated with treatment will provide the best chance of preventing infection in the exposed pediatric patient. There is no evidence that a 3-drug regimen is more effective than a 2-drug regimen, but a single drug regimen is not recommended under any circumstances. The recommended duration of nPEP for either a 3-drug or 2-drug regimen is 28 days.

65 TICK-BORNE INFECTIONS

Scott A. Heinrich

INTRODUCTION

- Ticks are capable of transmitting numerous diseases to children including bacterial, viral, and parasitic pathogens.
- As most patients do not recall a tick bite, tick-borne illnesses should be included in the differential diagnosis of nonspecific febrile illnesses, especially in the presence of an associated rash.
- The best ways to prevent tick-borne diseases are to avoid tick exposure and ensure prompt tick removal if bites do occur.
 - Children that are going to be in rural, wooded areas in endemic regions should wear long-sleeved shirts buttoned at the cuff and long pants tucked into their socks.
 - Clothing may be sprayed with 0.5% permethrin.
 - DEET, a commercially available insect repellent, can also be used at concentrations of 20% to 30%.
- Attached ticks should be removed as soon as possible to lessen the risk of infection.
 - First, viscous lidocaine may be applied to kill the tick and anesthetize the site.
 - Next, gentle, continuous traction is applied with a pair of fine forceps to the tick's head, ensuring that the tick is not crushed.
 - Lastly, ensure that all parts of the tick have been removed.

LYME DISEASE

- Lyme disease is the most common vector-borne illness in the United States and Europe.
- Three-fourth of patients have the onset of illness between May and August.
- Caused by gram-negative *Borrelia burgdorferi* in the United States.
- Transmitted by ticks of the *Ixodes ricinus* complex: *Ixodes scapularis* and *Ixodes pacificus* in the United States and *I ricinus* in Europe.
- For a substantial rate of transmission to occur, the tick has to be attached for 48 hours or more.
- Three stages of Lyme borreliosis infection: early localized, early disseminated, and late Lyme disease.
 - Erythema migrans, the “bulls eye rash”, is seen in up to 85% of patients and usually occurs within 7–10 days.
 - The second stage of Lyme infection, early disseminated disease, is characterized by multiorgan disease resulting from the hematogenous spread of *B burgdorferi*.
 - Erythema migrans is treated with amoxicillin, cefuroxime, or doxycycline for 14–21 days.
 - Lyme meningitis and carditis are treated with ceftriaxone, cefotaxime, penicillin G, or doxycycline if beta-lactam allergy is present.

ROCKY MOUNTAIN SPOTTED FEVER

- RMSF is the most common tick-borne rickettsial disease in the United States and is one of the most virulent human infections identified.
- RMSF is found only in North and South America, and in the United States is found in all 48 contiguous states, except Vermont and Maine.
- RMSF is caused by *Rickettsia rickettsii*, an obligate intracellular gram-negative coccobacillus that typically infects the host's vascular endothelial cells.
- Symptoms of RMSF usually appear approximately 7 days after tick exposure, although a history of tick bite is only elicited in 50% to 60% of patients.
- The initial phase of RMSF infection is characterized by sudden onset of fever, malaise, and severe headache, with accompanying nonspecific symptoms such as myalgias, nausea and vomiting, abdominal pain, anorexia, and photophobia.
- The rash of RMSF usually begins as blanching, erythematous macules on the wrists and ankles, and spreads centripetally to the arms, legs, and trunk within hours.
- Some patients may develop neurologic symptoms such as meningismus, altered mental status, amnesia,

coma, seizures, cranial nerve palsies, central deafness, and cortical blindness. Other manifestations are less commonly reported but include conjunctivitis, periorbital and peripheral edema, congestive heart failure, arrhythmias, myocarditis, shock, hepatomegaly, and jaundice.

- Because RMSF has a 20% mortality rate if untreated and up to a 5% rate if treated, and rapid confirmatory diagnosis is not easily achieved, antibiotic therapy should be started when the disease is suspected.
- Doxycycline is the drug of choice for treating RMSF regardless of the child's age, usually for 7–10 days.

TULAREMIA

- Tularemia is caused by infection with the gram-negative coccobacillus, *Francisella tularensis*, and is found worldwide and in every state except Hawaii.
- Most patients in the United States acquire tularemia infection from tick bites or from contact with infected mammals, usually rabbits.
- More than half of all reported tularemia cases are from just four states: Arkansas, Missouri, South Dakota, and Oklahoma, in descending order.
- Tularemia infection is commonly classified into six categories: ulceroglandular, glandular, oculoglandular, oropharyngeal, pneumonic, and typhoidal.
- Tularemia infection presents as an acute febrile illness with an incubation period averaging 3 to 5 days (range 1–21 days), and disease severity ranges from mild to severe
- Aminoglycosides are currently first-line therapy for tularemia, with doxycycline and chloramphenicol as alternative choices.

BABESIOSIS

- Babesiosis, a malaria-like disease, is caused by an intraerythrocytic protozoa, *Babesia microti* and *Babesia equi*.
- Babesiosis is transmitted by *Ixodes* ticks, although it may also be spread by blood transfusions and transplacental or perinatal transmission.
- The initial disease is marked by nonspecific symptoms such as fevers, chills, diaphoresis, fatigue, myalgia, malaise, headaches, nausea, and abdominal pain, although most cases are asymptomatic.
- Treatment options include atovaquone and azithromycin or clindamycin and quinine, which should be used for more serious cases, and lasts for 7–10 days.
- RBC exchange transfusion may be necessary for children with severe babesiosis as evidenced by

high-grade parasitemia ($\geq 10\%$), significant hemolysis, or pulmonary, renal, or hepatic compromise.

HUMAN MONOCYTE EHRLICHIOSIS

- Human monocyte ehrlichiosis (HME) is caused in the United States by *Ehrlichia chaffeensis*, a small, intracellular, gram-negative coccobacillus that infects circulating leukocytes.
- HME causes a clinical syndrome in children very similar to RMSF with almost all children presenting with fever and headache; myalgia, anorexia, nausea, abdominal pain, and vomiting are frequently seen.
- A tick-bite history is obtained in up to 90% of children within the preceding 3 weeks of illness onset and two-thirds of children will have a rash that commonly affects the trunk, extremities, or both.
- Rarely, the disease may lead to pneumonitis, respiratory failure, encephalitis, meningitis, shock, or disseminated intravascular coagulation.
- The treatment of choice for children with HME is doxycycline for 3 days after defervescence or for a minimum of 5 to 10 days. Rifampin in an alternative treatment.

HUMAN GRANULOCYTTIC ANAPLASMOSIS

- Human granulocytic anaplasmosis (HGA), formerly human granulocytic ehrlichiosis, is caused by infection with the gram-negative, intracellular coccobacillus, *Anaplasma phagocytophila*.
- Clinical manifestations of HGA, similar to HME, are usually nonspecific and include fever, chills, headache, and myalgias.
- The disease is usually mild and self-limited with symptoms resolving within 30 days in most patients, regardless of antibiotic treatment.
- Treatment is with doxycycline and if concomitant Lyme disease, amoxicillin or cefuroxime is added at the duration of treatment.

COLORADO TICK FEVER

- Colorado tick fever occurs when an RNA virus is transmitted by the bite of the wood tick *Dermacentor andersoni*.
- The disease usually presents as an acute, self-limited, febrile illness with headaches, chills, myalgias, and photophobia.
- Treatment for the disease is supportive.

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QUESTIONS

- Erythema migrans, the “bull’s-eye rash,” is sufficient for the diagnosis of which disease in endemic areas?
 - Rocky mountain spotted fever
 - Lyme disease
 - Tularemia
 - Human monocyte ehrlichiosis
 - Babesiosis
- A 5-year-old female presents to the ED in North Carolina with her parents. They state that she was walking in the woods about 2 weeks ago and then a week later developed sudden onset headaches, fevers, myalgias, nausea and vomiting, and photophobia. Four days later she developed a reddish rash on her wrists and ankles that quickly moved towards her trunk, arms and legs. She now has a petechial rash on her arms and legs, including her palms and soles. What is the most likely diagnosis?
 - Rocky mountain spotted fever
 - Lyme disease
 - Tularemia
 - Human granulocytic anaplasmosis
 - Colorado tick fever
- In regards to the above patient, what is the treatment of choice?
 - Amoxicillin
 - Azithromycin
 - Doxycycline
 - Penicillin G
 - Sulfamethoxazole-trimethoprim
- An 8-year-old male presents to the ED complaining of a lesion to her right leg with a very painful surrounding area. She states that she was visiting her cousin in Arkansas 4 days ago. She first developed abrupt-onset fevers, chills, myalgias, headache, and vomiting. On examination, she has an ulcerated eschar of her right upper thigh with surrounding painful lymphadenopathy. What is her most likely diagnosis?
 - Rocky mountain spotted fever
 - Lyme disease
 - Tularemia
 - Colorado tick fever
 - Babesiosis
- In regards to the above patient, what is the treatment of choice?
 - Amoxicillin
 - Ceftriaxone
 - Azithromycin
 - Gentamicin
 - Sulfamethoxazole-trimethoprim
- A 10-year-old male presents to your ED complaining of gradual onset fevers, chills, diaphoresis, fatigue, myalgia, malaise, headaches, nausea, and abdominal pain. He was hiking with his Boy Scout troop 2 weeks ago and noted a small “bug” on his arm. On physical examination, he has hepatomegaly and splenomegaly. On laboratory data, he has anemia, thrombocytopenia, mild liver dysfunction, and evidence of hemolysis. What is the most likely diagnosis?
 - Rocky mountain spotted fever
 - Lyme disease
 - Tularemia
 - Colorado tick fever
 - Babesiosis
- The clinical syndrome associated with HME most closely resembles which other tick-borne illness?
 - Rocky mountain spotted fever
 - Lyme disease
 - Tularemia
 - Colorado tick fever
 - Babesiosis

8. What is the treatment of choice for Colorado tick fever?
 - A. Amoxicillin
 - B. General supportive care
 - C. Cefuroxime
 - D. Doxycycline
 - E. Acyclovir
 9. How are the three stages of clinical Lyme borreliosis classified?
 - A. Early disseminated disease, early local disease, and late local disease.
 - B. Early local disease, late local disease, and late Lyme disease.
 - C. Early local disease, early disseminated disease, and late Lyme disease.
 - D. Early Lyme disease, late local disease, and late disseminate disease.
 - E. Early disseminated disease, late disseminated disease, and late Lyme disease.
 10. A 10-year-old girl has a rash characteristic of erythema migrans after hiking in the woods with her family. What percentage of patients with erythema migrans will go on to have either monoarticular or oligoarticular arthritis if left untreated?
 - A. 5%
 - B. 20%
 - C. 40%
 - D. 60%
 - E. 80%
4. C. Tularemia infection presents as an acute febrile illness with an incubation period averaging 3 to 5 days. Children present with abrupt-onset fever, myalgia, chills, vomiting, fatigue, and headache. Ulceroglandular tularemia presents with an ulcerating eschar at the inoculation site with painful regional lymphadenopathy, whereas the glandular form lacks the inoculation eschar.
 5. D. Aminoglycosides are currently first-line therapy for tularemia. Doxycycline and chloramphenicol are alternative choices, although they are bacteriostatic and are associated with increased relapses. Fluoroquinolones have been effective in treating tularemia, but their efficacy is not yet proven.
 6. E. Although most cases of babesiosis are asymptomatic in North America, when symptomatic it produces a malaria-like illness. The initial disease is marked by nonspecific symptoms such as fevers, chills, diaphoresis, fatigue, myalgia, malaise, headaches, nausea, and abdominal pain. Laboratory findings may reveal thrombocytopenia, anemia, liver dysfunction, renal dysfunction, and evidence of hemolysis.
 7. A. HME causes a clinical syndrome in children very similar to RMSF. Almost all children present with fever and headache; myalgia, anorexia, nausea, abdominal pain, and vomiting are frequently seen. Two-thirds of children will have a rash that commonly affects the trunk, extremities, or both.
 8. B. Colorado tick fever usually presents as an acute, self-limited, febrile illness with headaches, chills, myalgias, and photophobia. Treatment is supportive.
 9. C. The most common manifestation of early-localized disease is erythema migrans. The second stage of Lyme infection, early-disseminated disease, is characterized by multiorgan disease resulting from the hematogenous spread of *B burgdorferi*. The third stage, late Lyme disease, involves recurring large joint arthritis and neuroborreliosis that manifests as peripheral neuropathy, paresthesias, radicular pain, or encephalopathy.
 10. D. The majority of patients with erythema migrans who are not treated will go on to have complications of late-stage Lyme disease—60% will develop monoarticular or oligoarticular arthritis, 10% will have neurologic manifestations (facial nerve palsy being most common), and approximately 5% will have cardiac complications.

ANSWERS

1. B. Erythema migrans is seen in up to 85% of patients with Lyme disease and is classically described as an erythematous macular or papular rash that spreads centrifugally to form an erythematous annular plaque. The diagnosis of erythema migrans in endemic areas is purely clinical, and is the only manifestation of Lyme disease that is sufficient to make the diagnosis without laboratory data.
2. A. The initial phase of RMSF infection is characterized by sudden onset of fever, malaise, and severe headache, with accompanying nonspecific symptoms such as myalgias, nausea and vomiting, abdominal pain, anorexia, and photophobia. A rash usually begins between 3 and 5 days as blanching, erythematous macules on the wrists and ankles, and spreads centripetally to the arms, legs, and trunks within hours. By the end of the first week, the characteristic petechial rash will develop in 35% to 60% of patients.
3. C. Doxycycline is the drug of choice for treating RMSF regardless of the child's age. The optimal duration of treatment has not yet been established, but the current recommendations are to treat for 3 days after the fever subsides and until there is evidence of clinical improvement.

66 COMMON PARASITIC INFESTATIONS

Steven Lelyveld
Gary R. Strange

NEMATODES (ROUNDWORMS)

ASCARIASIS

- *Ascaris lumbricoides* is the largest and most prevalent human nematode (one billion cases worldwide).
- It is most common in preschool and early school age children.
- The egg measures 65 by 45 μm ; the adult grows to a length of 30 cm.
- Upon ingestion, the egg hatches in the small intestine; the larvae burrow through the gut mucosa, enter the bloodstream, and migrate to the lungs causing shortness of breath, hemoptysis, eosinophilia, fever and Löffler's pneumonia.
- The larvae break through the alveoli, migrate up the bronchial tree, and are swallowed.
- *A lumbricoides* can live freely in the small intestine as an adult for up to a year, shedding eggs in the stool.
- It can be asymptomatic or can cause gastrointestinal symptoms, including pain, protein malabsorption, biliary duct or bowel obstruction, and appendicitis (Table 66-1).
- If multiple infestations are present, treat *Ascaris* first to avoid causing obstruction from a large worm burden.
- Diagnose with stool testing for ova or do serologic hemagglutination and flocculation tests.
- Treatment: Albendazole (400 mg orally as a single dose) or ivermectin (150–200 $\mu\text{g}/\text{kg}$ orally as a single dose) is curative. Piperazine salts (50–75 mg/kg for 2 days) are recommended for ascariasis complicated by intestinal or biliary obstruction, since they cause rapid expulsion of the worms.

ENTEROBIASIS

- *Enterobius vermicularis* (pinworm) affects all ages, socioeconomic levels, and regions in the United States.
- The most common presentation is a toddler with anal itch (Table 66-1).
- The egg is inhaled or ingested and hatches between the ileum and ascending colon.
- The adult is 3 to 10 mm and thread shaped. It lives and copulates in the colon for 1 to 2 months.

- The gravid female migrates to the anus, where it deposits embryonated eggs during early morning hours (Fig. 66-1). When the host stirs, the adult migrates back into the body, causing pruritus ani, dysuria, enuresis, and vaginitis. Scratching and hand–mouth behavior reinoculates the host and the cycle repeats.
- Sticky tape placed against the perianal skin when the child first awakens and then viewed under low power is diagnostic.
- Treatment: albendazole, 400 mg orally, pyrantel pamoate (11 mg/kg) or mebendazole (100 mg) as a single dose, with a repeat given 2 weeks later to remove secondary hatchlings.

TRICHURIS TRICHIURA

- *Trichuris trichiura* (whipworm) is found in southern Appalachia and other warm rural areas.
- The life cycle mimics that of *E vermicularis*.
- The eggs are of similar size and configuration, with the addition of a rounded cap at each pole (Fig. 66-2).
- The adult resembles *E vermicularis*, with a long whip-like projection at one end.
- It lives in the cecum and can either be asymptomatic or cause malabsorptive symptoms, pain, bloody diarrhea, fever, rectal prolapse, anemia, and developmental and cognitive deficits (Table 66-1).
- Treatment: albendazole (400 mg daily for 3 days) or mebendazole (100 mg bid for 3 days).

TRICHINOSIS

- *Trichinella spiralis* is found throughout the United States, but predominantly in the Northeast and Mid-Atlantic states.
- Less than 100 cases of clinical disease are reported annually but cysts are found at autopsy in the diaphragms of 4% of patients.
- Digestive enzymes liberate the encysted larvae, which lodge in the duodenum and jejunum, grow and, within 2 days, mature and copulate.
- The females give birth to living larvae that bore through the mucosa, become blood-borne and migrate to striated muscle, heart, lung, and brain.
- Symptoms relate to the inflammation caused by the larvae.
- A classic triad of fever, myalgia, and periorbital edema has been described, but gastroenteritis, pneumonia, myocarditis, meningitis and seizures can occur (Table 66-1).

TABLE 66-1 Symptoms of Parasitic Diseases

SYMPTOM	POSSIBLE CAUSE
Abdominal pain	<i>Ascaris</i> , <i>Clonorchis</i> , <i>Diphyllobothrium</i> , <i>Entamoeba</i> , <i>Fasciola</i> , <i>Giardia</i> , hookworm, <i>Hymenolepsis</i> , <i>Schistosoma</i> , <i>Taenia</i> , <i>Trichuris</i>
Anemia	<i>Babesia</i> , <i>Diphyllobothrium</i> , hookworm, <i>Leishmania donovani</i> , <i>Plasmodium</i> species, <i>Trichuris</i>
Asthma	<i>Ascaris</i> , <i>Strongyloides</i> , <i>Toxocara</i>
Conjunctivitis and keratitis	<i>Filariae</i> (<i>Onchocerca volvulus</i>), <i>Taenia</i> , <i>Trichinella</i> , <i>Trypanosoma</i>
Diarrhea	<i>Dientamoeba</i> , <i>Entamoeba</i> , <i>Fasciola</i> , <i>Fasciolopsis</i> , <i>Giardia</i> , hookworm, <i>Hymenolepsis</i> , <i>L. donovani</i> , <i>Palantidium</i> , <i>Schistosoma</i> , <i>Strongyloides</i> , <i>Taenia</i> , <i>Trichinella</i> , <i>Trichuris</i>
Edema	<i>Fasciolopsis</i> , <i>Filariae</i> (<i>Wuchereria bancrofti</i>), <i>Trichinella</i> , <i>Trypanosoma</i>
Eosinophilia	<i>Ascaris</i> , <i>Dracunculus</i> , <i>Fasciola</i> , <i>Filariae</i> (<i>W. bancrofti</i> , <i>Brugia malayi</i>) fluke (<i>Paragonimus westermani</i> , <i>Chlonorchis sinensis</i> , <i>Fasciolopsisbuski</i>), <i>Hymnenolepsis</i> , hookworm, <i>Schistosoma</i> , <i>Strongyloides</i> , <i>Taenia</i> , <i>Toxocara</i> , <i>Trichinella</i> , <i>Trichuris</i>
Fever	<i>Ascaris</i> , <i>Babesia</i> , <i>Entamoeba</i> , <i>Fasciola</i> , <i>Filariae</i> (<i>W. bancrofti</i>), fluke (<i>C. sinensis</i>), <i>Giardia</i> , <i>L. donovani</i> , <i>Plasmodium</i> species, <i>Toxocara</i> , <i>T. trichiura</i> , <i>Trypanosoma</i>
Hemoptysis	<i>Ascaris</i> , <i>Echinococcus</i> , <i>Paragonimus</i>
Hepatomegaly	Fluke (<i>C. sinensis</i> , <i>Opisthorchis viverrini</i> , <i>Fasciola</i>), <i>L. donovani</i> , <i>Plasmodium</i> species, tapeworm (<i>Echinococcus</i>), <i>Schistosoma</i> , <i>Toxocara</i> , <i>Trypanosoma</i>
Intestinal obstruction	<i>Ascaris</i> , <i>Diphyllobothrium</i> , fluke (<i>Fasciolopsis buski</i>), <i>Strongyloides</i> , <i>Taenia</i>
Jaundice	Fluke (<i>C. sinensis</i> , <i>O. viverrini</i>), <i>Plasmodium</i> species
Meningitis	<i>Acanthamoeba</i> , malaria (<i>Plasmodium falciparum</i>), <i>Naegleria</i> , primary amebic meningoencephalitis, <i>Toxocara</i> , <i>Trichinella</i> , <i>Trypanosoma</i>
Myocardial disease	<i>Taenia</i> , <i>Trichinella</i> , <i>Trypanosoma</i> (<i>T. cruzi</i>)
Nausea and vomiting	<i>Ascaris</i> , <i>Entamoeba</i> , <i>Giardia</i> , <i>Leishmania</i> , <i>Taenia</i> , <i>Trichinella</i> , <i>Trichuris</i>
Pneumonia	<i>Ascaris</i> , <i>Filariae</i> (<i>W. bancrofti</i> , <i>B. malayi</i>), fluke (<i>P. westermani</i>), <i>Strongyloides</i> , <i>Trichinella</i>
Pruritus	<i>Dientamoeba</i> , <i>Enterobius</i> , <i>Filariae</i> (<i>O. volvulus</i>), <i>Trichuris</i>
Seizures	<i>Hymenolepsis</i> , <i>Trichinella</i> , <i>Paragonimus</i> , tapeworm (<i>Echinococcus</i> , <i>Cysticercus</i>)
Skin ulcers	<i>Dracunculus</i> , hookworm, <i>L. donovani</i> , <i>Trypanosoma</i>
Splenomegaly	<i>Babesia</i> , <i>Toxoplasma</i> , <i>Plasmodium</i>
Urticaria	<i>Ascaris</i> , <i>Dracunculus</i> , <i>Fasciola</i> , <i>Strongyloides</i> , <i>Trichinella</i>

- Tests: elevation of muscle enzymes and eosinophilia are suggestive. Muscle biopsy is confirmative. Serologic tests are available from the Centers for Disease Control and Prevention.
- Treatment: aspirin and steroids initially to reduce the inflammatory symptoms. Mebendazole (200–400 mg tid for 3 days and then 400 mg tid for 10 days) or albendazole (400 mg bid for 8–14 days) is indicated for severe disease but may not be effective after encystment.

HOOKWORMS

- The hookworms, *Necator americanus* and *Ancylostoma duodenale*, are found between 36-degrees north and 30-degrees south latitude.
- One billion individuals affected.
- The eggs hatch in the soil, releasing rhabditiform larvae that feed on organic debris.
- They survive as filariform larvae for several weeks and can migrate up to 4 feet from the site where the first host defecates.

- Upon contact, they burrow through the skin, causing pruritus (ground itch), enter the blood, travel to the lung, and are ingested, like *A lumbricoides*.
- The hallmark of hookworm infestation is the microcytic, hypochromic, iron deficiency anemia (Table 66-1). Children develop a characteristic yellow–green pallor called chlorosis.
- Along with the dog and cat hookworms (*Ancylostoma braziliense*), they cause the serpentine track of cutaneous larva migrans.
- Test: stool for ova and parasites.
- Treatment: Albendazole (400 mg qid for 2–3 days), mebendazole (100 mg bid for 3 days), or pyrantel pamoate (11 mg/kg–maximum 1 g qid for 3 days) is recommended.

TREMATODES (FLUKES)

- Flukes are oval, flat worms with a ventral sucker for nutrition and attachment.

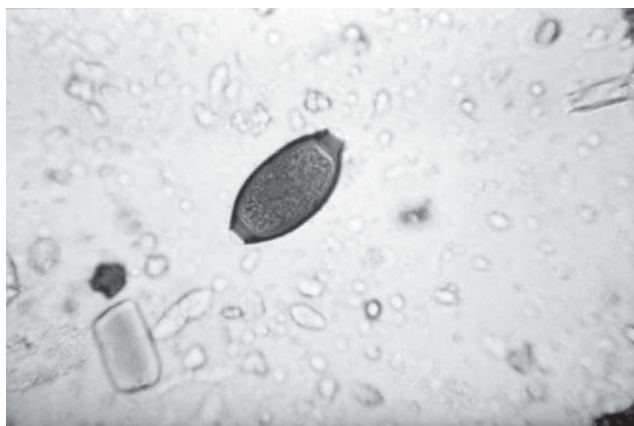


A

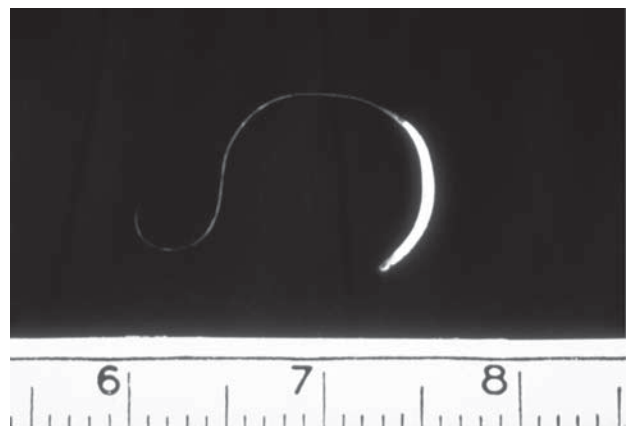


B

FIG. 66-1. (A) Pinworms. Multiple tiny pearly white worms are seen at the anus. (B) This photomicrograph depicts the eggs of the nematode, or round worm, *Enterobius vermicularis*, mounted on cellulose tape. (Courtesy of the Centers for Disease Control Public Health Image Library.)



A



B

FIG. 66-2. (A) This micrograph depicts an egg from the “human whipworm,” *Trichuris trichiura*, the causal agent of “Trichuriasis.” (B) This micrograph of an adult *Trichuris* female human whipworm, reveals that its size in centimeters is approximately 4 cm (A and B.) (Courtesy of the Centers for Disease Control Public Health Image Library.)

- Eggs are shed in the stool, hatch into miracidia and enter an intermediate host such as a snail, fish or bird.
- They develop into cercariae, leave the intermediate host, and become free living prior to infesting the definitive host on contact with contaminated water.
- There are no flukes producing human disease that are endemic in the contiguous United States. However, the avian schistosome, *Trichobilharzia ocellata* is spread by migratory birds to freshwater lakes in the northern United States.
- The cercariae cause dermatitis, known as swimmer's itch (Table 66-1).
- It is treated with heat and antipruritics or, in severe cases, thiabendazole cream.

CESTODES (FLATWORMS AND TAPEWORMS)

- Cestodes attach to the gut with hooks or suckers at the head (scolex), from which grow segmented proglottids.
 - Proglottids produce large volumes of eggs that, along with terminal proglottids, pass in the stool and are ingested by the intermediate host.
 - The eggs hatch into a larval stage, either cysticercus (larva with a single scolex in a fluid-filled cyst), cysticercoid (larva with the single scolex filling the cyst), coenurus (a cyst with grapelike daughter cysts internally containing protoscolices), or hydatid cysts (mature cyst filled with numerous immature cysts), depending on the species.
 - Symptoms occur when larvae act as space-occupying lesions or cause inflammation.
- Four cestodes produce most clinical disease seen in the United States.
 - *Taenia solium* (pork tapeworm) and *Taenia saginata* (beef tapeworm) infestations are generally asymptomatic and are diagnosed when a parent brings a proglottid for identification.
 - When *T solium* enters the cysticercus phase, it may migrate to the heart, brain, breast, eye, skin, or other solid organs.
 - Subcutaneous nodules, visual field defects, focal neurologic findings, acute psychosis, and obstructive hydrocephalus may develop years after infestation (Table 66-1).
 - Identified as calcified cysts on plain radiographs or a ring of calcification on computed tomography.
 - *Diphyllobothrium latum* (fish tapeworm) absorbs >50 times more vitamin B₁₂ than *Taenia* and causes pernicious anemia.

- *Echinococcus granulosus* (sheep tapeworm) is found in the southeastern United States. Symptomatology is secondary to hydatid cyst formation with mass effect.
- Treatment: most tapeworms respond to praziquantel (5–25 mg/kg once).
- *Echinococcus* infestation and cysticercosis respond best to albendazole (15 mg/kg/d divided tid for 28 days).

PROTOZOA

ENTAMEOBA HISTOLYTICA

- *Entamoeba histolytica* is a water-borne single-cell organism.
- It is spread by ingestion of contaminated water, sexual contact and in breast milk.
- Amebas live asymptotically in the cecum and large intestine at the base of large flask-shaped ulcers.
- Heavy infestations produce a colitis like picture (“gay bowel”) and pseudopolyps of normal tissue on a base of ulcerative disease (Table 66-1).
- *E histolytica* can invade the blood, causing abscess formation in the liver, lung, brain, and breast.
- Treatment: metronidazole (35–50 mg/kg/d divided tid for 10 days) followed by iodoquinol (40 mg/kg/d divided tid for 20 days).

GIARDIA LAMBLIA

- Giardia lives in the alkaline pH of the duodenum and proximal small bowel.
- Infestation occurs with ingestion of contaminated water or fecal–oral behavior.
- It is found in day care centers, immunocompromised children, and patients with cystic fibrosis.
- It causes flatulence, nonbloody diarrhea or constipation, abdominal distention, pain, fever, weight loss and fat, carbohydrate and vitamin malabsorption (Table 66-1).
- Diagnosis is made with duodenal aspiration.
- Treatment: metronidazole (15 mg/kg/d divided tid for 5 days).

PNEUMOCYSTIS

- *Pneumocystis jiroveci* (formerly *carinii*) has low virulence and is latent in a large percentage of the American population.
- Over 60% of patients with HIV have Pneumocystis.

- When the host is immunocompromised, trophozoites replicate in alveolar spaces and spread through the vascular and lymphatic beds.
- Symptoms are respiratory distress, fever, and non-productive cough with limited auscultatory findings (Table 66-1).
- X-rays are normal or have symmetric interstitial ground glass infiltrates in the middle and lower lung fields.
- The mortality rate in children is 40%, rising to 100% once radiographic changes occur in untreated non-AIDS patients.
- Diagnosis is confirmed by silver nitrate methenamine stain of a lung biopsy specimen.
- Treatment: trimethoprim (15–20 mg/kg/d) and sulfamethoxazole (75–100 mg/kg/d) in 3 or 4 divided doses orally or pentamidine (3–4 mg/kg/d) intravenously for 2 to 3 weeks.

ARTHROPODS

PEDICULOSIS

- The parasites *Pediculus humanus capitis* (head louse), *Pediculus humanus corporis* (body louse), and *Phthirus pubis* (pubic or crab louse) attach eggs (nits) firmly to hair shafts, close to the skin.
- Lice are transmitted by direct human contact or the sharing of clothing.
- They require a hair-bearing surface.
- The adult is viable for only 2 days and the nit for 10 days off the host.
- *P pubis* may produce blue-colored macules (maculae ceruleae).
- Eyelid lice in prepubescent children indicates possible sexual abuse.
- Treatment: near the eyes petrolatum is recommended to suffocate the lice. Dead nits can be removed from hair shafts with a fine-toothed comb (nit picking). The combination of 1% permethrin with trimethoprim/sulfamethoxazole has been reported to be 95% effective, compared with 80% for permethrin alone.

SCABIES

- *Sarcoptes scabiei* (scabies) is transmitted by direct, close, and prolonged human contact.
- After a 3- to 6-week incubation, the mite burrows between the fingers and toes, in the groin, external genitalia and axillae, depositing eggs in the tunnel as she goes.
- Small red, raised papules are formed that progress to vesicles and pustules.

- The itch is worse at night.
- Test: the diagnosis is done clinically or by scraping burrows or papules overlaid with mineral oil and looking for adults, eggs, and excreta.
- Treatment: 5% permethrin cream is curative for children older than 2 months.

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QUESTIONS

1. A family goes on a camping vacation through the southeastern United States. They sleep in tents, forage for some of their food and water and cook over a wooden campfire. While their travel, they have waded through slow moving streams, walked around several lakes, traveled through low farmland and over wooded hills. At the end of the first week, several of the group develop bloating, abdominal cramps, and diarrhea. These symptoms could have been avoided by doing which of the following options?
 - A. Wearing high laced boots
 - B. Everyone using a designated latrine spot
 - C. Not sharing eating utensils
 - D. Boiling all water before consumption
 - E. Sleeping under the stars rather than in the tents
2. A 7-year-old boy is on a camping trip to northern Minnesota when he reports that his legs itch above the ankles. He reports no other symptoms. He most likely has which of the following?
 - A. Chafing from dirt getting into his shoes

- B. A fungal infection from not changing wet stockings
 - C. MRSA from self-innoculation
 - D. Scabies acquired from a sibling
 - E. *T. ocellata* from walking in the water.
3. While helping her 11-year-old daughter wash her hair, a mother notes lice on her eyebrows and lashes. What is the most appropriate and effective treatment?
- A. Treat the affected area and the whole body with over-the-counter permethrin.
 - B. Get the Vaseline from her emergency kit and apply to the affected area.
 - C. Shave the eyelashes and eyebrows.
 - D. Remove nits with a fine-toothed comb and wash with baby shampoo.
 - E. Apply hydrocortisone cream to the affected area.
4. Parents note that their 2-year-old has developed anal itching. They increased diaper changes and frequent washing without a change in behavior. The anal area appeared normal to them. The toddler was noted to attempt to stool more frequently, but without success. Reinspection revealed a very small amount of mucosa that slid back into the anus spontaneously. There were no other findings. This child most likely has acquired which of the following?
- A. The tapeworm, *Taenia solium*.
 - B. The hookworm, *Necator americanus*.
 - C. The amoeba, *Entamoeba histolytica*.
 - D. The whipworm, *Trichuris trichiura*.
 - E. The pinworm, *Enterobius vermicularis*.
5. A 15-year-old boy develops an acutely painful abdomen. He has vomited twice, had one nonbloody loose stool and denies dysuria. The family seeks medical attention, at which time the pain is beginning to localize in the right lower quadrant. The doctor suspects appendicitis but should include which of the following parasites in his differential diagnosis?
- A. The amoeba, *Entamoeba histolytica*.
 - B. The pinworm, *Enterobius vermicularis*.
 - C. The hookworm, *Necator americanus*.
 - D. The roundworm, *Ascaris lumbricoides*.
 - E. The protozoa, *Giardia lamblia*.
6. You are working with a relief agency caring for people displaced by years of civil war, drought and famine. You are presented with a severely malnourished 6-month-old with a cough and fever. The mother reports that, except for the poor nutritional status, she and her child had both been previously healthy. In addition to fluid resuscitation and beginning to feed this child, you suspect pneumonia and wish to begin to treatment. What is the most appropriate empiric antibiotic choice in this clinical situation?

- B. IV or IM ceftriaxone
- C. IV vancomycin
- D. Oral chloramphenicol
- E. Oral trimethoprim/sulfamethoxazole

ANSWERS

1. D. Amoebae, Giardia, and other parasites are frequently water borne and could cause these symptoms within 1 week of infestation. The party is traveling in an area where hookworm is endemic; however, abdominal symptoms with hookworm do not occur until the infestation is heavy and the timing of the complaint does not fit a heavy worm burden. High-laced boots might prevent hookworms from burrowing through the skin, but would not help prevent the spread of waterborne agents. Using a designated latrine area is also helpful in preventing the spread of diseases such as hookworm, but is unlikely to help prevent waterborne infections. While the use of individual eating utensils may help in preventing person-to-person spread of disease, there is no evidence that sleeping under the stars will improve health.
2. E. The cercariae of the avian schistosome, *T. ocellata*, causes swimmer's itch. The area affected on the body is correct and the migratory birds that transmit this are native to the northern United States. Chafing is possible but unlikely to present as itching. Scabies is also possible but the location on the body is atypical. Fungal infections and MRSA require a point of entry and a longer time to establish themselves.
3. B. Vaseline is the appropriate treatment for arthropods on the face. Permethrin should not be used in proximity to the eyes. Shaving of hair will eliminate the nits, but is not recommended for eyelashes and eyebrows. Removal of nits alone is not sufficient treatment and baby shampoo would be ineffective, as is hydrocortisone.
4. D. The whipworm and pinworm both produce anal itch and are easily acquired by the oral exploratory behavior of toddlers. The eggs can also be inhaled and swallowed in dust. While heavy infestations of both produce cramping, rectal prolapse is more common with whipworm, *T. trichiura*, which is endemic to warm rural areas of the southern United States. There is no history of pork consumption, which would suggest trichinosis, or walking barefoot through the soil, which would suggest hookworm. *E. histolytica* can be acquired from groundwater, but does not present as anal itch.
5. D. Abdominal pain from large worm burdens or appendiceal obstruction and rupture is most common

with *Ascaris lumbricoides*. *Ascaris* can grow up to 30 cm long and commonly occurs simultaneously with other parasitic infestations. It is necessary, after the surgical intervention, to treat the *Ascaris* before other parasites. *Giardia* presents with nonbloody, loose stool and flatus and, as with the hookworm, has a component of malabsorption. *E. histolytica* more commonly presents with bloody stool from necrotic bowel. Abdominal pain and symptoms other than pruritis ani are uncommon with pinworm.

6. E. While *Pneumocystis jiroveci* has low virulence in the healthy population, it occurs frequently in hosts that have low resistance, either by being immunocompromised by disease or malnutrition. The mortality rate depending on the timing of treatment in this infant with *Pneumocystis* is between 40% and 100%. Therefore, any treatment regimen should include trimethoprim/sulfamethoxazole. While amoxicillin is a good choice for many simple pediatric infections, it frequently produces diarrhea that could compromise further this child's tenuous nutritional/fluid status. Ceftriaxone covers most major pediatric bacterial illnesses, except for certain strains of resistant pneumococci and staphylococci, which respond better to vancomycin or chloramphenicol, and could be a reasonable choice for this child's presumed infectious process, but it does not provide coverage for *Pneumocystis*.

67 IMPORTED DISEASES/DISEASES IN THE TRAVELING CHILD

Thomas L. Hurt

INTRODUCTION

- The evaluation and treatment of the ill-child traveler is aided by
 - an awareness of possible pathogens,
 - knowledge of the geographic distribution of specific pathogens,
 - knowledge of incubation periods of specific pathogens,
 - knowledge of the patient's immunization and chemoprophylaxis status,
 - recognition of signs and symptoms of specific illnesses,
 - application of a systematic approach to diagnosis and treatment,
 - awareness of sources of information about travel-related diseases.

POSSIBLE PATHOGENS

- Table 67-1 provides a partial list of imported diseases.

TRAVEL LOCATIONS

- Disease exposure differs according to the region of the world. Knowledge of the distribution of various diseases can help include or exclude the likelihood of a specific disease in a returning traveler.
- Table 67-2 provides a list of infections found in specific locations.

INCUBATION PERIODS

- Knowledge of incubation periods can help exclude some diseases entirely.
- Table 67-3 lists the incubation periods for a variety of illnesses.

IMMUNIZATIONS AND CHEMOPROPHYLAXIS

- Information about the patient's immunization and chemoprophylaxis status may help narrow the possible causes of a febrile illness.
- The most important immunizations a child should receive prior to travel are the routine immunizations.
- Arriving adoptees from developing countries comprise a patient population that is unique since they often are only partially immunized or have received no immunizations at all.
- Immunization records for these patients may not be accurate.
- Table 67-4 lists some of the recommended travel vaccines.

SOURCES OF INFORMATION FOR INTERNATIONAL TRAVEL-RELATED DISEASES

- A variety of sources can provide region-specific and disease-specific information.
- A partial list of Web sites providing information on international travel-related infections is provided in Table 67-5.

PHYSICAL SIGNS AND SYMPTOMS

- The physical examination can provide clues to the diagnosis.

TABLE 67-1 Imported Diseases

DISEASE	ORGANISM VECTOR	CLINICAL MANIFESTATION	TREATMENT
Plasmodium protozoa	Malaria (mosquito)	Fever, hepatosplenomegaly	Antimalarials: quinine, quinidine, doxycycline, clindamycin
Spotted fevers	Rickettsiae (tick)	Fever, headache, malaise, rash	Doxycycline
Yellow fever	Group B arbovirus (mosquito)	Fever, headache, vomiting, jaundice	Supportive
Dengue shock syndrome	Arbovirus (mosquito)	Fever, severe body aches, purpura	Supportive
Enteric fever	<i>S. typhi</i> or <i>paratyphi</i> (fecal)	Fever, abdominal pain, rash	Ceftriaxone or fluoroquinolones
Hemorrhagic fevers (Ebola, Marburg)	Filoviruses, ebola, and Marburg (direct contact with infected host)	Fever, DIC, shock	Supportive
Leptospirosis	Leptospira interrogans (contaminated freshwater)	Fever, icterohemorrhagic “Weil’s disease”	Cephalosporins, penicillins, doxycycline
Rabies	Rabies virus (animal bite)	Fever, pain, paresthesias, encephalitis, death	Postexposure vaccine, supportive care
Leishmaniasis	Parasite from sand flea bite	Cutaneous sores, abdominal visceral swelling and pain	Antimonials
Strongyloides	Helminthic parasite – cutaneous entry	Rapidly spreading serpiginous rash—larva currens	Ivermectin, thiabendazole
Schistosomiasis	Intestinal and urinary freshwater flatworms	Fever, back pain, paresthesias	Praziquantel
Cysticercosis	Pork tapeworm <i>Taenia solium</i>	Seizures	Praziquantel
Amebiasis	<i>E. histolytica</i>	Abdominal pain, bloody diarrhea	Mebendazole, iodoquinol
Roundworm (ascariasis)	<i>Ascaris lumbricoides</i>	Abdominal pain, Loeffler syndrome	Mebendazole, pyrantel pamoate

TABLE 67-2 Infections and Geographic Locations^a

Caribbean: malaria (Haiti), dengue fever, leishmaniasis
South and Central America: malaria, dengue fever, hepatitis A, cholera, leishmaniasis, yellow fever, amebiasis, viral encephalitis, schistosomiasis
Pacific Islands: malaria, dengue fever, hepatitis A, Japanese encephalitis
Southeast Asia, Far East, South Asia (India, Pakistan, Bangladesh, Nepal, and Sri Lanka): malaria, dengue fever, hepatitis A, Japanese encephalitis, cholera, rickettsial diseases, leishmaniasis
Middle East and North Africa: malaria, hepatitis A, leishmaniasis, tick typhus, schistosomiasis, brucellosis
Subsaharan Africa: malaria, hepatitis A, cholera, yellow fever, amebiasis, typhus, schistosomiasis
Europe and former Soviet Union: hepatitis, tick typhus, Lyme disease, leishmaniasis, diphtheria, polio, brucellosis, tick-borne encephalitis
Australia/New Zealand/North America: rocky mountain spotted fever, Lyme disease, viral encephalitis

^aDysentery, typhoid and tuberculosis are worldwide in distribution.

TABLE 67-3 Incubation Periods for Travel-Related Infectious Diseases

INCUBATION PERIOD	INFECTIOUS DISEASE
Short (<2 wk)	Malaria (especially <i>P. falciparum</i>)
	Viral hemorrhagic fevers—including dengue fever
	Rickettsioses: louse, flea-borne, typhus
	Enteric fever
	Influenza
Mid (2–6 wk)	Plague
	Anthrax
	Rabies
	Malaria (especially <i>P. falciparum</i>)
	Rickettsioses: spotted fever group
Long (>6 wk)	Enteric fever
	Leptospirosis
	Brucellosis
	Viral hepatitis
	Strongyloides
Long (>6 wk)	Rabies
	Malaria
	Tuberculosis
	Viral hepatitis
	Enteric protozoal infections and helminthes infections
	Schistosomiasis
	Amebic liver abscess
Leishmaniasis	
Rabies	

TABLE 67-4 Recommended Travel Vaccines

VACCINES	COMMENTS
Hepatitis A	Efficacy 94%–100%
Rabies	Antibodies exist for at least 2 y after the primary series.
Preexposure vaccines	
Human diploid cell vaccine (HDCV)	
Rabies vaccine adsorbed (RVA)	
Purified chick embryo cell vaccine (PCEC)	
Postexposure treatment	
Passive and active immunization	
Meningococcus	Antibody concentrations decrease rapidly over 2–3 y.
Vaccine available for children >2 y	
Typhoid	Approximately 70%–80% effective
Oral vaccine and parenteral vaccine are available in the United States	
Yellow fever	Provides at least 10 y protection
Available at approved state health department centers	

TABLE 67-5 Internet References for Travel-Related Illness

1. CDC “Health Information for International Traveler” (Yellow Book)	
General travelers’ health information, region- and destination-specific recommendations, including vaccinations and malaria prophylaxis	http://www.cdc.gov/travel/yellowbook.pdf
2. Centers for Disease Control and Prevention: Travelers’ Health Information	
Recommendations for specific travel destinations, links to other sites, travel advisories, and outbreak notices	http://www.cdc.gov/travel/
3. World Health Organization (WHO) Yellow Book, “International Travel and Health”	
General travelers’ health recommendations; with country-specific malarial risks. Very useful for current outbreaks.	http://www.who.int/ith/
4. Morbidity and Mortality Weekly Report, International bulletins	
Surveillance summaries & outbreak investigations.	http://www.cdc.gov/mmwr
5. American Society of Tropical Medicine and Hygiene (ASTMH)	
Current information on tropical disease prevention and control and worldwide outbreaks.	http://www.astmh.org
6. World Health Organization (WHO) Infectious Disease Health Topics	
Maps of travel-related diseases & general information.	http://www.who.int/health-topics/

- Table 67-6 provides a limited differential diagnosis for some travel-related diseases based on physical findings.

SIGNIFICANCE OF EOSINOPHILIA

- Eosinophilia (definition): peripheral eosinophil count > 400–500 cells/mm³.
- Eosinophilia is seen especially during the early acute tissue invasive phase of infestation as compared to the later chronic phase of illness.
- Diagnoses to be considered in febrile travelers with eosinophilia: ascariasis, strongyloides, acute schistosomiasis, toxocariasis, trichinosis, lymphatic filariasis, coccidiomycosis, echinococcus, tapeworm (cestode infestation), and lung and liver flukes.

- The more common intestinal protozoa, *G lamblia* and *Entamoeba histolytica*, do not cause eosinophilia.
- The positive predictive value of eosinophilia for helminth infection is only 18.9%.
- If eosinophilia is extreme ($\geq 16\%$ of the total WBC count), the probability of finding a definite diagnosis is > 60% and the positive predictive value for a helminth infection is 46.6%.

DIAGNOSIS AND TREATMENT OF RETURNING CHILD TRAVELER

- Recommended basic laboratory tests include malaria smears (if travel to areas endemic to malaria), CBC with differential, chemistry panel that includes liver

TABLE 67-6 Physical Findings for Some Important Diseases

PHYSICAL FINDING	JAUNDICE	PETECHIAE/ ECCHYMOSIS	HEPATOMEGALY	SPLENOMEGALY	WHEEZING	NEUROLOGIC COMPLAINTS
Disease	Hepatitis	Meningococcus	Malaria	Malaria	Loeffler syndrome	Rabies
	Malaria	Dengue fever	Hepatitis	Typhoid	Tropical pulmonary eosinophilia	Neurocysticercosis
	Relapsing fevers	Yellow fever	Typhoid	Dengue fever	Tuberculosis	Neuroschistosomiasis
	Leptospirosis	Rickettsial diseases	Leishmaniasis	Leishmaniasis		
	Dengue fever	Viral hemorrhagic fevers	Leptospirosis	Brucellosis		
	Liver abscess		Amebic liver abscess	Typhus		

function tests, urinalysis, urine culture, and stool studies.

- It is very practical to obtain a tube of serum that can be stored for future paired (acute and convalescent) serologic tests.
- See Fig. 67-1 for a systematic approach to evaluating the returning child traveler.

MALARIA

- Endemic to the tropics.
- 2.7 million deaths per year.
- Transmitted by night-biting *Anopheles* mosquito.
- Protozoal infection caused by four species of genus *Plasmodium* (*vivax*, *ovale*, *malariae*, and *falciparum*).
- 90% of global cases caused by *falciparum*.
- Most common symptoms: nonspecific flu-like syndrome with high fever, chills, rigors, sweats, and headache. Frequently misdiagnosed as viral syndrome.
- Can present with gastrointestinal symptoms and can be misdiagnosed as acute gastroenteritis.
- Most common complaint in pediatric patients is daily fever but only 1/3 have paroxysmal fevers.
- Hepatosplenomegaly in up to 2/3 of patients.
- Common laboratory studies show anemia, thrombocytopenia, moderate LFT elevation, and hypoglycemia.
- Thick and thin Giemsa stains of peripheral blood smears remain the accepted diagnostic technique for malaria.
- Thick smears give higher sensitivity of detecting malaria than thin smears by factor of 10:1.
- Thin smears primarily used for speciation and determination of degree of parasitemia.
- Treatment options depend on severity of illness, species, and whether or not resistant to chloroquine.

- Most practical approach in the emergency department is to assume that all malaria is resistant to chloroquine.
- In a critically ill child with malaria, *falciparum* should be presumed until proven otherwise and parenteral treatment with quinine or quinidine should be started.
- Quinidine, quinine, and malaria itself can cause hypoglycemia—frequent serum glucose checks should be made.
- If neither quinine nor quinidine is available, clindamycin can be used as a temporizing medication.
- Table 67-7 is a guide to treatment of malaria.

DENGUE FEVER

- The most significant arboviral infection of humans.
- Most commonly transmitted by the day-biting female *Aedes aegypti* and *Aedes albopictus* mosquitoes.
- Dengue virus causes a range of illness from benign minor fever to hemorrhagic condition (dengue hemorrhagic fever) that may lead to circulatory collapse (dengue shock syndrome) and death.
- Most common signs and symptoms in infants and children include fever, vomiting, and hepatomegaly.
- Most common signs and symptoms in older children and adolescents include rash, fever, headache, retro-orbital pain, myalgias, and arthralgias.
- Arthralgias and myalgias can be so severe that dengue fever has been nicknamed “breakbone fever.”
- Laboratory findings of thrombocytopenia (platelets <100,000) and hemoconcentration (hematocrit > 20% above baseline) indicate worsening disease.
- Treatment is symptomatic and supportive.
- Definitive diagnosis cannot be made in emergency department but acute and convalescent serologies can help identify dengue viral infection.
- Currently no vaccine for dengue fever.

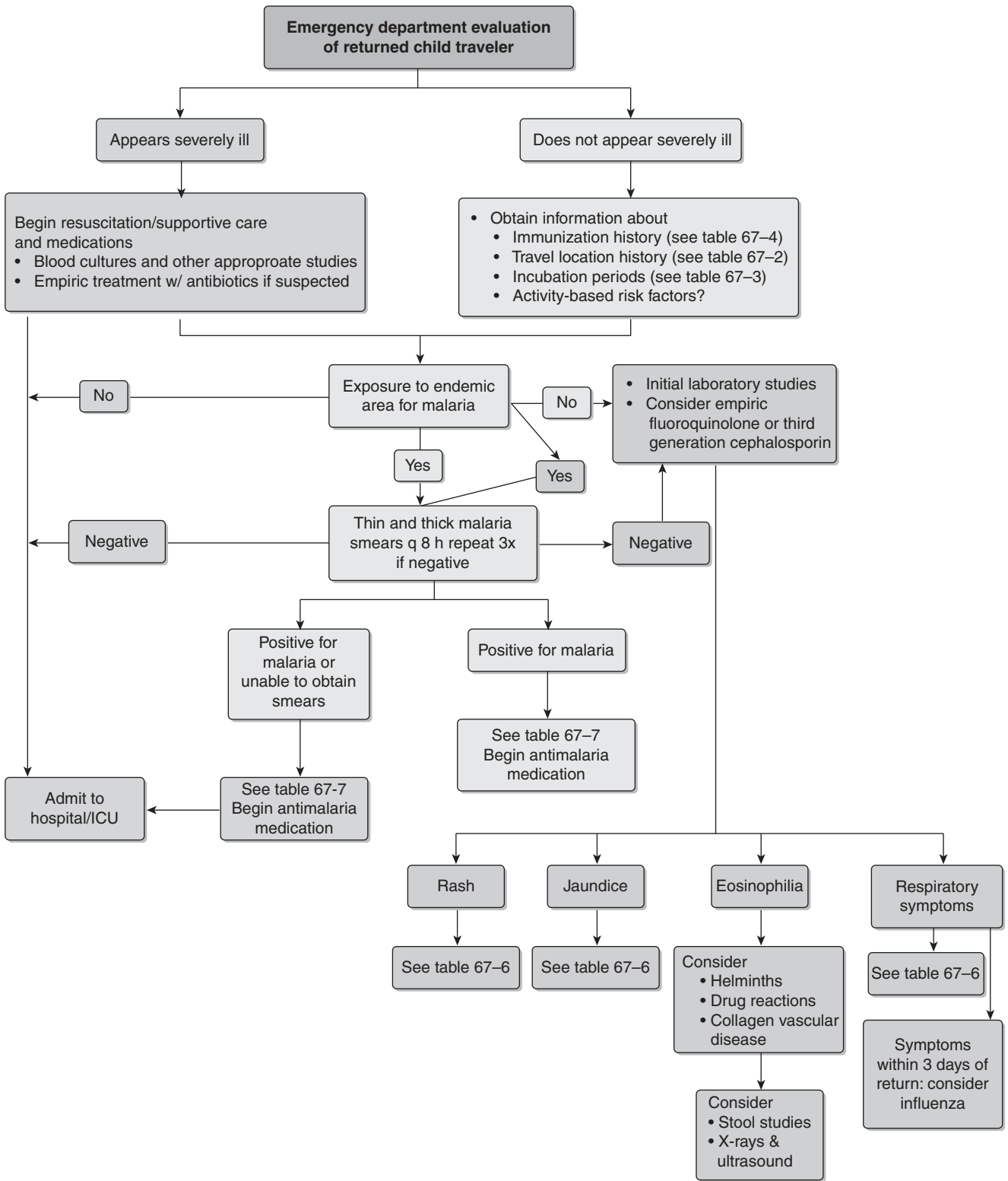


FIG. 67-1. General guide to the evaluation and management of the returning child traveler.

TABLE 67-7 Treatment of Malaria

MEDICATION	DOSE	
Patients with malaria who are seriously ill or cannot take oral medications (all species of plasmodium)		
IV quinidine gluconate	10 mg/kg IV load in NS over 1–2 h, then infuse 0.02 mg/kg/min with cardiac monitoring	
Patients with uncomplicated chloroquine-resistant <i>P. falciparum</i> or <i>Plasmodium vivax</i> (90% of patients)		
Oral quinine sulfate	25 mg/kg/d (max. 650 mg/dose) ÷ tid × 3–7 d	
Plus		
Doxycycline	2 mg/kg/d (max. 100 mg) × 7 d	
Or		
Pyrimethamine sulfadoxine	<1 y, 1/4 tablet 1–3 y, 1/2 tablet 4–8 y, 1 tablet 9–14 y, 2 tablets >14 y, 3 tablets	Note 1. Single oral dose 2. Sulfa allergy risk
Or		
Clindamycin (if <i>P. falciparum</i>)	20–40 mg/kg/d ÷ tid × 5 d (max 900 mg/dose)	
Suspected or proven malaria from areas without <i>P. falciparum</i> or <i>P. vivax</i>		
Chloroquine phosphate	10 mg/kg of base (max. 600 mg), then 5 mg/kg of base at 6, 24, and 48 h	
Plus		
Doxycycline	3 mg/kg (max. 200 mg) once daily × 3 d	
Or		
Pyrimethamine sulfadoxine	<1 y, 1/4 tablet 1–3 y, 1/2 tablet 4–8 y, 1 tablet 9–14 y, 2 tablets >14 y, 3 tablets	Note 1. Single oral dose 2. Sulfa allergy risk

TABLE 67-8 Antibiotic Therapy for Travellers Diarrhea and Enteric Fever

ANTIBIOTIC	DOSE	ADVANTAGES	EFFECTIVENESS IN CHILDREN
Ciprofloxacin	30 mg/kg/D IV or po ÷ bid	Oral or parenteral	Use is controversial in children younger than 12 y
Ceftriaxone	50–75 mg/kg/d maximum 2.5 g/d	Parenteral Once a day dosing	Highly effective
Cefixime	10–15 mg/kg/d ÷ bid	Oral dosing	Highly effective
Azithromycin	12 mg/kg/d on day 1 (max. 1g) followed by 6 mg/kg/d	Oral dosing	Highly effective
Trimethoprim/sulfamethoxazole (TMP/SMX)	8 mg/kg/dose (TMP) ÷ bid	Oral dosing	Significant resistance worldwide

TRAVELERS' DIARRHEA (TD)—FOOD AND WATERBORNE INFECTIONS

- Infectious causes usually occur due to ingestion of fecally contaminated food or water.
- The most common organisms found in children with TD include: *Rotavirus*, *Cryptosporidium*, *Salmonella*, *Campylobacter*, *Shigella*, enterotoxigenic, enteropathogenic, and enterohemorrhagic *E. coli*.
- The most common signs of TD are diarrhea, abdominal cramping (67%) nausea and vomiting (18%), fever (14%), and mucous and blood in stool (5%).
- The most important intervention in TD is fluid and electrolyte replacement.

- In the nontraveler child in a developed country, antibiotics have no significant role in empiric treatment of acute diarrhea.
- Oral rehydration in conjunction with antibiotic treatment decreases pediatric diarrhea duration in developing countries.
- The traditional empiric antibiotic treatment of choice for TD in children has been 3–5 days of trimethoprim-sulfamethoxazole (TMP-SMX), but resistance patterns are developing.
- Because of emerging TMP-SMX resistance, macrolides, quinolones, cefixime, and ceftriaxone are now also recommended.

- There is theoretical concern that antimotility agents may prolong the course of disease caused by invasive enteropathogens. The American Academy of Pediatrics still recommends against the use of loperamide in the management of acute diarrhea in children, whether TD or not.
- See Table 67-8 for empiric antibiotic treatment recommendations for TD.

TYPHOID AND PARATYPHOID ENTERIC FEVERS

- Typhoid fever (TF) is caused by *Salmonella enterica*, serotype *typhi*.
- *Salmonella paratyphi* causes an identical clinical syndrome.
- The term enteric fever is sometimes used in place of typhoid fever to describe either syndrome.
- The risk for acquiring TF varies by geographic region visited. The CDC reports that 76% of TF cases are associated with travel to only six countries—India (30%), Pakistan (13%), Mexico (12%), Bangladesh (8%), the Philippines (8%), and Haiti (5%).
- The typical incubation period is 5–21 days, but the range is broad from 3–60 days.
- Presenting symptoms include fever, chills, frontal headache, anorexia, nausea, and malaise.
- Infants and young children with TF frequently present with different symptoms when compared to older children and adults. Constipation is a feature of TF in adults and older children while diarrhea is a more common GI symptom in younger children.
- TF is notable for prolonged fevers, often continuing for 10–14 days.
- Relative bradycardia in response to fever is common (but not specific) in TF.
- Laboratory data commonly show leukopenia, anemia, and thrombocytopenia. Liver function tests may be moderately elevated. These findings are similar to those found in both malaria and dengue fever.
- Diagnosis of TF is based on identifying *S typhi* in blood or stool cultures.
- Blood cultures are most likely positive during the acute febrile phase in the first 2 weeks.
- Stool cultures have only a 10–15% yield in the first week with higher yields in weeks 3–4 (75%).
- Multidrug resistance has developed worldwide. Third-generation cephalosporins, fluoroquinolones are now considered the best choices for empiric treatment.
- See Table 67-8 (above) for empiric antibiotic treatment recommendations for TF.

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QUESTIONS

1. A 14-year-old male develops a fever 4 weeks after returning from a 2-week trip visiting relatives in India. He is LEAST likely to have which of the following diseases?
 - A. Malaria
 - B. Typhoid fever
 - C. Dengue fever
 - D. Viral hepatitis
 - E. Leishmaniasis
2. Which of the following is true regarding immunizations a child should receive before traveling?
 - A. The most important immunizations a child should receive prior to traveling abroad are the “routine” immunizations.
 - B. The malaria vaccine is contraindicated in children less than 2 years of age.
 - C. Typhoid vaccines are nearly 100% effective and the immunity lasts nearly 10 years.
 - D. Arriving adoptees must have proof of all childhood vaccinations before they are allowed to enter the United States.
 - E. The dengue fever vaccine is effective if given at least 2 weeks before travel to an endemic area.
3. A 16-year-old girl returned 3 weeks ago from a week-long class trip to Tanzania. Which of the following statements about malaria is true?

- A. It is unlikely that she has malaria because she was vaccinated against it prior to the trip.
- B. Close to 90% of malaria cases worldwide are caused by *Plasmodium ovale*.
- C. The thin smear is more likely than the thick smear to detect parasites.
- D. Quinidine and quinine, as well as malaria itself, can cause hypoglycemia.
- E. Malaria always presents with a distinct cyclic fever pattern.
4. A family recently adopted a 7-year-old girl who came from Nigeria. They were told she had malaria in the past, but it had been treated. She is now having cycles of high fevers with shaking chills. The most important laboratory test to obtain in this patient would be which of the following?
- A. CBC
- B. Liver functions
- C. CSF studies
- D. Stool for ova and parasites
- E. Thin and thick blood smears (Giemsa stain)
5. Common findings in a patient with malaria include all of the following except:
- A. Hepatomegaly
- B. Fever
- C. Petechiae
- D. Diarrhea
- E. Jaundice
6. A traveler to which country is least likely to contract yellow fever?
- A. Tanzania
- B. Venezuela
- C. Rwanda
- D. Cambodia
- E. Haiti
7. A 4-year-old girl is brought into the emergency department with fever and bloody diarrhea. The family recently returned from a trip to South Africa. In the course of evaluating her, a CBC reveals a moderately elevated WBC with 19% eosinophils. The least likely cause of the eosinophilia is which of the following?
- A. Ascariasis
- B. Schistosomiasis
- C. *G lamblia*
- D. *Strongyloides*
- E. *Echinococcus*
8. Which of the following laboratory findings suggests your patient with dengue fever is worsening?
- A. Parasites noted on a thick smear
- B. Eosinophilia
- C. Stool is guaiac positive
- D. Hypoglycemia
- E. Thrombocytopenia and hemoconcentration
9. An 8-year-old boy is brought to the emergency room by his parents because he has severe abdominal pain and diarrhea. The family just came back from a week in Mexico. Which of the following is false regarding travelers' diarrhea in children?
- A. Common symptoms of travelers' diarrhea include diarrhea, abdominal cramping, nausea, vomiting, and fever.
- B. Infectious causes usually occur due to ingestion of fecally contaminated food and water.
- C. The American Academy of Pediatrics endorses the routine use of loperamide and other antimotility agents in children with travelers' diarrhea.
- D. Antibiotics that are currently recommended to treat travelers' diarrhea include fluoroquinolones, azithromycin, cefixime, and ceftriaxone.
- E. The most important intervention for diarrhea is replacement of fluid and electrolyte losses.
10. A mother brings her 9-year-old child to your office and is worried that the child may have typhoid fever. He has a fever and nonbloody diarrhea. The child has a pet turtle and she recently read that turtles can transmit *Salmonella*. What would you tell her about the chances that her child has typhoid fever?
- A. Turtles can harbor typhoid fever, so she should be worried
- B. He should be started on empiric antibiotics
- C. Others in the family may develop typhoid fever, too, so they should be immunized against it
- D. The *Salmonella* infections transmitted by turtles are different from *Salmonella typhi*
- E. She should not worry since there are no longer any cases of typhoid fever in the United States.

ANSWERS

1. C. Dengue fever. This boy spent 2 weeks in India and returned 4 weeks ago. All of the illnesses listed in the question have moderate to long incubation periods except dengue fever. Dengue fever typically has an incubation period of less than 14 days.
2. A. Diseases such as diphtheria, polio, and measles occur in periodic outbreaks in many countries. For traveling children, "routine" vaccines against these diseases may not provide adequate immunity if a booster is not given before travel. There are no vaccines yet for malaria and dengue fever. Arriving

- adoptees do not require proof of immunizations. They may be only partially immunized and their records may not be accurate.
3. D. Quinidine and quinine as well as malaria itself can all cause hypoglycemia. It is important to perform frequent serum glucose checks when treating hospitalized patients with malaria. There is no vaccine yet for malaria. Close to 90% of worldwide cases of malaria are caused by *P falciparum*, not *P ovale*. Thick smears are more likely to reveal parasites than thin smears by a factor of approximately 10:1. Malaria most commonly presents as a non-specific flu-like syndrome. Distinct cyclic fevers occur far less frequently.
 4. E. This patient likely has malaria. Even though it had been treated in the past, it is quite likely that the parasites were not completely eradicated. Thin and thick blood smears remain the accepted diagnostic technique for malaria. One set of negative films is not enough to rule out disease and it is recommended that smears be repeated every 8–24 hours for at least three negative sets before malaria is ruled out.
 5. C. Petichiae are not common findings. The most common presentation for malaria is a nonspecific flu-like syndrome with high fever. It is frequently misdiagnosed as a viral syndrome. Gastrointestinal symptoms, including diarrhea, are common. Hepatosplenomegaly is noted in about 2/3 of cases. Laboratory studies frequently show moderately elevated liver functions with hyperbilirubinemia.
 6. D. Cambodia. Yellow fever is an arboviral infection which is transmitted by the *A aegypti* mosquito. It is found in sub-Saharan Africa and in Central and South America. It is not found in Southeast Asia.
 7. C. *Giardia lamblia*. Eosinophilia is defined as a peripheral eosinophil count of more than 400–500 cells/mm³. It is seen especially during the acute tissue invasive phase of infestation. The more common intestinal protozoa, *G lamblia* and *E histolytica*, do not cause eosinophilia.
 8. E. Thrombocytopenia and hemoconcentration are indicators of worsening disease in dengue fever. Parasites on a peripheral smear suggest malaria. Eosinophilia is not typically seen in dengue fever. Eosinophilia is more indicative of a helminth infection. Dengue fever may progress to a hemorrhagic state with oozing and bleeding, but a positive stool guaiac test alone is not an indication of worsening disease. Hypoglycemia can be seen in malaria but is less likely in dengue fever.
 9. C. The American Academy of Pediatrics still recommends against the use of loperamide or other antimotility agents for the management of diarrhea in children, whether travel-related or not. The routine use of antibiotics for the treatment of diarrhea in children who have not travelled out of the is not recommended. Trimethoprim-sulfamethoxazole used to be the empiric antibiotic of choice for the treatment of TD in children, but because of emerging resistance, other antibiotics are now recommended, including macrolides, quinolones, and third-generation cephalosporins.
 10. D. Typhoid fever is caused by *S enterica*, serotype *typhi*. There is no animal reservoir. Salmonella infections which are transmitted from animals such as turtles and poultry are caused by other serotypes such as *S typhimurium* or *S enteritidis*. It may be reasonable to obtain a stool culture in this patient, but antibiotics would not be indicated at this point. In children that are otherwise healthy, salmonella enteritis tends to be a self-limited disease and treatment with antibiotics is usually not indicated. Oral and parenteral vaccines against typhoid fever are 70–80% effective, but since this case is not typhoid fever, the vaccines would not be indicated. There are still about 500 cases of typhoid fever annually in the United States. Nearly two-thirds of these are acquired while abroad.

68 BIOTERRORISM—A PEDIATRIC PERSPECTIVE

Janet Lin

Timothy B. Erickson

BIOLOGICAL AGENTS

- Children remain potential victims of biological terrorism. In recent years, children have been specific targets of terrorists' acts.
- Specific to bioterrorism, more fulminant infectious diseases are possible in children because of immunologic immaturity and a more permeable blood–brain barrier.
- In contrast to a chemical attack, a covert biological attack will simulate a natural outbreak, with an incubation period rather than producing immediate mass casualties.

TABLE 68-1 Critical Biological Agent Categories for Public Health Preparedness

BIOLOGICAL AGENT	DISEASE
Category A	
<i>Bacillus anthracis</i>	Anthrax
<i>Yersenia pestis</i>	Plague
<i>Variola major</i>	Smallpox
<i>Clostridium botulinum</i> (<i>botulinum toxins</i>)	Botulism
<i>Francisella tularensis</i>	Tularemia
Filoviruses and arenaviruses (e.g., <i>Ebola virus</i> , <i>Lassa virus</i>)	Viral hemorrhagic fevers
Category B	
<i>Coxiella burnetii</i>	Q fever
<i>Brucella spp.</i>	Brucellosis
<i>Burkholderia mallei</i>	Glanders
<i>Burkholderia pseudomallei</i>	Melioidosis
<i>Alphaviruses</i> (<i>Venezuelan equine, eastern equine, western equine encephalomyelitis viruses</i>)	Encephalitis
<i>Rickettsia prowazekii</i>	Typhus fever
Toxins (e.g., ricin, staphylococcal enterotoxin B)	Toxic syndromes
<i>Chlamydia psittaci</i>	Psittacosis
Food safety threats (e.g., <i>Salmonella spp.</i> , <i>Escherichia coli</i> O157:H7)	
Water safety threats (e.g., <i>Vibrio cholerae</i> , <i>Cryptosporidium parvum</i>)	
Category C	
Emerging threat agents (e.g., <i>Nipah virus</i> , <i>hantavirus</i>)	

With permission from Centers for Disease Control and Prevention (CDC). Bioterrorism website. www.bt.gov/bioterrorism/factsheets. 2008. Bethesda, MD.

- The Center for Disease Control (CDC) has developed a list of critical agents for health preparedness (Table 68-1).

ANTHRAX

- Anthrax occurs in nature following contact with infected animals or animal products. The site of entry into the body is typically the skin, lungs, or GI tract.
- Ninety-five percent of naturally occurring anthrax cases are cutaneous disease. Cutaneous has less than 1% mortality if treated, but about 20% mortality if untreated. The risk is mostly occupational from close contact with sheep, goats, and cattle.
- The historical mortality rate of inhalational anthrax is up to 90%. The mortality from the U.S. mail-associated outbreak in 2001 was 45%.
- Naturally occurring GI anthrax accounts for less than 5% of reported cases.
- Release of exotoxins, edema toxin, and lethal toxin can lead to cell death and necrosis. The resultant release of

TNF (tumor necrosis factor) contributes to shock and death. There can be a large release of anthrax bacilli into the blood stream causing septicemia.

- The hallmark of cutaneous disease is a single lesion or a few lesions that are painless. Several days later, this progresses to a black depressed eschar at the base of the ulcer.
- *Bacillus anthracis* spores are highly stable and highly infectious upon inhalation. This is the form that has been manufactured for use in biological warfare. Illness resulting from an aerosolized release of anthrax spores is associated with an incubation period of 1 to 60 days, followed by fever, myalgias, cough, and chest pain.
- Children, particularly infants and toddlers, present with nonspecific symptom complexes primarily limited to fever, vomiting, cough, and dyspnea.
- Children with inhalational anthrax have abnormal chest roentgenograms; however, children with other forms of anthrax usually have normal roentgenograms.
- With inhalational anthrax there is abrupt onset of sepsis, hypotension, and death within 24 to 36 hours. Chest radiograph findings include mediastinal widening from lymphadenitis with pulmonary infiltrates or effusions.
- Hemorrhagic meningitis is expected in 50% of cases with a high overall case fatality rate. Hallmarks of the illness include gram-positive bacilli on tissue biopsy and blood smear or spinal fluid.
- Early antibiotic administration is likely to be the most important determinant of outcome in the setting of anthrax infection. Cutaneous anthrax without systemic sequelae can be adequately treated with oral doxycycline or ciprofloxacin.
- Patients with any other form of anthrax should be admitted and treated with intravenous antibiotics. Patients exposed to anthrax spores should receive antibiotic prophylaxis for 60 days. Unlike plague, secondary transmission of inhalational anthrax does not occur.

PLAGUE

- *Yersinia pestis*, the bacteria that causes plague, spreads in nature from infected fleas to humans. Historically, plague was known as the Black Death during the middle ages. Transmission of bubonic plague occurs through a fleabite, bite, or scratch of an infected animal or direct contact from an infected carcass.
- Pneumonic plague occurs via inhalation of respiratory droplets from an infected animal or person-to-person spread. Pneumonic plague can be primary inhalational or secondary, resulting from spread via the bloodstream in bubonic or septicemic cases.

- Naturally occurring plague is most often the bubonic type and occurs in the southwest U.S.
- An aerosol-mediated bioterrorist attack would be associated with pneumonic plague.
- Clinical manifestations include tender lymphadenopathy, if associated with bubonic plague and multiorgan failure, if associated with septicemic plague.
- An intense cough with thin bloody sputum is a characteristic finding. Gastrointestinal signs and symptoms can be very prominent.
- Petechiae, purpura, and an overwhelming picture of DIC may occur. Rapid progression to death occurs in those not treated with antibiotics within 24 hours of symptom onset.
- A clue to the diagnosis of *Y pestis* is a classic bipolar, “safety pin”-staining bacillus on Gram’s staining of the sputum or lymph node aspirate.
- Treatment consists of antibiotic treatment with streptomycin, gentamicin, doxycycline, ciprofloxacin, or chloramphenicol. Doxycycline or ciprofloxacin are the antibiotic choices for postexposure prophylaxis.
- Secondary transmission of pneumonic plague occurs. Therefore, standard blood and bodily fluid, as well as droplet precautions, are necessary. Postexposure prophylaxis is indicated for those exposed to pneumonic plague victims.
- If used as a biological weapon, smallpox represents a serious threat to the general population. It carries a high case-fatality rate of 30% among unvaccinated persons.
- There is no specific therapy.
- Smallpox has long been feared as the most devastating of all infectious diseases; its potential for devastation today is far greater than at any previous time.
- In a highly susceptible, mobile population, smallpox would be able to spread widely and rapidly throughout the world. Smallpox also has a high potential for secondary spread from person-to-person. Transmission occurs primarily through close face-to-face contact via droplet nuclei.
- Smallpox can also be transmitted via an airborne route in the setting of an infected patient with a severe cough and from direct aerosol inhalation. There is potential for the disease to spread exponentially. Secondary attack rate is estimated to be 25–40% in unvaccinated contacts. There is also very high potential for nosocomial spread.
- Vaccination with the smallpox vaccine within 3–4 days of exposure may prevent disease. Historically, the vaccine was given to children. Currently, it is not being offered to children for pre-event prophylaxis and it is not recommended for postexposure use in children less than 1 year-of-age.
- In the United States, virtually all children, and adults under the age of 35 years, are unvaccinated, and have no immunity to smallpox.
- The only currently available vaccine has been tested on adults, but not children. A “ring vaccination strategy” is now being recommended by the CDC. Cases of smallpox are rapidly identified, infected individuals are isolated, and contacts of the infected individuals as well as their contacts are immunized immediately.
- There is no specific treatment for smallpox. The mainstay of treatment is supportive therapy plus antibiotics as indicated for treatment of any secondary bacterial infections.
- No antiviral substances have yet proven effective for the treatment of smallpox.
- Airborne, droplet, and standard blood and bodily fluid precautions are indicated when caring for victims until all their scabs separate.

SMALLPOX

- A global campaign, begun in 1967 under the World Health Organization (WHO), succeeded in eradicating smallpox in 1977. The last documented case was reported in Somalia.
- The last documented case of smallpox reported in the United States was in 1949. Routine smallpox vaccination in the United States was discontinued in 1972. Therefore, the majority of US citizens are susceptible to an outbreak.
- If a terrorist had access to stored smallpox virus, a release could produce a chaotic situation.
- Smallpox afflicts only humans, as there are no known animal hosts.
- The diagnosis of smallpox is made clinically, with CDC laboratory confirmation.
- Smallpox has an incubation period of 7 to 17 days. Clinical illness is characterized by a prodrome of high fever, rigors, vomiting, and headache. Classic exanthem begins 2–4 days later, on the face and distal portions of the extremities, consisting of macules progressing to papules, umbilicated pustules, and then scabs.
- The centrifugal onset and synchronous nature of the rash helps to distinguish it from chickenpox.

BOTULISM

- Three forms of naturally occurring human botulism exist: foodborne, wound, and infantile.

- Botulism is an obligate anaerobe, spore-forming, and gram-positive rod. The bacteria produce its effect through botulinum toxin.
- The toxin is taken up by skeletal muscle motor neurons where it irreversibly inhibits the release of acetylcholine, resulting in postsynaptic muscle paralysis. The paralysis persists until axonal branches regenerate.
- Wound botulism and intestinal botulism are infectious diseases that result from production of botulinum toxin by *C botulinum* either in devitalized (ie, anaerobic) tissue or in the intestinal lumen.
- A man-made form of botulism, results from aerosolized and purified botulinum toxin and is called inhalational botulism.
- This mode of transmission has been demonstrated experimentally in primates and has been attempted by bioterrorists.
- Patients with botulism present with a descending flaccid paralysis that always begins in the bulbar musculature. It always presents with multiple cranial nerve palsies. Patients may complain of difficulty seeing, speaking, or swallowing.
- Prominent neurologic findings include ptosis, diplopia, blurred vision, often enlarged or sluggishly reactive pupils, dysarthria, dysphonia, dysphagia, and descending paralysis.
- Because botulism is toxin-mediated, there is no transmission from person-to-person. Respiratory isolation is not needed.
- The mainstays of treatment are supportive with passive immunization with one of two antitoxins.
- The trivalent botulinum antitoxin is available through the CDC and may be used to treat children of any age with subtypes A, B, or E.
- Botulism immune globulin (BIG) is used to treat infant botulism.

TULAREMIA

- Tularemia is a zoonotic illness that occurs after exposure to diseased animal fluids or bites from infected deerflies, mosquitoes, or ticks. The classic animal reservoir is the lagomorph (rabbit).
- While not highly fatal, its extremely high infectivity and its ability to escape laboratory detection make it an agent of potential use for bioterrorism.
- Symptoms include sudden fever, chills, diarrhea, dry cough, muscle aches, joint pain, and progressive weakness or prostration.
- An aerosolized release (as would occur in a bioterrorist attack) would likely result in clinical findings similar to community-acquired pneumonia.
- Tularemia can be treated with antibiotics. Treatment of choice is streptomycin administered by intramus-

cular injection. Alternatively, gentamicin can be given intravenously. Other antibiotics that are used include tetracyclines and chloramphenicol.

- Mass casualty prophylaxis can be achieved with doxycycline or ciprofloxacin.
- Person-to-person transmission does not occur and respiratory isolation is not required.

VIRAL HEMORRHAGIC FEVERS

- The two most noteworthy viral hemorrhagic fevers are caused by the filoviridae and the arenaviridae. They are both highly contagious and are associated with a high mortality rate.
- Filoviruses cause Ebola and Marburg hemorrhagic fevers. Arenavirus causes Lassa fever.
- Rodents, ticks, and mosquitoes are common vectors; however, the vectors for Marburg and Ebola are unknown.
- Documented cases of human-to-human transmission occur with Ebola, Marburg, and Lassa hemorrhagic fever viruses.
- Symptoms most often include fever, dizziness, fatigue, exhaustion, and muscle aches. Severe forms of the disease may present with bleeding under the skin, in internal organs and from body orifices. Fulminant illnesses will present with shock and, oftentimes, multiorgan system failure.
- Supportive care and blood product replacement are the mainstays of therapy.
- Ribavirin (an antiviral medication) is possibly effective in treating some patients with an arenavirus illness (Lassa fever).
- Secondary transmission is likely with Ebola, Marburg, and Lassa fever, necessitating airborne isolation with standard blood and bodily fluid precautions.
- A unique feature of previous Ebola outbreaks has been the relative sparing of children.

PEDIATRIC BIOLOGICAL AGENT EXPOSURE MANAGEMENT

- Children may respond more readily to therapeutic intervention, but they are more susceptible to various agents and more likely to deteriorate if they are not monitored carefully.
- Many of the highest-threat bioterrorist agents are appropriately treated in adults with ciprofloxacin or doxycycline.
- The fluoroquinolone and tetracycline classes of antibiotics have each been considered as relatively contraindicated in young children. The fluoroquinolone concern has primarily stemmed from arthropathy and growth abnormality.

- The tetracyclines are known to stain children's teeth, particularly in those children less than 8 years old taking prolonged or repeated courses.
- Clinical use of ciprofloxacin in young children demonstrates no evidence of arthropathy or slower rates of bone growth in these children.
- The Federal Drug Administration (FDA) has approved ciprofloxacin for use in bioterrorist-related anthrax exposures for children, as well as for adults.
- The pediatric healthcare professional must be in contact with their local, state, and federal public health infrastructure as soon as a potential biological agent is perceived clinically.
- The key component is identifying a biological agent in the community and then moving quickly to isolate those that may be at risk of spreading the infection.

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QUESTIONS

1. Which of the pathogens listed below is considered a Class A critical agent for health preparedness according to the CDC?

- A. *Clostridium perfringens*
- B. *Bacillus anthracis*
- C. *Brucella melitensis*
- D. *Escherichia coli* O157:H7
- E. *Botulinum toxicosis*

2. Unlike a chemical agent attack, a covert biological attack will simulate a natural outbreak with, oftentimes, an incubation period. Which of the following heighten suspicion that the pediatric patients presenting to your hospital may be related to a bioterrorist act?

- A. There has been a rodent die off that has occurred in your region prior to patients presenting with cervical lymphadenopathy.
- B. A patient who presents with flu-like symptoms responds to oral antibiotic treatment.
- C. Many patients who were on vacation in the same area in southwestern part of the United States come in with the same complaints of cough and lymphadenopathy.
- D. More severe cases of an infectious disease than expected have presented to your emergency department.
- E. A dairy farmer who traveled to the city to visit family presents with a rash and cough.

3. You suspect that botulism is the agent that was released in a bioterrorist attack at a local elementary school. Which of the following management strategies identified below is correct?

- A. Isolation of patients admitted with botulism is not needed.
- B. Decontamination need not to be performed.
- C. Supportive therapy is secondary as there is definitive treatment.
- D. Respiratory failure is not the primary concern with this form of botulism.
- E. Intravenous botulinum immunoglobulin (BIG-IV) is indicated in this setting.

4. A pediatric patient is brought into your ED with fever and a severe cough. Mom had noted some shortness of breath and noted a little blood with one of her child's coughing episodes. They had traveled to an amusement park during their vacation last week. On examination, lungs are clear and you note some tender lymphadenopathy. The most likely agent responsible for the presentation is which of the following?

- A. Anthrax
- B. Brucellosis
- C. Plague
- D. Q fever
- E. Tularemia

5. What is a reason that fluoroquinolone and tetracycline class medications can be used in the pediatric population?

- A. The FDA has approved widespread use of these antibiotics for infectious indications.
- B. Previous reports of toxicity or adverse effects have not been founded, especially for the tetracyclines.
- C. Benefit of treatment outweighs the risks associated with administration of these medications for bioterrorist-related indications.
- D. Dose adjustments can eliminate the associated risk of these antibiotics in the pediatric population
- E. Newer formulations of these two classes of medications have eliminated the previous known toxicities and/or adverse affects associated with administration in children.

ANSWERS

1. B. *B anthracis* is a class A critical agent. *Brucella* spp and *E coli* are class B. *C perfringens* is not on the list.
2. D. A rodent die off prior to presentation of plague typically implies something that may be naturally occurring. A patient who is responding to routine treatment for pneumonia is less likely to have a virulent, purified, or manufactured strain of infectious disease that would be more suspicious of a bioterrorist agent. The Southwest U.S. is known to harbor endemic plague. Victims who were in the same area at the same time are more likely to be exposed to same infectious disease. Dairy farmers may have cattle, sheep, or goats on the farm. Cutaneous anthrax is commonly diagnosed with occupational exposure.
3. A. As botulism is not spread person-to-person respiratory isolation is not needed. B is incorrect. Decontamination should still be performed prior to definitive diagnosis. Toxin that may be on persons can still be inhaled by responders or healthcare providers. C is incorrect because supportive therapy is the mainstay of treatment. Antitoxin is definitive treatment, but should not preclude supportive therapy. Antitoxin binds free toxin in blood, it cannot bind those that have already entered the neuron. D is incorrect as respiratory failure due to paralysis of respiratory muscles and/or diaphragm muscles is the cause of mortality. E is incorrect as BIG-IV is indicated in infantile botulism, age <1year but there is no role for immunoglobulin in any other form of botulism.
4. C. Classic presentation of plague is initially nonspecific flu-like symptoms. However, bloody sputum is common. Bubonic plague presents with tender lymphadenopathy and can progress into pneumonic plague via hematogenous spread.
5. C. These medications are considered relatively contraindicated. The FDA has approved the use of cipro for bioterrorist-related anthrax exposures in adults and children. There are studies that show that growth abnormality and arthropathy have not been seen in certain pediatric population treated for certain diseases. However, tooth staining is still an adverse effect to tetracyclines, especially in children under the age of 8. While prolonged or repeated use of tetracyclines may increase the risk of tooth staining, generally dose adjustments have not been found to decrease risks of toxicity/adverse effects. No pharmaceutical has been developed that has eliminated these risks.

Section 9

IMMUNOLOGIC EMERGENCIES

69 COMMON ALLERGIC PRESENTATIONS: ALLERGIC CONJUNCTIVITIS/RHINITIS

William R. Ahrens

HIGH-YIELD FACTS

INTRODUCTION

- Common allergic presentations in the emergency department (ED) include asthma, eczema, allergic conjunctivitis, and allergic rhinitis. Current literature emphasizes the interrelationship and overlap between these illnesses, especially allergic conjunctivitis and rhinitis: the two diseases are often discussed together as allergic rhinoconjunctivitis.

PATHOPHYSIOLOGY

- Allergic illnesses are most commonly seen in patients with a history of atopy, which is defined as a genetically determined hypersensitivity to environmental antigens. The most common form of hypersensitivity, type 1, is associated with IgE antibody and is the cause of most of the allergic presentations seen in the ED. Contact of IgE antibody with mast cells triggers mast cell degranulation, which results in the release of multiple inflammatory mediators. Histamine released on mast cell activation can cause pruritis, bronchospasm, increased vascular permeability, and vasodilation. Asthma, hay fever, allergic conjunctivitis, and allergic rhinitis can all be triggered by type 1 responses to environmental antigens.
- The most severe manifestation of a type 1 reaction is anaphylaxis, which can be fatal.

- Common indoor allergens include house dust mites, cockroaches, and pet allergens; these often cause persistent, perennial symptoms. Outdoor allergens include pollen from grass and trees—these tend to cause seasonal symptoms.

CLINICAL PRESENTATION

ALLERGIC CONJUNCTIVITIS

- The conjunctiva of the eyes is connected to the nose via the lacrimal ducts; in this sense, it is part of the airway. The association of allergic conjunctivitis with allergic rhinitis is such that some authors group the two together under the heading “allergic rhinoconjunctivitis.” There are several specific classifications of allergic conjunctivitis.
- Seasonal allergic conjunctivitis (SAC) implies that patients have symptoms for a defined period of time. Patients with perennial conjunctivitis (PAC) have symptoms year round. Together, SAC and PAC account for approximately 98% of allergic eye disease in patients. Patients with SAC and PAC usually have atopic symptoms and a family history of atopy. A fairly high percentage complains specifically of concomitant nasal symptoms. Itching of the eyes is an important symptom; without itching, the diagnosis of allergic conjunctivitis is unlikely. Physical findings include injection of the conjunctival vessels, chemosis, and eyelid edema (Fig. 69-1). Increased vascular permeability can result in conjunctival edema. Corneal involvement is uncommon and vision is rarely affected.
- Vernal keratoconjunctivitis (VKC) is a chronic bilateral inflammation of the conjunctiva that predominantly occurs in children (Fig. 69-2). It is most common in hot dry environments and typically occurs during spring months. As with SAC and PAC, itching is an important

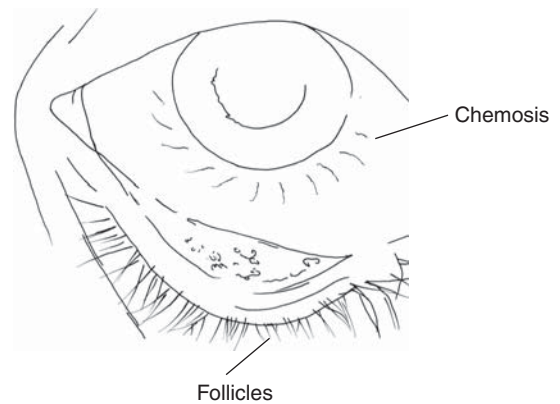
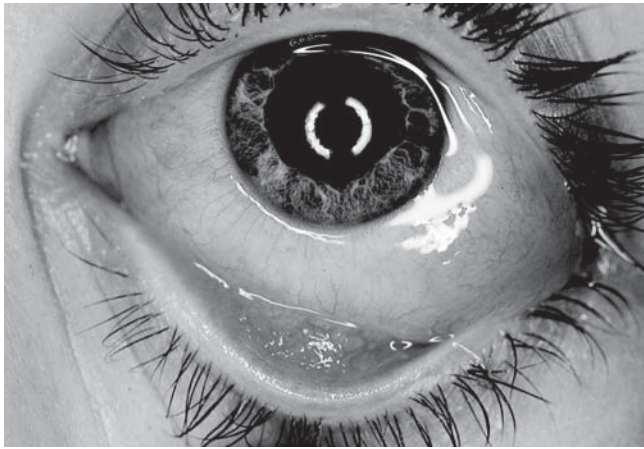


FIG. 69-1. Allergic conjunctivitis conjunctival injection, chemosis, and a follicular response in the inferior palpebral conjunctiva in this patient with allergic conjunctivitis secondary to cat fur.

symptom; photophobia and a foreign body sensation can be present. The eyelid skin is usually not involved. Eye discharge tends to be thick and mucoid.

- There are two types of VKC. In the palpebral variety, giant papillae are commonly found on the superior tarsal conjunctiva; the inferior tarsal conjunctiva is generally unaffected. The limbal variety of VKC is more common in dark-skinned individuals; papillae occur at the limbus. VKC can be complicated by punctate epithelial keratopathy, which can lead to a corneal shield ulcer, especially in very young patients.
- Giant papillary conjunctivitis (GPC) is an immune-mediated inflammatory disorder of the superior tarsal conjunctiva; it is characterized by the presence of “giant” papillae, characteristically greater than 0.3 mm in diameter. GPC may be a reaction to ocular irritants: contact lenses are associated with this

disorder. Ocular itching is associated with a mucoid or watery discharge.

DIFFERENTIAL DIAGNOSIS

- The differential diagnosis of conjunctivitis includes acute bacterial or viral infection and allergic disorders. Bacterial conjunctivitis is usually an acute illness associated with a purulent discharge. Viral conjunctivitis is also usually an acute process; while the discharge tends to be more watery than in bacterial conjunctivitis, the two can be difficult to differentiate. A careful history can help distinguish infectious from allergic conjunctivitis, since patients with the allergic conjunctivitis usually have a personal or family history of atopic illnesses, and allergic symptoms are often seasonal.

TREATMENT

- The treatment of each type of ocular allergy begins with trying to avoid the allergic stimulus or stimuli that trigger the illness. Artificial tears provide some relief. Treatment of SAC and PAC can be initiated in the ED with a topical antihistamine alone or in combination with a mast cell stabilizer (cromolyn sodium).
- Antihistamines have the advantage of providing relatively rapid relief; mast cell stabilizers have a delayed effect. Treating ocular symptoms may decrease nasal symptoms. Topical steroids are reserved for patients with allergic conjunctivitis not responding to other therapy, since they can cause complications such as glaucoma and cataracts. Treatment of patients suspected of having VKC and GPC is best initiated by an allergist or ophthalmologist.

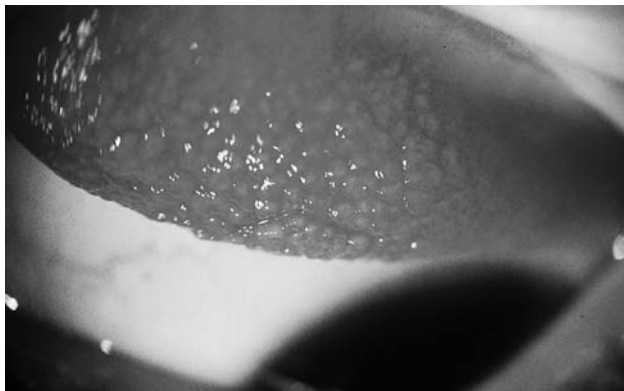


FIG. 69-2. Vernal conjunctivitis—the tarsal conjunctiva demonstrates giant papillae and a cobblestone appearance pathognomonic for vernal conjunctivitis.

TABLE 69-1 Classification of Allergic Rhinoconjunctivitis in Children

Intermittent: symptoms less than 4 d/wk and present less than 4 wk
Persistent: symptoms more than 4 d and present for more than 4 wk
Mild severity: no sleep disturbance, impairment of school or daily activities, symptoms well tolerated
Moderate to severe: sleep is disturbed, school and daily activities are disturbed, symptoms are well-tolerated

With permission from de Groot H, Brand PL, Wytse FF, et al. Allergic rhinoconjunctivitis in children. *BMJ*. 2007;335:985.

ALLERGIC RHINITIS OR ALLERGIC RHINOCONJUNCTIVITIS

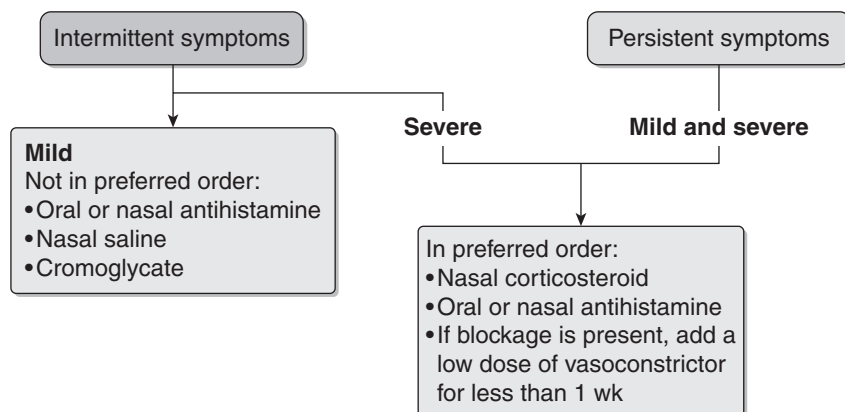
- The prevalence of allergic rhinoconjunctivitis has doubled in the past 20 years. The diagnosis is often missed, which is not surprising given the prevalence of rhinorrhea in the pediatric population.
- Untreated or undertreated, allergic rhinoconjunctivitis is associated with sleep disturbances and impairment of daily activities and school performance. Generalized fatigue and malaise is common. Allergic rhinitis is the second most common predisposing factor for sinusitis after upper respiratory infection. Postnasal drip can cause chronic cough and exacerbate the symptoms of asthma.
- The same allergens responsible for the ASC and PAC cause allergic rhinitis; however, not all children with ocular and nasal symptoms are sensitized to an identifiable antigen. Affected children often have a history of asthma, eczema, chronic sinusitis, and otitis media with effusion. Children with prolonged nasal symptoms or those who “always have a cold” are likely to be suffering from allergic rhinitis. Allergic rhinitis is characterized as intermittent or persistent; it is further characterized as mild or moderate/severe (Table 69-1).

TABLE 69-2 Partial List of the Differential Diagnosis of Rhinorrhea

Viral upper respiratory infections
Sinusitis
Allergic rhinitis
Vasomotor rhinitis
Nasal foreign bodies
Nasal polyps
Nasal septum deviation
Immunologic or inflammatory disorders

Reprinted from Ferdman RM, Linzer JF. The runny nose in the emergency department. *Clin Pediatr Emerg Med*. 2007;8:123, Copyright 2007, with permission from Elsevier.

- The symptoms of allergic rhinitis are largely the result of histamine released from mast cells. They include nasal congestion, itching, sneezing, and discharge. Complaints of itchy and watery eyes are common; fever is not. Symptoms can be seasonal or perennial. Classic physical findings associated with allergic rhinitis include “allergic shiners,” darkening of the lower eyelids due to suborbital edema; the “allergic salute,” upward rubbing of the nose with the hand to relieve nasal itching; and the “allergic crease” (a transverse line across the nasal bridge due to the “nasal salute”). Nasal discharge from AR tends to be more watery and clear than in patients with acute upper respiratory infection. Physical examination may reveal conjunctival injection and watery eye discharge from concomitant allergic conjunctivitis.
- Assessment depends on history and physical examination, of which the most important components are the presence of other manifestations of atopy. An abbreviated list of the differential diagnosis of rhinitis is given in Table 69-2.

**FIG. 69-3.** Algorithm for the management of allergic rhinoconjunctivitis in children.

TREATMENT

- Treatment of allergic rhinitis can begin in the emergency department. (Fig. 69-3). The most effective medications for allergic rhinitis are intranasal glucocorticoids; they reduce symptoms and improve quality of life. Second-generation antihistamines such as cetirizine and loratadine are also effective in children. Disodium cromoglycate is less effective than glucocorticoids or antihistamines. Intranasal decongestants can improve symptoms in the short term, but rebound mucosal swelling can occur with prolonged use.

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QUESTIONS

1. A 3-year-old girl is referred to the ED by her preschool for “red eyes.” The mother reports several days of nasal congestion and noticed the child rubbing her eyes this morning. You suspect allergic conjunctivitis. Which of the following statements are correct?
 - A. Does not require treatment because morbidity is rare.
 - B. Usually occurs as an isolated entity.
 - C. Almost always causes photophobia.
 - D. Almost always causes itchy eyes.
 - E. Should never be diagnosed in the emergency department
2. A 5-year-old boy is brought to the ED for possible exacerbation of his “seasonal allergies.” Per his mother, every spring he develops itching eyes associated with a foreign body sensation and a thick eye discharge. Which of the following would be an expected symptom or finding associated with this condition?
 - A. Is associated with giant papillae on the tarsal conjunctiva
 - B. Is always perennial

- C. Never causes an eye discharge
 - D. Is rare in children
 - E. Is usually associated with iritis
3. A 7-year-old girl is brought to the ED for evaluation of possible “pink eye.” Based upon the history you obtain as well as your physical examination, you conclude the patient has seasonal or perennial allergic conjunctivitis. The appropriate treatment for this patient would be?
 - A. Complex and should not be initiated in the ED
 - B. Should begin with ophthalmic corticosteroids
 - C. Commonly involves ocular antihistamines and mast cell stabilizers
 - D. Has no effect on concomitant allergic rhinitis
 - E. Is rarely effective

ANSWERS

1. D. Allergic conjunctivitis is associated with considerable morbidity, including malaise, fatigue, and lost school days. It is often accompanied by allergic rhinitis. Photophobia is uncommon. It can usually be diagnosed in the emergency department. A hallmark of allergic conjunctivitis is itchy eyes.
2. A. Vernal conjunctivitis tends to be seasonal, and is common in children. Iritis is rare. It is often accompanied by an eye discharge; giant papillae on the tarsal conjunctiva are common.
3. C. Treatment of allergic conjunctivitis should start in the ED with ocular antihistamines and/or mast cell stabilizers, which are usually effective treatment. Concomitant allergic rhinitis will often improve if allergic conjunctivitis is effectively managed. Ophthalmic corticosteroids are effective treatment, but due to possible side effects are best left to an ophthalmologist or allergist.

70 ANAPHYLAXIS

E. Bradshaw Bunney

- Anaphylaxis is a severe, potentially life-threatening hypersensitivity reaction characterized by skin or mucosal manifestations that include a pruritic rash, urticaria, or angioedema, respiratory compromise associated with airway edema and bronchospasm, and/or cardiovascular compromise that can result in distributive shock. It occurs within minutes to hours

after exposure to an offending allergen. Food allergy is the most common cause of anaphylaxis.

ALLERGENS

- Ninety percent of food-related anaphylaxis is caused by exposure to nuts and shellfish.
- In infants, cow's milk and eggs are the most common causes.
- Insect stings are a common cause of a localized allergic reaction and can also cause anaphylaxis.
- Anaphylaxis from medications is relatively rare.
- The most common medications causing anaphylaxis are aspirin, NSAIDs, and β -lactam antibiotics.
- IV administration of medication has a higher incidence of anaphylaxis and more rapid onset of the anaphylactic symptoms.
- Latex allergy is becoming an increasingly common cause of allergic reaction in children

PRESENTATION

- Mild anaphylaxis consists primarily of skin and mucosal manifestations
 - Erythema, flushing, urticaria (hives), itching, and angioedema.
 - Wheezing or throat irritation and mild tachycardia, without respiratory or cardiovascular compromise.
- Moderate anaphylaxis has the same cutaneous manifestations, with the addition of difficulty breathing, barky cough, stridor, light-headedness, and tachycardia.
- Severe anaphylaxis can cause severe respiratory distress and cardiovascular manifestations that range from hypotension and tachycardia to fulminant shock.
- Gastrointestinal symptoms such as vomiting, diarrhea, and crampy abdominal pain frequently accompany moderate-to-severe anaphylaxis.
- The onset of symptoms ranges from minutes to a few hours after contact with an allergen.

- Considers anaphylaxis in a hypotensive child with respiratory distress.
- Up to 10% of children with anaphylaxis will present without a rash.
- Insect stings have been associated with hypotension and a relative bradycardia.

ANCILLARY TESTS

- Laboratory tests or radiographic tests will not help to diagnose anaphylaxis.

TREATMENT

- Treatment begins with assuring airway patency.
 - The airway may be difficult to secure in the child with severe mucosal edema.
 - Immediate endotracheal intubation is done in any child at risk of respiratory arrest.
 - Always consider rescue airway techniques that may be needed, such as cricothyrotomy and jet insufflation.
- See Table 70-1 for medications used to treat anaphylaxis.
- Epinephrine is the primary drug of choice for treating moderate-to-severe forms of anaphylaxis, 0.01 mL/kg of 1:1000 IM every 15 minutes, or 0.01 mL/kg of 1:10 000 IV.
 - IM is preferred over SC because of more efficient absorption.
 - Inhaled epinephrine has been shown NOT to achieve the serum epinephrine levels needed to treat anaphylaxis.
- Isotonic IV fluids are the second mainstay of treating anaphylaxis in patients who are hypotensive.
- IV crystalloids are given as a 20 cc/kg bolus rapidly for hypotension; multiple boluses may be necessary.
- If hypotension persists, then an epinephrine or dopamine drip should be considered.
- Bronchospasm may respond to inhaled β -agonists.
- H-1 antihistamines such as diphenhydramine are often used in treating the histamine-related rash and pruritus.

TABLE 70-1 Medications used for the Treatment of Anaphylaxis

Epinephrine	0.01 mL/kg 1:1000 0.01 mL/kg 1:10 000	IM or SC IV	Every 15 min Every 15 min
Albuterol	0.03–0.05 mL/kg 0.5% solution	Nebulized	Every 15 min
Diphenhydramine	1–2 mg/kg	PO or IV or IM	Every 4–6 h
Cimetidine	5–10 mg/kg	PO or IV or IM	Every 6 h
Methylprednisolone	1–2 mg/kg	IV or IM	Every 6 h

- H-2 antihistamines such as cimetidine have not been proven to be of benefit.
- Steroids, such as methylprednisolone, are used to prevent recurrence of symptoms or a delayed reaction, the evidence for this is weak, and they are not first-line medications in the treatment of anaphylaxis.
- Keep the patient in the Trendelenburg position until symptoms have completely resolved.

DISPOSITION

- All severe anaphylaxis involving vascular collapse should be admitted to the ICU for close monitoring.
- Any child with respiratory symptoms that have completely resolved should be observed for 6 to 8 hours.
- Patients being discharged are given strict instructions to return if symptoms recur.
- Patients with first time anaphylaxis are referred to an allergist.
- Autoinjectable epinephrine is used by patients for immediate treatment of anaphylaxis symptoms outside the hospital.
 - 0.15 mg dose is used in children weighing 10 to 25 kg.
 - 0.3 mg dose is used in children weighing 25 kg and over.
 - Autoinjectable epinephrine is given to all patients with cardiovascular or respiratory reaction, idiopathic anaphylaxis, and persistent asthma with a food allergy.

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QUESTIONS

1. A 5-year-old male with a known allergy to peanuts accidentally received a dish at a Chinese restaurant, which contains peanuts. He arrives in severe respiratory distress, BP 40/P, HR 160. There is no IV. After securing the airway, the first line of treatment is
 - A. IM diphenhydramine
 - B. SC epinephrine
 - C. IM epinephrine
 - D. Nebulized epinephrine
 - E. IM methylprednisolone
2. The hypotension in the above case is best treated with which of the following?
 - A. Dopamine drip
 - B. Epinephrine drip
 - C. IV crystalloids
 - D. Dobutamine drip
 - E. IV colloids
3. An 8-year-old girl who developed a marked urticarial rash, moderate expiratory wheezing, and tachycardia without hypotension after eating scallops has complete resolution of her wheezing, tachycardia and improvement in her rash. How long should she be observed in the ED?
 - A. 1–2 hours
 - B. 2–4 hours
 - C. 4–6 hours
 - D. 6–8 hours
 - E. 8–10 hours

ANSWERS

1. C. Epinephrine is the first medication that should be used in any child with severe anaphylaxis. IM has more efficient absorbance compared to SC. Nebulized epinephrine has no proven benefit for anaphylaxis. Both diphenhydramine and methylprednisolone are second line agents in severe anaphylaxis.
2. C. The shock in anaphylaxis is due to dilation of the vasculature and leaking at the capillary beds. This is a distributive shock and therefore has to have the fluids lost to redistribution replaced. Pressors are second line treatment after crystalloid fluids at 20 cc/kg boluses have been given. Colloids have no proven benefit in anaphylaxis.
3. D. The delayed phase reaction in anaphylaxis can occur up to 6–8 hours after the initial symptom onset.

71 ABDOMINAL PAIN

Philip H. Ewing

THE HISTORY AND EXAMINATION FOR ABDOMINAL PAIN

- Abdominal pain can be either visceral (poorly localized, difficult to describe) or somatic (intense, readily localized).
- The history and physical for a patient should be conducted in an age appropriate manner that may require adult caregivers to leave the exam room for certain parts of the history or exam.
- Referred pain syndromes should alert the practitioner to include an abdominal process in the differential diagnosis.
 - Table 71-1 lists extraabdominal and systemic conditions which may present as abdominal pain.
- A complete examination of the patient with abdominal pain includes the genitalia and perineum, in addition to a rectal examination when appropriate.
 - Do not miss diagnoses such as an incarcerated inguinal epididymitis, hernia, or testicular torsion.

DIAGNOSTIC TESTS FOR ABDOMINAL PAIN

- Young infants and children are particularly vulnerable to the long-term effects of ionizing radiation, and the widespread use of CT for diagnostic imaging may increase the risk of cancer in the future.
 - Table 71-2 shows the exposure to ionizing radiation from common diagnostic imaging.
- Contrast CT scans are an important tool that should be used judiciously due to the risk of contrast-induced nephropathy. A detailed past medical history can be

helpful in discovering patients who are at risk, but a serum creatinine level is a quick screening tool.

- Ultrasound is emerging as a diagnostic adjunct for many abdominal emergencies that are addressed in the ED. Diagnostic ultrasound can be used at the bedside and does not expose the patient to ionizing radiation.
 - Table 71-3 lists conditions which ultrasonography may be used as the first imaging of choice.

OBSTRUCTION

- Bilious emesis in a newborn infant must be considered malrotation with midgut volvulus until proven otherwise. An upper gastrointestinal series should be ordered to confirm the diagnosis.
- Air contrast enemas are the therapy of choice for intussusception. Special consideration should be given to older patients who develop intussusception. Patients at and beyond the age of 2 to 5 years have an increased incidence of a pathologic lead point as the cause of prolapse.
- Incarcerated hernias that are not reducible through traditional means (cold packs, Trendelenburg positioning, or massage) require surgical attention because of the risk of bowel ischemia and perforation.

PERITONEAL IRRITATION, INFLAMMATION

- Acute appendicitis classically produces abdominal pain that is initially vague and periumbilical and within hours of onset localizes to the right lower quadrant. Vomiting, anorexia, and low-grade fever are usually present. Table 71-4 reviews helpful clinical bedside signs of appendicitis and peritoneal irritation.

TABLE 71-1 Extrabdominal and Systemic Causes of Abdominal Pain

- Asthma
- Pneumonia
- Heart disease
- Toxin exposure, ingestion, or overdose
- Collagen vascular disease
- Diabetic ketoacidosis
- Black widow spider bite
- Hemolytic–uremic syndrome
- Inborn error of metabolism
- Sepsis
- Abdominal epilepsy/migraine
- Henoch–Schönlein purpura
- Mononeucloisis
- Pharyngitis

TABLE 71-2 Ionizing Radiation from Common Radiological Imaging

• Two-view chest radiography	• 0.16 mSv
• Adult abdominal CT	• 10 mSv
• Neonatal abdominal CT	• 20 mSv
• Barium enema	• 15 mSv

The estimated lifetime attributable risk of death from any cancer after an abdominal CT in a pediatric patient ranges from 0.06% to 0.14%. Six to fourteen patients in 10 000 are estimated to develop fatal cancer after a single exposure.

- Spontaneous or primary peritonitis is a condition that is associated with nephrotic syndrome, characterized by ascites and relative immunodeficiency. Antibiotic therapy should be directed at *Streptococcus* species and gram-negative rods, but other organisms should be considered especially if the patient has an indwelling dialysis catheter.
- Antibiotics and decompression of the bowel may be indicated to stabilize a patient with suspected Hirschsprung's disease. The diagnostic workup may include a barium enema demonstrating a "transition zone," rectal manometry, or a suction biopsy of the rectum and colon.

NONSURGICAL CAUSES OF ABDOMINAL PAIN

- Acute gastroenteritis—vomiting and diarrhea frequently accompany the chief complaint of abdominal pain, and commonly present together. Be particularly concerned with the patient who presents with vomiting and abdominal pain without diarrhea. Surgically correctable disease must be ruled out.
- Urinary tract infections—patients may have fever, nausea, emesis, abdominal pain, and urinary symptoms such as frequency, urgency, and dysuria.

TABLE 71-3 Ultrasound as First Imaging of Choice

- Appendicitis
- Cholelithiasis
- Intussusception
- Pregnancy and ectopic pregnancy
- Renal stones
- Ovarian/testicular torsion
- Abdominal trauma

TABLE 71-4 Bedside Signs of Peritoneal Irritation (Appendicitis)

- McBurney's sign: rebound RLQ pain
- Rosving's sign: rebound tenderness referred from LLQ to RLQ
- Psoas sign: pain with passive extension of the thigh while the knee is extended
- Obturator sign: right knee pulled laterally while hip flexed 90 degrees

Screening urinalysis, culture, and Gram stain can identify affected patients. An oral course of antibiotics will suffice for treatment of patients who appear nontoxic; parenteral antibiotics are required for infants younger than 2 to 6 months and ill patients where the risk of pyelonephritis, bacteremia, and urosepsis is greater.

- Constipation—some patients will present with recurrent UTIs secondary to obstruction. Often the patient will have encopresis, loose, or liquid stools resulting from leakage around impacted stool in the distal colon and rectum.
- Colic—it is important to rule out pathologic causes for the infant's irritable behavior. Efforts should be directed to supporting the parents and empowering them with knowledge about the benign nature of the symptoms and giving them ample resources for stress relief. An emergency provider must help them partner with their primary care physician to ensure that the family has follow-up after discharge.
- Bleeding and pain
 - Anal fissures can be seen on careful examination of the perianal region and is a common cause of bleeding in infants.
 - Henoch–Schönlein purpura (HSP) may cause blood in the stools. The characteristic rash of HSP may also present with abdominal pain, arthralgias, and arthritis. Obtain a urinalysis for proteinuria and hematuria and especially look for RBC casts.
 - Hemolytic uremic syndrome (HUS) presents with anemia, thrombocytopenia, and renal failure. Abdominal pain with bloody diarrhea can be part of the prodrome; the causative agent is often *Escherichia coli* O157:H7.

- Liver/pancreatic pain—patients with ongoing hemolytic disease (ie, sickle cell anemia), enteric infections, and Kawasaki disease are at risk for biliary disease. Pancreatitis may present with abdominal pain radiating to the back after high-fat meals in conjunction with pale stools. Obesity appears to be increasing the incidence of pancreatitis in children and adolescents, but some children will develop the disease as a result of an infection or exposure, while others will develop pancreatitis as a manifestation of a systemic or genetic disorder such as cystic fibrosis or hereditary pancreatitis. Jaundice and abdominal pain may be the first manifestations of hepatitis, which can be related to infection, drug exposure (especially acetaminophen), systemic disease, or diseases inherent to the liver and biliary tree.
- Inflammatory bowel disease—the undiagnosed IBD patient may present repeatedly to the ED for complaints of abdominal pain. The subset of patients with Crohn's disease may have early manifestations of the disease in the perianal area, including fistulae and skin tags. At a minimum, the patient should have a CBC, liver function tests, and assessment of inflammatory markers.

GYNECOLOGIC/UROLOGIC CAUSES OF ABDOMINAL PAIN

- Gynecologic causes of abdominal pain include ovarian cysts and tumors, ovarian torsion, dysmenorrhea, endometriosis, pelvic inflammatory disease, pregnancy, and ectopic pregnancy.
- Genital problems in males with abdominal pain include testicular torsion, torsion of the appendix testes, orchitis, and epididymitis.

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QUESTIONS

1. A 3-week-old infant presents with vomiting and what the parents think is abdominal pain. During the exam, the infant vomits, producing a bright yellow-green emesis. What is the most appropriate diagnostic study?
 - A. Two view chest radiograph
 - B. Two view abdominal radiograph
 - C. Barium enema
 - D. Abdominal ultrasound
 - E. Upper gastrointestinal series
2. A 4-year-old girl who was recently diagnosed with nephrotic syndrome presents to the ED with two days of fever and now abdominal pain. You note that she is edematous and has a slightly distended and tender belly with no surgical scars. What is the most likely causative organism?
 - A. *Staphylococcus aureus*
 - B. *Pseudomonas*
 - C. *Streptococcus viridians*
 - D. *E coli*
 - E. *Streptococcus pneumonia*
3. You are evaluating a child with abdominal pain in whom you are concerned about appendicitis. You have decided that the child needs diagnostic imaging, but the parents are concerned about the potential effects of radiation. When compared in millisieverts, how many times more radiation does an abdominal computed tomography scan deliver than a two-view chest radiograph?
 - A. 10
 - B. 50
 - C. 100
 - D. 500
 - E. 1,000
4. A 13-year-old female is transferred from an outside facility with persistent suprapubic abdominal pain after a workup that included a normal urinalysis, negative abdominal series, and a negative urine pregnancy test. She describes 1-day history of colicky lower abdominal pain that does not radiate. Which study would be helpful in making the diagnosis?
 - A. Complete blood count with differential
 - B. Ultrasound
 - C. Computed tomography of the abdomen
 - D. Stool guaiac
 - E. Endocervical cultures
5. A 3-year-old male presents with recent onset of colicky abdominal pain, vomiting, and irritability. You strongly suspect intussusception, but plain radiographs of the abdomen do not suggest

- obstruction or free air. What is the next appropriate study to order?
- Air contrast enema
 - Computed tomography of the abdomen
 - Abdominal ultrasound
 - Upper gastrointestinal series
 - pH probe
- A 4-year-old male presents with a rash to his buttocks and legs; he also complains of joint pain. On examination, you note the characteristic rash of HSP. There is no history of diarrhea or bloody stools. Which test(s) should you recommend?
 - Inflammatory markers
 - Radiographs of the affected joints
 - Complete blood count
 - Coagulation panel
 - Urinalysis
 - After months of abdominal pain, a 7-year-old female gets relief after being given an enema. Her mother is not satisfied with your diagnosis of constipation because her daughter has been having diarrhea. How do you explain this symptom?
 - Concurrent UTI
 - Acute gastroenteritis
 - Chronic laxative use
 - Encopresis
 - Lactose intolerance
- choice because it can be diagnostic and therapeutic. Abdominal ultrasound is highly sensitive and specific for intussusception; it should be considered as an initial study, for atypical cases, and when an experienced radiologist is not available to perform and air contrast enema. In very equivocal cases, an abdominal ultrasound may be considered.
- E. A late sequela of HSP is nephritis, which may present with hematuria or proteinuria at the onset of HSP. Patients with significant findings on urinalysis should be followed closely for development of complications.
 - D. The child's symptoms stem from leakage of liquid stool around an impacted stool, known as encopresis. This symptom can often be a barrier to the parent's acceptance of constipation as the true source of a child's abdominal pain.

ANSWERS

- E. Bilious emesis in a newborn infant must be considered malrotation with midgut volvulus until proven otherwise. This is the gold standard test because it can show whether or not there is a malrotation of the intestine.
- E. Strep species are the most common cause of spontaneous bacterial peritonitis in children with nephrotic syndrome. Children with dialysis catheters are at risk for a broader range of pathogens, including staphylococcus species.
- C. An abdominal CT scan delivers between 10 and 20 millisieverts of radiation depending upon the protocol. This is about 100 times more than a chest radiograph, which delivers about 0.16 millisieverts of radiation.
- B. Ultrasound could be used to evaluate the appendix and, most importantly in this case, the ovaries. A Doppler ultrasound study can also establish whether there is inadequate blood flow to the ovaries due to torsion.
- A. In the patient where intussusception is strongly considered, air contrast enema is the study of

72 GASTROINTESTINAL BLEEDING

Cristina M. Estrada

INTRODUCTION

Gastrointestinal (GI) bleeding is a common and anxiety-provoking experience for both parents and children. Although the exact incidence of GI bleeding in children is unknown, hematemesis and hematochezia are common emergency department complaints. In healthy children, most GI bleeding is minor and self-limited, but occasionally the bleeding can be severe and even life threatening.

GENERAL APPROACH TO GASTROINTESTINAL BLEEDING

- Rapid assessment and stabilization.
- Secure the airway if there is profuse bleeding with aspiration of blood, or decreased mental status from blood loss.
- Place a nasogastric tube to decompress the stomach and minimize vomiting.
- Assess circulation, and if compromised place two large-bore IVs and push isotonic fluid boluses or blood products.

HISTORICAL FEATURES

- Ask about the color, timing, and volume of bleeding to accurately assess the volume of blood loss.

- Associated symptoms such as abdominal pain, preceding vomiting, fever, and stool patterns are also helpful.
- Some patients have a history of conditions such as coagulopathy, liver, or bowel disease that may put them at risk for GI bleeding.
- Inquire about medications such as NSAIDs, corticosteroids, or anticoagulants that are known to increase the risk for bleeding.

THE PHYSICAL EXAMINATION

- Pulse rate and quality, capillary refill, and skin color help determine if a patient has experienced a hemodynamically significant bleed.
- Examination of the nose and pharynx may reveal a non-GI bleeding source.
- Perform an abdominal examination to assess for signs of liver disease: jaundice, hepatosplenomegaly, or ascites.
- Perform a rectal and anal examination to assess for a lower GI bleeding source.

DIAGNOSTIC TESTING

- Complete a guaiac test to confirm the presence of blood in vomit or stool.
- Check a hematocrit to assess for anemia and platelets to check for thrombocytopenia.
- Liver and kidney function may be required in some patients as well as tests of coagulation such as prothrombin time and partial thromboplastin time.
- Perform a type and screen if there is excessive or continued blood loss.
- Gastric lavage can be helpful in determining if blood is upper GI in origin, but cannot rule out continued bleeding, particularly if bleeding is duodenal in origin.
- Upper and lower endoscopies can identify a bleeding source.

FACTITIOUS BLEEDING

- All that is red is not necessarily blood.
- Ingestion of red food dyes, medications such as cefdinir or rifampin, and foods such as beets or blueberries can all color the vomitus and stool.
- Bismuth salicylate, iron, and spinach can also turn stools very dark and be mistaken for melena.
- False positive guaiac tests may be seen if patients have ingested red meat or peroxidase-containing fruits and vegetables such as cauliflower, broccoli, turnips, or radishes.

TABLE 72-1 Causes of Upper Gastrointestinal Bleeding (by Age)

AGE	SMALL VOLUME	LARGE VOLUME
Newborn	Gastritis Stress ulcers Esophagitis	Ingested maternal blood Vitamin K deficiency A-V malformation Bleeding disorders/DIC
Infant	Gastritis Esophagitis Mallory–Weiss tear	Peptic ulcer disease A-V malformation
Child	Gastritis Esophagitis Foreign body Mallory–Weiss tear	Esophageal varices Peptic ulcer disease A-V malformation

UPPER GASTROINTESTINAL BLEEDING: OVERVIEW

- Defined as occurring proximal to the ligament of Treitz
- Carries a higher mortality rate compared to lower GI bleeding.
- Originates in the esophagus, stomach, or duodenum.
- Table 72-1.

UPPER GASTROINTESTINAL BLEEDING: ESOPHAGUS

- Gastroesophageal reflux is the most common cause of esophageal inflammation and may present with regurgitation, abdominal or chest pain, cough, and food refusal.
- Esophageal infections from *Candida*, cytomegalovirus, and herpes occur more commonly in immunocompromised children, and if bleeding occurs, it is typically small in volume.
- Lacerations of esophageal mucosa (Mallory Weiss tears) can occur in patients with repeated retching, vomiting, or paroxysmal cough.

UPPER GASTROINTESTINAL BLEEDING: STOMACH/DUODENUM

- Gastritis and peptic ulcer disease are common etiologies for upper GI bleeding.
 - Fig. 72-1.
- Medications such as NSAIDs, corticosteroids, and iron are known to increase the risk for gastric and duodenal inflammation.
- Infection with *Helicobacter pylori* is a common cause of gastric and duodenal ulcers in healthy children.



FIG. 72-1. Deep crater peptic ulcer seen in the fundus of the stomach on endoscopy. (Photo courtesy of Dr. Brian Riedel, Pediatric Gastroenterology, Vanderbilt University Medical Center.)

- Critically ill children are particularly at risk for development of “stress gastritis,” and prophylaxis with acid-suppressing drugs is essential.
- Vascular malformations are a rare but important cause of GI bleeding and can be present anywhere along the GI tract.
- The Dieulafoy malformation results from bleeding submucosal vessels occurring in the stomach without overlying ulceration.
- Although ingestion of foreign bodies is a common event in pediatrics, rarely this can result in upper GI bleeding.

LOWER GASTROINTESTINAL BLEEDING: OVERVIEW

- Lower GI bleeding is a common complaint, encompassing 0.3% of pediatric emergency department visits; however, most causes are relatively benign and self-limited.
- Lower GI bleeding can present as melena or hematochezia.
 - Melena typically indicates a more proximal source and occurs when blood has been present in the GI tract for a prolonged period of time, resulting in breakdown of hemoglobin.
 - Small volume hematochezia is typically from the distal colon or anus, although large volume rectal bleeding can result from lesions any place along the GI tract if bleeding is brisk.
- Lower GI bleeding can occur in the small intestine, colon, rectum, or anus.
- Table 72-2.

TABLE 72-2 Causes of Lower Gastrointestinal Bleeding (by Age)

AGE	SMALL VOLUME	LARGE VOLUME
Newborn	NEC Anal fissure Volvulus Duplication	Ingested maternal blood DIC AVM
Infant	Allergic colitis Enterocolitis Nodular lymphoid hyperplasia Duplication Intussusception Anal fissure Rectal prolapse	Meckel’s diverticulum AVM
Child	IBD Hemorrhoid HSP Toxic megacolon Enterocolitis	Juvenile polyp Meckel’s diverticulum AVM

LOWER GASTROINTESTINAL BLEEDING: INTESTINE

- Enterocolitis occurs among children of all ages and can result in abdominal pain, fever, and bloody stools.
- Common pathogens implicated in bloody diarrhea include *Salmonella*, *Shigella*, *Campylobacter jejuni*, *Yersinia enterocolitica*, *Escherichia coli*, *Clostridium difficile*, and *Entamoeba histolytica*.
- Necrotizing enterocolitis (NEC) is a rare but serious cause of lower GI bleeding in young infants often with a history of prematurity, significant anoxic stress at birth, or cyanotic congenital heart disease.
- Children with Hirschsprung’s disease can develop toxic megacolon and present acutely ill with abdominal distension, fever, explosive diarrhea, hematochezia, and abdominal pain.
- Intussusception most commonly occurs in children younger than 2 years and typically presents with intermittent colicky abdominal pain and vomiting, although some children present with only generalized illness and malaise.
 - Intussusception can occur anywhere within the bowel, but most commonly at the ileocecal junction.
 - Bleeding from an intussusception is described as “currant-jelly,” and occurs late in the course, after bowel ischemia has occurred.
- Volvulus can present with rectal bleeding from bowel ischemia.
 - It is most common in neonates and typically presents with bilious vomiting, abdominal distension, and feeding problems.

- Meckel's diverticulum occurs when the omphalomesenteric duct is incompletely obliterated early in fetal life.
 - Two percent of the population has this 2-inch long diverticulum within 2 ft. of the ileocecum.
 - Patients typically present before age 2 and are well appearing with painless rectal bleeding.
 - Meckel's diverticulum can also act as a lead point for intussusception.
- Juvenile polyps are a common cause of GI bleeding outside the neonatal period.
 - Bleeding is typically painless, recurrent, and small in amount.
 - Most polyps in children are solitary, benign, and occur within the left colon, often in the rectosigmoid region.
 - Fig. 72-2.
- Inflammatory disorders of the intestine are also a common cause of lower GI bleeding.
 - Infants most commonly have milk protein allergic colitis.
 - These infants are well appearing, but present with a history of bloody stools and occasionally vomiting or failure to thrive.
 - Cow milk and soymilk are the most commonly implicated allergens, although in a significant number of infants the allergen is unknown.



FIG. 72-2. A 0.7-cm pedunculated polyp was identified in the sigmoid colon of a child presenting with painless rectal bleeding. (Photo courtesy of Dr. Brian Riedel, Pediatric Gastroenterology, Vanderbilt University Medical Center.)

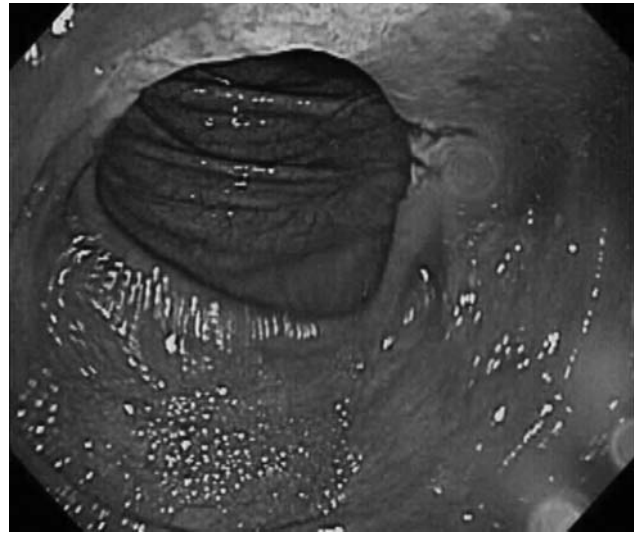


FIG. 72-3. Edema, friability, and ulceration of colon seen on endoscopy in a child with ulcerative colitis. (Photo courtesy of Dr. Brian Riedel, Pediatric Gastroenterology, Vanderbilt University Medical Center.)

- Inflammatory bowel disease including ulcerative colitis and Crohn's disease commonly results in rectal bleeding.
 - Fig. 72-3.
 - Inflammatory bowel disease often mimics other causes of colitis, causing crampy abdominal pain, frequent bloody stools, tenesmus, and weight loss.
 - While ulcerative colitis typically only involves the colon, Crohn's disease can involve any portion of the GI tract.
- Henoch-Schönlein Purpura (HSP) is a common immune-mediated vasculitis, which can involve the entire GI tract.
 - Children with HSP often have GI manifestations including abdominal pain, vomiting, and bloody stools or melena.
 - Most commonly, bleeding is due to mucosal hemorrhage, but intussusception is also common among children with HSP.
- Colonic lymphonodular hyperplasia from protein allergy or infection can result in asymptomatic, small volume bleeding.

LOWER GASTROINTESTINAL BLEEDING: RECTUM/ANUS

- The most common cause of rectal bleeding in infants is an anal fissure.
 - These infants typically pass a painful, hard stool with bright red blood seen on the surface of the stool.



FIG. 72-4. Rectal prolapse was seen on examination of this child presenting with rectal pain and bleeding. (Photo courtesy of Dr. Thomas J. Abramo, Pediatric Emergency Medicine, Vanderbilt University Medical Center.)

- Hemorrhoids are uncommon in young children but can occur in constipated adolescents, and may present with blood on the outside of stool.
- Young children with constipation are also predisposed to rectal prolapse, which may result in scant rectal bleeding.
- Fig. 72-4.
- Trauma from sexual abuse may also present as rectal bleeding.

CONCLUSIONS

- GI bleeding is a common complaint in the pediatric emergency department and may result in considerable anxiety for families but is only rarely associated with significant morbidity or mortality.
- Although the etiology can sometimes be challenging to identify, close attention to the volume, timing, and color of the bleeding as well as other associated signs and symptoms will often point to a diagnosis.

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QUESTIONS

1. A 3-year-old previously healthy child is brought to the emergency department for bloody stools. Mother reports two episodes over the last 24 hours. On examination, the child appears well with normal vital signs. Examination is consistent with hematochezia. Which of the following is can cause lower GI bleeding?
 - A. Foreign body
 - B. Esophagitis
 - C. Gastritis
 - D. Mallory Weiss tear
 - E. Meckel's diverticulum
2. A 6-year-old female presents with red stools which tests guaiac positive. False positive guaiac tests may be seen if patients have ingested peroxidase-containing fruits and vegetables. Which of the following vegetables may cause a false positive guaiac test?
 - A. Broccoli
 - B. Cabbage
 - C. Cucumbers
 - D. Squash
 - E. Sweet potatoes
3. A 3-year-old child is brought to emergency department for abnormally colored stools. Upon examination, the child is playful, appears well hydrated, and has normal vital signs. In taking a dietary history, which of the following foods would provide a possible explanation?
 - A. Blueberries
 - B. Carrots
 - C. Eggplant
 - D. Plums
 - E. Tomatoes
4. A 6-year-old presents to the emergency department with fever, abdominal pain, and bloody stools. Many infections in otherwise healthy children do not require treatment, and in some cases, treatment

- with antimicrobials may worsen the clinical course. Treatment with appropriate antimicrobials is recommended, however, for patients with
- C jejuni*
 - E coli*
 - Rotavirus*
 - Salmonella*
 - Y enterocolitica*
- A child presents with abdominal distension, fever, explosive diarrhea, hematochezia, and abdominal pain. Toxic megacolon can occur after surgical resection of the aganglionic segment, particularly in children with the following syndrome:
 - Down's syndrome
 - Noonan's syndrome
 - Short gut syndrome
 - Sweet syndrome
 - Turner's syndrome
 - A 4-year-old child presents to the emergency department with abdominal pain and currant jelly stools. An x-ray reveals an obstructive pattern. The most common cause of lower GI bleeding from an obstruction is
 - Bowel duplication
 - Intussusception
 - Meckel's Diverticulum
 - Polyp
 - Volvulus
 - A 2-year-old child presents with painless rectal bleeding that is described as "brick colored." Which of the following statements is true regarding features of a Meckel's Diverticulum?
 - It affects 20% of the population.
 - It consists of a 2-inch long diverticulum.
 - It occurs within 2 inches of the ileocecum.
 - Endoscopy is diagnostic.
 - Treatment involves banding.
 - A 3-week-old male presents with acute onset of rectal bleeding. The most common cause of rectal bleeding in an infant is
 - Anal fissure
 - Hemorrhoids
 - Lymphonodular hyperplasia
 - Milk protein allergy
 - Sexual abuse
 - A. Broccoli. Cauliflower, broccoli, turnips, and radishes may all cause false positive guaiac tests, as these are peroxidase-containing fruits and vegetables.
 - A. All that is red is not necessarily a GI bleed. Foods such as beets or blueberries can all color the vomitus and stool. Spinach can also turn stools very dark and be mistaken for melena.
 - A. Treatment with appropriate antimicrobials is recommended for patients with documented *Shigella*, *Campylobacter*, *C difficile*, and *E histolytica* infections.
 - A. Toxic megacolon can be the presentation of Hirschprung's disease, but can also occur after surgical resection of the aganglionic segment, particularly in children with longer segment disease or Down syndrome. Intestinal dilatation with air-fluid levels is often seen on plain abdominal x-ray, often with an intestinal cutoff sign (abrupt cutoff of intestinal distension at the pelvic brim). Treatment involves bowel decompression, hydration, and broad-spectrum antibiotics.
 - B. The most common cause of bleeding from obstruction is intussusception. Intussusception most commonly occurs in children younger than 2 years. These children typically present with intermittent colicky abdominal pain and vomiting, although some children present with only generalized illness and malaise. Intussusception can occur anywhere within the bowel, but most commonly at the ileocecal junction. A sausage-shaped abdominal mass may be palpated in the right lower quadrant or anywhere along the ascending or transverse colon, depending on the extent of bowel telescoping. Bleeding from an intussusception is described as "currant-jelly," and occurs late in the course, after bowel ischemia has occurred. Abdominal radiographs may show a "target sign" or paucity of bowel gas in the right lower quadrant, but may also be normal. Ultrasound is a common modality to diagnose intussusception. Air-contrast enema can be both diagnostic and therapeutic for intussusception, but complications such as failure to reduce the intussusception or bowel perforation can occur, and surgical consultation may be warranted.
 - B. Meckel's diverticulum occurs when the omphalomesenteric duct is incompletely obliterated early in fetal life. Two percent of the population has this 2-inch long diverticulum within 2 ft of the ileocecum. If the diverticulum contains ectopic gastric mucosa, ulceration and massive bleeding can occur. Patients typically present before age 2 and are well appearing with painless rectal bleeding. A radionuclide scan with technetium-99 m pertechnetate is diagnostic. This radioisotope binds preferentially to

ANSWERS

- E. Lower GI bleeding can occur anywhere from the ligament of Treitz to the anus. A-V malformations, Intussusception, Meckel's diverticulum, and polyps are all sources of lower GI bleeding.

gastric mucosa. Treatment involves surgical consultation and resection.

8. A. The most common cause of rectal bleeding in infants is an anal fissure. These infants typically pass a painful, hard stool with bright red blood seen on the surface of the stool. Diagnosis is made on physical examination, and treatment involves dietary modifications or medications to soften the stool.

73 GASTROESOPHAGEAL REFLUX

Jamie N. Deis

Thomas J. Abramo

INTRODUCTION

Gastroesophageal reflux (GER) is the most common esophageal disorder in children of all ages and a frequent reason for visits to the pediatric emergency department. It occurs when gastric contents pass into the esophagus through transient relaxation of the lower esophageal sphincter (LES). While the pathophysiology of GER in infants, children, and adults is similar, the symptoms and clinical presentation can be quite different (Table 73-1).

GASTROESOPHAGEAL REFLUX IN INFANTS

- Occurs in up to 67% of healthy infants by 5 months of age.
- Contributing factors: immaturity of the esophagus and LES, short length of the abdominal esophagus, and liquid diet.
- Great majority of infants are relatively unaffected
 - Gain weight appropriately.
 - Outgrow symptoms by 12 months of age.
- A small number of infants develop complications of reflux, known as gastroesophageal reflux disease (GERD).

CLINICAL PRESENTATION OF GERD IN INFANTS

- Excessive crying, irritability, arching, sleep disturbance, feeding aversion, and poor weight gain.
- Respiratory symptoms: chronic cough, stridor, wheezing, and laryngospasm.
- Apparent life threatening events (ALTE) with cyanosis.

TABLE 73-1 Common Symptoms of Gastroesophageal Reflux

INFANTS	OLDER CHILDREN AND ADOLESCENTS
Regurgitation	Abdominal pain
Feeding aversion	Heartburn
Excessive crying	Chest pain
Irritability	Dysphagia
Arching	Chronic cough
Sleep disturbance	Wheezing
Cough, wheezing, stridor	Sinusitis
Apnea or ALTE	

DIFFERENTIAL DIAGNOSIS OF REFLUX IN INFANTS

- Important to distinguish reflux from true vomiting.
- Reflux does not involve forceful muscle contraction.
- Vomiting involves forceful expulsion of stomach contents by contraction of the abdominal and chest wall muscles.
- Differential diagnosis of vomiting in infants is shown in Table 73-2.

EVALUATION OF REFLUX IN INFANTS

- Most cases are diagnosed by history and physical alone.
- Extensive work-up is not indicated in the emergency department unless metabolic or anatomic abnormality is suspected.
- Upper GI is nonspecific for reflux but can identify anatomic abnormalities (vascular rings, webs, strictures, hiatal hernia, pyloric stenosis, intestinal atresias, and malrotation).

TABLE 73-2 Differential Diagnosis of Vomiting in Infants

Obstruction	Neurologic
Pyloric stenosis	Hydrocephalus
Esophageal web	Subdural hematoma
Duodenal web/atresia	
Malrotation	
Intussusception	
Infection	Allergic
Sepsis	Milk protein allergy
Urinary tract infection	Food allergy
Meningitis	
Otitis media	
Toxic/metabolic	Other
Inborn errors of metabolism	Overfeeding
Urea cycle defects	Post-tussive emesis
Lead poisoning	

TABLE 73-3 Complications of Gastroesophageal Reflux

ESOPHAGEAL COMPLICATIONS	PULMONARY COMPLICATIONS	ENT COMPLICATIONS
Erosive esophagitis	Apnea or ALTE	Sinusitis
Esophageal stricture	Chronic cough	Otitis media
Barrett's esophagus	Asthma	Laryngitis
Adenocarcinoma	Aspiration pneumonia	Dental erosions

TREATMENT OF REFLUX IN INFANTS

- Conservative therapy is treatment of choice for mild reflux
 - Smaller feeding volumes
 - Frequent burping
 - Prone positioning upright at 45° to 60° after feeds
 - Thickening feeds with cereal
- If medications are needed, H₂ receptor antagonists (famotidine and ranitidine) are treatments of choice. Excellent safety profile.
- Use of prokinetic agents like metoclopramide is declining due to side effects and lack of proven clinical efficacy.

REFLUX IN CHILDREN AND ADOLESCENTS

CLINICAL PRESENTATION

- May present with epigastric pain, heartburn, noncardiac chest pain, and dysphagia.
- Extraesophageal symptoms: chronic cough, recurrent otitis media, sinusitis, laryngitis, and stridor.

DIFFERENTIAL DIAGNOSIS OF REFLUX IN CHILDREN AND ADOLESCENTS

- Increased ICP, rumination, bulimia, intestinal obstruction, peptic ulcer disease, infectious esophagitis, eosinophilic esophagitis, pill esophagitis, and achalasia.
- Eosinophilic esophagitis (EE) due to allergic response to food antigens
 - Symptoms of EE similar to GER, but medications used to treat GER generally not effective in treating EE.
 - Diagnosis requires endoscopy with biopsy.

EVALUATION OF GER IN CHILDREN AND ADOLESCENTS

- Majority of cases can be diagnosed by history and physical alone.
- Empiric trial of acid suppression may be the most simple test.

- If anatomic abnormalities suspected, upper GI series may be helpful.
- Esophageal pH monitoring and endoscopy can be performed on an outpatient basis.
- Upper endoscopy is the test of choice to document mucosal injury and esophagitis.

COMPLICATIONS OF GER

- Most common in children with neurological impairment and swallowing dysfunction.
- See Table 73-3.

TREATMENT OF GER IN CHILDREN AND ADOLESCENTS

- Conservative therapy: weight loss, dietary modifications, elevating head of the bed during sleep.
- Pharmacologic therapy: H₂ blockers and proton pump inhibitors (PPIs).
 - RTCs in adults indicate that PPIs are superior to H₂ blockers.
 - Omeprazole and lansoprazole approved for use in children.
- Surgical therapy:
 - Indicated in patients with severe reflux who fail medical management and develop complications of GERD.
 - Fundoplication: stomach either partially or completely wrapped around the esophagus above the just above the gastroesophageal junction.

SUMMARY

- GER common in children of all ages.
- Most infants with GER can be managed with conservative therapy alone.
- Most infants will outgrow symptoms by 12 months.
- Older children with chronic GER may develop both esophageal and extraesophageal complications.
- Further evaluation is indicated when diagnosis is uncertain or when complications occur.

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QUESTIONS

- A 3-month-old female is brought in by her mother for evaluation of intermittent “spitting up” after feeds. You learn that the infant is taking 4oz per feed, having normal stools and urine output, gaining weight well, and is having small volume reflux after feeds that is the same color as her formula. She is afebrile and has a normal physical exam. You inform the mother that
 - Reflux is abnormal in a 3-month-old infant and that radiographs should be performed.
 - Urine studies should be performed to exclude a urinary tract infection.
 - Blood should be sent to the lab to assess for hypernatremia.
 - Reflux is very common in infants and that most infants outgrow symptoms by 12 months of age.
 - She should increase the feeding volume to decrease the episodes of reflux.
- A 2-month-old male with reflux cries during feeds and arches his back for up to 30 minutes after feedings. He is well between feedings. Conservative measures including smaller volume feeds, frequent burping, and upright positioning after feeds have not resulted in any improvement. The mother would like to discuss a trial of medication for her son’s reflux. You inform her that
 - There are no safe medications that can be used to treat her son’s reflux and that she should continue conservative measures.
 - H₂ receptor antagonists, such as famotidine and ranitidine, have an excellent safety profile and can be used to treat GERD in infants <6 months of age.
 - Prokinetic agents, like metoclopramide, have proven clinical efficacy in infants <6 months.
 - She should introduce solid foods into his diet as quickly as possible.
 - Erythromycin is the medical treatment of choice for GERD in young infants.
- A mother brings her 5-week-old son in for evaluation of forceful vomiting after every feed. He was diagnosed with reflux by his pediatrician last week but has lost 8oz since that time. You observe a feed. He takes the bottle eagerly but then has a large volume episode of nonbilious emesis 1 minute after the feeding. Your next step is to
 - Try another feed, encouraging the mother to feed him more slowly this time.
 - Obtain a blood culture and a CBC.
 - Change his formula to soy and send him home with close follow-up with his pediatrician.
 - Obtain a urine culture.
 - Place an IV, give IVF, check electrolytes, and order an abdominal ultrasound.
- An abdominal radiograph is performed on the infant in question 3. You would expect to see
 - A normal abdominal radiograph with normal bowel gas pattern.
 - A dilated, air-filled stomach with little distal air.
 - An appendicolith in the right lower quadrant.
 - A double bubble sign.
 - Crescent sign in the right upper quadrant.
- A 4-week-old female is brought in by her mother for evaluation of irritability. You learn that she is having dark green colored emesis after all feeds. On exam, she is ill appearing with a firm, distended abdomen. There is bile stained emesis on her shirt. Your next step in management is to
 - Order an abdominal x-ray
 - Try to feed her to see if emesis recurs

- C. Order an air enema
 - D. Establish IV access, place an NG tube to intermittent suction, and call the pediatric surgeon
 - E. Obtain a urinalysis
6. A 12-year-old male presents for further evaluation of vomiting. His mother states that he has been vomiting every morning before school for the past 3 weeks. Vomiting resolves by 9 am, and he has no further vomiting throughout the rest of the day. His father has a history of peptic ulcer disease, and his mother is worried he may be developing an ulcer as well. On exam, his abdomen is soft and nontender, and he has normal bowel sounds, but he has dysconjugate gaze on extraocular motility testing. Your next step in evaluation is
- A. Arrange for follow-up with the ophthalmologist in 24 hours.
 - B. Encourage the family to seek assistance from the school counselor about school avoidance.
 - C. Order a head CT.
 - D. Start an empiric trial of omeprazole.
 - E. Obtain an abdominal radiograph.
7. A 15-year-old male presents with painful swallowing, heartburn, and intermittent epigastric pain. A trial of famotidine has not resulted in any improvement. There is a strong family history of allergic symptoms, and you suspect eosinophilic esophagitis. You inform your patient and his family that
- A. The medicines used to treat reflux are very effective in treating EE.
 - B. Children with EE have high levels of circulating eosinophils but low levels of eosinophils in the esophagus.
 - C. EE can be diagnosed by blood tests.
 - D. EE can be diagnosed by endoscopy with biopsy.
 - E. Metoclopramide is an effective treatment for EE.
8. Your resident wants to know more about the use of PPIs in children. You inform her that
- A. PPIs are effective antireflux medications in children that decrease gastric acid secretion by activating the H⁺K⁺ ATPase pump.
 - B. All PPIs on the market are currently FDA approved for use in children.
 - C. Multiple case series have shown symptomatic and endoscopic improvement in children with GERD following use of PPIs.
 - D. PPIs are first line therapy for treatment of reflux in infants <6 months.
 - E. Randomized controlled trials in adults indicate that H₂ antagonists are superior to PPIs in healing esophagitis.

ANSWERS

1. D. Reflux is very common in infants, occurring in up to 67% of all infants by 5 months of age. Most infants with reflux are “happy spitters” and are relatively unaffected by it. This infant is feeding well, gaining weight and has a normal physical exam. Tests such as radiographs, urine studies, and blood work are not indicated unless anatomic or metabolic abnormalities are suspected. Increasing the feeding volume is not indicated as this may increase reflux.
2. B. H₂ receptor antagonists have an excellent safety profile and can be used to treat reflux in young infants. Use of prokinetic agents is declining due to lack of proven clinical efficacy and risk of side effects, including dystonic reactions. Solid foods should not generally be introduced until 4 months of age and should be introduced gradually (one new food every 1–2 weeks). Erythromycin is not the treatment of choice for GERD in young infants as there has been an association with pyloric stenosis.
3. E. This 5-week-old has projectile vomiting after every feed, weight loss, and dehydration. His age and symptoms strongly suggest pyloric stenosis. These infants frequently develop hypochloremic hypokalemic metabolic alkalosis due to persistent vomiting. Further feeding should be avoided because his vomiting will persist until surgical pyloromyotomy is performed to relieve the gastric outlet obstruction. The most important step in management is to provide IV hydration, check electrolytes, and order an abdominal ultrasound, which is the definitive test to document hypertrophy of the pylorus muscle. CBC, blood culture, and urine culture are not indicated, as he is afebrile with no signs of infection.
4. B. This infant has pyloric stenosis. An abdominal radiograph would likely show a distended air filled stomach with little distal air due to the gastric outlet obstruction caused by the hypertrophied pyloric muscle. An appendicolith would not be seen in an infant with pyloric stenosis but may be seen in a child with appendicitis. The classic double bubble sign is seen in infants with duodenal atresia. The crescent sign can be seen in children with intussusception.
5. D. This is an ill appearing infant with an acute surgical abdomen and bowel obstruction. The first step in management is to establish IV access and provide gastric decompression by placing an NG tube to intermittent suction. Additionally, the pediatric surgeon should be consulted early. These steps should be performed before further diagnostic studies are performed. This infant likely has malrotation with

a mid-gut volvulus. Air enemas are used to reduce intussusception in children without peritoneal signs. Intussusception is unlikely in a 5-week-old, and this child has a surgical abdomen with peritoneal signs so air enema would be contraindicated.

6. C. Early morning vomiting in the setting of new dysconjugate gaze is worrisome for increased intracranial pressure. A head CT should be performed to assess for an intracranial mass and signs of increased ICP. Follow-up with an ophthalmologist or an empiric trial of omeprazole without first ruling out an intracranial mass would be inappropriate. As his abdominal exam is normal, an abdominal radiograph would be unrevealing as well. If an MRI could be obtained in a timely fashion, this would be even more appropriate.
7. D. Medicine used to treat reflux and gastritis are not very effective in treating EE. Children with EE have high levels of eosinophils in the esophagus. EE can only be diagnosed by endoscopy with biopsy. Metoclopramide does not have proven clinical efficacy in treating EE.
8. A. PPIs decrease gastric acid secretion by inhibiting the H⁺K⁺ATPase pump. There are currently only two PPIs approved for use in children, omeprazole and lansoprazole. While PPIs have been used to treat infants with reflux, safety and efficacy have not been clearly established in infants <6 months. H₂ receptor antagonists have an excellent safety profile and are still first line therapy for treatment of reflux in infants <6 months. RCTs in adults indicate that PPIs are superior to H₂ antagonists in healing esophagitis and maintaining remission.

74 GASTROINTESTINAL FOREIGN BODIES

Philip H. Ewing

ETIOLOGY

- According to the American Association of Poison Control Centers Toxic Exposure Surveillance System, in excess of 92,000 pediatric foreign body ingestions occurred in 2003.
- 24 mm disk batteries are becoming a significant cause of morbidity and mortality. The diameter of these batteries is between that of a nickel and a quarter. Prompt removal within two hours of ingestion is recommended to prevent complications.

TABLE 74-1 Conditions Increasing the Risk of Esophageal Obstruction

Esophageal atresia/tracheoesophageal fistula
Esophageal stricture
Congenital esophageal stenosis
Esophageal web
Previous esophageal injury—button battery erosion, caustic ingestion
Cancer requiring radiation therapy to the head and neck

- Multiple magnets in different parts of the bowel may attract one another and trap the tissues from the bowel wall in between, causing irritation, ischemia, and erosion through the bowel wall.

ANATOMIC SITES FOR OBSTRUCTION

- Objects that are smaller than 5 cm are likely to navigate the GI tract without obstruction.
- Table 74-1 reviews conditions increasing the risk of esophageal abnormality.
- Objects trapped at the thoracic inlet may produce vague discomfort of the throat and chest, drooling, and coughing with gagging.
- Foreign bodies trapped in other parts of the esophagus may cause chest or abdominal discomfort, but are less likely to produce other pulmonary symptoms.
- Objects that have passed into the stomach are likely to pass through the rest of the GI tract without incident.
- Gastric bezoars may be composed of a conglomeration of substances that are difficult for the stomach to digest, for example, milk, hair, and pill fragments (iron, aspirin).
- Long or sharp objects may become lodged at the pylorus or in the duodenum where they may cause localized pain.
- The ileocecal junction is a common place for impaction of difficult to digest foods, such as seeds and nuts.

DIAGNOSIS

- Metal detectors can locate an ingested, metallic foreign body and can track their movement through the GI tract. Metal detectors can be used as a screening tool to determine whether a child that may require diagnostic imaging.
- Patients with esophageal pain and secondary airway symptoms require imaging of the soft tissues of the neck and chest.
- Chest radiographs to include the upper abdomen may suffice for a child with only throat or epigastric discomfort.

- Barium swallow may be helpful to help locate a non-radiopaque object that is causing chest or epigastric discomfort.

THERAPEUTIC OPTIONS

- The management of swallowed foreign bodies that pass the thoracic inlet is conservative for the vast majority of patients.
- An asymptomatic object should be left to pass through the GI tract at its own pace. This “watchful waiting” approach has been documented to be safe, although it is important to provide the caregivers with detailed discharge instructions describing signs and symptoms of intestinal obstruction.
- Sharp objects should always be retrieved if they are trapped in the esophagus or stomach. If a sharp object passes through the pylorus, it will probably continue through the GI tract without incident.
- There is no data to support the use of medical adjuncts to aid the passage of foreign body through the gastrointestinal tract, including glucagon, 5-HT agonists, and cathartics.
- For the rare occurrence of lead foreign bodies trapped in the stomach, proton pump inhibitors may be considered to reduce acid while arranging endoscopic removal.

PROCEDURAL/SURGICAL APPROACHES

- Blind procedural approaches to esophageal foreign bodies may increase risk of mechanical injury to the esophagus and its complications.
- The use of fluoroscopy-guided endoscopic forceps may be effective, but needs further study.
- If a foreign body cannot be removed in the emergency department, the patient should be referred to a surgeon or endoscopist for removal under sedation or anesthesia.
- Once a foreign body has passed into the duodenum, it is probably not accessible by non-invasive procedural methods, eg, endoscopy, and surgical intervention becomes the only option for symptomatic foreign bodies until they pass through the ileocecal junction.

SMALL BOWEL

- Foreign bodies impacted in the small bowel should be removed by a surgeon.
- “Stuffed” drug packets may leak or rupture, as they were not intended to protect the contents during passage through the gut; rather, the packets are stuffed

to hide evidence from law enforcement. Rarely is the amount of drug in a stuffed packet enough to be toxic.

- Body “packers” may be carrying a lethal amount of drug in secure small packets.
- Whole bowel irrigation and activated charcoal should be considered for both “stuffers” and “packers” to ensure that the ingested drugs are not absorbed.

COLONIC/RECTAL FOREIGN BODIES

- A surgeon should remove impacted objects that do not release with gentle traction or those that cannot be visualized.

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QUESTIONS

1. A 3-year-old male presents having swallowed a screw. He is asymptomatic and radiographs demonstrate that the screw has passed into his stomach. What is the maximum length foreign body that has been demonstrated to pass through the gastrointestinal tract without incident?
 - A. 0.5 cm
 - B. 1 cm
 - C. 5 cm
 - D. 10 cm
 - E. 50 cm
2. A father brings in his 20-month-old daughter who swallowed his watch battery immediately before their arrival; she has no respiratory distress but she is drooling continuously. Radiographs demonstrate

- that battery is at the thoracic inlet. Which is your first step?
- Glucagon
 - Watchful waiting
 - Extraction using a Foley catheter
 - Advancing the battery using bougienage
 - Surgical consultation
- A 2-year-old girl has been playing with her older sibling and their mother found her with multiple toys in her mouth. She is worried that her daughter may have swallowed one of the toys and so she comes to the emergency department. If she has swallowed more than one object, which would be the most worrisome?
 - Coins
 - Beads
 - Magnets
 - Blocks
 - Puzzle pieces
 - A 2-year-old boy is transferred from another hospital for care after he swallowed a coin earlier in the day. New radiographs demonstrate that the coin has not moved from the thoracic inlet where it was seen by the transferring physician. Which treatment option do you recommend?
 - Glucagon
 - 5-HT agonist
 - A proton pump inhibitor
 - Cathartics
 - Surgical consultation
 - Three days ago, you discharged a child who had swallowed a foreign body which was identified in the stomach on radiograph. She has been asymptomatic until today when she began to complain of right lower quadrant pain. How do you plan her management?
 - Cathartics
 - Consultation for endoscopy
 - Make her NPO and start IV fluids
 - Glucagon
 - Surgical consultation
 - C. Magnets may affix to one another with bowel trapped between. This could cause bowel ischemia, necrosis, and perforation. While multiple foreign bodies are more concerning than a single object from a technical standpoint, only magnets present an added danger to the patient.
 - E. No medical therapy has demonstrated efficacy in encouraging esophageal foreign bodies to pass. A PPI should be considered only if there is a foreign body made of lead that does not pass through the stomach.
 - E. Impacted foreign bodies in the small bowel are not amenable to endoscopy. The foreign body will likely need to be removed surgically.

75 LIVER AND GALL BLADDER DISEASE

Susan M. Scott

Ashley Kumar

HYPERBILIRUBINEMIA

- The patient's age and the type of hyperbilirubinemia, whether direct or indirect, are important factors in determining the cause and treatment.
- Hyperbilirubinemia indicates either increased production of or impaired excretion of bilirubin and is the result of numerous etiologies, some pathologic and some physiologic.
- Increased red blood cell destruction may lead to unconjugated hyperbilirubinemia.
- Impairment of bile secretion from the liver or excretion from the gallbladder may lead to conjugated hyperbilirubinemia.
- Hyperbilirubinemia within the first day of life is never normal.
- Hyperbilirubinemia within the first week of life can be concerning and serious bacterial infection must be considered. Prompt diagnosis and aggressive intervention is imperative.
- Hyperbilirubinemia in older children and adolescents is concerning for hemolysis, extra hepatic obstruction, or hepatocellular injury.

ANSWERS

- C. 5 cm foreign bodies have been shown to pass without incident, including sharp objects.
- E. Surgical consultation is required to remove the battery, which may cause tissue injury within as little as two hours of ingestion. Medications do not enhance the clearance of foreign bodies and watchful waiting is not appropriate when the foreign body is a battery. Blind procedural methods may cause further injury to the esophagus.

NEONATAL HYPERBILIRUBINEMIA, UNCONJUGATED

- The most common causes of indirect hyperbilirubinemia in the first week of life are physiologic jaundice, breast milk jaundice, and hemolysis.

- Physiologic jaundice
 - Self-limited
 - Bilirubin levels peak by day of life 3
 - Levels return to normal over two weeks
- Breast feeding jaundice
 - Bilirubin levels peak by 2–3 weeks.
 - May remain elevated for weeks to months.
 - Decreased hepatic excretion and intestinal resorption of unconjugated bilirubin.
- Hemolysis
 - Maternal-fetal blood group incompatibility.
 - Associated with high risk for kernicterus.
 - Prompt recognition and initiation of treatment important.
- Presence of jaundice with fever, lethargy, and/or poor feeding should alert physician to the possibility of infection and performance of sepsis workup should be considered
- Initial studies should include bilirubin levels with fractions, CBC, reticulocyte count and coombs test.
- Bilirubin level, chronological and gestational age, as well as clinical status are considerations for initiation of phototherapy.
- AAP guidelines for initiation of phototherapy in infants of 35 or more weeks of gestation are available at www.aap.org.
- Bilirubin level >20 mg/dl can be lead to neurotoxicity, encephalopathy and kernicterus which is associated with long-term impairment.

NEONATAL HYPERBILIRUBINEMIA, CONJUGATED

- Direct hyperbilirubinemia is defined by a direct concentration of >2 mg/dl or if the direct concentration is greater than 20% total bilirubin.
- Neonatal direct hyperbilirubinemia is never normal and is indicative of hepatobiliary dysfunction.
- Most common causes include biliary atresia, extrahepatic biliary obstruction, neonatal hepatitis, and alpha-1-antitrypsin deficiency.
- Any infant with direct hyperbilirubinemia requires consult with pediatric gastroenterologist to aid with diagnostic workup and management.

HYPERBILIRUBINEMIA BEYOND THE NEONATAL PERIOD

- Unconjugated hyperbilirubinemia
 - Most commonly caused by hemolysis including sickle cell disease, hereditary spherocytosis, and G6PD deficiency.

- If no hemolysis present consider primary liver dysfunction including viral and drug-related hepatitis.
- Less common causes include autoimmune hepatitis and Wilson's disease.
- Conjugated hyperbilirubinemia
 - Caused by biliary obstruction or hepatocellular injury including cholelithiasis, tumor, or choledochal cyst.

HEPATITIS

- Important to establish a diagnosis and etiology since postexposure prophylaxis is available for some causes.
- Clinical presentation includes malaise, anorexia, nausea, vomiting, fever, and abdominal pain followed by the onset of scleral icterus and jaundice.
- A high index of suspicion must be maintained by the clinician as these patients are often diagnosed with viral gastroenteritis.
- Serodiagnostic testing should be obtained for hepatitis A, B, C, CMV, and EBV with further testing as indicated.
- Management is supportive with the majority of patients recovering completely.

FULMINANT HEPATIC FAILURE

- Fulminant hepatic failure is an acute or chronically progressive event with the loss of vital hepatic functions resulting in hyperbilirubinemia, hypoglycemia, coagulopathy, hypoproteinemia, and altered mental status from encephalopathy.
- In neonates, it is thought to have an infectious (such as HSV) or metabolic (such as Wilson's disease) source.
- In children, fulminant hepatic failure is most commonly a complication of viral hepatitis or secondary to toxin (mushrooms) or pharmacologic exposure (acetaminophen, anticonvulsants).
- The majority of patients have nonspecific complaints including nausea, vomiting, anorexia, and abdominal pain, particularly right upper quadrant. As liver failure progresses, jaundice develops and is often the symptom prompting medical attention.
- With the onset of coagulopathy, bruising and bleeding may occur. Hemorrhage can be significant, especially if it results from gastric and esophageal varices from portal hypertension.
- Hypoglycemia is a common complication and serum glucose must be monitored closely.
- Liver enzymes and bilirubin levels may be elevated but can return to normal as the number of viable hepatocytes decreases.

- Markers of liver function including albumin, clotting studies, and ammonia, are abnormal and are useful indicators of hepatic function.
- Prognosis is based on the underlying cause of liver failure and stage of hepatic encephalopathy.
- Poor prognosis is associated with:
 - Jaundice >7 days prior to onset of encephalopathy
 - Prothrombin time >50 seconds
 - Serum bilirubin >17.5 mg/dL

BILIARY TRACT DISEASE

- Biliary tract abnormalities seen in infancy include biliary atresia, choledochal cyst, and a potentially life-threatening disorder associated with both, acute cholangitis
- Acute biliary tract disease in childhood includes cholelithiasis, cholecystitis, and hydrops of the gallbladder

BILIARY ATRESIA

- Biliary atresia is a cause of neonatal conjugated hyperbilirubinemia and occurs in 1:10,000 to 1:18,000 births in European Americans.
- Hyperbilirubinemia usually occurs within 2 to 3 weeks of age. Parents will report stools that are light yellow, gray, or acholic. Hepatomegaly is also present.
- Visualization of the biliary tract, by either ultrasound or nuclear medicine hepatobiliary studies (DISIDA) scan, can determine presence of obstruction and define abnormalities.
- Treatment is both supportive care and surgical correction.
- Consultation with a pediatric gastroenterologist is imperative.
- Acute cholangitis is a complication often associated with surgical correction of biliary atresia.
 - Results from an ascending biliary infection or obstruction.
 - Patients present with fever, worsening jaundice, and elevated serum transaminases.
 - Prompt recognition and institution of broad-spectrum antibiotics is imperative, as the morbidity and mortality associated with this disorder is significant.

CHOLEDOCHAL CYST

- Choledochal cysts are congenital anomalies of the biliary tract.
- An extrahepatic cause of conjugated hyperbilirubinemia from cholestasis.

- Patients present with recurrent episodes of jaundice, pancreatitis, and abdominal pain. A right-sided abdominal mass may be palpated.
- In addition to liver function tests, an ultrasound of the abdomen is indicated.
- Referral to a pediatric gastroenterologist and pediatric surgeon is indicated.

CHOLELITHIASIS AND CHOLECYSTITIS

- Acute cholecystitis is a relatively uncommon entity in children and indicates gallbladder inflammation in the presence of gallstones.
- Gallstones develop as a result of abdominal surgery, sepsis, necrotizing enterocolitis, parenteral nutrition, hemolysis, or disorders with enhanced biliary enterohepatic circulation such as cystic fibrosis or Crohn's disease.
- Acalculous cholecystitis or inflammation of the gallbladder in the absence of stones is more common.
 - Caused by bacterial infections (eg, *Streptococcus*, *Salmonella*, *Klebsiella*), viral infections, and parasitic infestations (eg, giardia, ascaris).
 - Classic clinical presentation is upper-right quadrant pain, fever, and leukocytosis.
 - Patients with adequate pain control with oral medications can be managed as an outpatient with surgical follow-up.
 - Febrile patients may need admission and IV antibiotics.

HYDROPS OF THE GALLBLADDER

- Gallbladder hydrops is an acute noninfectious process leading to an enlarged gallbladder without gallstones.
- Can occur as a complication of viral gastroenteritis, Kawasaki disease, streptococcal pharyngitis, mesenteric adenitis, or nephrotic syndrome.
- Patients will present with abdominal pain, elevation in liver enzymes, and possibly hepatomegaly.
- Ultrasound is diagnostic.

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QUESTIONS

1. An 8-year-old female presents with fever, shaking chills, and jaundice. Past history is significant for sickle cell disease. The most common etiology for this type of presentation is
 - A. Ascending cholangitis
 - B. Cholecystitis
 - C. Hepatitis
 - D. Pancreatitis
 - E. EBV infection
2. A 15-year-old female with cystic fibrosis presents to the emergency department (ED) with fever and abdominal pain. Vital signs are normal except for fever. On physical examination, she has equal bilateral breath sounds, right-sided splinting, and upper-right quadrant abdominal pain with deep palpation.

Laboratory studies are remarkable for leukocytosis, and hyperbilirubinemia. Which of the following do you suspect?

- A. Gastroenteritis
 - B. Cholecystitis
 - C. Hepatitis
 - D. Pancreatitis
 - E. Pneumonia
3. A 14-day-old infant presents to the ED with poor feeding, lethargy and low-grade fever. Vital signs: HR 180, RR 45, BP 70/45, T 37.2C. On examination, the patient has a poor suck, clear breath sounds bilaterally, RRR without murmur, abdomen distended but soft, jaundice to legs, refill 3 seconds, distal pulses 1–2+. The most important next step is
 - A. Ultrasound of liver
 - B. Serum glucose
 - C. IV bolus
 - D. Sepsis workup
 - E. Antibiotic administration
 4. A 1-week-old infant is referred to the ED by the primary care physician for follow-up of a bilirubin level obtained in the office the prior day. The mother states she thinks the nurse said it was 22. The infant appears normal except for significant jaundice including scleral icterus. Birth History is significant for gestational age of 35 weeks and poor feeding for the first two days. The infant was discharged on day of life #3 and mother reports intermittent poor feeding. The next best step is
 - A. Sepsis workup
 - B. Send bilirubin level with CBC
 - C. Establishment of IV for hydration
 - D. Begin phototherapy
 - E. Call lab to check level drawn yesterday
 5. A 12-year-old male presents with a 2-day history of vomiting, diarrhea, and fever. Dad was treating him at home until he noticed that his son's eyes were yellow just like when he had hepatitis as a toddler. On examination, the patient is well hydrated with scleral icterus and no hepatomegaly. Total bilirubin is elevated to 10 with a direct fraction of 6; liver function tests are normal. The differential diagnoses for this patient includes
 - A. Hepatitis
 - B. Choledochal cyst
 - C. Hepatic tumor
 - D. Cholelithiasis
 - E. B, C, and D
 6. A 7-year-old male with no significant past history presents with vomiting, diarrhea, and abdominal pain. On examination, you note that he is jaundiced.

- Mom states that he was at Boy Scout Camp in the country east of town when the symptoms began very suddenly after a nature walk. None of the other boys are ill. One of your concerns should be
- A. Infectious diarrhea
 - B. Acute intoxication from mushroom ingestion
 - C. Hemolysis from spherocytosis
 - D. Acute hepatitis
 - E. None of the above
7. A 3-week-old male infant presents for evaluation of jaundice. Mom reports no fever, vomiting, diarrhea, or poor feeding. The infant is exclusively breast-fed and eats for 20 minutes every 2 hours. An indirect bilirubin level at the primary care office 2 days ago was 16 mg/dl. A level in the ED is 18 mg/dl. The best advice to give this mother is
- A. Supplement with water
 - B. Supplement with formula
 - C. Stop breast feeding for several days
 - D. Return for repeat bilirubin in 24 hours
 - E. Recheck with primary care provider
- cystic fibrosis is at increased risk for enhanced enterohepatic circulation and malabsorption leading to the development of gall bladder disease.
3. B. The patient is at risk for many serious complications including sepsis, hyperbilirubinemia, metabolic disorders, liver disease all of which can put the patient at risk hypoglycemia from poor intake or abnormal metabolism. Once proper glucose level is established further workup can continue.
 4. D. The initiation of phototherapy is imperative in this patient to reduce the risk of neurotoxicity. In addition, the infant has several other risks factors including prematurity and poor feeding.
 5. E. With normal liver function tests, the possibility of hepatitis is unlikely unless the patient is in liver failure. The next step would be an ultrasound of liver and gall bladder and consultation with pediatric gastroenterology service.
 6. B. The absence of fever or illness in the other campers makes infectious etiology less likely. The diagnosis of hereditary spherocytosis is usually made as a young child. The location of the camp makes the ingestion of a wild mushroom possible.
 7. E. Breast-feeding jaundice is due to poor intestinal reabsorption of unconjugated bilirubin and is a benign disorder usually requiring no treatment. Indirect bilirubin levels peak in week 2–3, usually at levels less than 20 mg/dl. Cessation of breast-feeding or supplementation is not indicated in this well infant.

ANSWERS

1. A. With presence of sickle cell disease the patient is at increase risk for hemolysis and the development of cholelithiasis. The classic presentation for cholangitis is fever, leukocytosis, and upper-right quadrant pain (Charcot's triad).
2. B. With bilateral clear breath sounds and normal vital signs, pneumonia is unlikely. The patient with

76 DISORDERS OF GLUCOSE METABOLISM

Nicholas Furtado

INTRODUCTION

- Diabetic ketoacidosis (DKA) is a complex endocrine condition caused by an absolute or relative lack of insulin. It is characterized by hyperglycemia, dehydration, ketosis, and metabolic acidosis.
- Hypoglycemia is seen in newborns and young infants who are asymptomatic or may manifest nonspecific symptoms. Older children exhibit more classic symptoms of hypoglycemia, including sweating, tachycardia, tremor, anxiety, tachypnea, and weakness.

DKA EPIDEMIOLOGY

- The annual incidence of DKA in the United States ranges from 4.6 to 8 episodes per 1000 patients with diabetes.

DKA PATHOPHYSIOLOGY

- Lack of insulin and stress lead to increase in the levels of counter regulatory hormones and gluconeogenesis.
- Lipolysis occurs in fatty tissues, forming ketoacids.
- Hyperglycemia and ketoacidosis cause diuresis that results in dehydration and metabolic acidosis.

DKA CLINICAL MANIFESTATIONS

- Often insidious in onset, with slow progression of the illness.

- Symptoms of fatigue and malaise, nausea/vomiting, abdominal pain, polydipsia, polyuria, polyphagia, and significant weight loss.
- Altered mental status characterized by drowsiness, progressive obtundation and loss of consciousness.
- Acidosis: tachypnea or hyperventilation or deep, rapid, sighing respirations called Kussmaul's respirations.
- Dehydration: poor peripheral perfusion and delayed capillary refill.
- Fever is present if infection precipitated the episode.

DKA LABORATORY STUDIES

- Initial laboratory studies include a complete blood count, serum electrolytes, glucose, calcium, phosphorus, and serum acetone. An arterial blood gas and bedside tests for blood sugar and urine ketones can be done for rapid diagnosis of DKA. Blood and body fluid cultures for fever.
- An initial electrocardiogram can be performed to assess for T-wave changes.
- Definition of DKA (biochemical criteria)
 - Hyperglycemia: blood glucose >200 mg/dL
 - Venous pH <7.3 or bicarbonate <15 mmol/L
 - Ketonemia and ketonuria

DKA MANAGEMENT

- Treatment of DKA consists of rapid assessment, replacement of the patient's fluid and electrolyte deficit and reversal of the central pathophysiologic process by the administration of insulin.
- Perform a quick clinical assessment and bedside tests to confirm the diagnosis.
- Assess the level of dehydration.

- Assess the level of consciousness using the Glasgow coma scale.
- Perform an ECG.
- Airway management for obtunded or comatose patients. Oxygen at 100% concentration is to be administered to patients in respiratory or circulatory failure and shock.
- Continuous cardiac monitoring is to be used for assessment of T-wave changes.
- As soon as hemodynamic stability is achieved, the child should be transferred to an intensive care unit.

FLUID RESUSCITATION

- Children with DKA are at least 5% to 10% dehydrated.
- The initial fluid resuscitation is with normal saline at a dose of 10 to 20 mL/kg over 1 to 2 hours. After the initial bolus, the patient's cardiovascular status is reevaluated and a second bolus may be administered.
- Subsequently, the remaining fluid deficit should be replaced slowly over 48 hours with a solution of tonicity greater or equal to 0.45% saline with added potassium chloride, potassium phosphate, or potassium acetate.
- In addition to assessment of dehydration, calculation of effective osmolarity can guide fluid and electrolyte therapy.

$$\text{Serum osmolality (mOsm/L)} = 2[\text{serum Na (mEq/L)}] + \text{blood glucose (mg/dL)}/18.$$

INSULIN THERAPY

- Normalization of blood glucose and suppression of lipolysis requires low-dose, continuous, intravenous insulin infusion.
- The starting dose is 0.1 U/kg/h and this should continue till resolution of DKA (pH >7.3, bicarbonate >15 mmol/L).
- Occasionally, the infusion may have to be decreased to 0.05 unit/kg/h when there is marked sensitivity.
- The goal of therapy is to decrease the serum glucose by 75 to 100 mg/dL/h.
- When the serum glucose reaches 250 mg/dL, 5% glucose is added to the infusing fluid. If the serum glucose is dropping precipitously, a glucose solution of $\geq 10\%$ may need to be administered.
- When the serum glucose normalizes, metabolic acidosis improves, and serum ketones decrease to trace, the insulin infusion is discontinued, subcutaneous (SC) insulin is started and oral intake of liquids or solids begun.

- If low-dose IV insulin cannot be administered because of the circumstances, then SC or intramuscular intermittent doses of short- or rapid-acting insulin analog may be used.

POTASSIUM

- Children with DKA are potassium-depleted with a deficit of 3 to 6 mEq/L/kg.
- Hypokalemia is most common after several hours of rehydration. Both severe hypo- and hyperkalemia can cause life-threatening cardiac arrhythmias hence ECG monitoring is critical.
- Serum potassium levels should be checked every 2 to 4 hours.
- Replacement therapy is started once normal or low serum potassium is ensured and urine output is established.
- The usual dose of potassium is twice-daily maintenance or 3 to 4 mEq/kg per 24 hours provided as 40 mEq/L in the IV fluids, with half as potassium chloride and half as potassium phosphate. The maximum recommended rate of IV potassium is usually 0.5 mEq/kg/h.

SODIUM

- The osmotic diuresis usually induces sodium depletion in patients with DKA.
 - In DKA, both the hyperglycemia and hyperlipidemia cause pseudohyponatremia.
 - Corrected serum sodium should be used for monitoring changes during therapy. The formula for corrected serum sodium is as follows
- $$\text{Corrected serum sodium (mEq/L)} = \text{Measured Na} + 0.016 (\text{Serum glucose} - 100)$$
- Corrected serum sodium should not be allowed to drop faster than 10 to 12 mEq/L per 24 hours.

PHOSPHATE

- Depletion of phosphate during DKA occurs as a result of osmotic diuresis.
- Clinically significant hypophosphatemia can cause impaired cardiac function and insulin resistance.
- Supplementation is indicated if the serum level is <2 mEq/L and can be administered with potassium replacement as potassium phosphate alone or with potassium chloride.
- During phosphate replacement, monitor serum calcium for development of hypocalcemia.

ACIDOSIS

- The acidosis that is fundamental to DKA is usually reversible with fluid resuscitation and insulin therapy.
- Bicarbonate causes paradoxical CNS acidosis, rapid onset hypokalemia, and increase in serum osmolality and hence is not generally recommended for routine use in DKA.
- Despite these adverse effects and lack of clinical benefit, its cautious use may be considered in patients with severe acidosis (pH <6.9 or serum bicarbonate <5 mEq/L) and hyperkalemia, which are associated with insulin resistance and cardiac arrhythmias.
- If bicarbonate is considered necessary, administer 1 to 2 mEq/kg over 60 minutes.

DKA COMPLICATIONS

- Inadequate rehydration, hypoglycemia, hypokalemia, hyperchloremic acidosis, and cerebral edema.
- Hypoglycemia is common, especially in young diabetics, who are extremely sensitive to insulin and labile.
- Adjusting the insulin infusion and providing supplemental intravenous and oral glucose will successfully correct this.
- Hypokalemia occurs within several hours of initiation of therapy and can lead to arrhythmias. Treatment is with potassium replacement.
- Cerebral edema occurs in 0.5% to 0.9% of DKA patients and the mortality rate is 21% to 24%.
- The warning signs of cerebral edema are headache, slowing of the heart rate, change in neurological status, and rising blood pressure.

TREATMENT OF CEREBRAL EDEMA

- IV mannitol 0.5 to 1 g/kg over 20 minutes to be repeated if there is no response in 30 minutes.
- Fluid restriction by one-third.
- Hypertonic saline 3%, 5 to 10 mL/kg over 30 minutes.
- Elevate the head of the bed.
- Controlled ventilation to maintain a PCO₂ 32–36 mm Hg.
- After treatment of cerebral edema, a CT scan should be performed to rule out other causes of neurological deterioration such as thrombosis or hemorrhage.

DKA DISPOSITION

- All patients presenting with DKA as the initial presentation of diabetes are hospitalized at a center where a pediatric endocrinologist is available for consultation.
- Patients with severe acidosis are best treated in a pediatric intensive care unit for reasons of close monitoring and need for repeated blood sampling.
- Occasionally, children with recurrent and mild DKA, with good family support, may be treated in the emergency department (ED) and discharged and followed up as an outpatient in consultation with their endocrinologist.

HYPOGLYCEMIA

- Hypoglycemia is most common in early neonatal life and may reflect a maladaptive response to extrauterine life.
- Hypoglycemia is pathological when low blood glucose levels are recurrent and persistent leading to acute systemic effects and long-term neurological sequelae.
- In childhood and adolescence, hypoglycemia usually presents as a complication of aggressive treatment for insulin-dependent diabetes mellitus.

PATHOPHYSIOLOGY OF HYPOGLYCEMIA

- The homeostasis of glucose is maintained by a complex balance between the exogenous supply of food and the body's regulatory hormones.
- The definition of hypoglycemia varies with age 3–24 hours <40 mg/dL, over 24 hours <45 mg/dL and infants and children <50 mg/dL.
- Since glucose is the main energy substrate for the central nervous system, hypoglycemia causes autonomic and central nervous system dysfunction.

SIGNS AND SYMPTOMS OF HYPOGLYCEMIA

- Newborns and young infants may be asymptomatic or may manifest nonspecific symptoms.
- Older children exhibit more classic symptoms of hypoglycemia, including sweating, tachycardia, tremor, anxiety, tachypnea, and weakness.
- Neuroglycopenia, which is a condition of prolonged and severe hypoglycemia manifested on the CNS, can result in permanent neurologic sequelae.

DIAGNOSTIC EVALUATION OF HYPOGLYCEMIA

- Evaluation of an infant or child with hypoglycemia should include a detailed history of the past, including perinatal history, acute or recurrent symptoms and physical examination.
 - Perinatal history, birth weight, maternal diabetes, dietary relationship to acute symptoms: time after food or association with starvation.
 - Past history and family history: symptoms, mortality.
 - Toxic ingestion.
 - Growth and development.
- Signs and symptoms of the current episode.
- Physical examination
 - Anthropometrics: short stature, macrosomia, macroglossia, hepatomegaly, jaundice.
 - Midline defects: single central incisor, cleft lip/palate, microphallus, undescended testis.
 - Skin pigmentation due to adrenal insufficiency.
- Laboratory tests
 - A rapid screen for the plasma glucose level at the bedside.
 - A critical sample which is tested for repeat blood glucose and other important studies such as insulin, C-peptide, growth hormone, cortisol and glucagon levels.
 - Bedside urine test for ketones.

HYPOGLYCEMIA MANAGEMENT

- Maintenance of normal plasma glucose levels is a must for preserving CNS function.
- If the patient is alert, oral glucose 15 g, formula feeds, or juice can be given.
- If the patient is not alert, start an IV and take the critical blood sample simultaneously.
- In newborns, give 10% dextrose 2 mL/kg (0.2 g/kg) as a bolus, followed by infusion at 6 to 9 mg/kg/min.
- In children, give 10% dextrose at 5 mL/kg (0.5 g/kg) as a bolus, followed by continuous infusion at 6 to 9 mg/kg/min.
- If an IV line is not possible, then give glucagon 0.03 mg/kg (maximum dose 1 mg) subcutaneously.
- If the history, physical examination and laboratory tests suggest a specific cause for hypoglycemia, then the following drugs may be given as appropriate:
 - Growth hormone 0.1 mg/kg/dose.
 - Hydrocortisone 5 mg/kg/dose orally
 - Diazoxide 10 to 20 mg/kg/day orally
 - Somatostatin (Octreotide) 2 to 4 g/kg/d divided into 2 to 4 doses SQ/IV.
 - Carnitine 100 mg/kg/d.

HYPOGLYCEMIA DISPOSITION

- Admission of the patient is indicated when there is no obvious cause, toxic ingestion as with oral hypoglycemic agents is suspected, administration of long-acting insulin was the cause and if there are persistent neurological deficits.
- Discharge may be considered after a high carbohydrate meal if an obvious cause is found and treated with rapid reversal of symptoms. For all insulin-dependent diabetics with hypoglycemia, discharge should be coordinated with a pediatric endocrinologist and the child's family after appropriate adjustment of insulin dose.

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QUESTIONS

1. A 5-year-old boy has been admitted for DKA about 12 hours ago. Which of the following tests would be useful at this point?
 - A. Calcium concentration.
 - B. Venous pH value.

- C. White blood cell count.
 - D. Creatinine level.
 - E. Phosphorus concentration.
2. A 7-year-old girl with DKA, who has received intravenous therapy for 4 hours, develops a headache, drowsiness and dilated pupils. Which of the following would be appropriate treatment at this time?
- A. Elevation of the head of the bed.
 - B. Controlled ventilation.
 - C. Restriction of fluids.
 - D. Mannitol.
 - E. All of the above.
3. You are evaluating a 12-year-old girl who has a 5-year history of type 1 diabetes. She is known to be noncompliant with her insulin therapy. She complains of abdominal pain and she appears mildly dehydrated. A serum glucose level is 450 mg/dL. Her urinalysis is positive for glucose and ketones and she has a venous pH of 7.19. Of the following, the most appropriate initial management step is to
- A. Administer a bolus of 10 to 20 mL/kg normal saline.
 - B. Administer an intravenous bicarbonate infusion.
 - C. Begin an insulin drip at a rate of 0.5 U/kg per hour.
 - D. Obtain a glycosylated hemoglobin level.
 - E. Start two times maintenance fluid requirements with 1/2 normal saline and potassium.
4. You are managing a 14-year-old boy who has DK in the ED. He had an initial blood glucose level of 560 mg/dL and so far has received only a normal saline bolus. Which of the following statements regarding the further management of this patient is true?
- A. Bicarbonate should be added to the fluids if signs of cerebral edema develop.
 - B. Glucose should be added to the fluids once the blood glucose levels are 100 mg/dL .
 - C. Insulin initially should be administered subcutaneously as a combination of regular and intermediate acting forms.
 - D. Potassium should be added to the intravenous fluids only if the potassium levels decrease below 3.5 mEq/L.
 - E. The blood glucose should decrease by 80 to 100 mg/dL per hour.
5. You are evaluating a child with postprandial hypoglycemia. The child is afebrile. Which of the following findings is least likely on physical examination?
- A. Weakness
 - B. Tremor

- C. Pallor
- D. Bradycardia
- E. Cold sweat

ANSWERS

1. E. Phosphorus levels begin to decrease about 12 hours into treatment of DKA. Extremely low phosphorus levels can cause weakness and exacerbate coma. Calcium concentration is measured when phosphorus replacement is considered for low serum levels because of the risk for hypocalcemia. Blood pH, creatinine, and white count levels are monitored only if indicated by a worsening of the patient's condition, fever or decrease in urine output.
2. E. The 7-year old has developed cerebral edema secondary to rapid fluid shifts into the brain. This type of cerebral edema is osmotic in nature. Treatment of this cerebral edema is accomplished by elevation of the head of the bed, restriction of fluids, controlled ventilation and intravenous mannitol. Steroids are not useful to treat osmotic cerebral edema.
3. A. The first step in the treatment of DKA is always to correct hydration. Most children with DKA are 5–10% dehydrated; therefore, starting a bolus of normal saline is indicated. Intravenous bicarbonate is not routinely indicated in DKA, as it is associated with harm in patients with pH above 6.9. Without initiation of hydration, insulin infusions may be associated with rapid drop in blood sugar leading to fluid shifts across the blood brain barrier and possible cerebral edema. Glycosylated hemoglobin is only useful for the long-term management of diabetes mellitus. Starting maintenance fluids is indicated only after the correction of initial fluid deficits and may not offer sufficient hydration for the acute management of DKA.
4. E. The rate of blood glucose decrease should not exceed 80–100 mg/dL/hour because of increased risk of development of cerebral edema and hypoglycemia. Bicarbonate is not routinely indicated for the management of DKA. Glucose should be added to the fluids when the blood sugar is below 250 mg/dL. This prevents further ketosis. Initial insulin therapy is usually rapid acting insulin. Potassium is added to the fluids only after ensuring normal renal function and urine output and usually if the potassium level is below 5 mEq/L.
5. D. During hypoglycemia, the signs and symptoms are the result of increased adrenergic activity, such as tachycardia, pallor, tremor, and cold sweats. Bradycardia is rarely seen during hypoglycemia.

77 ADRENAL INSUFFICIENCY

Nicholas Furtado

INTRODUCTION

- The adrenal cortex produces two main hormones: glucocorticoid (cortisol) and mineralocorticoid (aldosterone). Adrenal insufficiency (AI) is a clinical state that results from the inability of the adrenal cortex to produce these hormones in response to stress.

PATHOPHYSIOLOGY

- Primary AI results from congenital or acquired adrenal gland dysfunction.
- Secondary and tertiary AI results from pituitary or hypothalamic under-function, respectively.
- Glucocorticoid deficiency impairs gluconeogenesis and glycogenolysis, and decreases the sensitivity of the vascular system to angiotensin II and norepinephrine.
 - This results in hypoglycemia, tachycardia, and mild hypotension.
- Aldosterone deficiency causes decreased sodium retention by the kidney, osmotic diuresis, hyponatremia, hypovolemia, and dehydration.
 - In addition, it causes a decreased distal renal tubular exchange of potassium and hydrogen ions for sodium ions, leading to hyperkalemia and acidosis.
- Androgen deficiency in primary AI leads to ambiguous genitalia and underdeveloped secondary sexual characteristics in prepubertal children.
- In addition, in primary AI, the lack of negative feedback from cortisol on the anterior pituitary causes over-secretion of ACTH and proopiomelanocortin that stimulates skin hyperpigmentation.

ETIOLOGY AND EPIDEMIOLOGY

- The commonest cause of AI in North America is the abrupt withdrawal of glucocorticoids while on chronic treatment.
 - Children who have been on glucocorticoid therapy for 2 to 4 weeks tend to have prolonged suppression of the hypothalamo-pituitary axis leading to secondary AI after treatment is stopped.
- The most common cause of primary AI in children is congenital adrenal hyperplasia (CAH), with an incidence of 1 in 10,000 to 18,000 live births.
 - CAH results from a deficiency in the enzymatic activity of one of the enzymes in the cor-

tisol biosynthetic pathway, the commonest being 21-hydroxylase deficiency.

- Mortality for CAH is five times that of the general population.
- Acquired causes of primary AI in children are less common than congenital disorders and result from autoimmune, infectious, infiltrative, hemorrhagic, or ablative disorders of the adrenal cortex.
- Secondary AI that is not because of pharmacologic glucocorticoid withdrawal can result from any process that interferes with the pituitary's ability to secrete ACTH, such as tumors, craniopharyngioma, infections, infiltrative diseases of the pituitary, lymphocytic hypophysitis, head trauma, and intracranial aneurysms.
 - Most of these coexist with other pituitary hormone deficiencies and there is history of pituitary insult or abnormality of the hypothalamopituitary axis on MRI.

CLINICAL PRESENTATION

- AI can present with vague and nonspecific symptoms.
 - As a result diagnosis is delayed in many cases.
- Acute insufficiency or Addison's crisis is typically encountered in a previously undiagnosed child who has been subjected to the stress of an acute illness.
 - Inadequate administration of stress steroid dosing in known cases or abrupt withdrawal in the context of prolonged steroid therapy may precipitate this illness.
- In all these clinical situations, the presentation is characterized by dehydration, hypotension, hypoglycemia, or altered sensorium.
- Hypoglycemia occurs most commonly in infants and young children.
- Physical clues to AI include hyperpigmentation of the face, neck, hands, areas subject to friction, such as elbows, knees, and knuckles, the buccal mucosa, areolae, or scars.
 - Other skin findings are vitiligo, secondary to autoimmune melanocyte destruction.
 - In the absence of history of steroid withdrawal, secondary AI is usually associated with signs of other pituitary hormone deficiencies such as growth failure, delayed puberty, secondary hypothyroidism, and diabetes insipidus.
 - In the rare case of missed neonatal screening for CAH, ambiguous genitalia may be present.

LABORATORY FINDINGS

- Laboratory findings and common abnormalities of AI are based on the etiology and pathophysiology.

- The diagnosis of primary AI is confirmed by the documentation of an elevated plasma ACTH level (>100 picogram/mL) and a low serum cortisol level (<10 g/dL).
- Mineralocorticoid deficiency is confirmed by documentation of low aldosterone levels with or without hyperkalemia and hyponatremia.
- Secondary AI is diagnosed by documenting simultaneously low blood ACTH and cortisol levels.

DIFFERENTIAL DIAGNOSIS

- The differential diagnosis of acute AI must include all causes of hyponatremia, hyperkalemia, ketotic hypoglycemia, and shock in children.

MANAGEMENT

- Recognizing adrenal crisis immediately requires a high index of suspicion and is warranted in children presenting with unexplained shock and hypoglycemia.
- The patient's airway should be stabilized, the child placed on continuous cardiac monitoring and rapid fluid resuscitation is started with 5% dextrose and normal saline.
- If sepsis is suspected, appropriate culture samples should be taken and antibiotics administered.
- If there is no previous history of AI, prior to specific treatment, a critical blood sample must be collected for ACTH, cortisol, aldosterone, and plasma renin activity.
- If CAH is suspected, 17-hydroxyprogesterone and androgen levels should also be requested.
- Correction of specific disturbances, such as hypoglycemia and hypokalemia, is implemented.
- Glucocorticoid replacement.
 - Hydrocortisone 50–75 mg/m² IV or IM.
 - Methylprednisolone 10–15 mg/m² or dexamethasone 1.5–2 mg/m² are comparable but not ideal because of poor mineralocorticoid effect.
 - Prednisone is not used for acute therapy.
- Mineralocorticoid replacement is achieved with fludrocortisone (Fluorinef) 0.05–0.1 mg PO when the patient is able to tolerate oral fluids.
- Maintenance cortisol and mineralocorticoid therapy.
 - Hydrocortisone 9–12 mg/m²
 - Florinef 0.05–0.2 mg/day

DISPOSITION

- All patients presenting to an emergency department (ED) in acute adrenal crisis must be admitted

to the pediatric intensive care unit for continued parenteral fluid replacement and steroid maintenance therapy.

- A pediatric endocrinologist must be consulted for further inpatient evaluation, management and continuity of care.
- Children with known AI and mild symptoms may be managed as an outpatient with consultation and close follow-up with their pediatric endocrinologist.

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QUESTIONS

1. A 3-day-old female infant is brought to the ED. Her mother reports that she has decreased activity and is bringing up most of her feedings for the last 24 hours. On examination, you notice significant dehydration and clitoromegaly. Which of the following is

- the most appropriate step in the initial management of this patient?
- Immediately order a karyotype study.
 - Rapidly establish IV access and give a bolus of normal saline, 20 mL/kg.
 - Collect a critical blood sample and order ACTH, cortisol, aldosterone and plasma renin, as well as a complete metabolic profile.
 - Administer broad-spectrum antibiotics.
 - Consult a pediatric endocrinologist prior to initiating treatment.
- A 3-week-old infant is brought to the ED for evaluation of decreased feeding. The infant has a diagnosis of CAH. He has ambiguous genitalia and classical hyperpigmentation. Patients with CAH produce too little _____ and too much _____.
 - Epinephrine, norepinephrine
 - Androgens, cortisol
 - Cortisol, aldosterone
 - Cortisol, androgen
 - None of the above
 - A 2-week-old male infant is brought to the ED for persistent vomiting. On examination, you notice the infant to be hyperpigmented and virilized. What is the critical laboratory abnormality that will need treatment on an emergent basis?
 - Hyponatremia
 - Hyperkalemia
 - Hyperglycemia
 - Hyperchloremia
 - Hypomagnesemia
 - A 4-week-old infant is brought to the ED for a 1-week history of progressively increasing vomiting. The emesis is nonbilious, nonprojectile and there is no abdominal distension. He is moderately dehydrated but stable. He is unable to keep down oral fluids. You start an IV line, send labs and start basic fluid resuscitation. You suspect adrenal insufficiency. Which of the following laboratory values are most consistent with that diagnosis?
 - Hyperkalemia, hypernatremia, and hyperchloremia
 - Metabolic alkalosis and hypokalemia
 - Hemo-concentration
 - Low BUN and dilute urine
 - Hyperkalemia, hyponatremia, and hypoglycemia
 - A 9-year-old girl who has nephritic syndrome has been given glucocorticoid therapy in the form of prednisone 20 mg daily for a year. She requires an emergency surgery. The most appropriate approach to the immediate management of her adrenocortical status is
 - Administer adrenocorticotrophic hormone 1 unit/kg intramuscularly preoperatively and repeat every 12 hours for the next 48 hours.
 - Cover possible glucocorticoid and mineralocorticoid deficiencies by administering calculated physiologic replacement doses of both hormones.
 - Determine adequacy of adrenal function by measuring the cortisol/creatinine ratio on a spot urine sample to guide replacement needs.
 - Give a methylprednisolone preparation 1 mg/kg IV preoperatively and every 6 hours for the next 48 hours.
 - Substitute treatment with an aerosolized glucocorticoid in twice the usual dose preoperatively and repeat every 3 hours for the next 48 hours.
 - A 5-year-old girl is on chronic steroid therapy for chronic lung disease. She developed a fever 1 day ago and her parents report that for the last 24 hours she has been vomiting and not able to keep down her usual doses of medication. On examination in the ED, you notice that she is in early shock and you diagnose her to be in adrenal crisis. After establishing IV access and correcting the shock, what is the appropriate fluid for replacement of her deficit?
 - Normal Saline solution.
 - Ringers Lactate solution.
 - 10% Dextrose solution.
 - 5% Dextrose in 0.5N Saline.
 - Distilled water.

ANSWERS

- B. The most likely diagnosis is CAH, which presents in an emergency room usually due to symptoms of acute adrenal insufficiency. The most important immediate step to be taken is to establish IV access and correct dehydration and associated metabolic derangements. Karyotyping is not useful in the acute setting. A critical blood sample must be collected before specific treatment is initiated, but is not essential before initiation of fluid resuscitation. Antibiotics are administered early on if sepsis is suspected, but again after fluid resuscitation is begun. An endocrinology consult may be helpful but should not delay initial resuscitation.
- D. CAH is due to enzyme deficiencies in the adrenal cortex leading to decrease in cortisol production. Precursors of cortisol production are shunted toward increased production of androgens and hence virilization is a common clinical sign. Epinephrine and norepinephrine are products of the adrenal medulla and therefore are unaffected in CAH.
- B. Male infants with CAH usually present with hyperpigmentation and virilization. This infant has a severe deficiency and thus presented in the early neonatal period with pernicious vomiting due to metabolic instability. The most critical abnormal-

ity is hyperkalemia, which can lead to cardiac arrhythmias and mortality. Hyponatremia is usually corrected slowly along with correction of dehydration. Hyperchloremia, hyperglycemia, and hypomagnesemia are usually not seen in CAH.

4. E. Because of the effect of low cortisol, the triad of hyperkalemia, hyponatremia, and hypoglycemia is most commonly seen in acute adrenal insufficiency. Hypochloremia may also be seen. Metabolic acidosis is common, as are anemia and eosinophilia. Prerenal azotemia is also common.
5. D. The patient who has been on glucocorticoid therapy for over 4–6 weeks will have significant adrenocortical suppression. When subjected to a stressful event like major surgery, her adrenal cortex is unlikely to respond and produce adequate levels of cortisol. Therefore, stress doses of glucocorticoids are indicated for this patient. ACTH will not stimulate a suppressed adrenal cortex. Physiologic doses and inhaled glucocorticoids are not effective for treatment of acute adrenal insufficiency.
6. D. AI is usually accompanied by hypoglycemia. Hence, after correction of the hemodynamic instability and shock with the use of normal saline boluses, the recommended IV solution is D5 0.5NS over 24 hours. Ringers lactate solution contains potassium and is not used in resuscitation routinely. 10% Dextrose alone and distilled water will result in excess free water and exacerbate the electrolyte disturbances that are seen in AI and are not recommended for resuscitation.

78 HYPERTHYROIDISM

Nicholas Furtado

INTRODUCTION

- Hyperthyroidism is a state of increased production and secretion of thyroid hormones resulting in the hypermetabolic clinical syndrome of thyrotoxicosis.
- The term thyroid storm refers to an extreme state of decompensated thyrotoxicosis and is a thyroid emergency that can potentially be fatal.

EPIDEMIOLOGY

- The most common cause of hyperthyroidism in children is Graves' disease.
 - This disease occurs in 1 in 5000 children with a peak incidence between 11 and 15 years of age.
 - The male to female ratio is 1: 5.1.

- Although the true incidence of childhood thyrotoxicosis is unknown, 5% of all thyrotoxicosis occurs in childhood and 0.6% to 10% of neonates born to mothers with Graves' disease will show signs of thyrotoxicosis.
- The reported mortality in neonatal thyrotoxicosis is as high as 20%.
- Because childhood hyperthyroidism occurs mostly in adolescents, thyroid storm also occurs more frequently in this group.

PATHOPHYSIOLOGY

- Thyrotoxicosis results from thyroid hormone excess either caused by overproduction of thyroid hormone by the thyroid gland or by administration of synthetic hormone.
- Increased concentration of serum free thyroid hormone is almost always found in thyrotoxicosis.
- In Graves' disease, activated B-lymphocytes produce antibodies against antigens shared by the thyroid gland and eye muscle.
- Thyrotropin receptor-stimulating antibodies (TRSAb) bind to TSH receptors to increase thyroid hormone production.
- In congenital hyperthyroidism, transplacental transfer of TRSAb from the mother with Graves' disease stimulates the thyroid gland to cause hyperthyroidism or thyrotoxicosis.
- The actions of thyroid hormone at the cellular level include calorogenesis, acceleration of substrate turnover, amino acid and lipid metabolism, and stimulation of water and ion transport.
- Thyroid hormones also activate the adrenergic system by up-regulation of β -adrenergic receptors causing symptoms of sympathetic nervous system overactivity, including hyperthermia.
- In thyroid storm, the clinical manifestations of thyroid hormone excess are thought to be because of an uncoupling of oxidative phosphorylation secondary to the illness, resulting in an enhanced rate of lipolysis, with fatty acid oxidation, increased oxygen consumption, calorogenesis, and hyperthermia.
- Specific conditions, such as thyroid surgery, withdrawal of antithyroid medications, radioiodine therapy, palpation of a generous goiter and iodinated contrast dyes, are known to precipitate thyroid storm in patients with hyperthyroidism.

ETIOLOGY

- The causes of thyrotoxicosis may be divided into conditions in which the source of excess thyroid hormone is endogenous or exogenous.

- The most common disorder causing thyrotoxicosis in children, as in adults, is the autoimmune disorder, Graves' disease.
- In 5% to 10% of thyrotoxicosis, the cause is autoimmune thyroiditis or hashitoxicosis.
- Autonomously functioning thyroid nodules (toxic adenoma) are sometimes encountered in children.
- Rarely, hyperthyroidism is secondary to TSH oversecretion from a pituitary tumor or because of isolated pituitary resistance to negative feedback control by thyroid hormones on a genetic basis.
- The possibility of a molar pregnancy, which produces a thyroid-stimulating hormone, must be considered in adolescent females with thyrotoxicosis.
- Administration of iodine-containing medications, such as dyes, to patients with a nodular goiter may rarely induce hyperthyroidism in Hashimoto's thyroiditis, endemic goiter, multinodular goiter, and nontoxic diffuse goiter.
- Finally, thyrotoxicosis can occur as the result of intentional excess thyroxine or triiodothyronine intake or it can be iatrogenic because of overtreatment.

CLINICAL PRESENTATION

- Children who present with thyrotoxicosis have symptoms of nervousness, palpitations, weight loss, muscle weakness, and fatigue.
- A history of developmental delay and declining school performance may be found.
- Other symptoms include tremulousness, anxiety, excessive sweating, temperature intolerance, and emotional lability.
- Gastrointestinal over-activity with symptoms of frequent stools is common.
- An increased appetite is classically present. However, an apathetic state, including decreased appetite, occasionally occurs.
- The signs of Graves' disease are similar to those seen in adults but the ophthalmologic signs are usually milder in children.
- Signs of sympathetic and cardiac over-activity are common.
 - These include tremor, brisk deep tendon reflexes, tachycardia, hyperactive precordium, and a widened pulse pressure.
 - CHF may develop because of the inability of cardiac function to meet metabolic demands and papillary muscle dysfunction, causing mitral valve prolapse.
 - Except in neonates and children with underlying cardiac disease, CHF is uncommon in childhood thyrotoxicosis.

- In thyroid storm, severe hyperpyrexia, atrial dysrhythmia and CHF, delirium or psychosis, severe gastrointestinal hyperactivity and hepatic dysfunction with jaundice are present.
- A history of a precipitating event, illness or major stress should be identified.

DIFFERENTIAL DIAGNOSIS

- Conditions that cause tachydysrhythmias (atrial flutter, atrial fibrillation, and ventricular tachycardia) must be differentiated from hyperthyroidism.
 - These include electrolyte disturbances and cardiac disease.
 - The murmur of mitral valve prolapse in association with tachycardia may lead to a mistaken diagnosis of CHF and cardiac valvular disease.
- The patient who is febrile and appears "toxic" may have sepsis alone or as a precipitating factor in thyroid storm.
- Intoxication with adrenergic and anticholinergic drugs may also mimic the hyper-metabolic state seen in thyrotoxicosis.
- Gastrointestinal hyperactivity may imitate an acute abdomen in thyroid storm.

MANAGEMENT

- Treatment of severe thyrotoxicosis or thyroid storm is directed at preventing further thyroid hormone synthesis, alleviation of the acute peripheral effects of excess thyroid hormone and general supportive measures.
- Initial laboratory tests should include the measurement of total and free T₄, T₃, and TSH levels, along with a complete metabolic profile.
- Blockade of thyroid hormone synthesis should be initiated and continued until the crisis resolves: PTU at a dosage of 175 mg/m²/d or 4 to 6 mg/kg/d divided and given at 6- or 8-hour intervals or 200 mg every 4 hours or methimazole 30 mg every 6 hours.
- Blockade of release of thyroid hormone: start 1 to 3 hours after antithyroid medication is initiated. Sodium iodide 0.05 mg IV every 12 hours, Lugol's solution (5% iodine) 3 to 5 drops orally every 8 hours or Lithium 600 mg oral loading dose followed by 300 mg every 6 hours (do not use with CHF, renal failure or arrhythmia).
- Inhibition of peripheral T₄ and T₃ conversion: dexamethasone 2 mg IV followed by 2 mg orally every 6 hours.
- β -adrenergic antagonists: propranolol 10 to 20 mg orally every 8 hours in children and adolescents.

- Hyperthermia: acetaminophen, cooling blankets and ice packs. Salicylates must be avoided because they can displace thyroid hormone from binding sites, potentially worsening the hypermetabolic state.
- Fluid resuscitation: normal saline, 20 mL/kg, is administered; then the fluid deficit is calculated and replaced in the form of half normal saline with 5% dextrose over the next 24 to 48 hours.
- Cardiovascular complications: arrhythmias and CHF are treated with antiarrhythmics, digoxin, and diuretics.
- In all cases, the precipitating event causing severe thyrotoxicosis must be sought and treated.

TREATMENT OF NEONATAL THYROTOXICOSIS

- Neonatal hyperthyroidism is characterized by growth failure, microcephaly and wide-eyed stare.
 - Tachycardia and irritability are usually seen a few days after birth.
 - Occasionally, the onset can be delayed by weeks.
- Neonatal thyrotoxicosis is usually seen in newborns whose mothers are on antithyroid drugs.
- Antithyroid treatment: PTU 5 to 10 mg every 8 hours, iodide drops (sodium or potassium salt) one drop every 8 hours orally.
- β -adrenergic blockade: propranolol 2 mg/kg/d every 12 hours.
- Supportive measures: treatment of CHF, arrhythmia, airway management, fluid, and caloric replacement.

DISPOSITION

- Treatment and disposition of children with thyrotoxicosis should always be undertaken in consultation with a pediatric endocrinologist.
- Children with severe thyrotoxicosis, thyroid storm, and those with cardiovascular complications, such as arrhythmia, CHF and shock, should be admitted to a pediatric intensive care unit for further management.

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QUESTIONS

1. The mother of a 14-year-old girl brings her to the ED because her daughter has become progressively restless and irritable over the last several weeks and she has also reported palpitations and a rapid pulse. No abnormalities are found on physical examination of the patient except for undue restlessness, a pulse of 128 beats/min and a fine tremor. The thyroid gland is of normal size and consistency. There are no signs of ophthalmopathy. Of the following findings, which would rule out a diagnosis of Graves' disease *most* conclusively?
 - A. Absence of ophthalmopathy.
 - B. A low level of TSH.
 - C. A low uptake of radioactive iodine.
 - D. Presence of antithyroid antibodies.
 - E. Frequent stools.
2. The findings noted for the patient described above are *most* suggestive of
 - A. Hashimoto thyroiditis.
 - B. Ingestion of thyroid hormone.
 - C. Pituitary adenoma.
 - D. Thyroid adenoma.
 - E. Thyroid storm.
3. A 13-year-old girl presents with a nontender, asymmetrically enlarged thyroid gland. She is otherwise asymptomatic. Her growth and development have been normal. The laboratory results are: normal T4 concentration, mildly elevated TSH level and moderately elevated antimicrosomal and antithyroglobulin antibody titers. The *most likely* diagnosis is
 - A. Chronic lymphocytic thyroiditis.
 - B. Graves' disease.
 - C. Autoimmune polyglandular syndrome, type II.
 - D. Colloid goiter.
 - E. Subacute thyroiditis.

ANSWERS

1. C. In Graves' disease, uptake of radioiodine is usually high due to overstimulation of the thyroid gland. In children with Graves' disease, the presence of ophthalmopathy is very variable and can be completely absent in many cases. Low TSH and the presence of circulating thyroid-stimulating antibodies typically characterize Graves' disease, as does frequent stools.
2. A. A smooth thyroid gland, low TSH, increased peripheral T4 levels and low radioiodine uptake typically characterize Hashimoto's thyroiditis. Ingestion of thyroid hormone would not cause low radioiodine uptake. Pituitary adenoma produces TSH in excess; therefore circulating levels of TSH would be high. Thyroid adenoma would be diagnosed by a hot spot on the nuclear scan with surrounding low uptake and also an adenoma is sometimes picked up on the clinical examination.
3. A. Chronic lymphocytic thyroiditis or Hashimoto's thyroiditis is the commonest thyroid disease in childhood. These patients can be transiently hyperthyroid, like patients with subacute thyroiditis, but are usually euthyroid like this patient. In Graves' disease, T4 is highly elevated and the thyroid stimulating antibody titres are elevated. Subacute thyroiditis usually causes transient hyperthyroidism secondary to viral infections. Autoimmune polyglandular syndrome is usually characterized by Graves' disease associated with other autoimmune diseases, such as diabetes mellitus, and occurs at an earlier age.

79 RICKETS*Carla Minutti***INTRODUCTION**

- Rickets is defined as the failure of osteoid to calcify in a growing person. Osteoid is the collagen-containing organic matrix of the bones.
- Rickets results from vitamin D deficiency or the abnormal metabolism of vitamin D.
- Vitamin D deficiency is currently a highly prevalent condition among infants, children, and adolescents in the United States, especially in minority ethnic groups and immigrants.
- Vitamin D deficiency is the major cause of rickets around the world.
- The majority of the reported cases occur in individuals with dark skin and breast-fed infants who do not receive vitamin D supplementation.
- Vitamin D deficiency in childhood and rickets continue to be public health problems in the Middle East, North Africa, and some parts of Asia.
- Vitamin D (cholecalciferol or vitamin D₃) is formed in the skin under the stimulus of ultraviolet (UV) light. UV light was the only significant source of vitamin D until ergosterol (vitamin D₂), contained in fish liver oil, was introduced as a dietary supplement in 1918.

ETIOLOGY

- The causes of rickets are summarized in Table 79-1.

VITAMIN D DEFICIENCY

- Studies suggest that vitamin D intake in adults is inadequate to prevent a state of deficiency.
- Deficiency in adults is defined by most experts as a level less than 20 ng/mL (Table 79-2).
- Among infants and young children, vitamin D deficiency has been defined as a level below 11 ng/mL. However, children with skeletal abnormalities characteristic of vitamin D-deficient rickets have been found to have levels between 11 and 15 ng/mL.
- The definition for adult vitamin D deficiency (<20 ng/mL) is being increasingly applied for children.
- Recent studies have shown that vitamin D deficiency and insufficiency are highly prevalent among pregnant women, even when prenatal vitamins are being administered.
- Infants born to and breast fed by vitamin D deficient mothers can have a relative vitamin D deficiency resulting from decreased maternal transfer and have a higher risk of developing rickets in infancy and childhood.

DECREASED SUNLIGHT EXPOSURE

- In comparison to sunlight, diet provides less than 10% of the body's vitamin D requirements.
- A lightly pigmented adult, with 10 to 15 minutes of full body sunlight exposure during summer, will generate between 10,000 to 20,000 IU of vitamin D in a day.
- Many organizations in recent years have launched major public health campaigns to decrease the

TABLE 79-1 Etiology of Rickets

Vitamin D Deficiency
Dietary deficiency
Maternal vitamin D deficiency (congenital rickets)
Breast-fed infants
Infants and children who are fed macrobiotic diets
Deficient Endogenous Synthesis
Deficient exposure to UV light
Sunscreen use
Skin pigment
Pollution
Season/latitude
Decreased Bioavailability
Gastrointestinal tract disorders
Severe intestinal malabsorption
Obesity (sequestration of vitamin D in body fat)
Disorders of Vitamin D Metabolism
Decreased synthesis of calcidiol
Hepatobiliary disease
Accelerated catabolism of calcidiol
Use of anticonvulsants: phenobarbital and phenytoin
Hereditary
Vitamin D-resistant (absent or abnormal receptors)
Vitamin D-dependent (defect in 1 α -hydroxylase)
Acquired
Chronic renal disease
Decreased synthesis of calcitriol
Calcium Deficiency
Nutritional deprivation
Hypercalciuria
Phosphate Deficiency
Nutritional deprivation or use of antacids
Hereditary
X-linked hypophosphatemic rickets
Hypophosphatemic rickets with hypercalciuria
Acquired
Sporadic hypophosphatemic osteomalacia
Oncogenic osteomalacia
Neurofibromatosis and fibrous dysplasia
Other Rare Causes
Primary mineralization defects
Defective osteoid
Toxicities, such as aluminum

incidence of skin cancer and photoaging by recommending the limitation of exposure to UV light.

- Many factors limit the amount of sunlight exposure for a given individual: season of the year, latitude, pollution, lifestyle or cultural practices and skin

TABLE 79-2 Vitamin D Levels in Adults^a

	25-HYDROXYVITAMIN D (CALCIDIOL)	
	ng/mL	nmol/L
Deficiency	<20	50
Insufficiency	21–29	—
Sufficiency	>30	75

^aAmong infants and young children, the Institute of Medicine and the AAP have defined vitamin D deficiency as a level below 11 ng/mL (27.5 nmol/L); however, the definitions described above are increasingly being applied to children.

pigment. This results in increased dependence on dietary vitamin D to maintain vitamin D sufficiency.

BREAST FEEDING AND FORMULA FEEDING

- All infant formulas, evaporated milks, and almost all whole milk sold in the United States contain 400 IU of vitamin D per liter.
- Breast milk contains only 12 to 60 IU of vitamin D per liter.
- Breast-fed infants should be started on daily supplementation with 200 IU of vitamin D within the first 2 months of life.
- If an infant is ingesting at least 500 mL/d of formula, he or she will receive the recommended vitamin D intake of 200 IU/d.

PATHOPHYSIOLOGY

- In the human body, vitamin D can be either exogenous or endogenous.
 - Exogenous vitamin D₂ (ergosterol) can be acquired through a healthy diet or vitamin supplements. Dietary sources of vitamin D include fatty fish and fortified food products, including dairy, infant formula, juice, and cereals.
 - Endogenous vitamin D₃ (cholecalciferol) is synthesized in the dermis and epidermis from 7-dehydrocholesterol by exposure to the UV light fraction of sunlight (Fig. 79-1).
 - UV rays convert 7-dehydrocholesterol to precholecalciferol. Precholecalciferol is then, through thermal isomerization, converted to cholecalciferol.
 - Excessive exposure to sunlight degrades any excess vitamin D preventing any intoxication.
 - Vitamin D (either D₂ or D₃) can be deposited in and then released from fat cells.
 - Both vitamin D₂ or D₃ are activated by hydroxylation at two different sites.
 - The first hydroxylation occurs mainly in the liver by vitamin D-25 hydroxylase, producing calcidiol (25-hydroxyvitamin D or 25-hydroxycholecalciferol). This step is substrate dependent and measuring this metabolite is used to measure vitamin D status.
 - The second hydroxylation occurs primarily in the kidney, where 25-hydroxyvitamin D is converted to the active metabolite calcitriol (1,25-dihydroxycholecalciferol or 1,25-dihydroxyvitamin D), a hormone, by the enzyme 1 α -hydroxylase. Activity of 1 α -hydroxylase is stimulated by parathyroid hormone, hypocalcemia, and

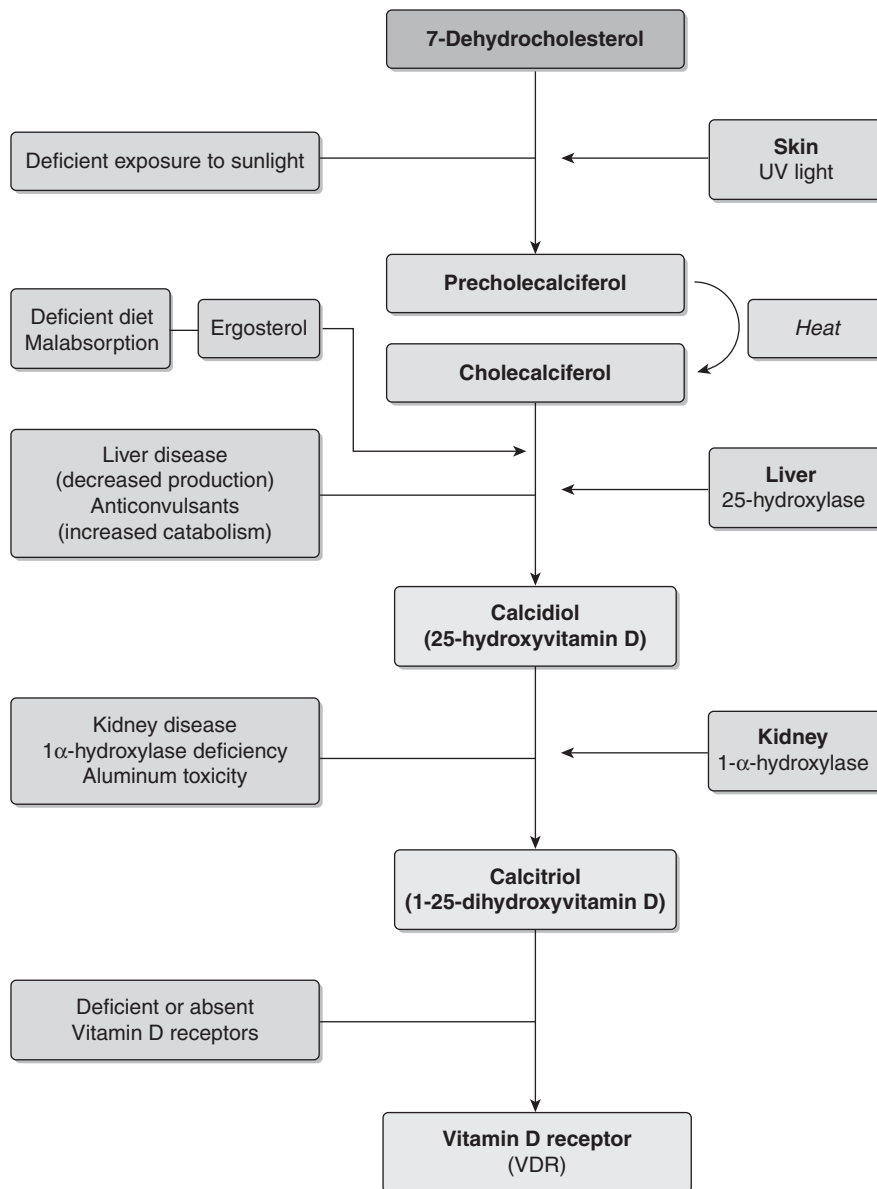


FIG. 79-1. Pathway of vitamin D production.

hypophosphatemia. Inhibitors of 1-hydroxylase are calcium, phosphate, and calcitriol. Deficient or underactive 1α -hydroxylase is the cause for vitamin D-dependent rickets.

CALCITRIOL ACTIONS

- Table 79-3

DIAGNOSIS

- Figure 79-2 provides an algorithm to guide the diagnosis of vitamin D-deficiency rickets.

CLINICAL MANIFESTATIONS

SIGNS AND SYMPTOMS

- Patients with rickets may present with hypocalcemic signs and symptoms.
 - Usually, signs of hypocalcemia are present when the ionized calcium concentration falls below 2.5 mg/dL.
 - Symptoms of hypocalcemia may include paresthesias, muscle cramping, carpopedal spasm, and laryngospasm with resultant stridor and apnea.
- Subclinical tetany may be induced by Chvostek's or Trousseau's signs.

TABLE 79-3 Actions of Calcitriol

BONE	SMALL INTESTINES
Increased mineralization (through calcium and phosphorus absorption)	Increased absorption of calcium
Increased osteoclastic activity (to release calcium and phosphorus)	Increased absorption of phosphorus
	Decreased absorption of magnesium
PARATHYROID GLANDS	KIDNEY
Decreased PTH synthesis and secretion	Autoregulation of calcitriol production
	Enhanced tubular reabsorption of calcium
	Decreased excretion of phosphorus
OTHER	
Decreases production of type I collagen	
Stimulates 24-hydroxylation of 25-hydroxyvitamin D	

- Cardiac contractility may be reduced, with prolongation of electrical systole as measured by the QT interval.
- Neurologic findings include seizures, irritability, memory loss, and affective disorders.
- Systemic findings observed in children with rickets may include hypotonia, muscular weakness, delay in walking, anorexia and increased susceptibility to infection, especially pneumonia.
- Site-specific findings of rickets are shown in Table 79-4.

LABORATORY EVALUATION

- In vitamin D-deficient rickets, calcium is low in the early and late stages, but it can be normal.
- The phosphorus level is low in the vast majority of cases.
- Alkaline phosphatase levels are almost always elevated.
- Calcidiol levels are low and PTH levels are elevated.
- Calcitriol levels are normal or high because of PTH activity.

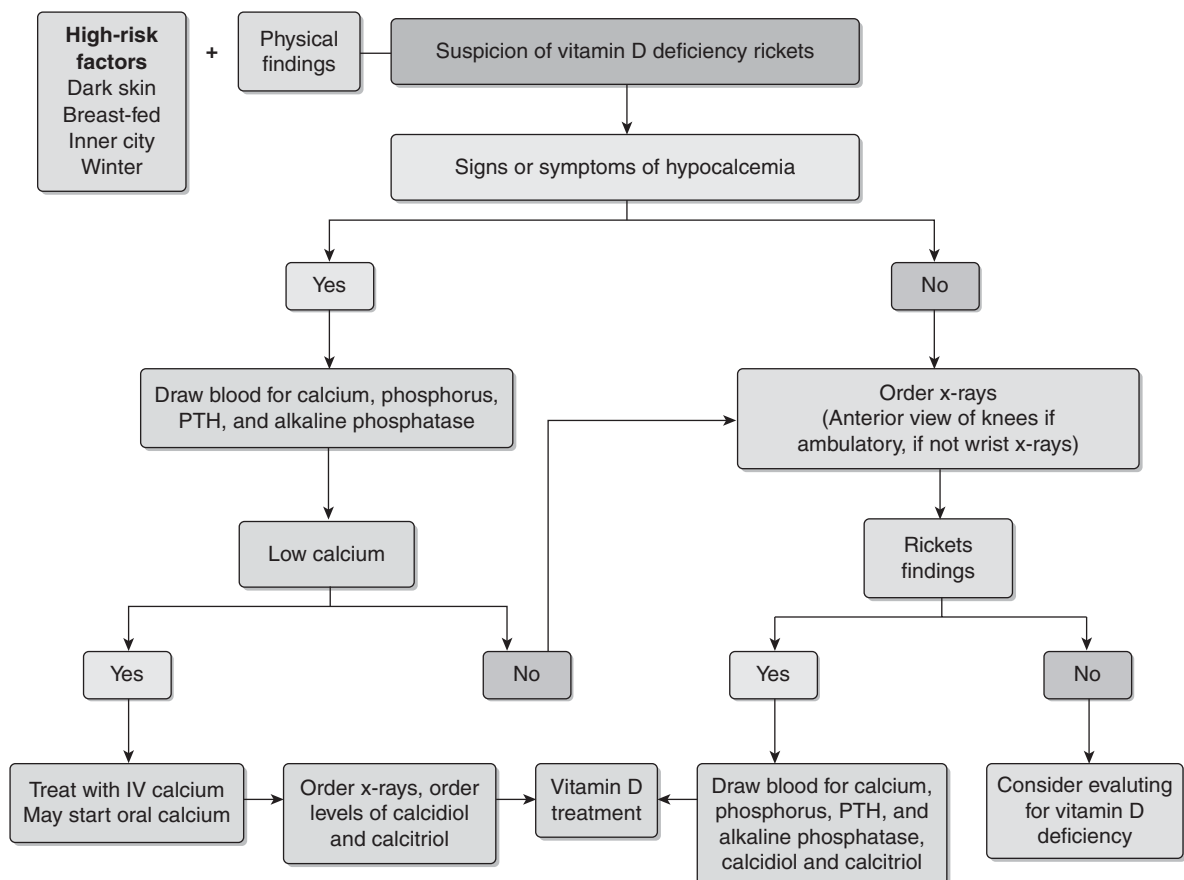
**FIG. 79-2.** Algorithm for diagnosis of vitamin D-deficient rickets.

TABLE 79-4 Site Specific Findings of Rickets

Head
Craniotabes (posterior flattening of the skull)
Frontal bossing and square forehead (caput quadratum)
Widened cranial sutures
Teeth
Delayed dental eruption
Enamel hypoplasia
Thorax
Rachitic rosary—bulging of costochondral junction
Prominent sternum
Harrison groove—indentation of the lower anterior thoracic wall
Arms
Bowing of the long bones
Thickening of the wrist at the level of the epiphysis
Legs
Bowing of the long bones
Genu varum, because of weight bearing
Anterior bowing of the tibia (saber shin deformity)
Knock-knees (genu valgum)
Thickening at the level of the ankle

- Calcidiol, calcitriol, and PTH levels may take days to be reported and will not be available during emergency department evaluation.
- Table 79-5 summarizes the laboratory evaluation of the suspected rickets patient.

IMAGING

- The most rapidly growing bones show the most striking abnormalities.
- The best radiograph for infants and children younger than 3 years is an anterior view of the knee showing the metaphyseal ends and epiphyses of the femurs and tibiae.
- Radiologic findings in rickets may include osteopenia with visible coarsening of trabeculae (Fig. 79-3) and cortical thinning.
- The metaphyses may show fraying, widening, and cupping because of their exaggerated normal concavity and irregular calcification (Figs. 79-3 and 79-4).

- When healing occurs, the metaphysis may have a brush border appearance (Fig. 79-4).
- Along the shaft, the uncalcified osteoid may cause the periosteum to appear separated from the diaphysis.

TREATMENT

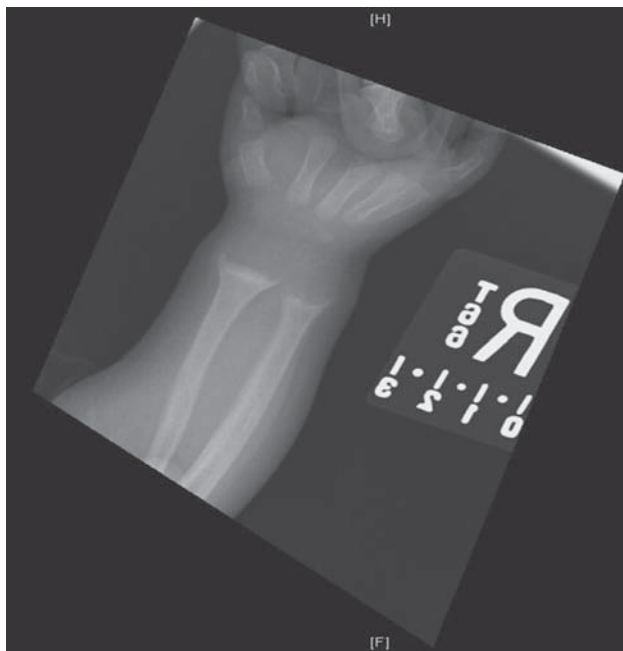
- Vitamin D is stored in the body and released gradually over many weeks.
- Treatment of vitamin D deficiency may be administered gradually or in a single day.
- Vitamin D can be given as a single oral dose of 300,000 to 600,000 IU or a single intramuscular injection at a dose of 600,000 IU.
- Vitamin D can be given as a monthly intramuscular injection of 10,000 to 50,000 IU of vitamin D for 3 to 6 months; oral vitamin D at a dose of 50,000 IU once a week for 8 weeks, followed by 50,000 IU every 2 to 4 weeks; or oral vitamin D at a dose of 1,000 to 2,000 IU/d for several weeks.
- Some sources recommend up to 4,000 IU daily in older children.
- Some authors also recommend 400 IU of vitamin D daily for 6 to 12 months after treatment for rickets.
- Calcium must be given along with vitamin D to avoid hypocalcemia because of “hungry bone” syndrome.
- Calcium is given at a daily dose of 40 mg/kg of elemental calcium.
- For symptomatic hypocalcemia, intravenous calcium may be administered cautiously.
 - Calcium gluconate 10% (50–100 mg/kg/dose) or calcium chloride 10% (10–20 mg/kg/dose) is infused slowly over at least 12 to 15 minutes.

MONITORING

- Serial measurements of alkaline phosphatase and appropriate x-rays are recommended to evaluate healing and resolution of the rachitic lesions.

TABLE 79-5 Rickets Because of Vitamin D Deficiency or Abnormal Metabolism of Vitamin D

LABORATORY EVALUATION	CALCIUM	PHOSPHORUS	CALCIDIOL	CALCITRIOL	PTH	ALKALINE PHOSPHATASE
Vitamin D deficiency	↓	↓	↓	N or ↓	↑	↑
Liver disease	↓	↓	↓	↓	↑	↑
Renal disease	↓	↑	N	↓	↑	↑
Vitamin D-dependent rickets (1 α -hydroxylase deficiency)	↓	↓	N	↓	↑	↑
Vitamin D-resistant rickets (abnormal or absent vitamin D receptor)	↓	↓	N	↑	↑	↑



A



B

FIG. 79-3. Frontal views (A and B) of the wrists show metaphyseal widening, cupping, and fraying consistent with rickets.

- Treatment is continued until healing is well established and the alkaline phosphatase concentration approaches the normal range.
- Serial blood samples for levels of calcium, phosphorus, PTH, 25-hydroxyvitamin D and urine calcium are not always recommended, but may be useful in specific cases.



FIG. 79-4. AP view of the knees show widening of the metaphysis. Brush border appearance of the metaphysis is consistent with healing rickets. There are unsharp, smudged-appearing trabecular markings consistent with osteomalacia.

PREVENTION

- To prevent vitamin D deficiency and rickets in healthy infants and children, the National Academy of Sciences recommends a supplement of 200 IU of vitamin D per day for the following:
 - Breast-fed infants unless they are weaned to at least 500 mL/d of vitamin D-fortified formula or milk, beginning during the first 2 months of life.
 - All nonbreast-fed infants who are ingesting less than 500 mL/d of vitamin D-fortified formula or milk, beginning during the first 2 months of life.
 - Children and adolescents who do not get regular sunlight exposure, do not ingest at least 500 mL/d of vitamin D-fortified milk or do not take a daily multivitamin supplement containing at least 200 IU of vitamin D.
- Vitamin D supplementation is critically important for breast-fed infants and for infants and children living in an inner-city area and those with increased skin pigmentation.
- These guidelines are based on data, which show that an intake of at least 200 IU/d of vitamin D will prevent physical signs of vitamin D deficiency and maintain serum 25-hydroxy-vitamin D at or above 11 ng/mL.
- Other organizations and individuals recommend a daily intake of 400 to 2,000 IU of vitamin D daily as a preventive dose, for infants through 18 years of age.

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QUESTIONS

- A 4-month-old African boy is brought to the emergency department by his mother who is a recent immigrant from North Africa for evaluation of irritability and decreased po intake. The infant is exclusively breast fed. Which of the following factors increases his risk for the development of rickets?
 - A family history of rickets
 - Prematurity
 - Breast feeding
 - Darker skin
 - Cultural practices
- You suspect rickets in the above-mentioned patient. Your suspicions are confirmed with laboratory studies. In addition to starting treatment, you counsel the mother about Vitamin D supplementation. Vitamin D can be acquired by diet from which of the following?
 - Fish oil
 - Supplemented dairy products
 - Supplemented juice
 - Multivitamins
 - All of the above
- A 10-month-old African-American female is brought to the ED because she has just experienced a seizure. She has a low-grade fever and no previous history of seizures. Parents report that she has been exclusively breast fed and does not receive any vitamin supplementation. Which of the following is the *most* likely etiology of the seizure?
 - Hypomagnesemia
 - Hyponatremia
 - Fever
 - Hyperphosphatemia
 - Hypocalcemia
- A 7-year-old boy is diagnosed with recurrent pneumonias. His ethnicity and cultural practices places him at increased risk of rickets. Which of the following is recommended as optimal treatment of vitamin D deficiency rickets?
 - Single oral dose of 600,000 IU
 - Single intramuscular dose of 600,000 IU
 - Monthly intramuscular injections of 10,000–50,000 IU for 3 months
 - Oral dose of 50,000 once a week for 2 months
 - All of the above
- A 2-year-old girl is brought to the emergency department by his parents for evaluation of delayed walking. The child was exclusively breast fed until 1 month ago and did not receive any supplementation. In addition to blood work, you obtain x-rays. What would be the best radiograph to diagnose rickets in an infant or young child?
 - Bilateral wrists
 - Chest x-ray
 - KUB
 - Anterior view of the knees
 - Bone survey
- Which of the following laboratory abnormalities would you expect to see in this child if your suspicion of rickets is correct?
 - High calcium
 - Hyperphosphatemia
 - High alkaline phosphatase
 - High vitamin D-25 (calcidiol)
 - Low PTH (parathyroid hormone)

ANSWERS

- D. The majority of the reported cases of rickets occurs in individuals with dark skin and breast-fed infants who do not receive vitamin D supplementation. Other factors that limit the amount of sunlight exposure for a given individual can contribute to the development of rickets. These include season of the year, latitude, pollution, lifestyle or cultural practices, and use of sunblock. This results in increased dependence on dietary vitamin D to maintain vitamin D sufficiency.

2. E. Since the discovery of vitamin D in the early nineteenth century, many food products have been supplemented with vitamin D. Dietary sources of vitamin D include fatty fish and fortified food products, including dairy, infant formula, juice, and cereals.
3. E. Patients with rickets may present with hypocalcemic signs and symptoms. Signs and symptoms of hypocalcemia relate more to the velocity of the fall in calcium levels than to the actual serum concentration. The more acute the drop in calcium, the more likely clinical symptoms will be expressed. Usually, signs of hypocalcemia are present when the ionized calcium concentration falls below 2.5 mg/dL. Neurologic findings include seizures, irritability, memory loss, and affective disorders. There are several reports of congenital rickets presenting with hypocalcemic seizures.
4. E. Vitamin D deficient rickets can be treated on one day or gradually. Usually the 1-day approach is good for patients with poor compliance.
5. D. The best radiograph for infants and children younger than 3 years is an anterior view of the knee showing the metaphyseal ends and epiphyses of the femurs and tibiae.
6. C. Alkaline phosphatase levels are almost always elevated. In vitamin D-deficient rickets, calcium is low in the early and late stages, but it can be normal. The phosphorus level is low in the vast majority of cases. Calcidiol levels are low and PTH levels are elevated. Calcitriol levels are normal or high because of PTH activity. Calcidiol, calcitriol, and PTH levels may take days to be reported and will not be available during emergency department evaluation.

- TBW as a percentage of body weight and the distribution between compartments varies with age (Fig. 80-1).
- The electrolyte and protein content of the compartments is not the same (Fig. 80-2).

MOVEMENT OF FLUID

- Cell membranes form the barrier between the extracellular and intracellular compartments.
 - The cell membrane is freely permeable to water, and impermeable to electrolytes and proteins, except by active transport.
 - Although the specific osmoles differ in the two compartments, the osmolality is equal.
 - Water flows across this barrier by osmotic pressure.
- The vascular endothelium forms the barrier between the intravascular and interstitial spaces.
 - It is permeable to water and electrolytes, but not to protein.
 - Two forces regulate fluid movement.
 - Hydrostatic pressure, created by the propulsion of blood through vessels, favors movement of fluid from the intravascular space to the interstitial space.
 - Oncotic pressure, exerted primarily by albumin found in the vascular space, favors water movement from the interstitium into the vascular space.
- Normally, movement of water and electrolytes from the vascular space to the endothelium at the arteriolar side and in the reverse direction at the venous side are balanced.

FLUID DISTRIBUTION

- Free water added to the vascular space distributes proportionally to all three compartments.
- Isotonic crystalloid distributes throughout the extracellular space.
- Iso-oncotic fluid will remain in the vascular space, with the exception of a small distribution to the interstitial space because of the increase in hydrostatic pressure.

FLUID REQUIREMENTS

- Maintenance Fluids
 - Maintenance fluids replace routine daily fluid losses
 - Insensible losses
 - Routine output of urine and stool

80 FLUID AND ELECTROLYTE DISORDERS

Susan A. Kecskes

FLUIDS

FLUID COMPARTMENTS

- Total body water (TBW) is divided into two parts.
 - Intracellular (approximately two-third of TBW).
 - Extracellular (approximately one-third of TBW).
 - Intravascular (approximately one-fourth of extracellular water).
 - Extravascular (approximately three-fourth of extracellular water).

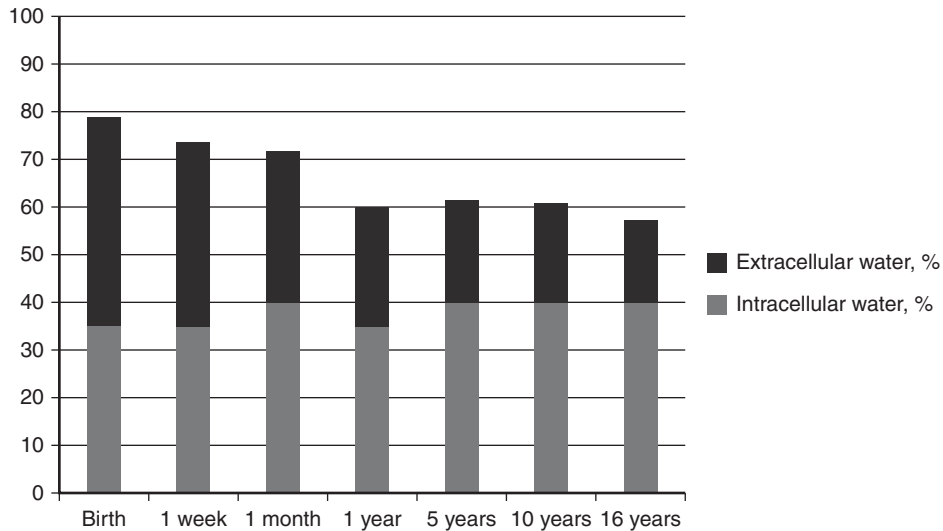


FIG. 80-1. Distribution of total body water (TBW) based on body weight at various ages. Both TBW and ECW decline significantly over the first year of life.

- Maintenance fluids are proportional to body surface area (BSA)
 - Infants and children have a higher BSA per kilogram, so they have proportionally higher fluid requirements.
- There are four common methods to calculate maintenance fluids (Table 80-1).
- Fluid Choices
 - Infants: D₅ 0.2 NaCl with 20 mEq/L of KCl
 - Children and adults: D₅ 0.45 NaCl with 20 mEq/L of KCl
- Consider higher sodium content (isotonic) in postoperative patients and those with hypovolemia, sepsis, pneumonia, asthma, congestive heart failure, meningitis, or head injury.
- Adjust for underlying electrolyte disorder or organ system malfunction
- Deficit fluids
 - Estimate deficit
 - ◻ Assess circulation: signs and symptoms (Table 80-2).
 - ◻ Mild (water loss <5% TBW)

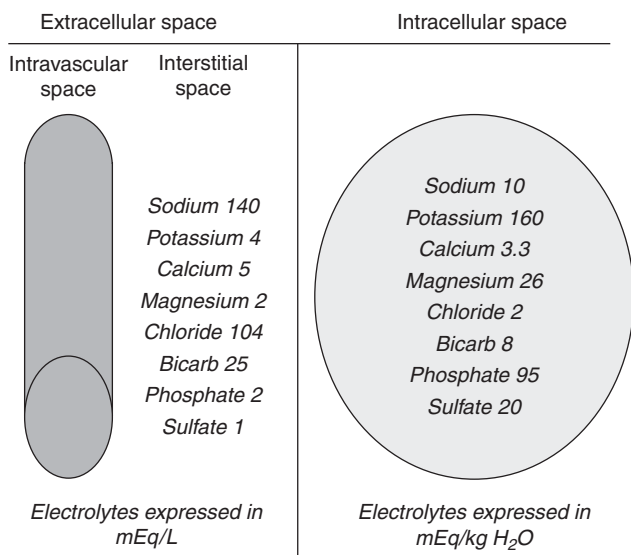


FIG. 80-2. Electrolyte composition of intracellular space versus extracellular space.

TABLE 80-1 Four Methods for Maintenance Fluid Calculations

Body Surface Area Method	
1500 mL/BSA (m ²)/d	
100/50/20 Method	
Weight (kg)	Fluid
0–10	100 mL/kg/day
11–20	100 mL + 50 mL/kg/day for every kg > 10 kg
>20	1500 mL + 20 mL/kg/day for every kg > 20 kg
4/2/1 Method	
Weight (kg)	Fluid
0–10	4 mL/kg/h
11–20	40 mL + 2 mL/kg/h for every kg > 10 kg
>20	60 mL + 1 mL/kg/h for every kg > 20 kg
Insensible + Measured Losses Method	
400–600 mL/BSA (m ²)/d + urine output (mL/mL) + L other measured losses (mL/mL)	

TABLE 80-2 Signs and Symptoms of Dehydration

	MILD (5% TOTAL BODY WEIGHT)	MODERATE (10% TOTAL BODY WEIGHT)	SEVERE (15% TOTAL BODY WEIGHT)
Mental status	Alert	Irritable; drowsy	Lethargic
Skin turgor	Brisk retraction	Mild delay	Prolonged retraction
Anterior fontanel	Normal	Minimally sunken	Sunken
Eyes	Moist; + tears	Dry; – tears	Sunken; – tears
Mucous membranes	Moist	Dry	Very dry
Pulses	Normal	Rapid; weak peripherally	Rapid; weak centrally
Capillary refill	< 2 s	2–5 s	> 5 s
Respiration	Normal	Rapid	Deep and rapid
Urine output	>1 mL/kg/h	< 1 mL/kg/h	Minimal or absent
Blood pressure	Normal	Low normal	Hypotension

- Moderate (water loss 5% to 10% TBW)
- Severe (water loss >10% TBW)
- Alternatively, calculate deficit from changes in body weight
 - [premorbid body weight (kg)-morbid body weight (kg)] × 1000 = fluid loss (mL)
- Helpful laboratory values: serum electrolytes, blood urea nitrogen, creatinine, acid-base status, urinalysis, and urine sodium concentration.
- Resuscitation fluid
 - If perfusion is inadequate, fluid resuscitation should be initiated.
 - Initial bolus: 20 mL/kg of isotonic crystalloid [0.9% NaCl or lactated Ringer's (LR) solution] IV over <20 minutes.
 - Reassess
 - Further boluses are given until perfusion is adequate.
 - Pediatric patients commonly require >60 mL/kg of resuscitation fluid to restore perfusion.
 - Consider inotropes or pressors if the circulatory failure is not solely related to fluid deficit.
- Remaining deficit after resuscitation
 - Subtract resuscitation fluids from the calculated deficit and replace the remainder over 24 hours if the patient is in a normal osmotic state.
 - Adjust replacement solutions to the electrolyte status of the individual patient.
 - Typical replacement fluid: D₅ 0.45% NaCl with 20 mEq/L of KCl.
 - Consider oral rehydration solution if the patient is tolerating PO.
- Ongoing fluid losses
 - Some patients may require replacement of ongoing fluid losses not included in normal maintenance requirements (Table 80-3).
 - The type of fluid should be tailored to the content of the fluid lost.
 - A standard solution with a composition close to the fluid being replaced is usually adequate to maintain homeostasis in patients with intact renal function.
 - If more precision is required, the electrolyte content of the fluid being lost may be measured and replaced.

TABLE 80-3 Adjustments to Maintenance Fluids

Fever	Increase maintenance fluids by 10% for each degree > 37.8°C.
Tachypnea (<i>nonhumidified environment</i>)	Increase maintenance fluids by 5%–10%.
Vomiting and gastric loss	Replace with 0.45% NaCl with 10 mEq/L KCl.
Stool loss	Replace with LR with 15 mEq/L KCl or 0.45% NaCl with 20 mEq/L KCl and 20 mEq/L NaHCO ₃ .
Cerebrospinal fluid	Replace with LR or 0.9% NaCl.
Pleural fluid, peritoneal fluid, wound drainage (serous)	Replace with LR or 0.9% NaCl; may need to replace albumin periodically—base replacement on measured serum albumin levels.
Blood	≤25% blood volume: replace with LR or 0.9% NaCl; assess hematocrit and physiologic status for administration of blood. >25% blood volume: replace one-half to two-thirds of loss as whole blood and reassess. Alternatively, use “3-for-1” and replace 3 × the blood loss with LR or 0.9% NaCl.
Third-space losses	Estimate based on patient's physiologic status. Replace with LR or 0.9% NaCl.

SODIUM

- Sodium is found in highest concentration in the extracellular compartment and is normally maintained between 135 and 145 mEq/L.
- In both hyponatremia and hypernatremia, the total body store of sodium may be high, low, or normal. TBW relative to total body sodium determines sodium concentration.
- *Hypernatremia* is defined as serum sodium >150 mEq/L.
 - It may result from intake of sodium in excess of water or, more commonly, from loss of water in excess of sodium.
 - Causes of primary sodium excess:
 - Inadequately diluted infant formula.
 - Excessive sodium bicarbonate administration.
 - Intravenous hypertonic saline administration.
 - Causes of hypovolemic hypernatremia, in which water loss exceeds sodium loss:
 - Gastroenteritis with diarrhea and vomiting.
 - Increased insensible water loss (ie, fever, use of radiant warming devices, burns).
 - Diabetes mellitus.
 - Inadequate access to free water.
 - Diabetes insipidus.
 - Clinical manifestations of hypernatremia
 - In primary sodium excess, the skin is often described as “doughy.”
 - In hypovolemic hypernatremia, the signs and symptoms are those of dehydration and decreased perfusion.
 - In both, CNS signs, including irritability, lethargy, coma, and seizures may be seen.
 - Therapy for hypernatremia (Fig. 80-3)
 - Correct circulatory failure, if present.
 - Gradually restore TBW, over ≥ 48 hours. Fatal cases of cerebral edema have occurred with correction over <24 hours.
 - The goal is to reduce the serum sodium at a rate of 0.5–1 mEq/L/h.
 - Begin with isotonic crystalloid and complete with hypotonic crystalloid, such as D₅ 0.45 NaCl.
 - Provide maintenance fluids and replace ongoing losses in addition to the deficit correction.
 - Plasma electrolytes and osmolality should be monitored frequently.
 - In patients in whom DI is suspected, vasopressin may be trialed:
 - Aqueous pitressin, via continuous infusion; start at 0.5 mU/kg/h and titrate every 30 minutes (maximum dose 10 mU/kg/h) to produce urine osmolality > serum osmolality.
 - Alternatively, desmopressin (DDAVP) may be administered intermittently. Titrate up to produce urine osmolality > serum osmolality.
 - Oral administration: the initial dose is 0.05 mg PO BID (titrate up to a maximum of 0.4 mg per dose TID).
 - Intranasal administration: the initial dose is 0.05 mL (5 mcg), once per day (titrate to a maximum of 0.4 mL per day over 1–3 doses).
 - Primary sodium excess is treated by removal of excess sodium.
 - First, sodium intake should be curtailed.
 - In patients with intact renal function, free water via the gastrointestinal tract or hypotonic fluids parenterally will help in correction.
 - Patients with renal failure require dialysis.
- *Hyponatremia* is defined as a serum sodium concentration <130 mEq/L. It is characterized by excess body water relative to body sodium.
 - Depending on etiology, total body sodium may be decreased, increased, or normal (Fig. 80-4).
 - Hyponatremia with decreased total body sodium occurs when sodium loss exceeds water loss. These losses may be extrarenal or renal.
 - Extrarenal etiologies are associated with renal sodium conservation (urine sodium <20 mEq/L).
 - Renal causes with decreased total body sodium are associated with ongoing urinary sodium loss (urine sodium >20 mEq/L).
 - Hyponatremia with increased total body sodium occurs when the increase in TBW exceeds sodium retention.
 - Hyponatremia with normal total body sodium is associated with:
 - Syndrome of inappropriate antidiuretic hormone (SIADH) which leads to a dilutional hyponatremia.
 - Urinary osmolality (>200 mOsm/L) and sodium concentration (>20 mEq/L) are inappropriately elevated for the hypotonicity and sodium concentration of the serum.
 - Water intoxication
 - WIC syndrome occurs when small infants are fed overly dilute formula or excess water.
 - Inappropriately hypotonic replacement of fluid losses.
 - Clinical manifestations of hyponatremia depend on the volume status of the patient, the rapidity of development and degree of hypo-osmolality.
 - In hypovolemic hyponatremia, symptoms of dehydration and acute circulatory failure prevail.

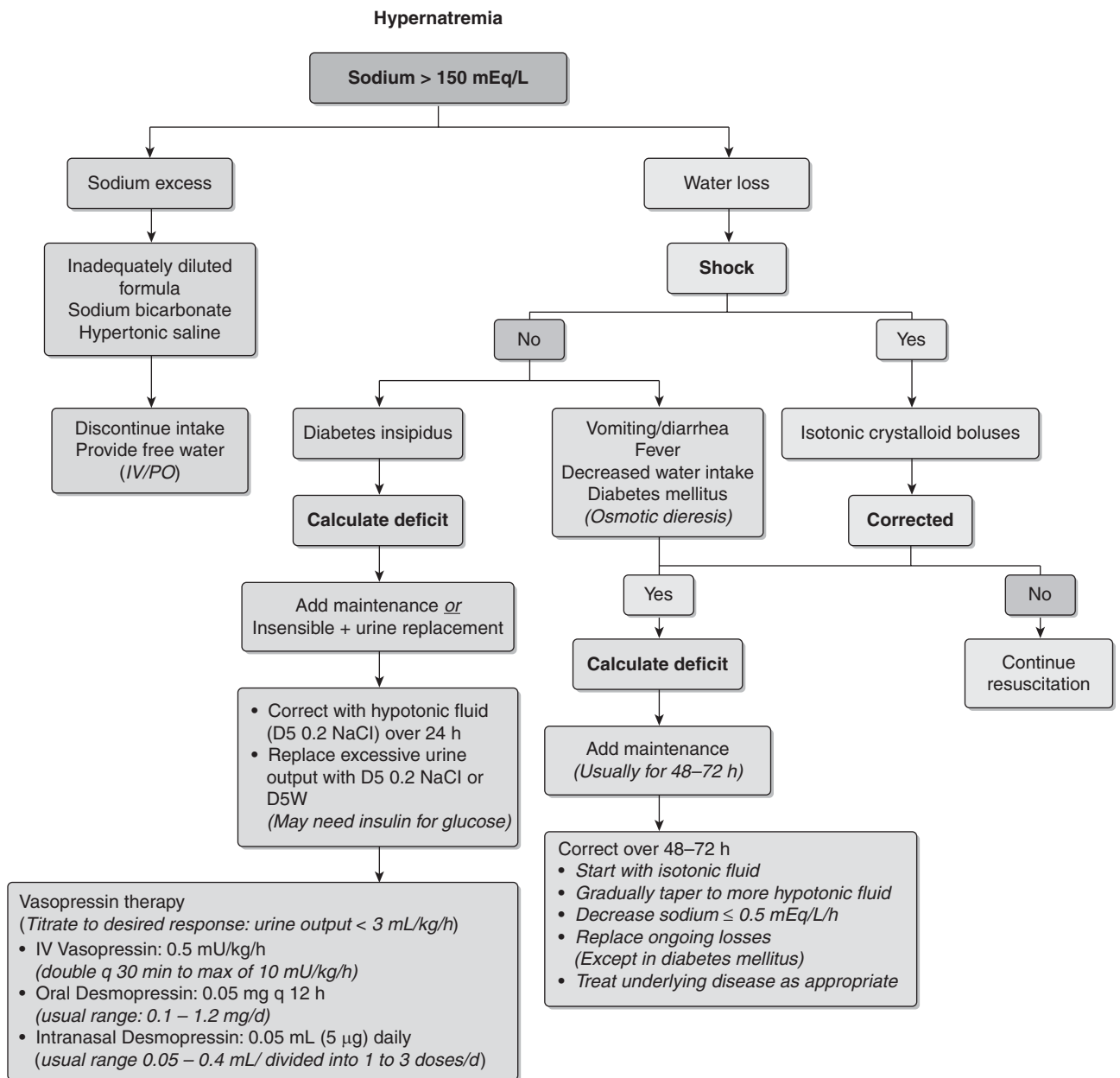


FIG. 80-3. Treatment of hyponatremia.

- Rapid onset may lead to brain edema and CNS pathology, ranging from lethargy to coma and brain herniation in the most severe cases. Hyponatremia is a common cause of afebrile seizures in children <6 months of age.
- Treatment begins with an assessment of the patient's volume status and correction of any hypovolemic shock (Fig. 80-5).
- Correction of the hyponatremia requires a loss of water in excess of sodium. This must be undertaken with care, as aggressive correction may lead to the osmotic demyelination syndrome.
- In hyponatremia of acute onset (<48 hours), it appears safe to correct the sodium over 24 hours.
- In more gradual onset, sodium correction should not exceed a rate of 0.5 mEq/L/h.

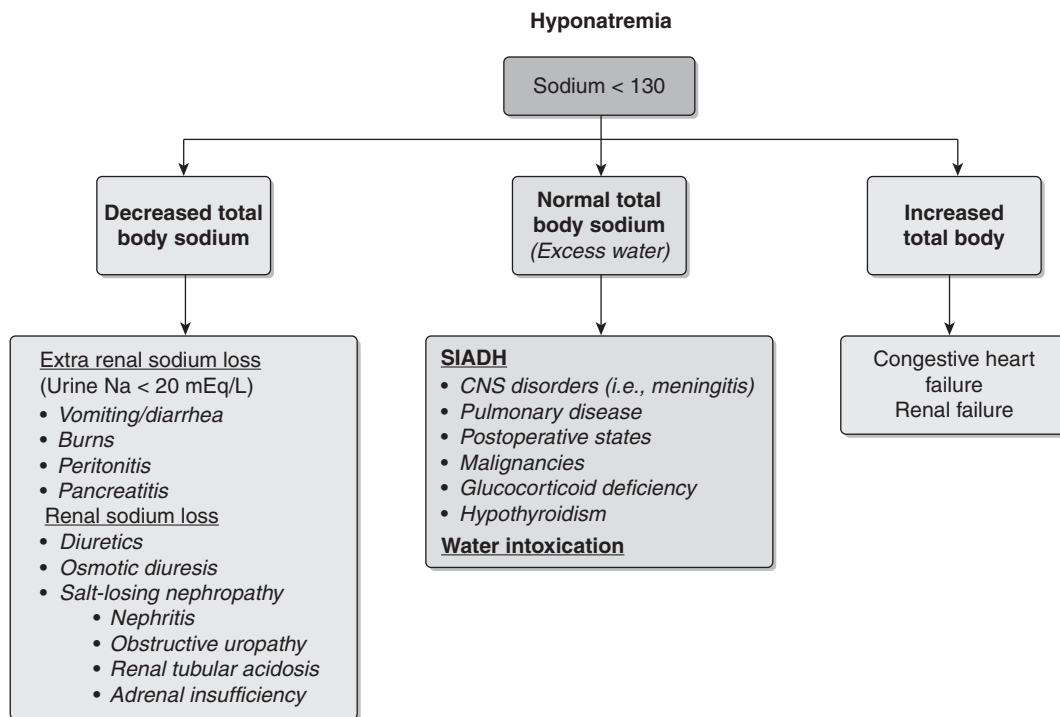


FIG. 80-4. Causes of hyponatremia.

- Initiate therapy with isotonic crystalloid at rates determined by the volume status of the patient.
- In euvolemic or hypervolemic patients, use maintenance or fluid-restricted rates.
- SIADH: fluid restrict to two-third maintenance, or even to insensible fluid loss.
- In hypovolemic patients, time the correction to the desired rise in sodium concentration (approximately 10 mEq/L/24 h).
- More aggressive partial correction may be needed when there are severe neurologic symptoms, such as seizures. Use IV infusion of 6 mL/kg of 3% sodium chloride over 1–2 hours to produce rise in serum sodium of 5 mEq/L. A single bolus is usually sufficient to reduce acute symptoms and the remainder of the correction should be undertaken more gradually.
- Loop diuretics, such as furosemide, have been used as an adjunct to therapy to increase free water clearance.
- In all types of hyponatremia, the underlying pathology should be identified and appropriate treatment initiated.

POTASSIUM

- Normal potassium concentration in the extracellular fluid (ECF) is 3.5 to 5.5 mEq/L, compared with 160 mEq/L in the intracellular fluid (ICF). Potassium homeostasis is managed through the use of both excretion and translocation.
 - The majority of potassium excretion occurs in the kidney.
 - The kidney can adjust urinary potassium excretion from <5 mEq to >1000 mEq/24 h.
 - Approximately 10% of daily potassium intake is lost in stool.
 - Translocation allows the body to maintain stable ECF potassium.
 - Potassium uptake is regulated by insulin, epinephrine, aldosterone, and acid-base balance.
 - Acidemia promotes movement of potassium to the ICF.
 - Alkalosis favors movement of potassium to the ECF.
- Hyperkalemia is defined as serum potassium >5.5 mEq/L and can result from increased potassium intake, decreased potassium loss, or from redistribution from the ICF.
 - Causes of hyperkalemia
 - Increased potassium intake rarely results in elevated serum potassium, unless iatrogenic, or simultaneously associated with decreased excretion.
 - Iatrogenic causes include excessive IV administration of potassium, administration of large quantities of cold stored blood, large doses of

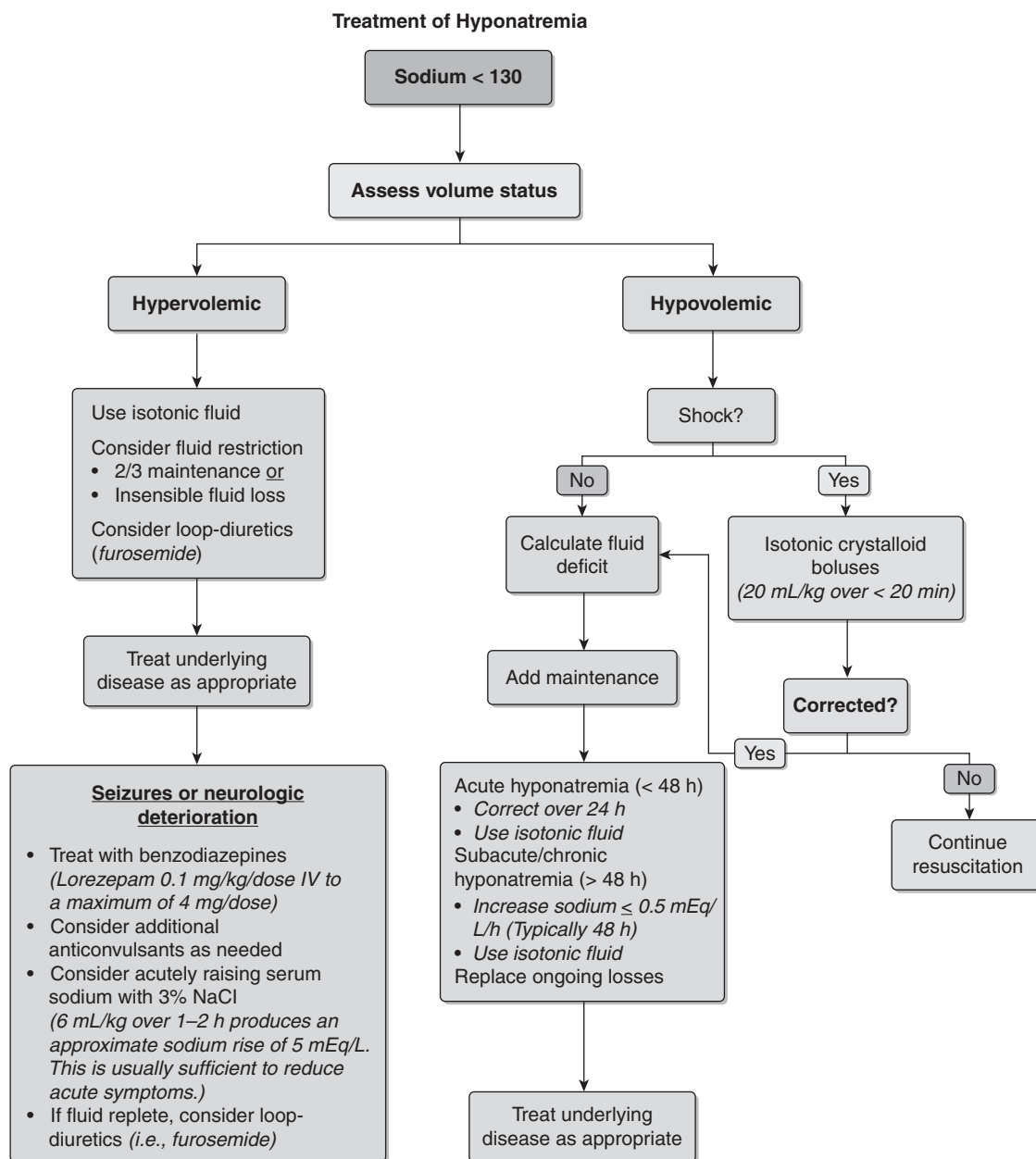


FIG. 80-5. Treatment of hyponatremia.

the potassium salts of penicillin or oral intake of potassium-containing salt substitutes.

- Acute renal failure is the primary cause of decreased excretion.
- Adrenal insufficiency may result in hyperkalemia due to decreased mineralocorticoid activity.
- Use of potassium-sparing diuretics.
- Redistribution of potassium from the ICF to the ECF may occur via cell destruction or translocation from intact cells.
 - Cell destruction: trauma, burns, rhabdomyolysis, massive intravascular coagulopathy, and tumor necrosis. Hematomas in the newborn

and gastrointestinal bleeding may result in large volumes of hemolyzing cells and elevated potassium levels.

- Potassium can be quickly shifted from the ICF to the ECF in response to metabolic acidosis.
- Pseudohyperkalemia must be considered in the differential diagnosis of hyperkalemia. It is associated with hemolysis from the blood draw. When pseudohyperkalemia is suspected, specimens should be redrawn with attention to avoiding such mechanical factors.
- Clinical findings of hyperkalemia:
 - Most patients with hyperkalemia are asymptomatic.

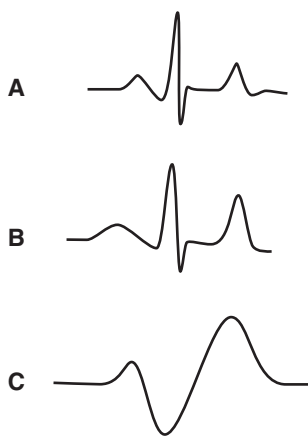


FIG. 80-6. ECG changes in hyperkalemia. (A) Normal ECG, (B) ECG with peaked T waves, prolonged PR interval, and widened QRS, seen in moderate hyperkalemia (potassium >7.0 mEq/L), (C) “Sine wave” ECG seen at potassium levels >8 mEq/L.

- Neuromuscular symptoms progress from paresthesias to muscle weakness and, ultimately, flaccid paralysis.
- Cardiac abnormalities may produce life-threatening situations: electrocardiogram (ECG) changes: peaked T-waves, prolongation of the PR interval and progressive widening of the QRS complex. As potassium rises further (typically >8 mEq/L), the classic “sine wave” of hyperkalemia appears (Fig. 80-6). This may rapidly degenerate to asystole or ventricular fibrillation.
- Diagnostic tests
 - Serum electrolytes, renal indices (BUN, creatinine, and urinalysis), a complete blood count (CBC) and acid-base status should be measured.
 - A 12-lead ECG should be performed.
 - Urinary potassium levels may help evaluate the cause of the hyperkalemia.
 - All patients with serum potassium levels >6.5 mEq/L should have continuous ECG monitoring.
- Treatment of hyperkalemia depends on the level of serum potassium, clinical symptoms, and the renal status of the patient.
 - Moderate hyperkalemia (<7 mEq/L)
 - ◻ Halt intake of potassium and potassium-sparing medications.
 - ◻ Administer potassium-binding agent, sodium polystyrene sulfonate (Kayexalate, 1 to 2 g/kg PO, NG, or PR).
 - ◻ Follow-up serum potassium levels.
 - ◻ Dialysis should be considered for patients with renal dysfunction.
 - Serum potassium levels >7 mEq/L or symptomatic patients require aggressive intervention to stabilize the cellular membrane, shift potassium intracellularly, and increase potassium elimination (Fig. 80-7).
- Hypokalemia is defined as a serum potassium level <3.5 mEq/L and can result from decreased intake, increased renal excretion, increased extrarenal losses, or a shift of potassium from the ECF to the ICF.
 - Causes of hypokalemia
 - A low-potassium diet, eating disorders such as anorexia nervosa and prolonged administration of intravenous fluids without potassium.
 - Increased renal excretion associated with diuretics, osmotic diuresis, hyperaldosteronism, Bartter syndrome, magnesium deficiency, or renal tubular acidosis.
 - Extrarenal losses such as vomiting, diarrhea and nasogastric losses may lead to hypokalemia, both from the direct loss of potassium and from secondary hyperaldosteronism associated with hypovolemia.
 - Movement of potassium into the cells from the ECF can occur with correction of acidosis, alkalosis, administration of insulin, administration of β_2 -agonists, or familial hypokalemic periodic paralysis.
 - Clinical manifestations of hypokalemia:
 - Muscle weakness, areflexia, and autonomic instability.
 - Ileus.
 - Respiratory insufficiency.
 - Rhabdomyolysis.
 - ECG changes: flattening of the T wave, ST segment depression, U waves, premature atrial and ventricular contractions and dysrhythmias.
 - Polyuria due to a reduced ability to concentrate urine.
 - Diagnostic tests:
 - Serum electrolytes, including magnesium, serum pH.
 - Urine potassium concentration of <15 mEq/L indicates renal conservation and suggests extrarenal loss.
 - 12-lead ECG
 - Treatment
 - If there are no life-threatening complications, correct hypokalemia gradually with oral supplementation or, if contraindicated, an increase in the maintenance potassium concentration in the intravenous fluids.
 - Correct underlying conditions that accompany the hypokalemia, such as alkalosis or hypomagnesemia.

Treatment of Hyperkalemia

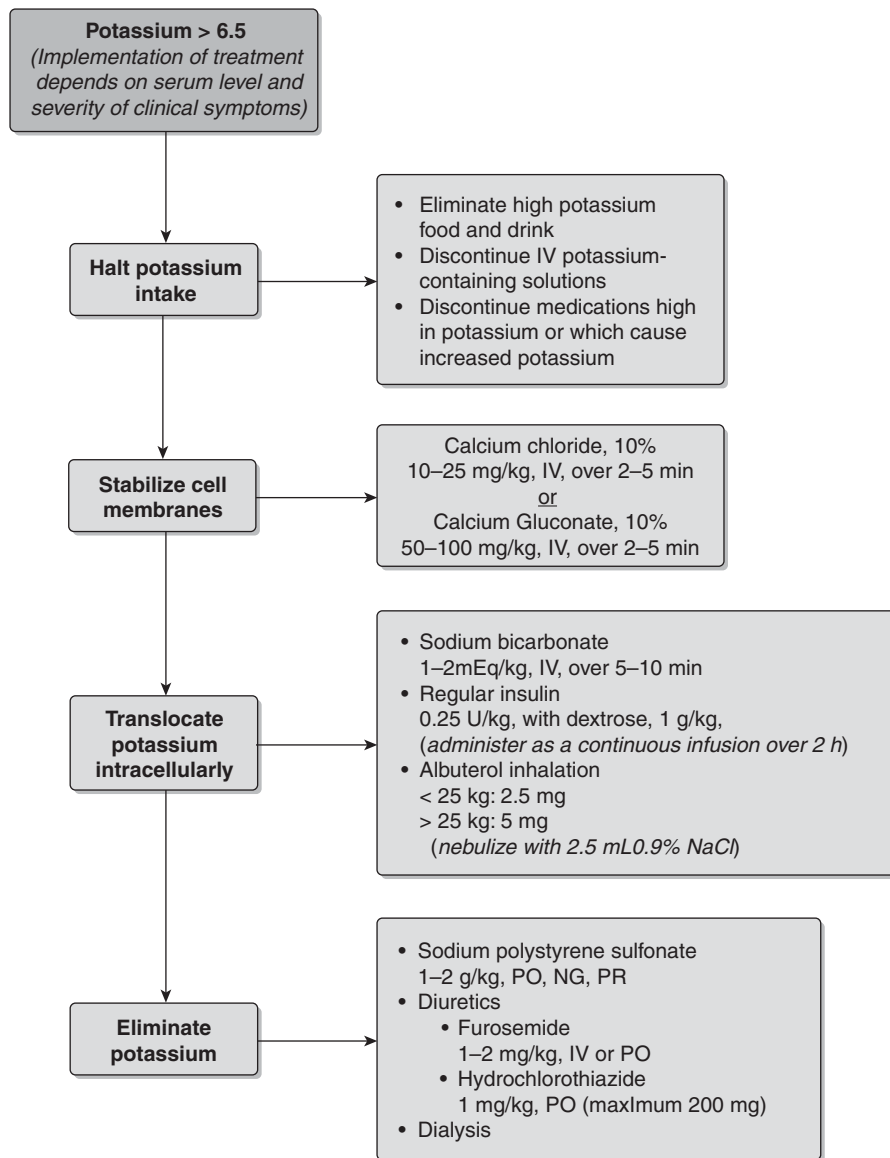


FIG. 80-7. Treatment of hyperkalemia.

- Identify sources of ongoing potassium loss; measure and replace as needed.
- Determine the cause of the loss and, if possible, treat it.
- If hypokalemia is associated with digoxin use or life-threatening complications, such as cardiac dysrhythmias, rhabdomyolysis, extreme muscle weakness, or respiratory arrest, intravenous therapy is required.
- Extreme care should be exercised in the ordering, preparation and administration of intravenous potassium. Potassium chloride may be administered intravenously:
 - 0.5 to 1 mEq/kg/dose (maximum dose: 40 mEq) to infuse at 0.3 to 0.5 mEq/kg/h (maximum rate: 1 mEq/kg/h).
 - Potassium must be diluted prior to intravenous administration. In peripheral lines, the maximum concentration is 80 mEq/L. The maximum recommended central line concentration is 200 mEq/L (usually reserved for severely fluid restricted patients).

- Continuous ECG monitoring, along with frequent assessment of serum potassium levels, is essential during intravenous correction of hypokalemia.

CALCIUM

- Calcium is one of the most abundant and important minerals in the body, with 99% of body calcium stored in bone.
 - Of the 1% present in the circulation, 40% is bound to proteins such as albumin, 15% is complexed with anions such as phosphate and citrate and 45% is physiologically free and ionized.
- Parathyroid hormone, vitamin D, and calcitonin interact to regulate calcium in a narrow range by controlling intestinal absorption, renal excretion, and skeletal distribution.
- Calcium is responsible for cellular depolarization, muscle excitation/contraction, neurotransmitter release, hormonal secretion, and the function of both leukocytes and platelets.
- A serum calcium level measures both ionized and protein-bound calcium. Since approximately half of serum calcium is bound to albumin, the serum calcium level may need to be adjusted for alterations in the albumin level.
 - For every 1 g/dL decrease in serum albumin, true serum calcium may be estimated by adding 0.8 mg/dL.
 - Alternatively, ionized calcium levels are widely available.
- Hypercalcemia is defined as a serum calcium level >10.5 mg/dL.
 - Symptoms
 - Often asymptomatic.
 - GI complaints may include constipation, anorexia, vomiting, abdominal pain or pancreatitis.
 - CNS: lethargy, depression, psychosis, or coma.
 - ECG changes include QT segment shortening, bradycardia, heart block, and sinus arrest.
 - Nephrolithiasis,
 - In children, hypercalcemia with malignancy is associated with bone metastasis or tumor lysis syndrome.
 - Other causes in children include primary or tertiary hyperparathyroidism, hyperthyroidism, vitamin D intoxication, immobilization, thiazide diuretics, milk-alkali syndrome, and sarcoidosis.
 - Laboratory investigation:
 - Total or ionized serum calcium, serum albumin, and total protein, electrolytes (including magnesium and phosphorus), BUN, creatinine, CBC, ECG, and urinalysis.
- Hypocalcemia is defined as serum calcium < 9 mg/dL.
 - Causes:
 - Hypoparathyroidism
 - Vitamin D deficiency
 - Hyperphosphatemia and magnesium deficiency
 - Massive transfusion of citrated blood
 - Phosphate enema toxicity
 - Sepsis
 - Pancreatitis
 - Clinical manifestations:
 - Nonspecific symptoms include nausea, weakness, paresthesias, and irritability.
 - Neuromuscular irritability: positive Chvostek's and Trousseau's signs.
 - In more severe cases, tetany, seizures, laryngospasm, and psychiatric manifestations may be seen.
 - ECG changes: prolongation of the QT interval, bradycardia, and dysrhythmias
 - Diagnostic tests:
 - Ionized and total calcium, magnesium, phosphorus, albumin and total protein, BUN, creatinine, and alkaline phosphatase.
 - Vitamin D and parathyroid hormone levels.
 - Urine calcium and phosphorus levels.
 - For significant or symptomatic hypocalcemia, intravenous calcium may be administered with continuous ECG monitoring.
 - Calcium gluconate, 10%, 50 to 100 mg/kg/dose, or calcium chloride, 10% (10 to 20 mg/kg/dose), may be administered over 3 to 5 minutes.
 - Intravenous calcium is very irritating to tissues and veins. It should be diluted prior to administration and is preferably administered through a central line. It should never be given intramuscularly, subcutaneously or via an endotracheal

route, as tissue necrosis and sloughing will occur.

- The maximum concentration of calcium gluconate should be 50 mg/mL and calcium chloride, 20 mg/mL.
- Intravenous calcium predisposes to digitalis toxicity and precipitates when mixed with bicarbonate.
- Hyperphosphatemic patients are at risk of metastatic calcium deposition with calcium administration and require treatment aimed at lowering phosphorus levels.
- When hypomagnesemia is present, it should be corrected.
 - Magnesium sulfate 25 to 50 mg/kg, IV, diluted to a maximum concentration of 200 mg/mL, over 2 to 4 hours.

- C. D5 0.45 NaCl with 20 mEq/L KCl at 44 mL/hr.
- D. D5 0.45 NaCl with 20 mEq/L KCl at 500 mL/m² BSA given over 24 hours.
- E. LR solution at 44 mL/hr.

2. A 4-month-old infant (3.5 kg) is brought to the ED with a 2-day history of vomiting and diarrhea. Patient has a HR of 200, fontanel is sunken, and capillary refill is 6 second. Appropriate fluid management would be
 - A. Bolus with 20 mL/kg D5 0.2 NaCl with 20 mEq/L KCl IV over < 20 minute.
 - B. Bolus with 10 mL/kg Normal Saline and then start D5 0.2 NaCl with 20 mEq/L KCl at 14 mL/hr.
 - C. Start on 2x Maintenance with D5 0.45 NaCl.
 - D. Bolus with 70 mL of Ringer's lactate over 1 hour and then start on D5 LR at 14 mL/hr.
 - E. Bolus with 70 mL of normal saline over <20 minutes; after reassessments, bolus repeated × 2. Patient then started on IV D5 0.45 at 28 mL/hr × 24 hours.

3. An 11-month-old male (10 kg) with acute febrile illness and respiratory distress is brought to the ED. Respiratory rate is 60. He has SpO₂ of 97% on oxygen, 2 lpm, via humidified nasal cannula. Temperature is 39°C. He has already received a bolus of normal saline and cap refill is 2 second. Appropriate maintenance fluids would be
 - A. D5 0.2 NaCl with 20 mEq/L KCl at 42 mL/hr
 - B. D5 0.45 NaCl with 20 mEq/L KCl at 46 mL/hr
 - C. Normal Saline 20 cc/kg over < 20 min.
 - D. D5 0.2 NaCl with 20 mEq/L KCl at 48 mL/hr
 - E. D5 0.45 with 20 mEq/L KCl at 83 mL/hr

4. A 9-year-old female (23 kg) with cerebral palsy presents to the ED with a history of vomiting and diarrhea for 1 day. She is fed via a gastrostomy tube. Heart Rate 160, Respiratory Rate 28, Blood Pressure 110/62. Laboratory: sodium 165, potassium 4.0, serum bicarbonate 10. blood glucose 110, capillary refill 2 second. Appropriate initial fluid management would be
 - A. Bolus with 20 mL/kg Ringer's lactate over 20 minute, then switch to 100 mL/hr for a planned 48 hours.
 - B. Bolus with 1 L normal saline, then switch to D5 0.45 NaCl at 63 mL/hr.
 - C. Bolus with 460 mL normal saline, then switch to 0.45 with 20 mEq KCl at 100 cc/hr.
 - D. Bolus with 20 mL/hr normal saline, then switch to 0.9 NaCl with 20–40 mEq/L at 63 mL/hr.
 - E. Run normal saline at 100 mL/hr.

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QUESTIONS

1. A 3-year-old child (12 kg) with a known seizure disorder is brought to the emergency department (ED) for intractable seizures. She is unable to take anything orally and you want to start her on maintenance IV fluids. Appropriate fluid orders would be
 - A. D5 0.2 NaCl with 20 mEq/L KCl at 48 mL/hr.
 - B. Normal saline 20 mL/kg over 20 min or less.

ANSWERS

1. C. Appropriate maintenance fluids would be: D₅ 0.45 NaCl with 20 mEq/L of KCl. If perfusion is inadequate, fluid resuscitation should be initiated with an initial bolus: 20 mL/kg of isotonic crystalloid (0.9% NaCl or LR solution) IV over <20 minutes. Consider higher sodium content (isotonic) in postoperative patients and those with hypovolemia, sepsis, pneumonia, asthma, congestive heart failure, meningitis, or head injury.
2. E. Pediatric patients commonly require >60 mL/kg of resuscitation fluid to restore perfusion. If perfusion is inadequate, fluid resuscitation should be initiated with an initial bolus: 20 mL/kg of isotonic crystalloid (0.9% NaCl or LR solution) IV over <20 minutes. Consider an oral rehydration solution if the patient is tolerating PO.
3. B. The appropriate maintenance fluids for this patient would be D₅ 0.45 NaCl with 20 mEq/L of KCl. The patient appears to have adequate perfusion.
4. A. Fluid resuscitation should be initiated in this patient with an initial bolus: 20 mL/kg of isotonic crystalloid (0.9% NaCl or LR solution) IV over <20 minutes. Resuscitation fluids are subtracted from the calculated deficit and the remainder replaced over 24 hours if the patient is in a normal osmotic state. Replacement solutions should be adjusted to the electrolyte status of the individual patient. In patients with vomiting and gastric loss, replacement with 0.45% NaCl with 10 mEq/L KCl is indicated

- In the case of an x-linked recessive disorder, there may be a history of affected males related through the maternal family.
 - An example of this situation would occur in a family with ornithine transcarbamylase deficiency resulting in affected male infants with hyperammonemia.
- Table 81-1 lists categories and examples of some IEM that may present in the ED.

PRESENTATION

- Neonatal presentation typically occurs when a severe metabolic defect is present.
 - Infants may present during the first weeks of life after an uneventful neonatal hospital course. However, when severe neonatal forms of these disorders are present, symptoms related to the disorder are expected to develop within hours or days after birth.
 - A number of organic acidemias, urea cycle defects and maple syrup urine disease (MSUD) can present in this manner.
- In some cases, the child is healthy during the first years of life and the initial presentation occurs much later in life.
- Intermittent forms of some disorders occur, where a patient may experience repeated episodes with vomiting and dehydration.
- In the majority of patients with an IEM, there is a relationship between onset of symptoms and a patient's health and nutritional status.
 - The presence of an acute illness, typically viral, is often associated with the development of metabolic problems related to the IEM.
 - A change in diet can precipitate biochemical change.
 - An infant with glycogen storage disease will present with hypoglycemia when the feeding interval increases.
 - Symptoms can develop in a patient with impaired protein metabolism who has been transitioned to cow's milk at the age of 1 year due to the higher protein content of milk.
 - Symptoms may develop after introduction of fructose or sucrose to the diet in a patient with hereditary fructose intolerance.
- Given the diverse nature of IEM with the varied symptoms, clinical history, course, and laboratory findings, it is important that biochemical genetic disorders be considered in the differential diagnosis of ED patients with findings consistent with an IEM.

81

INBORN ERRORS OF METABOLISM

George E. Hoganson

INTRODUCTION

- Inborn errors of metabolism (IEM) represent a diverse group of genetically determined diseases.
 - The majority of these conditions are inherited in an autosomal recessive pattern. The family history is usually negative, depending on an autosomal recessive inheritance pattern, with a 25% risk for affected siblings
 - A subset of these disorders has an x-linked recessive mode of inheritance.

TABLE 81-1 Examples of Inborn Errors of Metabolism

AMINO ACID DISORDERS	ORGANIC ACIDEMIAS	FATTY ACID OXIDATION DEFECTS (FAOD)	DISORDERS OF CARBOHYDRATE METABOLISM	MITOCHONDRIAL DISORDERS
Maple syrup urine disease (MSUD)	Isovaleric acidemia	Medium chain acyl-CoA dehydrogenase deficiency (MCAD)	Glycogen storage disease Ia, Ib, III (GSD)	Disorders of pyruvate metabolism
Tyrosinemia type I	Methylmalonic acidemia	Very long chain acyl-CoA dehydrogenase deficiency (VLCAD)	Hereditary fructose intolerance Galactosemia	Mitochondrial myopathies
Phenylketonuria does not cause acute metabolic problems	Propionic acidemia	Long chain hydroxyl acyl-CoA dehydrogenase deficiency (LCHAD)		Electron transport chain defects
Urea cycle defects Ornithine transcarbamylase deficiency Citrullinemia Argininosuccinic acidemia Carbamyl phosphate synthetase deficiency	Glutaric acidemia Type I	Carnitine palmitoyl transferase I, II deficiency (CPT I, CPT II) Carnitine deficiency		

TABLE 81-2 Presenting Symptoms of IEM and Associated IEM

	ORGANIC ACIDEMIA	FAOD	MSUD	GSD	UREA CYCLE DEFECT	AMINO ACIDOPATHY	TYROSINEMIA	FRUCTOSE INTOLERANCE	HOMOCYST-EINURIA/ COBALAMIN DEFECT
Hypoglycemia	×	×		×					
Anion gap acidosis	×		×						
Ketosis	×		×						
Vomiting	×		×		×				
Seizures	×		×	×	×				
Altered mental status/coma	×	×		×	×	×			
Hypotonia		×		×					
Hepatomegaly	×	×		×					
Hepatic failure		×					×	×	
Cardiomyopathy		×		×					
Thrombocytopenia	×								
Thrombotic events							×		×

FAOD, fatty acid oxidation deficiency; MSUD, maple syrup urine disease; GSD, glycogen storage disease.

- Patients with an IEM may present with common symptoms including vomiting, diarrhea, and febrile illness. In these situations, the severity and presence of other symptoms or laboratory abnormalities may suggest the presence of an IEM.
 - Examples of findings that are suggestive of IEM include unusual odors (present in an organic acidemia or MSUD) and more severe acidosis or ketosis than would be predicted based on the history or hypoglycemia.
- In other cases, patients may present to the ED with severe life-threatening problems including coma, severe acidosis, seizures, cardiomyopathy, or hypoglycemia.
- Table 81-2 lists symptoms associated with IEM.

DIAGNOSTIC TESTING

- Identification of which general laboratory tests to perform is critical not only to assist in the diagnosis

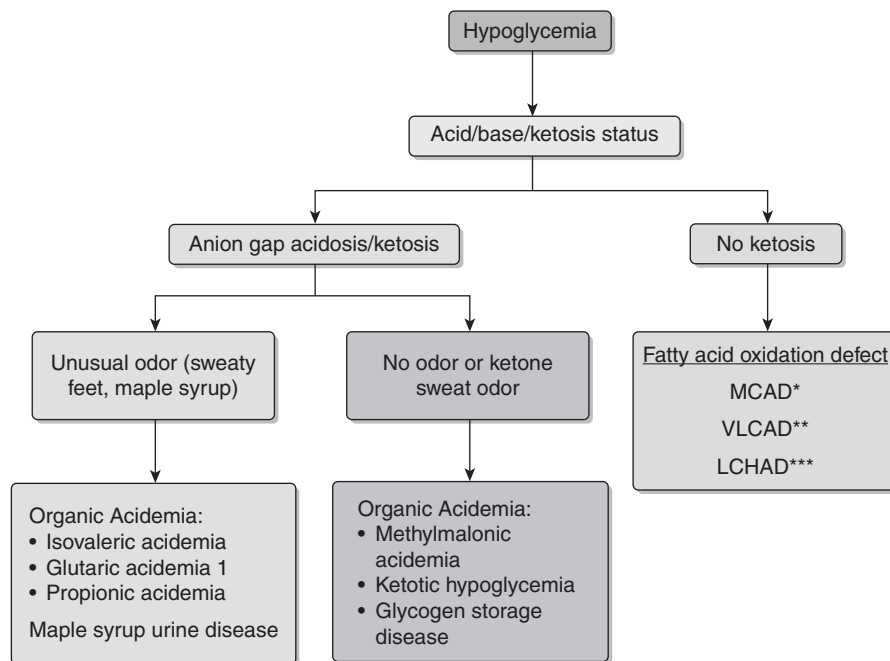
but, potentially of greater immediate importance, to identify special areas requiring treatment.

- It is important to recognize that complicating factors present at the time of the patient's initial presentation may impact test results and their interpretation. For example, a severely dehydrated child with poor perfusion may have a secondary lactic acidosis. The patient's clinical course, including laboratory results and response to treatment, usually allows for a determination as to whether the abnormalities are secondary or the consequence of an IEM.
- Table 81-3 lists some of the general laboratory testing that may be performed in this clinical setting.
 - Testing performed in most situations would include glucose levels, evaluation of acid/base status, urinalysis including urine ketones, complete blood count, and evaluation of renal and liver function.
 - Figure 81-1 outlines a possible approach for evaluation of hypoglycemia in the case of a suspected IEM. Hypoglycemia with negative ketone levels may indicate the presence of a FAOD; while the presence of acidosis, hypoglycemia, and ketosis is more suggestive of an organic acid disorder.
 - A lactate level, to assess for lactic acidosis, may be indicated for cases of acidosis.

TABLE 81-3 General Laboratory Testing to Consider in Case of Suspected IEM

Glucose: hypoglycemia
Blood gas: acid/base states
Comprehensive metabolic panel: renal/liver/electrolyte
Lactate: lactic acidosis
Uric acid: elevated in organic acidemia/MSUD
Ammonia: hyperammonemia in urea cycle defect, FAOD, organic acidemia
Creatine kinase: increased with rhabdomyolysis
Urinalysis: ketones, blood/myoglobin
Urine reducing substances: simple sugars, fructose, galactose
Complete blood count: neutropenia, thrombocytopenia

- An ammonia level for hyperammonemia is indicated in cases of altered mental status.
 - In urea cycle defects, organic acid disorders and FAODs, it is important to obtain the blood ammonia level to diagnose hyperammonia.
- If evidence of myopathy, muscle weakness or myalgia is present in an older patient, especially with an elevation in AST/ALT, a creatine kinase level is indicated to evaluate for a myopathic process and rhabdomyolysis as is present in some FAODs.



*MCAD: medium chain acyl-CoA dehydrogenase deficiency

**VLCAD: very long chain acyl-CoA dehydrogenase deficiency

***LCHAD: long chain hydroxyl acyl-CoA dehydrogenase deficiency

FIG. 81-1. Possible IEM explanations for hypoglycemia.

TABLE 81-4 Biochemical Genetic Testing Utilized in the Diagnosis and Management of IEM

Analyte determination
Plasma amino acids
Urine organic acids
Blood carnitine level
Blood acylcarnitine profile
Enzyme determination
Molecular diagnostics
Common mutation detection
Gene sequencing

- The presence of blood on urinalysis with normal urine RBC count in this setting may be indicative of myoglobinuria.
- Definitive diagnosis of an IEM typically requires special biochemical diagnostic testing which is not performed in many hospital laboratories.
 - However, initiating this testing following an ED visit or subsequent hospitalization can greatly facilitate the diagnosis of one of these conditions and assist in the patient's medical management.
 - Table 81-4 lists some of the specialized biochemical genetic testing considered in the evaluation of IEM.
 - Specific testing to be performed is often determined in consultation with the biochemical/clinical geneticist or other metabolic specialist.

NEWBORN SCREENING

- With the implementation of tandem mass spectrometry in the diagnosis of IEM through neonatal screening, many of the conditions referred to thus far can be diagnosed in the newborn period.
 - This allows for presymptomatic detection and the institution of a corresponding treatment regimen.
 - Three groups of biochemical genetic disorders are screened for using this technology: amino acid disorders, organic acidemias, and FAODs.
 - Galactosemia testing is also included on most newborn screening panels.
- It is important to note that some disorders cannot be detected with this screening, because either biochemical markers of the disorder are not present or newborn screening does not detect the disorder in all cases.
- Under optimal conditions, it is expected that newborn screen testing will be completed and reported to the birth hospital and primary MD within 7 days of birth.
- While guidelines recommend which disorders should be included on newborn screening disease panels, not all state programs test for all disorders. Therefore, it is important for physicians caring for newborns

and pediatric patients to know which disorders are included in the newborn screen-testing panel for their state and how to obtain the test results for an infant.

- Newborn screen ACT sheets are available from the American College of Medical Genetics; these sheets summarize information on newborn screening diseases and medical response to a report of a positive result.
- The early diagnosis of some IEM through newborn screening has resulted in another category of ER visits for patients with IEM. This group of diagnosed IEM patients is instructed by their treating physicians to take proactive and preventative steps at times of illness, or if there is a change in the child's condition. These instructions as well as testing recommendations and treatments are often summarized in an "emergency protocol." The emergency protocol typically provides the diagnosis, acute medical problems associated with the disorder and treatment recommendations. In some situations, hospitalization is required to continue the appropriate supportive treatment and to prevent worsening of the patient's metabolic disorder.
 - For many disorders, intravenous caloric support with dextrose can prevent or treat metabolic problems, such as ketosis in organic acid disorders and hypoglycemia in FAODs. In general, patients with these disorders cannot be discharged from the ED unless metabolic problems have been resolved and the patient can maintain an adequate oral caloric intake.
 - For some disorders, patients must also be able to take prescribed medications. Arginine and ammonia-conjugating medication (Buphenyl) is needed to control blood ammonia levels in urea cycle defects.

TREATMENT

- If the diagnosis of IEM is known, targeted evaluation can be initiated to determine the presence of specific problems associated with the specific IEM. Clinical and laboratory evaluation will identify areas requiring treatment. Additional general laboratory testing should be performed (Table 81-3) to evaluate the patient for associated medical problems. This approach will also increase the likelihood that medical problems, such as hyperammonemia or rhabdomyolysis, which require further evaluation and specialized treatment, will be detected.
- In general, the initial treatment of medical problems in patients with IEM follows recommendations outlined in the respective chapters.

- In some clinical situations, it is necessary to address multiple problems simultaneously. For example, treatment of dehydration and hypoglycemia in a patient with an FAOD requires administration of fluids with high dextrose content.
- Consultation with a metabolic specialist, such as a biochemical/clinical geneticist, may be indicated for determining the most appropriate course of treatment and diagnostic evaluation.
- When serious medical problems or complications are present, hospitalization, potentially in a PICU, is indicated. Hospitalization is often required to address medical problems resulting from the acute illness and to prevent worsening of the patient's condition.
- ED patients with an IEM who have responded well to initial treatment and have remained stable during a period of observation are capable of resuming their normal routine of diet and medication at home. However, if there is clinical evidence to suggest that problems might recur, hospitalization is indicated.

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QUESTIONS

1. A 3-week-old neonate is brought to the emergency department for lethargy. The parents report an uneventful neonatal hospital course. Per the mother, the infant was doing well. She reports 2 days prior to his decreased activity, he began sneezing and coughing and taking less po. There is no significant family history. Which of following statements correctly identifies the pattern in which the majority of inborn IEM are inherited?
 - A. Autosomal dominant
 - B. X-linked recessive
 - C. Unpredictable mutation
 - D. Autosomal recessive
 - E. X-linked dominant
2. A 3-month-old male infant is brought to the ED due to recurrent episodes of decreased responsiveness. Physical examination is essentially normal, but laboratory evaluation reveals an elevated ammonia level. Which of the following defects should be considered?
 - A. Ornithine transcarbamylase deficiency
 - B. Glycogen storage disease
 - C. Disorder of pyruvate metabolism
 - D. Galactosemia
 - E. Mitochondrial myopathy
3. A concerned mother brings her 4-year-old son for evaluation. She states that she recently read an article, which indicated that not all IEM are diagnosed in the neonatal period and the initial presentation can occur much later in life. She states that lately her son has become a “picky-eater” and appears malnourished to her. You attempt to reassure the mother. Which of the following, if present, would be a common presenting symptom of IEM?
 - A. Hypertonicity
 - B. Cardiomegaly
 - C. Thrombocytosis
 - D. Constipation
 - E. Altered mental status
4. A neonatal girl is born to a family with a history of ornithine transcarbamylase deficiency. The parents are extremely anxious and worried about whether or not their daughter will be affected with IEM as well. Which of the following IEM is usually covered by neonatal screening?
 - A. Amino acid disorder
 - B. Organic acidemia
 - C. Fatty acid oxidation defect
 - D. Galactosemia
 - E. All of the above

ANSWERS

1. D. The majority of these conditions are inherited in an autosomal recessive pattern. The family history is usually negative, depending on an autosomal recessive inheritance pattern, with a 25% risk for affected siblings.
2. A. Hyperammonemia is seen in urea cycle defects, including ornithine transcarbamylase deficiency, FAODs, and organic acidemias. The other options are not associated with hyperammonemia. Glycogen storage disease typically presents with hypoglycemia.
3. E. The most common presenting symptom of IEM in general is altered mental status. Other common presentations include hypotonia, cardiomyopathy, thrombocytopenia, vomiting, and seizures.
4. E. Three groups of biochemical genetic disorders are screened for using tandem mass spectrometry: amino acid disorders, organic acidemias, and FAODs. Galactosemia testing is also included on most newborn screening panels. Ornithine transcarbamylase deficiency is an x-linked recessive disorder, resulting in affected male infants with hyperammonemia. Therefore, it would be unlikely for their daughter to be affected with this particular disorder.

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82 MALE GENITOURINARY PROBLEMS

John W. Williams

TESTICULAR TORSION

- Testicular torsion is a urologic emergency and Doppler ultrasound is the test of choice.
- Testicular torsion may occur at any age but is more common in neonates and adolescence.
- Abnormal anatomy and horizontal lie of the testes allow the testes to twist within the scrotal sac, compromising flow through the associated artery (Fig. 82-1).
- Complete torsion may result in testicular necrosis in as little as 4 hours.
- Sudden onset of unilateral scrotal pain is typical, followed by vomiting. There may be a recent history of blunt trauma or epididymitis.
- Pain may be intermittent, which may represent episodes of spontaneous de-torsion.
- A swollen, erythematous, very tender hemiscrotum is typical; the testicle may be high-riding and have a horizontal lie.
- Doppler ultrasound is highly sensitive and specific.
- Urinalysis may suggest UTI but should not delay prompt urologic consultation.
- In some cases, manual detorsion by turning the testicle outward toward the thigh, like “opening a book,” may be indicated while awaiting the urologist (Fig. 82-2).
- A “blue-dot” sign may be noted with torsion of the appendix testis, which may be difficult to distinguish from testicular torsion, but is self-limited.

EPIDIDYMITIS

- Approximately one-third of children with scrotal pain have epididymitis, which is more common in adolescents and adults than young children.
- It can result from spread of infection from urethra or bladder, involving typical UTI pathogens in young children and sexually transmitted diseases in adolescents; viruses and chemical inflammation may also be implicated.
- Dull unilateral scrotal pain with fever and vomiting may precede swelling of the hemiscrotum; onset is usually gradual.
- Urinary symptoms may be present.
- Examination reveals an erythematous, warm, and swollen epididymis, perhaps at the superior pole of the testicle; the testicle itself should be nontender and have a normal lie.
 - Color Doppler ultrasound is necessary to rule out torsion and may show increased blood flow to the affected testis.
- Pyuria is present in less than half the cases, however urine should be cultured. Viral infections are common.
 - Children under 1 month with an associated UTI should be admitted for IV antibiotics; children under 3 months may be treated similarly, with ampicillin and an aminoglycoside or cefotaxime (Table 82-1).
- Older children may be treated with outpatient antibiotics (TMP-SMX or cephalexin) and analgesics, bed rest and scrotal elevation.
- Sexually active children can be treated with ceftriaxone IM and doxycycline PO (or erythromycin under 9 years of age).
- Urologic consultation and follow-up is recommended.

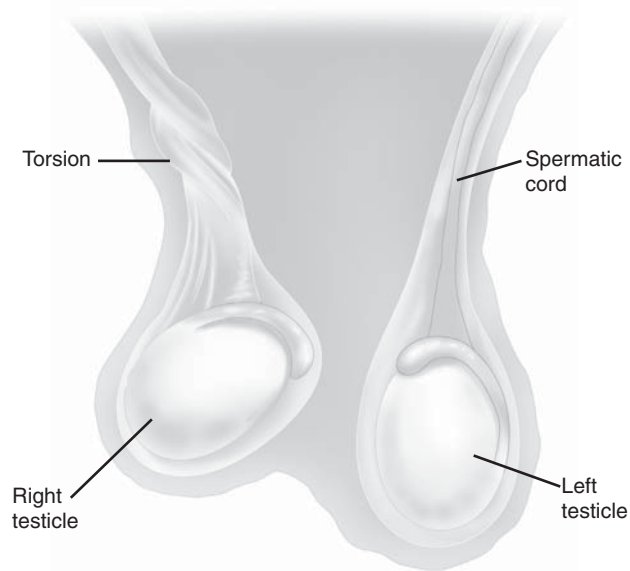


FIG. 82-1. Torsion of the right testicle.

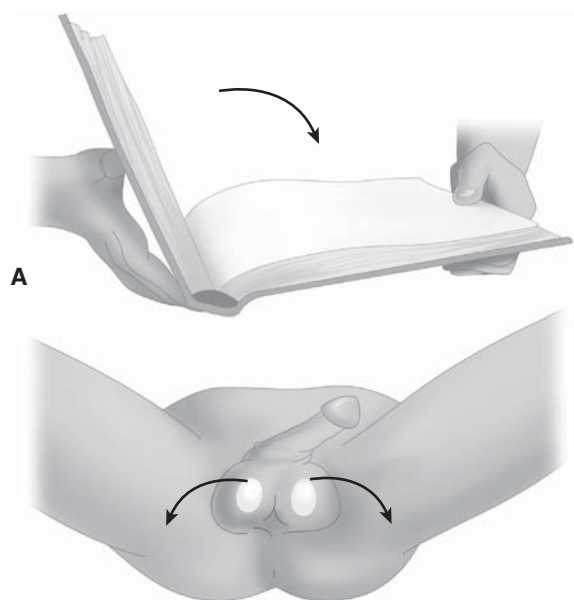


FIG. 82-2. Testicular detorsion.

SCROTAL TRAUMA

- Blunt trauma from a direct blow or a straddle injury can result in a scrotal hematoma.
- Testicular rupture may result in a scrotum tense with blood; ultrasound is necessary to evaluate followed by prompt urologic consultation.

TESTICULAR TUMORS

- Testicular tumors are rare in childhood and undescended testes are much more likely to contain a tumor than descended testes.

- Tumors are usually painless; patients may present only with a smooth, firm mass.
- Testing includes UA, CBC, fetoprotein, and β -HCG levels, and ultrasound evaluation.

INGUINAL HERNIA

- Indirect inguinal hernia repair is the most common pediatric surgery; this occurs in approximately 2% of children, usually right-sided and in males under 1 year.
- Peritoneal contents herniate through the processus vaginalis into the scrotal sac.

TABLE 82-1 Antibiotic Therapy for Epididymitis in Children

ANTIBIOTIC	DOSE	ROUTE
Outpatient management		
<i>Nonsexually active</i>		
Trimethoprim-sulfamethoxazole, or Cephalexin	8–10 mg/kg per 24 h 25–50 mg/kg per 24 h	PO bid for 10–14 d PO qid for 10 d
<i>Sexually active</i>		
Ceftriaxone, plus (if >8 y) Doxycycline, or Erythromycin	250 mg 100 mg 50 mg/kg per 24 h	IM PO bid for 10 d PO qid for 10 d
Inpatient management		
Ampicillin Plus gentamicin, or Cefotaxime, or Ceftriaxone	100 mg/kg per 24 h 7.5 mg/kg per 24 h 50–150 mg/kg per 24 h 50–100 mg/kg per 24 h	IV q 6 h IV q 8 h IV q 6 h IV q 12–24 h

PO, orally; bid, twice daily; qid, 4 times daily; IM, intramuscularly.

Note: ceftriaxone IV is incompatible and possibly lethal with calcium-containing parenteral solutions, and its use is being reconsidered for in-patient pediatrics; do not use it in hyperbilirubinemic neonates.

- Patients present with a bulge into the scrotal sac, which is often painless.
- Fever, vomiting, pain, or poor feeding may indicate incarceration of the hernia; examination may reveal a firm, tender, nonreducible mass.
- Incarcerated hernias can become strangulated with ensuing peritonitis and shock.
- In cases where the physical examination is inconclusive, ultrasound can differentiate among masses.
- 95% of incarcerated hernias can be reduced with firm finger pressure, analgesics, cold application, and Trendelenburg positioning.
- Prompt surgical consultation is necessary for all non-reducible hernias.

HENOCH-SCHÖNLEIN PURPURA

- Characterized by purpuric rash, nephritis, arthritis, hematuria and abdominal pain, with or without scrotal involvement; may be difficult to distinguish from torsion.

HYDROCELE AND VARICOCELE

- Hydroceles usually occur in infants and varicoceles in adolescent males.
- A hydrocele is a fluid-filled sac within the scrotum, usually nonpainful, which may be associated with (and can be difficult to differentiate from) an inguinal hernia.
- A varicocele is a painless scrotal swelling of veins described as looking like a “bag of worms.”
- Varicoceles are usually left-sided and benign, although they can represent an obstruction of the renal vein due to a tumor.

SCROTAL SWELLING

- Idiopathic scrotal edema occurs in prepubertal boys and involves thickening and erythema of the scrotum (not the testes), which is sometimes painful and pruritic; it may be preceded by trauma, insect bites or local irritation; treatment is with antihistamines.
- Benign scrotal swelling must be differentiated from cellulitis.

PHIMOSIS

- The normal prepuce (foreskin) usually cannot be retracted over the glans in infants; phimosis occurs in



FIG. 82-3. Paraphimosis.

children when the distal prepuce is inflamed and cannot retract normally over the glans penis.

- Painful swelling and erythema of the prepuce (posthitis) may be associated with infection of the glans also (balanoposthitis).
- Signs of urinary obstruction (unable to urinate or frequency, abdominal pain or fullness) should be evaluated with urinalysis and perhaps ultrasound.

PARAPHIMOSIS

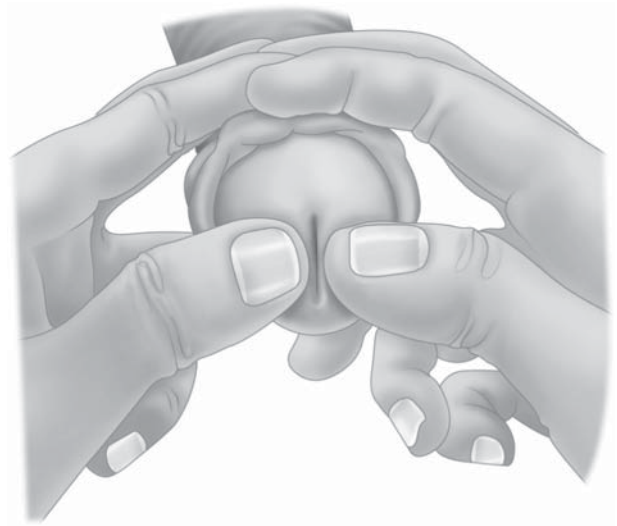
- The prepuce is retracted and trapped behind the glans, which may become swollen from venous congestion (Fig. 82-3).
- Patients must be evaluated for hair tourniquets or other foreign bodies.
- Manual reduction of prepuce over the head of the glans may require anesthesia (Fig 82-4).
- Inability to retract the prepuce is an indication for urgent urologic consultation.

BALANITIS AND BALANOPOSTHITIS

- Infection of the glans and prepuce are more common in uncircumcised preschool-age males (Fig 82-5).
- Signs and symptoms include swelling, erythema, penile discharge, dysuria, and bleeding.
- Etiology is related to inadequate hygiene or chemical irritation (soaps) and associated with both gram-positive and gram-negative organisms and candida.
- Testing is usually not necessary but urinalysis and culture may be useful.



A



B

FIG. 82-4. Reduction of paraphimosis.



FIG. 82-5. Balanoposthitis.

- Sitz baths with gentle cleaning and topical antibiotics are the treatment; severe cases may warrant an oral antibiotic such as cephalexin.

PRIAPISM

- Prolonged painful erection unrelated to sexual stimulation is uncommon in children except with sickle cell disease.
- Results from two mechanisms: low flow or high flow.
 - In low flow or ischemic priapism, there is little cavernous blood flow and the penis is painful and tender; typical in sickle cell disease and polycythemia.
 - In high flow or engorgement priapism, the penis is typically not fully rigid or painful and trauma is the most common etiology.
 - Both mechanisms result in corpus cavernosum engorgement with flaccid corpora spongiosum and glands, leading to stasis, acidosis, thrombosis (and eventually to fibrosis and impotence if untreated).
- Precipitating factors include drugs, trauma, infection, and sexual intercourse.
- Most cases can be treated medically with oxygen, hydration, and analgesics.

- Urinary retention is relieved by placement of a catheter.
- Urgent urologic consultation is necessary; the specialist may perform therapeutic aspiration with or without irrigation for cases of ischemic priapism, perhaps followed by injection of a vasoconstrictor (phenylephrine).

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QUESTIONS

1. A 13-year-old male presents with severe left scrotal pain that started suddenly an hour before while he was wrestling with his brother. The boy has vomited once. Examination shows a tender, swollen, erythematous left scrotum, which is made worse by elevation of the testes. What is the most appropriate next step?
 - A. Catheterize the patient for urinalysis.
 - B. Provide pain relief, call the urologist and wait for the consultation.
 - C. Emergent urologic consultation, attempt manual detorsion and obtain a stat color Doppler ultrasound to assess for blood flow.
 - D. Attempt manual detorsion and refer for urology follow-up if successful
 - E. Initiate antibiotics and observe.
2. A 12-year old is brought to the emergency department for intermittent scrotal pain. His mother reports that he began complaining of pain shortly after beginning summer workout sessions for gymnastics. Which of the following is true regarding a suspected diagnosis of torsion of an appendix testis?
 - A. An abnormal urinalysis rules out testicular torsion.
 - B. The “blue-dot” sign confirms torsion of a testicular appendage.
 - C. Emergent ultrasound is indicated to confirm the diagnosis and rule out testicular torsion.
 - D. The condition is usually painless.
 - E. The condition is of the same significance as testicular torsion.
3. An 11-month-old boy is brought for evaluation of scrotal swelling and pain. The parents describe previous swelling in his scrotum, which the doctor “pushed back in.” Now in the emergency department he begins vomiting. Upon examination, you find a firm, tender scrotal mass that does not transilluminate. The next most appropriate action is
 - A. Give the patient analgesics, place in Trendelenberg and apply gentle, firm pressure to the hernia.
 - B. Discharge on analgesics and refer to a pediatric surgeon.
 - C. Order an ultrasound to check vascular blood flow.
 - D. Consult with a pediatric surgeon.
 - E. Admit for observation.
4. A toddler is brought to the emergency department by his parents. The parents report that the child has been crying constantly for the last several hours. On examination, you note that the prepuce is retracted and trapped behind the glans. On closely evaluation, you discover a hair tourniquet. Which of the following is true about this condition?
 - A. It only occurs in circumcised males.
 - B. It is a nonpainful condition.
 - C. It may be caused by poor hygiene.
 - D. Treatment is prompt reduction of the prepuce over the head of the glans.
 - E. Should prompt urgent urologic consultation.

ANSWERS

1. C. Testicular torsion can occur at any age with peak incidence in adolescence. Torsion of the testes is a urologic emergency and should prompt emergent urologic consultation. Meanwhile, a stat Doppler ultrasound should be obtained to assess for blood flow. The child should be made more comfortable and manual detorsion may be attempted, but this should not delay the study or consultation.
2. C. Torsion of an appendix testis can be very painful and clinically indistinguishable from testicular

torsion. Thus, emergent Doppler ultrasound is indicated and urologic consultation for any cases in doubt. Treatment is bed rest, analgesics, scrotal elevation, and urologic follow-up.

3. A. Inguinal hernias usually occur on the right in males in their first year. It is appropriate to attempt manual reduction with firm finger pressure on the internal inguinal ring after analgesics or sedation. Systemic signs that a hernia has become incarcerated include fever, vomiting, or poor feeding, abdominal pain and a firm, tender and nonreducible mass in the inguinal area and scrotum. Patients with incarcerated hernias or signs of peritonitis must have immediate surgical consultation, fluid resuscitation, and antibiotics.
4. D. In paraphimosis the prepuce (foreskin) becomes trapped in its retracted position, constricting the distal penis proximal to the coronal sulcus. This causes venous congestion of the glans, which then swells, becomes more painful and the foreskin is more difficult to reduce. It only occurs in uncircumcised males and is not necessarily related to hygiene (as balanitis and posthitis can be). Prompt reduction is important to reduce ischemic injury to the glans; this requires analgesia and possible sedation of the child. Urologic consultation is necessary only if the reduction is unsuccessful.

83 URINARY TRACT DISEASES

John W. Williams

URINARY TRACT INFECTION

- Signs and symptoms of urinary tract infection (UTI) can be nonspecific and children may not complain of dysuria.
- Fever may be the only manifestation of a UTI and the temperature can be high.
- Up to 7% of infants and children with fever without a source may have a UTI.
- Males may have a higher risk of UTI than females in the early months of life, but by one year of age females predominate.
- 75% of infants under 3 months with fever and a UTI are bacteremic.
- Most UTIs in children are caused by *Escherichia coli*, spread via fecal contamination and retrograde ascent into the bladder.
- Anterograde flow of urine and complete bladder emptying is the main defense against UTIs.

- Nonspecific symptoms of UTI in infants and young children include fever, vomiting, anorexia, diarrhea, lethargy, oliguria, and failure to thrive.
- Older children may exhibit abdominal pain, flank or back pain, dysuria, frequency, or hematuria.
- Dysfunctional elimination is a risk factor for UTI.
- Patients with fever, vomiting, and abdominal or flank pain should be evaluated for pyelonephritis.
- While a urine culture is the gold standard for diagnosing a UTI, a urinalysis showing more than five WBCs per HPF or Gram stain showing bacteria is indicative of a UTI (Fig. 83-1).
- A “bagged” urine is the least reliable method of testing and is only useful if it is negative.
- Urinary tract catheterization is the preferred method for obtaining urine in febrile infants and young children.
- Antibiotic choice for treating UTIs depends on local resistance patterns and effectiveness against *E coli* (Table 83-1).
 - There may be less resistance to first-generation cephalosporins than to ampicillin, amoxicillin, and trimethoprim-sulfamethoxazole (TMP-SMX).
- All neonates and infants less than 2 months and patients with pyelonephritis, urinary obstruction, or immunocompromise should be admitted for IV antibiotics.
- Stable patients who can tolerate oral medications, who have reliable caretakers, may be discharged with 10 to 14 days of oral antibiotic therapy.
- Children with UTIs should be evaluated for urinary tract abnormalities; up to half of these patients will have abnormal tests in subsequent testing.
 - The most common urinary tract abnormality is vesico-ureteral reflux, usually detected by a voiding cystourethrogram (VCUG).
 - Renal ultrasonography (RUS) can detect anatomic abnormalities, hydronephrosis, or nephrolithiasis.

UROLITHIASIS

- Urolithiasis is uncommon in children and may be associated with metabolic disorders or developmental anomalies of the urinary tract.
- 90% of renal stones are radio-opaque.
- Urolithiasis may present with abdominal or flank pain, hematuria, and fever; urinalysis, culture, and renal function studies are essential and are often followed by an ultrasound or a noncontrast CT scan of the abdomen.
- Adequate hydration and pain control, further diagnostic studies and consultation with a pediatric urologist is usually indicated in these patients.

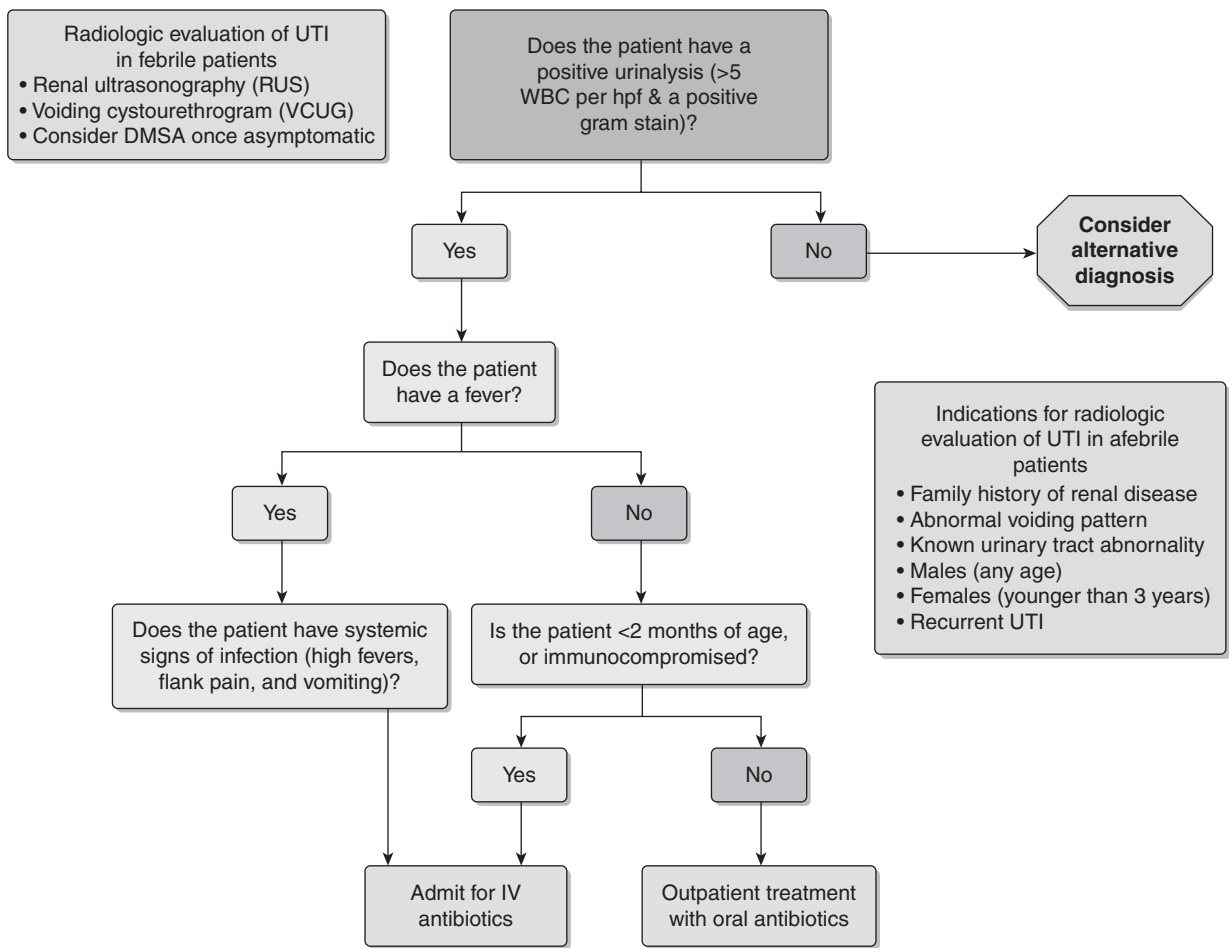


FIG. 83-1. Algorithm showing evaluation and management of suspected UTI.

TABLE 83-1 Antibiotic Therapy for Treatment of Urinary Tract Infections in Children

OUTPATIENT MANAGEMENT		INPATIENT MANAGEMENT	
Cotrimoxazole (trimethoprim-sulfamethoxazole)	10 mg/kg per 24 h (based on TMP component) divided bid	Ampicillin plus	100 mg/kg per 24 h divided qid
Cefixime	8 mg/kg per 24 h divided bid	Cefotaxime	150 mg/kg per 24 h divided tid
Cephalexin	50 to 100 mg/kg per 24 h divided qid	Ceftriaxone ^a	75 mg/kg per 24 h
		Ceftazidime	150 mg/kg per 24 h divided qid
		Gentamicin	5–7.5 mg/kg per 24 h divided tid

Bid, twice daily; qid, 4 times daily; tid, 3 times daily.

^aCeftriaxone should be used with caution in neonates and infants; read latest CDC guidelines.

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QUESTIONS

1. A 4-month-old male is brought to the emergency department (ED) for evaluation of fever for 1 day. His mother reports a temperature of 38.4 at home associated with decreased po intake. He received acetaminophen 2 hours prior to their arrival in the ED. He is afebrile with normal vital signs and an unremarkable physical examination. As part of your fever workup, you obtain an urinalysis which is nitrite positive with 50 WBCs. Which of the following statements regarding UTIs is most accurate?
 - A. Circumcision is a risk factor for UTIs in young males.
 - B. In febrile neonates younger than 2 months without a definite source of fever, the likelihood of a UTI is at least 5%.
 - C. A “bagged” urine sample and urine dipstick is adequate to diagnose UTI in infants.
 - D. Infants with UTIs are rarely bacteremic.
 - E. Females have a higher risk for UTI than males in all age groups.
2. Parents bring a 6-year-old girl for evaluation of dysuria and abdominal pain for the past 2 days. She has mild suprapubic tenderness upon examination. A urine dipstick is performed which shows only trace leukocyte esterase. The most appropriate action is
 - A. Obtain a catheterized urine specimen for urinalysis and culture, and start antibiotics.
 - B. Give analgesics and obtain a CT scan of the abdomen.
 - C. Obtain a “clean-catch” urine sample and attempt urine dipstick again.
 - D. Obtain an intravenous pyelogram.
 - E. Start empiric antibiotics without further testing and refer for follow-up.
3. A teenager complains of left abdomen and flank pain for 3 hours and now is nauseated. She has mild abdominal tenderness and marked left costo-vertebral tenderness. Her urine dipstick shows hematuria and she is not pregnant. The most useful test in establishing the diagnosis is
 - A. A formal urinalysis.
 - B. An intravenous pyelogram (IVP).
 - C. A voiding cystourethrogram (VCUG).
 - D. A noncontrast CT of the abdomen and pelvis.
 - E. Renal ultrasound.

ANSWERS

1. B. Uncircumcised boys have up to a 10-fold greater risk of UTI than circumcised males. Febrile neonates younger than 2 months have a 4.6% likelihood of a

UTI in general, higher in those patients without a definite source of fever (but less likely, if there is a definite source of fever). A “bagged” urine sample is usually contaminated and a urine dipstick is inaccurate; catheterized specimen and urine culture is the best test for diagnosing a UTI, particularly in infants. Up to 50% of infants under 3 months with a UTI are bacteremic.

2. A. Urine culture is the most accurate and reliable test to diagnose a UTI. Urine dipsticks (even those tests showing leukocyte esterase or nitrites) will miss at least 12% to 20% of UTIs. CT scan of the abdomen may be useful if appendicitis is suspected but has no place in the evaluation of UTIs. An IVP may be useful in the evaluation of nephrolithiasis. Empiric treatment without culture may lead to problems in cases of resistance.
3. D. Nephrolithiasis and urolithiasis, stone formation in the urinary system, is best shown by computed tomography, which will also demonstrate underlying abnormalities and related intraabdominal pathology. A formal urinalysis may establish a concomitant UTI but will not help evaluate for renal stones. VCUG is the test of choice for children with suspected urethral pathology and is useful in the workup of vesico-ureteral reflux. While renal ultrasound may be used to identify stones, it is less useful in identifying other pathology and in differentiating obstructive from nonobstructive causes of hydronephrosis.

84 SPECIFIC RENAL SYNDROMES

Roger M. Barkin

ACUTE GLOMERULONEPHRITIS

- Glomerulonephritis is a histopathologic diagnosis acutely associated with clinical findings of hematuria, edema, and hypertension.
- It commonly follows a pharyngitis caused by group A β -hemolytic *Streptococcus* in children between 3 and 7 years of age.
- Timely treatment of pharyngitis does not clearly decrease the incidence of acute glomerulonephritis.
- Initial findings may be mild facial or extremity edema only, with a minimal rise in blood pressure.
- Patients uniformly develop fluid retention and edema and commonly have hematuria (90%), hypertension (60–70%), and oliguria (80%). Fever, malaise, and abdominal pain are frequently reported. Anuria and

renal failure occur in 2% of children. Urinalysis reveals microscopic or gross hematuria. Erythrocyte casts are present in 60% to 85% of hospitalized children. Proteinuria is generally less than 2 g/m² per 24 hours. Hematuria and proteinuria may present independently and require a specific evaluation. Leukocyturia and hyaline and granular casts are common.

- The fractional excretion of sodium as a reflection of renal function may be reduced. The blood urea nitrogen (BUN) level is elevated disproportionately to the creatinine level.
- Total serum complement and specifically C₃ is reduced in 90% to 100% of children during the first 2 weeks of illness, returning to normal within 3 to 4 weeks. The antistreptolysin (ASO) level is elevated, consistent with a previous streptococcal infection. Fluid and salt restriction essential to normalize intravascular volume. Diuretics are often required.
- Elevated blood pressure may require specific pharmacologic management.
- Specific complications, such as congestive heart failure, renal failure, and hyperkalemia, must be anticipated and treated.
- Recovery is usually complete.
- Children without evidence of hypertension, congestive heart failure, or azotemia may be followed closely at home.
- A nephrologist is usually consulted.

NEPHROTIC SYNDROME

- Nephrotic syndrome is associated with increased glomerular permeability, which produces massive proteinuria. Hypoalbuminemia results, producing a decrease in the plasma osmotic pressure. Edema develops.
- The etiology is generally idiopathic, but has been associated with glomerular lesions. Intoxications, allergic reactions, infection, and other entities have also been associated with the syndrome (Table 84-1)
 - Primary nephrotic syndrome occurs more commonly in children younger than 5 years while secondary nephrotic syndrome is more common in older children.
- Patients frequently have edema, often with a history of a preceding flu-like syndrome. Edema initially is present periorbitally and may become generalized, associated with weight gain. Ascites may be caused by edema of the intestinal wall, often associated with abdominal pain, nausea, and vomiting. Pleural effusion or pulmonary edema may occur. Blood pressure may be decreased if the intravascular volume is

TABLE 84-1 Nephrotic Syndrome: Etiology

Primary renal disorders	Nil or minimal change disease Focal segmental glomerulosclerosis Membranoproliferative glomerulonephritis Membranous glomerulonephritis
Intoxication	Heroin Mercury Probenecid Silver
Allergic reaction	Poison ivy or oak, pollens Bee sting Snake venom
Infection	Bacterial Viral: Hepatitis B, cytomegalovirus, Epstein-Barr virus Protozoa: Malaria, toxoplasmosis
Neoplasm	Hodgkin's disease, Wilms' tumor, etc.
Autoimmune disorder	Systemic lupus erythematosus
Metabolic disorder	Diabetes mellitus
Cardiac disorders	Congenital heart disease Congestive heart failure Pericarditis
Vasculitis	Henoch-Schönlein purpura Wegener's granulomatosis

- depleted or increased in the presence of significant renal disease. Renal failure may develop.
- Infection is probably the most common complication, related to the increased risk of peritonitis and concomitant immunosuppression due to the steroid therapy. Immune protein levels, including IgG, are low due to urinary losses.
 - Children's blood is hypercoagulable, leading to an increased risk of thromboembolism. Renal vein thrombosis is probably underrecognized but should be suspected if hematuria, flank pain, and decreased renal function occur.
 - Hypoalbuminemia is common, as well as proteinuria and hyperlipidemia.
 - BUN and creatinine levels are elevated in 25% of children.
 - Serum complement is decreased.
 - Plasma cholesterol carriers (low-density lipoprotein and very-low-density lipoprotein) are increased. A renal biopsy should be considered if the following poor prognostic signs are present:
 - Age more than 10 years
 - Azotemia
 - Decreased complement
 - Hematuria persistent hypertension
 - No response to steroids
 - Other causes of edema should be excluded, including congestive heart failure, vasculitis, hypothyroidism, starvation, cystic fibrosis, protein-losing enteropathy, and drug ingestions (such as steroids or diuretics).

- The majority of patients should be hospitalized initially, usually in consultation with a nephrologist.
- Treat hypovolemia with albumin and fluids.
- Monitor closely and treat hypertension if it occurs.
- After diagnosis and stabilization, patients without complications (<10 years, normal complement, no gross hematuria, no large protein loss) are started on prednisone at a dose of 2 mg/kg per 24 hours up to 80 mg per 24 hours and tapered once a response is noted.
 - Nearly 75% of patients will respond within 14 days.
 - Limited response to initial steroid therapy is generally predictive of a poor outcome.
- Salt and water restriction should be initiated.
- Diuretics may be needed if there is pulmonary edema or respiratory distress. However, they must be used judiciously to avoid vascular volume depletion and electrolyte abnormality. Watch for signs of infection since these patients are considered immunocompromised.
- Avoid deep vein punctures if possible to avoid triggering a deep vein thrombosis.
- There is a tremendous spectrum of severity of clinical disease ranging from mild elevation of BUN with anemia to total anuria due to acute nephropathy with severe anemia and thrombocytopenia.
- Ultimately, patients may develop hypertension; anemia with pallor, petechiae, and easy bruising; hepatosplenomegaly; and edema.
 - Hypertension occurs in up to 50% of patients.
- Irritability or lethargy may develop. Seizures occur in 40% of the cases.
- Hyponatremia and hypocalcemia are common.
- Acute abdominal conditions including intussusception, bowel perforation, and toxic megacolon can occur. Hepatic and pancreatic injury can occur in HUS.
- There may be cardiac involvement with cardiomyopathy, myocarditis, or high-output failure.
- Recurrences may occur, often without a prodrome, and may be associated with a high mortality rate.
- Laboratory evaluation should include assessment of renal function including electrolyte, BUN, and creatinine levels and urinalysis. Hematologic studies reveal low hemoglobin with a microangiopathic, hemolytic anemia. Burr cells are common. Platelets are usually decreased below 50 000/mm³. C-reactive protein is commonly elevated. Coagulation studies are usually normal.

HEMOLYTIC UREMIC SYNDROME

- Nephropathy, microangiopathic hemolytic anemia, and thrombocytopenia are found in patients with hemolytic uremic syndrome (HUS).
- This syndrome commonly occurs in children younger than 5 years following an episode of gastroenteritis or respiratory infection.
- Siblings may also develop the disease due to a familial genetic component.
- The illness has an acute onset with rapid progression to renal failure and thrombocytopenia.
- Associated infections can be found.
 - *Escherichia coli* serotype 0157:H7 is the most commonly found organism, producing a cytotoxin that inhibits protein synthesis leading to cell death in gastrointestinal organs.
 - *Shigella*, *Salmonella*, and group A *Streptococcus* may be associated with HUS, as well as coxsackievirus, influenza, and respiratory syncytial virus (RSV).
- Patients usually have a history of gastroenteritis with vomiting, bloody diarrhea, and crampy abdominal pain within 2 weeks of the onset of HUS.
- Children who develop HUS without a prodrome of gastroenteritis have a poor prognosis.
- Low-grade fever, pallor, hematuria, oliguria, and gastrointestinal bleeding occur.
- Central nervous system deterioration can occur with a spectrum from irritability to seizures or coma.
- Initial stabilization is followed by admission to an appropriate medical center.
- Volume overload may occur secondary to anemia.
- Hypertension may occur and appears to be caused by increased renin levels.
 - Treatment is recommended if the diastolic pressure is above 120 mm Hg. A variety of agents may be used, including nifedipine, labetalol, captopril, and hydralazine.
- Renal failure requires meticulous balancing of intake and output with specific treatment of hyperkalemia, acidosis, hypocalcemia, hyperphosphatemia, and other metabolic abnormalities. Peritoneal dialysis may be required, especially when the BUN is more than 100 or when congestive heart failure, encephalopathy, or hyperkalemia are present. Peritoneal dialysis is also indicated when anuria has been present for 24 hours.
- A serum hemoglobin <5 g/dL or hematocrit <15% generally requires treatment with packed red blood cells, infused slowly.
- Platelet survival is shortened and platelet infusions may be required in children with active bleeding.
- Seizures require specific management and are usually caused by hypertension or uremia.

- Acute treatment includes stabilization and anticonvulsants, as well as a consideration of emergency dialysis.
- Heparin and streptokinase have been tried without significant success.

ACUTE RENAL FAILURE

- The etiology of acute renal failure may be categorized on the basis of the type of renal injury. It may be prerenal (decreased perfusion of the kidney), intrarenal (damage to the actual nephron), or postrenal (downstream obstruction of the urinary tract; Table 84-2).
- Patients may have oliguria with urine output less than 1 mL/kg/h or be nonoliguric with an output excessive for the volume status. Azotemia may be noted.
- Laboratory evaluation should include electrolytes, studies of renal function, and a search for the underlying pathology.
- The creatinine clearance is a good measure of glomerular filtration rate (GFR) and is useful in initial assessment and ongoing monitoring. A 24-hour urine is normally needed.
- Normal values are the following:
 - Newborn and premature: 40 to 65 mL/min/1.73 m².
 - Normal child: female, 109 mL/min/1.73 m² or male, 124 mL/min/1.73 m².
 - Adult: female, 95 mL/min/1.73 m² or male, 105 mL/min/1.73 m².
- Ultrasonography is also important in the evaluation of these patients.
- Combining data from serum, urine, and ultrasonography helps differentiate among prerenal, intrarenal, and postrenal failure (Table 84-2).

- Initial management must focus on stabilization with correction of fluid imbalance (Fig. 84-1).
- If the intravascular volume is adequate or overloaded, urine output may be enhanced with furosemide, usually in an initial dose of 1 mg/kg increased up to 6 mg/kg/dose.
 - Mannitol may be administered if there is no response to furosemide. The dose is 0.5 to 0.75 mg/kg/dose IV.
 - Do not use these agents if obstruction is present.
- In oliguric or anuric patients with decreased intravascular volume, fluid may be administered slowly, often in conjunction with monitoring of the central venous pressure.
- Low-dose dopamine occasionally may be used to increase renal blood flow and GFR.
- Those with high urine output must receive a significant amount of fluid to avoid hypovolemia.
- Children having acute hypertension with a diastolic pressure more than 100 mm Hg should be treated parenterally because of the risk of seizures, encephalopathy, and other sequelae.
- Specific and immediate treatment for a potassium level >7.0 mEq/L is required.
- Treatment may include calcium chloride, 20 to 30 mg/kg slowly; sodium bicarbonate, 1 to 2 mEq/kg/dose; or glucose and insulin infusion of 1 mL/kg of D₅₀ W followed by 1 mL/kg of D₂₅ W and 0.5 U/kg of regular insulin per hour to keep serum glucose between 120 to 300 mg/dL. Kayexalate at 1 g/kg/dose every 4 to 6 hours mixed with 70% sorbitol, orally or rectally, may be useful after initial stabilization. Dialysis may be required for unresponsive fluid overload, severe hyperkalemia, severe hyponatremia or hypernatremia, unresponsive metabolic acidosis, BUN > 100 mg/dL, or altered level of consciousness secondary to uremia.

TABLE 84-2 Evaluation of Renal Failure

PRERENAL	INTRARENAL	POSTRENAL
Ultrasound: normal	Ultrasound: can have increased renal density or slight swelling	Ultrasound: dilated bladder or kidney
Serum BUN:creatinine ratio >15:1		History and examination may be diagnostic
Urine Na ⁺ <15 mEq/L	Urine Na ⁺ > 20 mEq/L	Indexes not helpful
Urine osmolality >500 mOsm/kg H ₂ O	Urine osmolality <350 mOsm/kg H ₂ O	
Urine: plasma creatinine ratio > 40:1	Urine: plasma creatinine ratio < 20:1 (often <5:1)	
Fractional excretion of Na ⁺ < 1 (<2.5 in neonates)	Fractional excretion of Na ⁺ > 2 (>2.5 in neonates)	
$\text{Fractional excretion of Na}^+ = \frac{\text{Urine Na}^+(\text{mEq/L})}{\text{Plasma Na}^+(\text{mEq/L})} \times \frac{\text{Plasma creatinine (mg/dL)}}{\text{Urine creatinine (mg/dL)}}$		

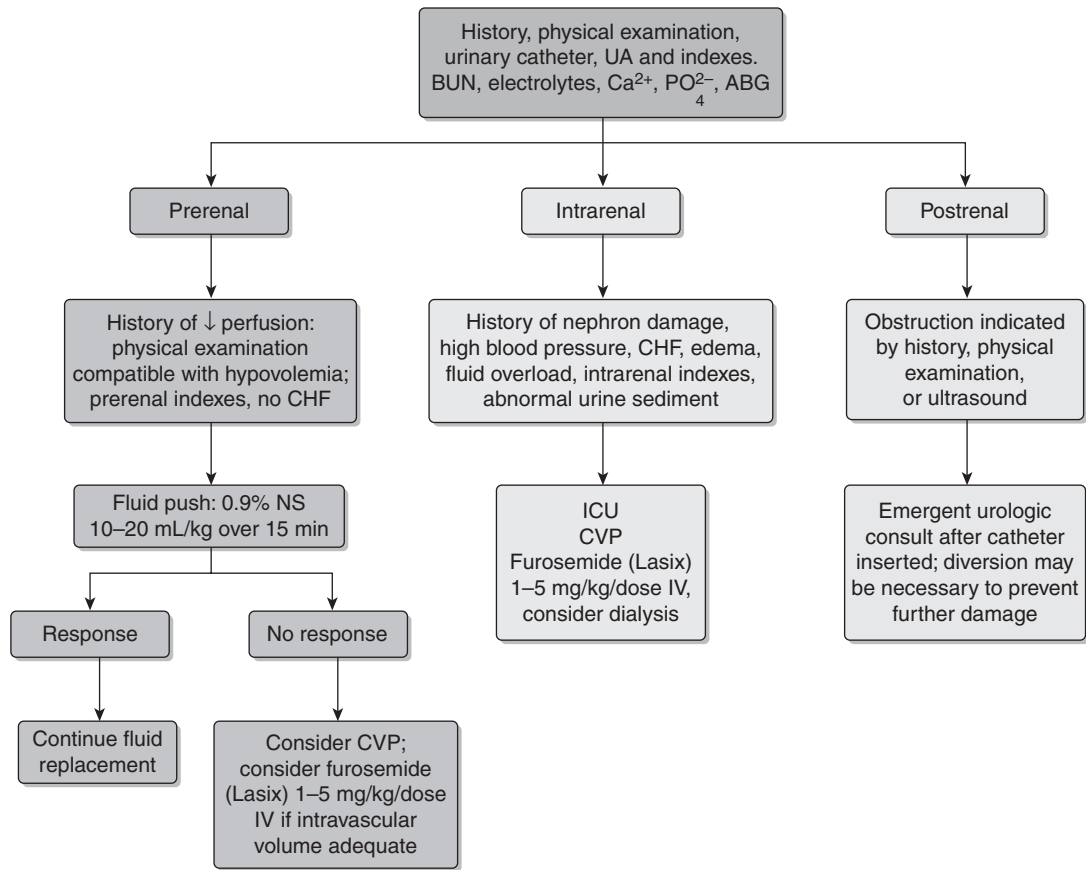


FIG. 84–1. Acute renal failure: initial assessment and treatment. UA, urine analysis; BUN, blood urea nitrogen; Ca, calcium; po, orally; ABG, arterial blood gas; CHF, congestive heart failure; NS, normal saline; ICU, intensive care unit; CVP, central venous pressure; IV, intravenous.

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QUESTIONS

- A 5-year-old girl is brought to the doctor's office with complaints of swelling of her hands and face. She has been well except for a sore throat 2 weeks ago, which was treated with penicillin. Her physical examination is unremarkable except for the edema and a mildly elevated blood pressure. Her urine is notable for microscopic hematuria and mild proteinuria. The most likely diagnosis is:
 - Nephrotic syndrome.
 - HUS.
 - Acute renal failure (idiopathic).
 - Urinary tract infection.
 - Acute glomerulonephritis.
- Which of the following is essential in the management of the patient described above?
 - Albumin
 - Steroids

- C. Fluid and salt restriction
 - D. Diuretics
 - E. Antihypertensives
3. A 3-year-old boy presents with low-grade fever, hematuria, and blood in the stool. He has been well except for an episode of gastroenteritis 2 weeks before which was associated with bloody diarrhea and crampy abdominal pain. On physical examination, he appears ill and his blood pressure is elevated. Laboratory evaluation shows anemia, azotemia, and thrombocytopenia. The most likely diagnosis is
 - A. Nephrotic syndrome
 - B. HUS
 - C. Acute glomerulonephritis
 - D. Acute renal failure
 - E. Intussusception
 4. In the case described above, an associated infection is found. What is the most commonly associated organism?
 - A. RSV
 - B. Shigella
 - C. Influenza virus
 - D. *E. coli* serotype 0157:H7
 - E. Group A *Streptococcus*

ANSWERS

1. E. This patient has acute glomerulonephritis. Patients uniformly develop fluid retention and edema and commonly have hematuria (90%), hypertension (60–70%), and oliguria (80%). Fever, malaise, and abdominal pain are frequently reported. Anuria and renal failure occur in 2% of children. Urinalysis reveals microscopic or gross hematuria and mild proteinuria. Nephrotic syndrome is associated with massive proteinuria and edema. HUS is usually associated with a prodrome of gastroenteritis and is typically very acute in onset and rapidly progressive. Progression to renal failure can occur with acute glomerulonephritis but is not usually an early finding.
2. C. Fluid and salt restriction is essential to normalize intravascular volume with acute glomerulonephritis. Albumin and steroids are used in the treatment of nephrotic syndrome. Diuretics and antihypertensives may be required in some cases of acute glomerulonephritis but are not essential in every case.
3. B. This boy has HUS. Nephropathy, microangiopathic hemolytic anemia, and thrombocytopenia are found in patients with HUS. This syndrome commonly occurs in children younger than 5 years following an episode of gastroenteritis or respiratory infection. The illness has an acute onset with rapid progression to renal failure and thrombocytopenia. The other renal syndromes listed are not associated with such an acute onset and rapid progression to severe illness. Intussusception may be associated with HUS but alone would not be associated with all these symptoms.
4. D. *E. coli* 0157:H7 is the most commonly associated infectious agent. However, all of these agents can be associated with HUS.

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85 PETECHIAE AND PURPURA

Malee V. Shah

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PETECHIAE CAUSED BY SEPSIS

- Petechiae found just over the face, neck, and upper trunk above the nipple line in a nontoxic patient with a history of significant coughing or forceful vomiting is likely due to increased venous pressure, and not an infectious cause.
- Purpura fulminans is a life-threatening condition characterized by hemorrhagic infarction of the skin and multiorgan dysfunction.
 - May be caused by acute severe gram-negative bacterial or other infections.
 - The organism most commonly implicated in pediatric patients is *Neisseria meningitidis* (>90%), followed by *Streptococcus pneumoniae* and group A and B streptococci.
 - Outbreaks occur in semiclosed communities, such as childcare centers, college dormitories, and military bases.
 - Transmission occurs by direct contact with secretions or fomites carrying the offending organism.
 - The sepsis-induced cutaneous lesions are similar regardless of the causative organism.
- Clinical course:
 - Skin necrosis begins with a region of dermal discomfort that quickly progresses to petechiae within minutes to hours.
 - The petechiae, which can be found anywhere on the body, will usually distribute acrally over the hands and feet. They then coalesce to form purpura.
 - Hemorrhagic infarction and subsequent skin necrosis can occur at this point without initiation of aggressive therapy.
 - Frank skin necrosis and gangrene are associated with more than 50% morbidity and mortality.
 - The majority of affected patients develop septic shock and DIC.
 - Early recognition of the disease state and timely treatment is crucial and will decrease mortality.
- Treatment:
 - Rapid systemic organ support is paramount for survival of these patients.
 - Supplemental oxygen and mechanical ventilation help reduce metabolic demands by taking away the work of breathing.
 - Aggressive fluid resuscitation is required to restore intravascular volume. Crystalloid solutions are the first-line choice for fluid resuscitation.
 - Hypotension may arise despite aggressive fluid resuscitation, and inotropic support may be necessary.
 - Administration of antibiotics should also be initiated soon after diagnosis. Empiric coverage should include a third-generation cephalosporin, since it provides good coverage against *N meningitidis* and streptococcus. Vancomycin should be added if methicillin-resistant *Staphylococcus aureus* is suspected.
 - Once a causative organism is identified with antibiotic sensitivities, the coverage can be narrowed.
- Complications:
 - Purpura fulminans almost always leads to full-thickness skin loss; thus, the treatment is similar to that of a burn patient.
 - The complications are also the same as a deep burn, including secondary infection and compartment syndrome.

- Debridement of the necrotic tissue and eventually grafting may be required. When the tissue necrosis is extensive, limb amputation may be necessary.
- A blood culture is valuable regardless of whether antibiotics have been given prior to obtaining it.
- Other cultures that should be obtained once the patient is stabilized are cerebrospinal fluid and urine cultures.
- Laboratory studies that may be useful are blood gas, DIC panel, lactate, and complete blood count.
- Highly sensitive rapid polymerase chain reaction (PCR) tests are available for detection of the most common serotypes of *N meningitidis*, which can be performed on blood, urine, and cerebrospinal fluid.

HENOCH–SCHONLEIN PURPURA

- The triggers of the immune complex cascade causing HSP are mostly infectious.
 - Mild viral respiratory infections (i.e. parvovirus)
 - Bacterial pharyngitis caused by group A β -hemolytic streptococcus
 - Certain vaccines have been implicated as possible offenders.
- Median age is 4 to 6 years. Boys are affected more often than girls.
- The severity of symptoms tends to be milder in patients younger than 2 years, and becomes more severe in older patients.
- A prodromal event can usually be identified a couple of weeks prior to the onset of symptoms.
- Clinical appearance:
 - Cutaneous involvement includes 1 to 10 mm palpable purpuric lesions and pinpoint petechiae distributed symmetrically over the lower extremities and buttock region.
 - Though the purpura is usually concentrated over the lower half of the body, they are not restricted to those areas.
 - Lesions are unrelated to any underlying coagulopathy.
 - Most patients with HSP will develop purpura during the course of the disease.
 - Joint manifestations are the second most common feature of HSP and can be seen in up to 80% of patients.
 - The vasculitis typically causes arthralgia and arthritis of the knees and ankles.
 - Arthritis may precede the onset of purpura by a week in 15% to 25% of patients.
 - Renal involvement is common and can be seen in up to 70% of children. It can manifest within a few days to 4 weeks after the onset of other symptoms.
- Although most patients have asymptomatic microscopic hematuria, some may present with an acute nephritic/nephritic syndrome. Of the patients who develop HSP nephritis, 1% to 7% will likely advance to end-stage renal disease.
- The risk factors for developing HSP nephritis are children older than 4 years, those with gastrointestinal bleeding, factor XIII activity less than 80% of normal, and/or those treated with factor XIII concentrate. The long-term morbidity of HSP is largely attributed to renal involvement.
- Orchitis may be present in 10% to 20% of males. Testicular torsion, though unlikely, should still be considered.
- Abdominal pain is the most common gastrointestinal symptom; however, patients may also complain of vomiting, hematemesis, and bloody stools. The abdominal pain can be very severe, but usually does not last more than 24 hours.
- The stool can have either occult or gross blood.
- Gastrointestinal manifestations may precede purpura by up to 2 weeks.
- Intussusception has been reported in 1% to 5% of patients.
- Rarely, there can be cerebral involvement causing seizures, paresis, or coma.
- HSP is usually a self-limited condition. Supportive care, such as analgesia, is required for patients with skin involvement only.
- When there is internal organ involvement, inpatient therapy may be necessary.
- Treatment is usually supportive, but for severe symptoms options include:
 - Pulse therapy with intravenous corticosteroids may be beneficial for children with joint pain to the point of being unable to walk or with considerable abdominal pain. The efficacy of corticosteroids is unknown for the long-term prognosis of renal disease.
 - Rarely blood transfusions may be needed in those patients with severe blood loss through the GI tract.
 - The use of plasmapheresis, immunosuppressive agents (cyclophosphamide, azathioprine), and intravenous immunoglobulin therapy may be of benefit when used in conjunction with corticosteroids for the more severe forms of nephritis.
- There are no specific diagnostic tests for HSP; however, certain laboratory findings will help the clinician with the diagnosis as well as the identification of specific organ system involvement.
 - A platelet count will reveal whether or not the cutaneous lesions are secondary to thrombocytopenia. In HSP, platelet counts are normal.

- Serum IgA concentrations may be elevated.
- A urine analysis with microscopy will show the extent of renal involvement.
- Serum C3 and C4 concentrations are low in a few patients.
- Stool guaiac will help to identify presence of gastrointestinal bleeding.
- Other laboratory and radiographic studies are useful to exclude other conditions that may mimic HSP before the purpuric lesions appear (i.e. septic joint, appendicitis, testicular torsion).

IDIOPATHIC THROMBOCYTOPENIC PURPURA

- Numerous viral infections including Epstein–Barr virus and cytomegalovirus have been implicated as a cause of ITP.
- The incidence of ITP is approximately 3 to 8 per 100,000 children per year, making it one of the most common acquired bleeding disorders in pediatrics. It affects males and females equally.
- Patients that present with ITP are typically healthy and only complain of easy bruising and spontaneous bleeding.
- About one-third of patients will have nosebleeds and gingival bleeding as part of their initial complaint. Other organ systems that may present with bleeding includes the gastrointestinal tract and urinary tract. Rarely, intracranial hemorrhage may occur. This is seen when the platelet counts are extremely low ($<10\,000/\text{mm}^3$). The mortality rate for intracranial hemorrhage is approximately 1% in children.
- Besides petechiae and bleeding of mucous membranes, the physical examination should be normal including the absence of hepatosplenomegaly and lymphadenopathy.
- ITP is classified as acute and chronic where the latter persists for more than 6 months. Patients with chronic ITP may have underlying autoimmune disorders and usually are in the adolescent age group.
- Acute ITP is more commonly seen in children younger than 10 years.
 - Approximately 80% to 90% of cases are self-limited and will demonstrate recovery within 6 months without treatment.
 - Patients who have platelet counts less than $20\,000/\text{mm}^3$ and/or have significant mucous membrane bleeding should have consultation and follow-up with a hematologist.
- Therapeutic options are controversial and may include intravenous gamma globulin and/or corticosteroids. Although both have advantages and disadvantages, they may improve total platelet counts within 24 to 72 hours.
- A second-line option is anti-Rh(D). It is only useful in patients who are Rh-positive.
- Splenectomy should be considered in those patients with chronic ITP refractory to pharmacologic measures. Long-term remission is only guaranteed in 60% to 90% of patients that undergo a splenectomy.
- Bone marrow examination is only needed when the diagnosis is uncertain and there are other systemic findings that are not typical of ITP.
- Laboratory findings on a complete blood count should only show an isolated thrombocytopenia ($<150\,000/\text{mm}^3$). When other cell lines are decreased, other diseases, specifically leukemia and aplastic anemia should be considered.
- Microscopic examination of a peripheral blood smear may demonstrate large young platelets and occasional megakaryocytes in response to the rapid platelet destruction.
- Coagulation studies are usually not needed.

CHILD ABUSE

- Bruising caused by maltreatment is a consequence of blunt trauma inflicted on the patient. This can occur when caretakers attempt to discipline the child or when their anger is displaced on the child as bodily harm. Children that are abused are usually too young to protect themselves or fight back.
- It is estimated that each year more than 3 million children are victims of abuse, and as a result, an estimated 2000 children die.
- Children who are younger than 4 years or those who have comorbid conditions such as learning disabilities, chronic illnesses, prematurity, conduct disorder, and mental retardation are at highest risk.
- Caretakers who are likely to abuse are those who were abused themselves as children, and young, single, nonbiological parents.
- Diagnosis of child abuse is achieved by a thorough history and physical exam as well as laboratory studies and imaging modalities.
- Interview caretakers individually because different stories may be given for the same injury.
- It is also important to be familiar with medical conditions and cultural practices (coining, cupping) that mimic child abuse.
- The pattern of distribution and location of the traumatic lesions will help differentiate nonintentional from intentional trauma.
 - Bruising over bony prominences often signify noninflicted trauma of a mobile child; however,

bruising over the back, earlobes, buttocks, or other protected areas may suggest abuse.

- Bruising in the shape of teeth marks, belt marks, or hands should also increase your suspicion of maltreatment as should burns.
- Human bites, compared to animal bites, are more often crush injuries.
- Animal bites usually produce a puncture wound or cause tearing of the skin.
- Siblings may be used as scapegoats for a bite mark, but a bite width of more than 4 cm is indicative of an adult bite.
- Fresh bruises suggest that abuse is ongoing. Aging bruises using standardized aging charts is no longer used.
- Unexplained bruising in infants younger than 6 to 12 months should raise suspicion of abuse (see Chapter 145 for a detailed discussion of child abuse).
- In all states, physicians and other professionals are mandated by law to report suspected child abuse to the child protective services or law enforcement.
- Consultation with other health professionals and child protective services may be needed in cases where the physician is uncertain about the likelihood of abuse.
- The main goal is to safeguard the child from further harm. If the patient cannot be immediately placed in another home, admission is warranted until disposition to a foster family is completed.
- Other children in the offender's home may require removal as well.
- Coagulation studies may help in ruling out a bleeding disorder.
- A skeletal survey helps identify subtle fractures that may need to be addressed.
- When a patient presents with decreased level of consciousness, a computed tomography of the head may reveal an intracranial bleed.

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QUESTIONS

1. A five year-old boy is seen for forceful cough and posttussive vomiting for the past two days. He is afebrile, alert, ambulating, and vital signs are normal for age. Physical exam is remarkable for scattered petechiae over the face, neck, and upper chest. The petechiae are *most likely* due to
 - A. Septic emboli
 - B. Thrombocytopenia
 - C. Capillary fragility
 - D. Increased venous pressure
 - E. Vasculitis
2. A febrile, ill appearing 4-month-old infant presents with a history of fever and vomiting for 3 hours, and onset of a generalized petechial/purpuric rash most intense over the extremities. *HR*=200/min; *BP*=60/30; *RR*=60/min and nonlabored; *T*=39.8°C. Your first and *most important* management priority is
 - A. Immediate blood, urine, and cerebrospinal fluid cultures.
 - B. Aggressive intravenous fluid resuscitation with isotonic crystalloid.
 - C. Broad-spectrum antibiotic administration.
 - D. Immediate inotropic support.
 - E. Blood gas, lactate, DIC panel to guide therapy.
3. An otherwise healthy 5-year-old child presents with palpable 0.5–1.5 cm purpuric lesions on her legs and buttocks. She had onset of vomiting, intermittent cramping abdominal pain, and dark blood in her stools for the past three hours. The source of her symptoms could best be determined by
 - A. Platelet count
 - B. Stool culture for *E coli* 0157
 - C. Laparoscopic surgery
 - D. Serum C3, C4 levels
 - E. Abdominal ultrasonography
4. The most consistent physical exam finding in a 7-year-old child with idiopathic thrombocytopenic purpura is
 - A. Easy bruising and spontaneous minor bleeding in an otherwise well appearing child.
 - B. Presence of splenomegaly.

- C. Purpuric lesions limited to the lower extremities and buttocks.
 D. Petechiae limited to the face and neck.
 E. High fever and toxicity.
5. A noncustodial parent brings his daughter to the emergency department for evaluation. He is concerned that she is being physically abused. Which of the following pattern bruises are most consistent with child abuse?
 A. Multiple fresh and old bruises on the anterior legs of a 16 month old.
 B. A 2 cm bite mark on the arm of a 2 year old.
 C. Bilateral bruising of the arms, and lateral chest wall of a 4-month-old infant
 D. A frontal contusion in a 14-month-old toddler.
 E. An isolated cheek contusion in a 6 year old.

ANSWERS

1. D. Petechiae localized to the head, neck, and upper thorax in an otherwise afebrile and non toxic child is most like due to stress and caused by increased venous pressure from forceful coughing and/or vomiting.
2. B. Although all of the answers may be a priority at some point in the resuscitation and management process, this patient is in septic shock, and the most important priority is fluid resuscitation with isotonic crystalloid.
3. E. Intussusception has been reported as a complication of Henoch–Schonlein Purpura in 1–5% of cases. Although abdominal cramps and blood in stools is common, intussusceptions cannot be missed. An abdominal ultrasound will help confirm the presence of intussusceptions which must be managed emergently.
4. A. Children with ITP rarely are ill appearing. ITP may follow an infectious process, but has minimal systemic symptoms. When splenomegaly is present, always consider leukemia in the differential diagnosis. A febrile or toxic appearing child has sepsis until proven otherwise. Purpuric lesions localized to the lower extremities and buttocks are HSP.
5. C. Infants who are not yet crawling or independently mobile rarely should have any unexplained bruises. Bilateral pattern bruises in infants under six months should always suggest abuse. As infants become mobile, (especially over 1 year old), bruising over bony prominences and in exposed areas is common.

86 PRURITIC RASHES

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ATOPIC DERMATITIS

- Patients with a personal history and/or a family history of asthma and other allergies are more prone to atopic dermatitis.
- Infants usually present with exudative lesions over the cheeks, trunk, and extensor surfaces. In children, the distribution is over the flexor surfaces, including the antecubital and popliteal fossae. They present with areas of hypopigmentation and diffuse scaly patches. By adolescence, the involvement is the same with the addition of the face, neck, hands, and feet.
- The clinical course is characterized by relapsing episodes of symptom flares.
- Lichenification and hyperpigmentation are findings characteristic of chronicity.
- Diagnostics:
 - Diagnosis is by clinical observation and a good history.
 - Although elevated serum IgE levels and eosinophilia are seen in 70% to 80% of patients, they are not specific to this disease process and need not be routinely used to diagnose atopic dermatitis.
 - Skin testing is also of little value and certain dietary restrictions are difficult in this age group.
- Treatment:
 1. Avoidance of nonspecific skin irritants such as synthetic fabrics, wool, and nonessential, highly fragranced toiletries.
 2. Keep the skin moisturized (i.e. limiting bathing time, using a mild moisturizing cleanser, and applying emollients frequently) to reduce inflammation and flare-ups.
 3. Low- and medium-potency topical corticosteroids should be used to control mild-to-moderate exacerbations.
 4. Severe flare-ups may require a short course of systemic steroids.
 5. Topical tacrolimus and pimecrolimus are nonsteroidal topical immunosuppressants and should be used as a second-line treatment for moderate-to-severe flare-ups.
 6. Sedative effects of H1 antihistamines help to manage pruritus.
 7. Oral and topical antibiotics are only indicated for active, secondary bacterial infection.

CONTACT DERMATITIS

- Common allergens include nickel (jewelry, watches), chemicals in elastic, latex, tanning chemicals in leather, fabric dyes, and rhus (i.e. poison ivy, oak, and sumac).
- Characterized by the onset of an erythematous, papulovesicular eruption in the distribution of where skin contact with the irritant occurred. Bullae may also be present.
- Chronic exposure to the allergen may cause scaling, fissuring, and lichenification.
- Diagnosis is made when there is pattern recognition at the site of exposure to the offending agent.
- Treatment:
 - Removal and avoidance of the causative material is most important.
 - Mild, localized dermatitis responds well to cool compresses.
 - For small areas of moderate contact dermatitis, potent topical steroids should be used twice daily for 2 to 3 weeks.
 - New topical immunomodulators such as tacrolimus and pimecrolimus are second-line therapies.
 - Antipruritic lotions, such as calamine, and oral antihistamines are also useful for pruritis and impaired sleep.
 - For severe cases with more than 20% to 30% body involvement, a 5- to 7-day course of systemic corticosteroids may be indicated.
- The rash may last several weeks despite treatment.
- Patch testing may be done to elicit a reaction for confirmation of the diagnosis, but it should be avoided during an acute episode.

PEDICULOSIS

- *Pediculus humanus capitus* (head louse), *Pediculus humanus humanus* (body louse), and *Phthirus pubis* (crab louse) are the three types that infect humans.
- Spread from person to person is achieved by close contact. Sharing of hats and combs facilitates the transmission.
- Pruritis to the scalp, especially behind the ears and at the nape of the neck, is highly suggestive of head louse infestation. Direct visualization of the live louse, not presence of nits alone, confirms active infestation.
- Treatment:
 - 1% permethrin, 0.3% pyrethrin cream rinse, or 0.5% malathion to destroy the louse. A second treatment is recommended 7 to 10 days after the initial course to eliminate any recently hatched lice.

- Viable eggs and nits may then be mechanically removed by combing through wet hair with a fine-toothed comb. Topical application of petroleum jelly or mayonnaise, both louse suffocants, may be considered.
- A course of trimethoprim-sulfamethoxazole is also an effective second-line treatment for resistant disease.
- Washing clothes and bedding are necessary to destroy nits that may be attached.
- All close contacts should be checked for lice and treated simultaneously.

SCABIES

- Caused by the arachnid, *Sarcoptes scabiei*
- Sudden onset of severe itching is the hallmark of scabies. Involvement of other family members should be another clue to diagnosis.
- Present as red, excoriated papular eruptions distributed in the folds of the body (beltline, gluteal folds, axillae), and the interdigital webs. Infants may also develop eczematous papules over the head and face. The classic linear burrows are considered to be diagnostic, but may not always be present.
- Treatment:
 - Head-to-toe application of permethrin 5% cream for 8 to 14 hours and then washed off with soap and water. A repeat application may be needed a week after the initial treatment.
 - Systemic therapy with oral ivermectin should be considered in widespread outbreaks in schools, nursing homes, or prisons for better disease control.
 - All caregivers and other close contacts of the household should be treated simultaneously.
 - Bedding and clothing should be washed in hot water.
- Pruritis may persist for 1 to 2 weeks after treatment, even if the infection has resolved.
- Scabies is a clinical diagnosis, but confirmation of the diagnosis is possible by direct microscopic visualization of the mite, eggs, or feces of the mite. Samples should be obtained by scraping the linear burrows caused by the mites.

PAPULAR URTICARIA

- Dog and cat fleas are common offenders. Other less common sources include mosquitoes, grass mites, gnats, and bedbugs.
- The lesions tend to be more evident during the summer and spring months.
- Predominantly seen in patients 18 months to 7 years of age.

- Characterized by crops of symmetrically distributed pruritic erythematous papules surrounded by an urticarial wheal. They are frequently arranged in clusters over the shoulders, upper arms, and other exposed areas.
- Crops last 2 to 10 days, but recurrence is common and the eruptions may persist for 3 to 9 months.
- Scratching may produce erosions and ulcerations, causing secondary impetigo or pyoderma.
- The management is mostly conservative. Topical medium potency corticosteroid creams and systemic antihistamines may be used to control itching.
- Dogs and cats should be treated for fleas.
- DEET-containing insect repellents are also useful.
- Carpets and furniture may require cleaning with the use of commercialized insecticides.

URTICARIA

- Causes include foods (i.e. berries, nuts, shellfish), drugs (i.e. penicillins, cephalosporins), bites and stings, infections (i.e. viral, streptococcal), and certain systemic diseases. In most cases, no causative agent is identified.
- They appear suddenly as erythematous, raised, pruritic wheals with a pale center and serpiginous borders over any part of the body. They tend to disappear and reappear on different parts of the body.
- *Acute urticaria*
 - Generally do not last more than 24 hours, but may persist.
 - Patients should be assessed for airway compromise or hemodynamic instability.
 - Treatment depends on the severity of the condition and presence or absence of airway involvement.
 - Mild cases without airway involvement will resolve with oral antihistamines.
 - For severe cases with airway involvement and/or hemodynamic instability, intramuscular epinephrine (1:1000) 0.01 mL/kg. Repeat doses may be required every 5 to 15 minutes. Adjunctive therapies include nebulized β -agonists, H1 and H2 antihistamines, as well as corticosteroids once the patient is stabilized.
 - It is also important to identify and avoid any precipitating agents in the future.
- *Chronic urticaria*
 - Persists longer than 6 weeks.
 - No concerns for airway compromise or hemodynamic instability.
 - Treatment consists of non- or low-sedating antihistamines. Stress and fatigue may exacerbate recurrent symptoms.
- Minor flares may also require a short course of oral corticosteroids.
- For uncontrollable disease, immunomodulators such as cyclosporine, methotrexate, tacrolimus, or intravenous immunoglobulins may be given.
- Plasmapheresis may be considered in the most severe cases.

ERYTHEMA MULTIFORME

- Linked to a variety of drugs and infections.
 - Herpes simplex virus and *Mycoplasma pneumoniae* infections are the most common precipitants.
 - The offending drugs include sulfonamides, penicillins, anticonvulsants, and nonsteroidal antiinflammatory agents.
 - Most cases will not have a causal agent that can be identified.
- Clinical appearance:
 - Characterized as round, fixed, and erythematous, appearing symmetrically on the skin.
 - Often described as “target” lesions because of their central dusky area.
 - The lesions are distributed acrally but are often found on the trunk.
 - Remain for a minimum of 1 week and can persist up to 2 to 3 weeks.
 - Although uncommon, EM can have mucosal involvement, and when present, it usually involves the oral mucosa only.
- Self-limited process and treatment usually is unnecessary
 - Supportive care with cool compresses and antihistamines suffices.
 - For painful oral lesions, patients may benefit from soothing mouthwash.
 - Acyclovir is not generally indicated but should be considered in patients who have recurrent EM secondary to HSV.
 - There is no evidence to support the use of corticosteroids.
 - Discontinue any recent medications or any possible causative agent.

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QUESTIONS

1. A 2-month-old infant is brought to the emergency department. The infant is afebrile and appears well. The parents are frustrated because of the reappearance of a rash. The father states that “comes and goes.” There is a strong family history of asthma and “skin problems.” You suspect atopic dermatitis. The clinical appearance most likely to describe the rash would be which of the following:
 - A. A scaly rash most intense on the flexor surfaces of the arms and legs.
 - B. Exudative lesions over the cheeks, trunk, and extensor surfaces of the arms.
 - C. Greasy yellow scales in the scalp.
 - D. Red papules with a surrounding wheal.
 - E. Crusting, weeping, lesions behind the ears.
2. A 12-year-old girl is brought to the emergency department by her father for evaluation of the sudden appearance of a rash associated with itching. On physical examination, you note erythematous, raised, pruritic wheals with a pale center diffusely spread her body. The family is extremely concerned because of the school dance in 1 week. The child has no significant past medical history and has been previously well. The most probable cause of acute urticaria in this patient would be which of the following?
 - A. Antibiotic use
 - B. Shellfish
 - C. Nuts
 - D. Insect bites/stings
 - E. Unknown causes
3. An otherwise asymptomatic 10-year-old presents with a one-day history of a generalized rash consisting of erythematous macules, papules, and circular erythematous lesions with a dusky center.

The lesions are most common on the trunk. He is asymptomatic with the exception of mild itching. Treatment should include

- A. Acyclovir, orally for 10 days.
- B. A five day course of oral corticosteroids.
- C. Epinephrine 1:1000 IM.
- D. Oral diphenhydramine as needed for itching.
- E. IV diphenhydramine.

ANSWERS

1. B. Infants with atopy usually present with exudative weeping lesions on the face, trunk, and extensor surfaces of the extremities. Older children will have more intense rash in the flexor creases. Seborrheic dermatitis often involves the scalp and has greasy scales.
2. E. Although all of the answers are precipitants of acute urticaria, most cases are from unknown causes.
3. D. This is descriptive of a typical case of erythema multiforme, and supportive care is all that is usually necessary. If pruritis is a problem, oral antihistamines may be indicated. No other therapy is necessary.

87 SUPERFICIAL SKIN INFECTIONS

Malee V. Shah

Robert A. Wiebe

IMPETIGO

- Superficial bacterial infection of the epithelium.
 - Initially present as small vesicles and pustules that eventually rupture.
 - Eventually, they become eroded and progress to areas of honey-crusted lesions.
- *Staphylococcus aureus* and group A β -hemolytic *Streptococcus* are the organisms most commonly isolated.
- Usually occurs in younger children over the exposed areas (i.e. face and extremities)
- Associated with poor skin hygiene. Good hand washing and other simple measures of personal hygiene may reduce the likelihood of spread.
- Topical mupirocin ointment can be used for uncomplicated, localized lesions.
- Areas with larger involvement require oral antibiotics.

- The choice of antibiotics should be determined by local resistance patterns
- Penicillin or cephalosporin may be sufficient coverage for gram-positive organisms.
- Erythromycin is used for penicillin or cephalosporin-allergic patients, but has little coverage for MRSA.
- Clindamycin is a good choice when resistance is recognized.
- In recurrent or persistent infections, lesions should be cultured to determine if an antibiotic-resistant organism is present.

STAPHYLOCOCCAL SCALDED SKIN SYNDROME (SSSS)

- Caused by an exfoliative exotoxin-producing strain of *S. aureus*
- Most commonly presents in infants and children younger than 5 years.
- Clinical presentation:
 - Prodromal symptoms may include pharyngitis and conjunctivitis followed by fever, malaise, and a blistering skin eruption
 - Initially, the rash resembles sunburn, which usually starts over the face, neck, axillae, and groin.
 - Large, superficial bullae then form over the erythematous areas and rupture. The skin is exquisitely tender and fragile.
 - Gentle rubbing of the skin results in desquamation of the epidermis, exposing a moist red base with the characteristic scalded skin appearance.
 - In infants and preschool children, the lesions are limited to the upper body.
 - In newborns, the entire cutaneous surface may be involved (Ritter's disease).
- Patients with severe SSSS are managed similar to burn patients with fluid rehydration, topical wound care, pain control, and parenteral antistaphylococcal antibiotics.
 - Intravenous nafcillin adequately treats infections caused by penicillinase-producing staphylococci.
 - Other options include cephazolin, clindamycin, and vancomycin
 - Corticosteroids are contraindicated.
- Blood cultures as well as cultures from the nose, nasopharynx, conjunctivae, and external ear canal help isolate the offending pathogen.

FUNGAL INFECTIONS

- *Tinea capitis* infection can present with the presence of dry, scaly patches resembling seborrhea or atopic dermatitis, or it may be irregular areas of scaling

alopecia with broken hairs, giving the appearance of "black dots."

- Can progress to folliculitis suppuration or kerion formation, which are erythematous, boggy masses with overlying hair loss
- A Wood's lamp may help differentiate tinea from other scaly lesions by the presence of blue-green fluorescence.
- First line treatment includes oral Griseofulvin for 6 to 8 weeks. The concomitant use of topical agents such as 2.5% selenium sulfide shampoo may be considered. Second-line oral therapy includes itraconazole, terbinafine, and fluconazole.
- *Tinea corporis* can be found anywhere on the body and are characterized as sharply circumscribed scaly patches with central clearing and an elevated border.
 - Treatment is with topical antifungal agents, such as clotrimazole, miconazole, econazole, terbinafine, and butenafine.
 - Improvement will be seen in about a week, but therapy should be continued for two weeks.
 - Consider adding a low-to-medium potency topical corticosteroid for the first few days.
 - If treatment is ineffective or there is extensive skin infection, oral griseofulvin or azoles should be considered.

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QUESTIONS

1. A 3-year-old male presents with a tender, fine erythematous rash of his face and neck that has progressed to peeling around the nares and extension to his chest. The skin is tender to the touch. The most appropriate choice for treatment would be
 - A. Topical corticosteroids
 - B. Topical mupirocin

- C. Penicillin V
 D. Clindamycin
 E. Erythromycin
2. A 9-year-old girl presents with circumferential dry, scaling patches of alopecia on her scalp with visible broken hairs. First-line treatment would be with
- A. Topical clotrimazole
 B. Topical terbinafine
 C. Topical corticosteroids
 D. Selenium sulfide shampoo only
 E. Oral griseofulvin

ANSWERS

1. D. This is a patient with staphylococcal scalded skin syndrome. Of the choices given, clindamycin is correct. This condition should be treated with systemic antibiotics to cover *Staphylococcus aureus*. Mupirocin is not useful since this is a toxin related rash and may come from a remote site. Corticosteroids are contraindicated.
2. E. This patient has tinea capitis, and needs systemic treatment with a griseofulvin preparation. Topical antifungal agents that can be used to treat tinea corporis are not helpful for tinea capitis. Selenium sulfide shampoo can be used as adjunctive therapy to reduce the risk of spread.

88 EXANTHEMS

Robert A. Wiebe
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INTRODUCTION

- The vast majority of childhood exanthems are a result of nonspecific viral illnesses. Most are benign and self-limited and resolve without any therapeutic measures. The enterovirus group including echovirus and coxsackievirus consist of nearly 80 human host infections that may cause childhood exanthemas. Although they rarely cause serious or life-threatening disease, some enterovirus infections can result in serious sequelae such as encephalitis and myocarditis. The clinician should always be vigilant to recognize associated symptoms that suggest life-threatening complications when examining children with exanthemas.

RUBEOLA (MEASLES)

EPIDEMIOLOGY/PATHOPHYSIOLOGY

- Most cases today are limited to immunocompromised patients and in developing countries where poverty and malnutrition is a factor; outbreaks still occur in developed nations.
- Most contagious diseases known to man with a 90% transmission rate to the unimmunized household contact.
- Significant cause of morbidity and mortality in developing countries.
- Infants are usually immunized between 12 and 15 months of age.
- Maternally acquired antibodies usually are sufficient to protect against clinical exposure in infants younger than 1 year of age.
- Incubation period ranges from 7 to 18 days after exposure, and patients are contagious for approximately 5 days starting with the onset of symptoms.

CLINICAL FINDINGS

- Rash associated with measles is preceded by 3 days of high fever. It is maculopapular, erythematous and first appears on the face and spreads cephalocaudal to involve the trunk and extremities.
- Intense mucoid nasal drainage, hacking cough, and marked scleral and papebral nonpurulent conjunctivitis are always present prior to the onset of rash.
- Koplik spots present during the early febrile period, but may be absent by the time the rash appears.
- The fever will resolve within 3 days of onset of rash, and in uncomplicated cases, all symptoms will resolve within 7 to 8 days.

COMPLICATIONS

- Otitis media, occurs in up to 25%.
- Encephalitis will occur in approximately 1:1000 cases.
- Pneumonia, diarrhea with dehydration, and blindness can be as high as 10% in poorly nourished children and 30% in the immunocompromised.
- Subacute sclerosing panencephalitis (SSPE) is a rare central nervous system degenerative disease.

MANAGEMENT

- Nonspecific supportive care for the uncomplicated patient.

- Vitamin A supplementation may be helpful in preventing corneal ulcerations and blindness in malnourished children.
- When given within 72 hours of exposure, live virus vaccine may be beneficial in modifying the disease course.
- Susceptible children exposed to measles should receive intravenous immunoglobulin 0.25 mL/kg within 6 days of exposure. For children with history of immunodeficiency syndromes and HIV, 0.5 mL/kg is recommended.

RUBELLA (GERMAN MEASLES)

EPIDEMIOLOGY/PATHOPHYSIOLOGY

- Postnatal rubella is transmitted by airborne droplets from the upper respiratory tract.
- Transmission of disease can occur for approximately 1 week before the onset of rash until up to 14 days after symptoms appear.
- The risk associated with serious morbidity and mortality is only from exposure of pregnant females in the first 20 weeks of gestation.
- Young mothers born outside the United States should be screened to assess immunity to rubella.

CLINICAL FINDINGS

- Postnatally acquired rubella is generally mild and self-limited.
- Presentation is with mild cough, coryza, conjunctivitis, and a maculopapular erythematous rash that starts on the face and progresses cephalocaudal.
- Low-grade fever is often present.
- Can be differentiated from rubeola by the presence of a rash appearing at or near the onset of fever and the mild nature of symptoms.
- Headache and transient polyarthralgias and polyarthritis are common in older children and adults.
- Symptoms usually resolve in 3 to 4 days.
- Pea-sized postauricular lymphadenopathy is usually seen when the rash appears.

COMPLICATIONS

- Complications from postnatal acquired rubella are rare.
- Thrombocytopenia occurs in approximately 1:3000 cases.
- Encephalitis occurs in 1:5000 cases with headache, vomiting, stiff neck, and lethargy. These symptoms are usually self-limited and not serious.

- Transplacentally acquired infections can result in fetal death or congenital rubella syndrome.

MANAGEMENT

- Symptomatic treatment is all that is necessary for postnatal rubella.
- Isolation from childcare or school should be recommended for 7 days after onset of rash.
- Intramuscular immunoglobulin (0.55 mL/kg) may reduce viral load and is beneficial for treating susceptible pregnant females who choose not to terminate pregnancy.

ROSEOLA INFANTUM (EXANTHEM SUBITUM)

EPIDEMIOLOGY/PATHOPHYSIOLOGY

- Caused by human herpesvirus 6 and 7.
- Virus is transmitted through respiratory secretions from asymptomatic individuals and during the febrile viremic phase of the illness.
- Maternally transferred antibodies provide protection for the first 3 to 6 months of life.
- By the age of 4 years, nearly all children have serologic evidence of prior infection from HHV-6.
- The incubation period is approximately 9 to 10 days.

CLINICAL FINDINGS

- Sudden onset of high fever to 40°C with fever spikes persisting for 72 hours followed by a transient erythematous maculopapular truncal rash in an infant from 6 months to 2 years of age.
- The rash may occur up to 24 hours before or after the fever resolves.
- Skin manifestations of roseola may be so transient that they escape detection.
- Infants with fever are rarely ill appearing and look remarkably well for the high fever.
- Large postoccipital lymph nodes are characteristic and their presence in an infant with high fever can predict roseola as the cause of fever.
- A full fontanelle is found in up to 25% of infants infected with HHV-6.

COMPLICATIONS

- Febrile convulsions are seen in up to 20% to 30% of infants with roseola.
- Fatigue, irritability, and anorexia are reported occasionally.

- Pneumonia, encephalitis, hepatitis, and hemophagocytic syndrome are rarely reported in immunocompromised individuals.
 - Management is supportive and fever may be treated for comfort.

FIFTH DISEASE (ERYTHEMA INFECTIOSUM)

EPIDEMIOLOGY/PATHOPHYSIOLOGY

- Caused by the human parvovirus B19.
- Humans are the only known host for carrying and transmitting the disease.
- Infection can occur at any age, but is most common in mid-to-late childhood.
- Transmitted through contact with respiratory secretions, but can be carried through blood products and from maternal–fetal transmission.
- Replication of virus in red cell precursors causes a transient reduction in red blood cell formation during viremic phase. This is unimportant in an otherwise healthy child with a normal red cell survival of 120 days, but can cause serious symptoms when the normal red cell lifespan is short as occurs in the child with hemolytic anemia, or in early fetal development.
- Viremia occurs approximately 1 to 2 weeks after exposure and lasts 3 to 5 days. By the time the rash appears, the patient is no longer contagious. This is important because patients with hemolytic disease when presenting with an aplastic crisis are shedding virus and can transmit disease to others. They are not, however, at risk for getting the virus from patients who exhibit the characteristic rash.

CLINICAL FINDINGS

- The rash consists of bright erythema of the cheeks giving a “slapped cheek” appearance, and a fine, lacy reticular rash on the extremities and trunk.
- The rash may resolve in a few days, only to reappear with exposure to sun or warm baths, and may recur for several weeks.
- Low-grade fever, arthralgia, headache, and myalgia may be seen in some children as a prodrome before the rash appears. Arthritis is rare in children but common in adults.

COMPLICATIONS

- Rare in otherwise healthy children and limited to transient arthritis or arthralgias.

- Children with hemolytic disease and short red cell survival are at risk for aplastic crises with critical drops in hemoglobin and no compensatory rise in reticulocyte counts during the viremic phase.
- Pregnant women, especially during the first trimester are at risk for hydrops fetalis with spontaneous abortion.
 - Management is supportive care only. Patients with hemolytic anemia and aplastic crisis may require blood transfusions.

VARICELLA (CHICKEN POX)

EPIDEMIOLOGY/PATHOPHYSIOLOGY

- Caused by varicella-zoster virus (human herpesvirus-3).
- Man is the only known reservoir for this highly contagious virus.
- The virus enters through the respiratory tract or conjunctivae, and is transferred from person-to-person by air or contact with fluid from vesicles.
- Incubation period is 10 to 21 days, and infected patients are most likely to be infectious to others from 2 days before the onset of rash until the lesions are crusted and dry.

CLINICAL FINDINGS

- A prodrome of nonspecific symptoms and low-grade fever precedes the rash by 1 to 2 days.
- The rash begins as erythematous macules that rapidly progress to papules and eventually form the characteristic vesicles on an erythematous base (dewdrops on a rose petal).
- Within 6 to 24 hours, the vesicles become pustules that break and dry into crusts. New crops of lesions develop with all stages of lesions present on the skin at the same time.
- In a normal host, the lesions resolve in 5 to 7 days.

COMPLICATIONS

- The most common minor complication is secondary bacterial infection of the skin lesions.
- Serious complications include varicella pneumonia, encephalitis, secondary bacterial pneumonia, cellulitis, necrotizing fasciitis, and sepsis.
- Symptoms are more severe in adolescents and adults.
- Immunocompromised children and those on chronic corticosteroids are also at greater risk.

TABLE 88-1 Antiviral Agents for the Treatment of Varicella-zoster and Herpes Simplex

HOST CONDITION	AGE	DRUG	ROUTE	DOSE
Varicella, immunocompetent with "at-risk" conditions	>2 y	Acyclovir	po	80 mg/kg/d in 3 divided doses for 5 d (max 3200 mg/d)
Varicella, immunocompromised host	All ages	Acyclovir	IV	30 mg/kg/d in 3 divided doses for 7–10 d
Zoster, immunocompromised host	<12 y	Acyclovir	IV	60 mg/kg/d in 3 divided doses for 7–10 d
	>12 y		IV	30 mg/kg/d in 3 divided doses for 7 d
Neonatal herpes simplex	Birth–3 mo	Acyclovir	IV	60 mg/kg/d in 3 divided doses for 14–21 d
Herpes encephalitis	3 mo–12 y	Acyclovir	IV	60 mg/kg/d in 3 divided doses for 14–21 d
	>12 y			30 mg/kg/d in 3 divided doses for 14–21 d
HSV, immunocompromised host	<12 y	Acyclovir	IV	30 mg/kg/d in 3 divided doses for 7–14 d
	>12 y			15 mg/kg/d in 3 divided doses for 7–14 d
HSV prophylaxis in immunocompromised host	>2 y	Acyclovir	po	600–1000 mg/kg/d in 3–5 divided doses for risk period
Genital HSV, primary infection	>12 y	Acyclovir or Valacyclovir or Famciclovir	po	400 mg 3 times daily for 7–10 d 1 g twice daily for 7–10 d 250 mg 3 times daily for 7–10 d
Genital HSV, recurrent (episodic treatment)	>12 y	Acyclovir or Valacyclovir or Famciclovir	po	400 mg 3 times daily for 5–10 d 1 g once daily for 5 d (1 g twice daily for 5–10 d for immunocompromised) 125 mg twice daily for 5 d (500 mg twice daily for 5–10 d for immunocompromised)

Adapted from: American Academy of Pediatrics. Herpes Simplex. In: Pickering LK, Baker CJ, Long SS, McMillan JA, eds. *Red Book: 2006 Report of the Committee on Infectious Diseases*. 27th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2006:361–371, and Fatahzadeh M, Schwartz RA. Human herpes simplex virus infections: epidemiology, pathogenesis, symptomatology, diagnosis and management. *J Am Acad Dermatol*. 2007;57(5):737–763.

- Congenital varicella syndrome with brain and eye anomalies, limb hypoplasia, and intrauterine growth retardation can occur in 0.4% to 2% of susceptible pregnant women who are infected during the first 20 weeks of gestation.

MANAGEMENT

- Uncomplicated varicella in the normal host requires only symptomatic treatment to control itching.
- In healthy children, virus replication ends within 72 hours of onset of the rash, making late treatment with antiviral agents of no value.
- Early acyclovir may be given in low-risk children in order to modify the disease course.
- For children who are at high risk of complications, specifically those on chronic corticosteroids and immunosuppressives, intravenous antiviral therapy should be started early.
- Varicella-zoster immunoglobulin may modify the disease if given early after exposure.
- Oral acyclovir is indicated in moderate-risk children with chronic cutaneous or pulmonary disease, those on long-term salicylate therapy, children on short,

intermittent oral or aerosol corticosteroids, and children older than the age of 12 years (Table 88-1).

- Varicella vaccine, when administered within 36 hours of exposure, can be useful in preventing disease and providing postexposure prophylaxis.

HERPES ZOSTER (SHINGLES)

- A common complication resulting from a prior varicella-zoster virus exposure.
- Virus remains latent in spinal sensory nerve root ganglia.
- Rare in children, but incidence increases with age and lifetime incidence is 10% to 50%.
- *Clinical findings:* Tingling pain may precede the onset of rash. Crops of vesicles occur, usually limited to a specific dermatome nerve distribution. In adolescents and adults, shingles can be extremely painful, but in children, pruritis rather than pain is often the chief complaint.
- *Complications:* A concern for immunocompromised patients and patients on chronic corticosteroids.
- *Treatment:* Guidelines for management of zoster is the same as for varicella. Famciclovir or valacyclovir are preferred for treatment of adults (Table 88-1).

HERPES SIMPLEX

EPIDEMIOLOGY/PATHOPHYSIOLOGY

- Clinical disease in humans is caused by two members of the Herpesviridae family, HSV1 and HSV2. There is overlap in clinical expression.
- Both can remain latent in cells after primary infection.
- Enters the host through epithelial cells and may undergo replication at the site of invasion.
- After the primary infection has occurred, HSV travels through the periaxonal sheath of sensory nerves to ganglia of the host nervous system.
- The virus replicates in ganglia and may persist for life. Periods of virus latency may be interrupted by reactivation.
- An asymptomatic person can shed virus, but during primary infection and reactivation, virus shedding is increased.
- Reactivation of virus can occur as a result of a variety of external and internal triggers including fever, immunosuppression, local trauma, menstruation, stress, fatigue, temperature changes, and exposure to sunlight.

CLINICAL FINDINGS

- HSV1 is mainly associated with infections involving the mouth, pharynx, and eyes, while HSV2 primarily has genital mucosal symptoms, but there is overlap and both viruses can enter the central nervous system and cause encephalitis and systemic disease.
- The most common manifestation of HSV1 is primary herpetic gingivostomatitis.
- Neonatal herpes has three distinct clinical presentations and occurs in 1:3000 to 1:20 000 births.
 - Disseminated herpes presents in the first 1 to 4 weeks of life with a clinical presentation similar to bacterial sepsis and a mortality rate of 25% even with antiviral treatment.
 - Herpes encephalitis may present with seizures, often focal in the neonatal period and if untreated, will progress to systemic disease.
 - Neonatal herpes with involvement limited to the skin, eye, and mouth can result in minimal morbidity if treated early with antiviral agents. Symptoms may occur as late as 6 weeks of life.
- Eczema herpeticum occurs in children with atopic dermatitis and breaks in skin integrity.
- See Chapter 97 for a review of genital herpes.
- Cutaneous herpes simplex occurs from contact of abraded skin with active oral or other herpes lesions actively shedding virus.

- Herpetic whitlow is caused by secondary inoculation of virus from mouth to fingers. This can be confused with a paronychia, and attempts at drainage can cause bad cosmetic results.
- Other manifestations include ocular herpes or herpes keratoconjunctivitis, a major cause of blindness worldwide. Conjunctivitis can be unilateral or bilateral and result in corneal ulcerations.
- Herpes meningoencephalitis may occur at any age and have no skin manifestations at presentation.
- The most common presentation of HSV is recurrent orofacial herpes, commonly referred to as “fever blisters.”
- Erythema multiforme (EM) has been associated with HSV reactivation and possibly with primary infections. In some series, presence of HSV with EM has been as high as 75%. Recurrent EM with recrudescence of HSV can occur repeatedly with attacks lasting up to 2 weeks (see Chapter 86).

ANCILLARY TESTS

- Uncomplicated HSV infection can be diagnosed clinically.
- DFA of skin lesions provides quick and reliable results.
- PCR of the cerebrospinal fluid is particularly useful in diagnosing neonatal HSV and herpes encephalitis.
- Use of the Tzanck smear for diagnosis should be discouraged. There is a high incidence of false-negatives.
- Culture from lesions or body fluids will grow in vitro in 1 to 15 days.

COMPLICATIONS

- Serious complications are mainly limited to the immunocompromised host, the neonate, and those with involvement of the eye or central nervous system.
- Scarring can occur with eczema herpeticum or children with chronic cutaneous conditions.

MANAGEMENT

- Antiviral agents can suppress virus replication and modify the clinical course but cannot eradicate the virus.
- Topical antiviral agents such as docosanol 10%, penciclovir 1%, and acyclovir 5% creams have been shown to reduce the symptoms of recurrent labial herpes when started early during the prodrome.

- Acyclovir is the mainstay for systemic disease in neonates, infants, and young children. Drugs with greater bioavailability and longer plasma half life, such as valacyclovir and famciclovir (see Table 88–1), may be helpful for the immune suppressed host.

ENTEROVIRUSES

- Although most enteroviral infections have nonspecific clinical presentations, a few are associated with clinically distinguishable exanthemas and/or enanthemas.

CLINICAL FINDINGS

- Hand-foot-mouth syndrome is associated with infection from coxsackievirus A16 and enterovirus 71.
 - Prodrome of low fever, malaise, and abdominal pain that may precede the skin and mucosal manifestations.
 - Small vesicles appear on the palms and soles and frequently rupture leaving fine erythematous macules.
 - The skin lesions occasionally may be found on the buttocks and trunk. Vesicles appear on the tongue, palate, and buccal mucosa.
 - The symptoms resolve in less than a week in a normal host.
 - The clinical picture can be distinguished from herpetic gingivostomatitis by a lack of intense gingivitis.
 - Complications are rare, but myocarditis, pneumonia, and meningoencephalitis have been reported.
- Herpangina is caused by coxsackievirus A1 to A8 and A12.
 - Examination of the oropharynx reveals small erythematous vesicles localized to the soft palate and anterior tonsillar fossae.
 - Mainly a disease of infants and toddlers with refusal to eat, low-grade fever, and irritability as the presenting symptoms.
 - Diagnosis is made by clinical examination and needs no further testing.

SCARLET FEVER

PATHOPHYSIOLOGY

- Caused by erythrogenic toxin producing types of *Streptococcus pyogenes*.

- The source of infection is usually from pharyngitis with group A β -hemolytic *Streptococcus* (GABHS), but rarely skin, soft tissue, and surgical wound infections can be a source for toxin production.
- Peak incidence is at 4 to 8 years of age, and it is rarely seen before 2 years of age or in adolescents.

CLINICAL FINDINGS

- Fever to 40°C, fatigue, sore throat, abdominal pain, vomiting, and headache will precede the rash by 12 to 48 hours.
- The exanthema is a fine, erythematous, rough (“sandpaper”) rash that blanches on pressure.
- Rash is generalized with increased intensity in the skin folds (Pastia’s lines) with circumoral pallor and facial flushing.
- An enanthem may include red macules on the hard and soft palate (Forchheimer spots) and a “strawberry” tongue.
- Rash fades over 3 to 4 days with desquamation that may continue for 1 to 6 weeks.
- Early treatment with antibacterial agents may reduce the desquamation.

ANCILLARY TESTS

- Culture of the pharynx or source of infection is recommended and can give results in 24 hours.
- Rapid screening tests are used as a surrogate to culture, but false-negatives occur.
- Specificity of rapid tests is high and if positive, does not require confirmation with culture.

COMPLICATIONS

- The risk of poststreptococcal sequelae, acute rheumatic fever, and acute glomerulonephritis is the same for scarlet fever as for GABHS pharyngitis.
- Sepsis, meningitis, pneumonia, and toxic shock are rare complications.

MANAGEMENT

- Penicillin V is the oral treatment of choice for both scarlet fever and GABHS pharyngitis. Oral therapy may be given 250 mg three times daily for children less than 27 kg, and 500 mg three times daily for children more than 27 kg.
- Intramuscular penicillin G at 25 000 U/kg to a maximum adult dose of 1.2 million units is an acceptable alternative.

- In the penicillin allergic patient, clindamycin or erythromycin may be used.
- Isolation for 24 hours after starting treatment will prevent spread.

PITYRIASIS ROSEA

- A benign, self-limited exanthema that appears in case clusters, but there is no evidence of person-to-person spread and the cause is unknown.
- Most common in adolescents and young adults.

CLINICAL PRESENTATION:

- Starts with the *herald patch*, an oval, scaly, 2- to 5-cm lesion that is often confused with a fungal dermatitis, and occurs 1 to 20 days before onset of a generalized rash.
- Exanthem consists of multiple rose-pink ovoid macules that have a “Christmas tree” distribution over the back and may involve the trunk and extremities to a lesser degree.
- Rash lasts 2 to 6 weeks
- May be associated with no symptoms or low-grade fever, headache, malaise, nausea, and pruritis.
 - *Treatment:* Supportive to control itching, if present. Limiting sun exposure may reduce symptoms.

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QUESTIONS

1. A toddler is brought to the emergency department (ED) for evaluation of a rash. The toddler recently immigrated to the United States. His vaccination history is unknown. Parents report high fevers, nasal discharge, and “red eyes.” Which of the following descriptions of the rash would confirm your suspicion in this patient?
 - A. Preceded by 3 days of high fever.
 - B. Begins on the arms and legs and moves to the trunk.
 - C. Has the onset on the first day of fever.
 - D. Mild and often escapes detection on exam.
 - E. Has petechiae, most intense in the skin folds.
2. A 6-month-old infant is brought to the emergency department for evaluation of fever. On examination, you note a febrile but non-ill appearing 6 month old infant with a full fontanelle. The physical examination is essentially unremarkable with the exception of postoccipital adenopathy. This infant is most likely to have an infection from
 - A. HSV-1
 - B. HSV-2
 - C. HHV-3
 - D. HHV-6
 - E. Coxsackie A-8
3. A 4-year-old child with sickle cell disease has been exposed to a friend in school with a diagnosis of erythema infectiosum. He is most likely to present to the ED with
 - A. Sequestration crisis
 - B. Vaso-occlusive crisis
 - C. Aplastic crisis
 - D. Dactylitis
 - E. Fever and a facial rash
4. A 3-year old is brought to the ED for evaluation of a rash. The rash is vesicular, and immunization status is unknown. You learn that the child is on chronic corticosteroids. The most common complication of a varicella infection is
 - A. Pneumonia
 - B. Impetigo
 - C. Necrotizing fasciitis
 - D. Encephalitis
 - E. Sepsis
5. A 2-week-old infant presents with a vesicular rash and low-grade fever. You suspect neonatal herpes simplex infection. The most reliable diagnostic laboratory tests to confirm the diagnosis would be
 - A. Culture of cerebrospinal fluid and skin lesions.
 - B. PCR of CSF and DFA of skin lesions.
 - C. Tzank prep of skin lesion and culture of CSF.

- D. Blood and CSF culture for herpesvirus.
- E. CT of head to demonstrate characteristic pattern of encephalitis.

ANSWERS

1. A. The characteristic rash associated with measles is preceded by 3 days of high fever. It is maculopapular, erythematous and first appears on the face and spreads cephalocaudal to involve the trunk and extremities. The fever will resolve within 3 days of onset of rash, and in uncomplicated cases, all symptoms will resolve within 7 to 8 days.
2. D. HHV-6 infection, or roseola infantum may have a full fontanelle in as high as 25% of infected infants. If the infant with high fever has a full fontanelle, but is smiling and not ill appearing, and playful on exam, it is likely due to roseola. The presence of large postoccipital adenopathy will provide further clinical evidence that roseola is the cause.
3. C. In patients with rapid red blood cell turnover such as sickle cell disease, other disorders of hemoglobin production, or red cell membrane disorders, aplastic crises are a common and serious complication seen with parvovirus B-19 (erythema infectiosum) infection.
4. B. Although all of the answers provided may on occasion occur, impetigo, or secondary bacterial superficial skin infections are by far the most common complication. The pruritus characteristic of varicella infections results in scratching, excoriations, and secondary impetigo. Chronic corticosteroids may put this child at risk for more serious complications.
5. B. A PCR of the CSF is the most rapid and specific method for confirming the presence of herpes simplex encephalitis. A DFA of the skin lesions may also be helpful in confirming HSV-1 or HSV-2 as a cause. Cultures for HSV may be obtained, but requires up to 15 days for diagnostic confirmation.

- As sebaceous and sweat glands in the skin adapt to the changes, transient and benign rashes commonly appear. Although clinically insignificant, these rashes cause high anxiety in young, new parents.
- *Milia*: Small, discreet white papules, sebaceous inclusion cysts, usually limited to the face and scalp. No treatment is necessary, and the lesions resolve spontaneously in weeks to months.
 - *Miliaria*: Lesions caused by obstruction of eccrine sweat glands.
 - *Miliaria crystallina* results from sweat being trapped in the intracorneal layer of the skin producing tiny clear vesicles.
 - *Miliaria rubra* or *heat rash* is common in febrile or overheated infants. These are erythematous small papules that are most commonly found on the upper trunk and head.
 - *Erythema toxicum*: Present in up to 50% of newborn infants.
 - Usually appear shortly after discharge from the hospital at 24 to 48 hours and last approximately 1 week.
 - Pinpoint, papulopustular lesions on an erythematous base that appear on the face, trunk, and extremities.
 - A scraping performed with Wright stain will reveal sheets of eosinophils.
 - *Pustular melanosis*: Found almost exclusively in African-American infants, and are usually present at birth. These are very superficial pustules that become scaly brown macules as they resolve.
 - A Wright stain of the contents will show a predominance of neutrophils.
 - Can be differentiated from neonatal pyoderma by the absence of bacteria on Gram stain. As they resolve, the subcorneal pustules may persist as hyperpigmented small brown macules for months.
 - There are a variety of vesiculopustular lesions that can be confused with pustular melanosis that may have serious complications (Table 89-1).
 - *Seborrheic dermatitis*:
 - Onset within the first 4 weeks of life.
 - Scalp is the first to show signs with the appearance of greasy yellow scales, occasionally with loss of hair,
 - Erythematous plaques develop and skin creases are often involved.
 - The retroauricular area may have weeping or scaly denuded areas of involvement.
 - Treatment consists of low-potency topical corticosteroids to treat inflammation. Mild tar shampoo and oatmeal baths will help resolution.
 - *Diaper dermatitis*: Encompasses all causes of skin irritation localized to the diaper area.

89 INFANT RASHES

Robert A. Wiebe

Malee V. Shah

BENIGN INFANT RASHES

- When the newly born infant leaves the protection of the intrauterine fluid environment, the skin and its complex organs must adapt quickly to the continually changing environment of the *real world*.

TABLE 89-1 Vesiculopustular and Bullous Lesions with Systemic Complications

CONDITION	CAUSE	DIAGNOSIS	COMPLICATIONS
Bacterial infections	<i>Staphylococcus</i> , <i>Streptococcus</i>	Gram stain of lesions	Sepsis; see Chapter 85
Neonatal herpes	Herpes Simplex, 1 or 2	DFA, PCR	See Chapter 86
Fetal, neonatal varicella	Varicella–Zoster virus	DFA, PCR	See Chapter 86
Incontinentia pigmenti	X-linked inherited	Skin biopsy	Neurologic sequelae, seizures
Mastocytosis	Unknown	Clinical, biopsy	Histamine release
Acrodermatitis enteropathica	Defect in zinc uptake from GI tract	Serum zinc level	Diarrhea, Irritability, FTT
Methylmalonic academia	Defect in valine, isoleucine metabolism	Plasma amino acids	Lethargy, FTT, hypotonia
Epidermolysis bullosum	Autosomal recessive and dominant forms	Skin biopsy	Multiple comorbidities

- *Irritant dermatitis* is the most common cause and may be related to prolonged skin contact with urine and feces, or from soaps and chemicals present in the diaper. Absorbable disposable diapers have decreased to incidence of this problem. Irritant dermatitis can usually be recognized by erythema and scaling of the skin with sparing of the skin folds.
- *Candida dermatitis*: Caused by secondary infection with *Candida*. It can be identified by involvement of the skin folds and presence of satellite lesions extending beyond the area of erythema. Diffuse erythema involving the perineal area with scaling and satellite lesions is the characteristic presentation.
 - Treatment includes topical antifungal agents such as nystatin, ketoconazole, or clotrimazole.
- When irritated by scratching, they release histamine, thereby, producing the characteristic wheals and occasionally blisters.
- With temperature changes, metabolic insults, such as fever or viral illnesses, large numbers of degranulating mast cells can produce flushing and systemic symptoms.
- Incontinentia pigmenti is an x-linked disorder occurring in females and is usually present at birth. It is a neurocutaneous syndrome with four overlapping stages of skin presentation.
 - The first stage is manifested by linear blisters surrounded by erythema.
 - The second stage presents usually before 6 months of age as the blisters evolve into warty plaques.
 - As the plaques disappear, they become whorles of hyperpigmentation during stage 3.
 - The final stage occurs later in adulthood with hypopigmentation and loss of hair and sweat glands.
 - Approximately 20% of patients will have seizures, various neurologic deficits, and mental retardation.

DISORDERS OF PIGMENTATION

- Dermal melanosis or Mongolian spots are diffuse blue–grey patches of melanocytes located in the dermis. They are most commonly found on the buttocks and sacral region of the lower back and slowly resolve over a few years. Mongolian spots should not be confused with bruising.
- *Neurofibromatosis* is a group of inherited neurocutaneous disorders manifested by multiple café au lait spots. Diagnosis is based on
 - Presence of six or more lesions measuring >5 mm in an infant.
 - Presence of axillary or groin “freckling.”
 - Lisch nodules (hammertomas of the iris).
 - These disorders are associated with a high incidence of optic gliomas.
- McCune–Albright syndrome is characterized by polyostotic fibrous dysplasia and multiple endocrine abnormalities.
- Urticaria pigmentosa can present in the first few months of life as red–brown macules that form a wheal and flare when scratched or rubbed.
 - This condition is caused by populations of mast cells in the cutaneous tissue.

VASCULAR LESIONS

- *Cutis marmorata* is a vascular instability caused by cold ambient temperature and may be present in an unclothed infant for the first few months of life. An erythematous, fine lacy, blanching rash is characteristic, and this may be confused with mottling. Acrocyanosis, cyanosis of the hands and feet, may also be present. Keeping the infant warm and in a neutral thermal environment will resolve the color changes in minutes.
- *Vascular malformations* are common skin presentations at birth or in the first few months of life. They can be secondary to abnormalities of the capillary, venous, arterial, or lymphatic systems. Most are benign and self-limited, but a few may herald serious systemic consequences.
 - The *salmon patch* is a common capillary malformation usually located on the forehead and upper eyelids and is present at birth. These usually resolve

within 1 to 2 years, but if present on the back of the neck, may persist for life.

- *Nevus flammeus, or Port wine stains*, are generally benign capillary malformations, but may persist for life. When present in the area innervated by the ophthalmic branch of the facial nerve (includes the upper eyelid and forehead), it is associated with *Sturge–Webber syndrome*, a neurocutaneous disorder with vascular malformations of the ipsilateral cerebral cortex and intractable seizures.
- Capillary hemangiomas are the most common vascular tumors in infancy. They may be unrecognized at birth, but appear early in the neonatal period as a small, red telangiectasia that rapidly grows to become the characteristic strawberry hemangioma over the first 6 months to 1 year, then slowly regress and disappear over the first decade. Multiple capillary hemangiomas may be associated with deep tissue and parenchymal involvement. Vascular malformations of the skin over the “beard distribution” of the face in an infant presenting with stridor or upper airway involvement suggests airway hemangiomas.

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QUESTIONS

1. A 2-day-old infant presents to the ED with a pinpoint papulopustular rash with surrounding erythema present on the face, trunk, and extremities. There is no fever, and exam is otherwise unremarkable. This finding would be most consistent with
 - A. Milia
 - B. Pustular melanosis
 - C. Miliaria
 - D. Erythema toxicum
 - E. Impetigo
2. A 6-month-old infant presents to the ED with multiple pigmented macules over his trunk that have been increasing in numbers since birth. When he takes a warm bath, they “swell up” and the patient becomes red, flushed, and irritable. This is most consistent with a diagnosis of
 - A. Pustular melanosis
 - B. Erythema toxicum
 - C. Urticaria pigmentosa
 - D. Neurofibromatosis
 - E. Dermal melanosis
3. A 2-month-old infant presents to the ED with progressively increasing difficulty breathing and stridor for 3 days. He has inspiratory stridor and moderate increased work of breathing. There is no croupy cough. Physical exam is otherwise unremarkable except for a streaky red nevus under the chin. The most appropriate next step is
 - A. Neck films to look for foreign body
 - B. Stat ENT consult for direct laryngoscopy
 - C. CT of neck
 - D. Epinephrine by aerosol
 - E. Immediate intubation
4. A 2-year-old toddler is seen in the ED for evaluation of fever. Mother is concerned about an “incidental finding” of multiple 0.5–2.0 cm pigmented oval macules on his trunk, and axilla. You suspect Type I neurofibromatosis. The most common serious complication would be
 - A. Astrocytoma
 - B. Acoustic neuroma
 - C. Neuroblastoma
 - D. Leukemia
 - E. Optic glioma

ANSWERS

1. D. Small pinpoint pustules on an erythematous base in an otherwise normal newborn are the typical presentation of erythema toxicum. If there is any question about the diagnosis, a simple scraping of a lesion with Wright stain will show sheets of eosinophils.
2. C. Pigmented lesions that urticate with rubbing, scraping, or with warm water is typical of urticaria pigmentosa. The diagnosis can be confirmed by gently scraping the pigmented lesion with a blunt object such as a tongue blade resulting in urtication.
3. B. Whenever an infant has stridor in the presence of a vascular nevus in the “beard distribution” of the face or neck, always worry about the presence of an airway hemangioma. This infant should be kept in a position of comfort, upright, and not manipulated until set up for a surgical airway if necessary.
4. E. Type 1 neurofibromatosis that has its onset in infancy has a 15% incidence of optic glioma.

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90 EAR AND NOSE EMERGENCIES

Evan J. Weiner

ACUTE OTITIS EXTERNA

- Otitis externa is a clinical diagnosis, with the hallmarks being pain upon manipulation of the tragus/pinna, redness and edema of the ear canal, and exudate.
- History of local trauma, swimming, and travel to a warm climate may or may not be present.
- Those with diabetes or who are immunocompromised may develop malignant otitis externa, a condition caused by *Pseudomonas* leading to severe cellulitis and an osteomyelitis of the underlying bone.
- Differential diagnosis includes abscess/folliculitis, atopic dermatitis, seborrheic dermatitis, contact dermatitis, Ramsay Hunt syndrome (herpes zoster), and acute otitis media with perforation.
- Treatment of uncomplicated cases consists of topical antibiotic preparations targeting *Pseudomonas* and external ear cleaning. Treatment with an antibiotic-steroid preparation may lead to faster cure rates.
- External ear cleaning consists of suctioning, irrigation, and dry swabbing. Placement of an ear wick may help deliver topical medications. It should be removed within 2 days.

ACUTE OTITIS MEDIA AND MASTOIDITIS

- Alteration of Eustachian tube function due to obstruction, inflammation, or excessive compliance leads to effusion and superinfection in the middle ear.

- Eustachian tubes of children are more horizontal in orientation, impeding drainage.
- Common conditions predisposing to otitis media include upper respiratory infections, allergic rhinitis, supine bottle-feeding, daycare attendance, and exposure to tobacco smoke. Breastfeeding may decrease the occurrence of otitis media.
- The major pathogens include *Streptococcus pneumoniae*, nontypeable *Haemophilus influenzae*, and *Moraxella catarrhalis*.
- Diagnosis is based on a history of fever, otalgia, nasal congestion, and fussiness as well as exam findings of a bulging tympanic membrane with decreased mobility. Redness of the membrane is less reliable due to confounding factors such as fever or crying; therefore, pneumatic otoscopy is imperative (Fig. 90-1).
- Tympanic membrane perforation is a common complication and manifests as purulent material within the ear canal (Fig. 90-2).
- Mastoiditis, one of the more serious suppurative complications, presents with postauricular erythema/tenderness and lateral displacement of the pinna. Computerized tomography is required to determine extent of bone involvement and intracranial extension.
- High-dose Amoxicillin (80–90 mg/kg/day) remains first-line treatment for acute otitis media. Amoxicillin/clavulanate and second and third-generation cephalosporins are second-line therapy for treatment failures. Macrolides are an option for those with penicillin allergy.
- Since many cases self-resolve, a 48-hour observation period without antibiotics may be reasonable for indeterminate cases and those without severe symptoms and are over 2 years of age.

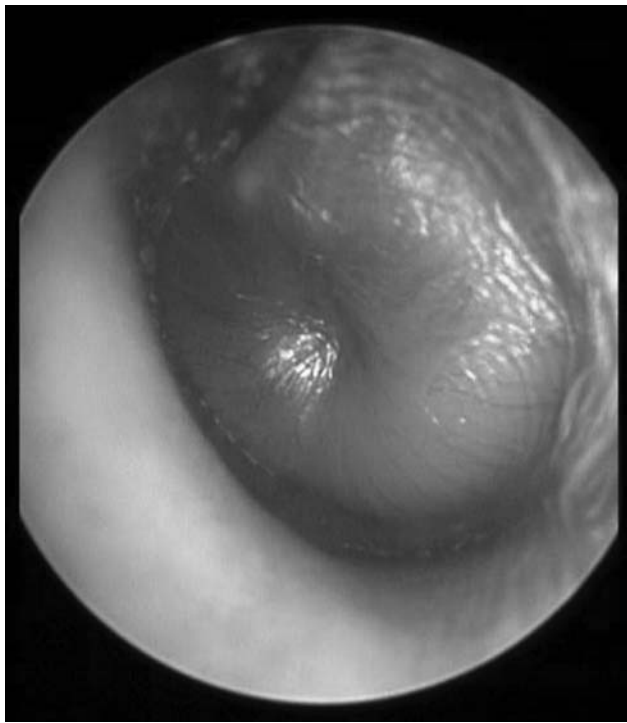


FIG. 90-1. Acute otitis media. Note the inflamed, bulging appearance of the tympanic membrane.



FIG. 90-2. Acute otitis media with tympanic membrane perforation. Note the otorrhea from the external auditory canal.

FOREIGN BODY OF THE NOSE AND EAR

- Beads, toy parts, eraser tips, food, and insects are common nose and ear foreign bodies.
- Retained nasal foreign bodies may lead to unilateral foul-smelling discharge, sinusitis, or periorbital cellulitis
- Depending on the object, options for removal include forceps removal, curettage, irrigation, and suctioning.
- Insects can be killed with mineral oil, antipyrine/benzocaine drops, lidocaine, or alcohol prior to removal.
- Wood and other vegetable matter tend to swell when wet and are best removed before irrigation.
- Nasal foreign bodies may be removed with positive pressure ventilation via a bag valve mask applied to the mouth while occluding the noninvolved nare. Alternatively, the caregiver can try mouth-to-mouth breathing.
- Button batteries can cause tissue necrosis, ossicular disruption, perforation, and facial nerve paralysis and need urgent removal.
- Unremoved foreign bodies should be referred to an otolaryngologist.

EPISTAXIS

- Epistaxis is most commonly caused by nose picking, mucosal irritation due to decreased humidity, and rhinitis.
- Bleeding disorders should be suspected in patients who have prolonged nosebleeds with minor trauma, frequent nosebleeds, or have a family history of bleeding disorders. The most common bleeding disorder associated with epistaxis is von Willebrand disease.
- Simple cases of resolved epistaxis can be treated with cool mist humidifiers, saline drops/spray, and petroleum jelly applied to nostrils.
- First-line treatment for active epistaxis is to apply continuous firm compression of the nasal alae for 5–10 minutes. A topical nasal decongestant such as oxymetazoline or phenylephrine may help as well.
- Second-line therapy includes silver nitrate application and electrocautery.
- Recently, the use of topical thrombin, fibrin, or hemostatic sealants was reported to be more effective, easier to use, and better tolerated than packing or cautery.
- Anterior nasal packing involves the use of nonresorbable (Xeroform™, Meroceel™) or resorbable agents (Gelfoam®, Surgicel) and should be removed in 1–2 days. A course of antibiotics should be prescribed.

SINUSITIS

- Two common predisposing factors of this bacterial superinfection include viral infection and allergic rhinitis. Other factors include tobacco smoke, dry air, foreign bodies, craniofacial abnormalities, septal deviation, adenoidal hypertrophy, polyps, and daycare attendance.
- Maxillary and ethmoid sinuses are present at birth, while the sphenoid sinuses appear after 5 years of age, and frontal sinuses appear around 7 years of age.
- The diagnosis is clinical consisting commonly of fever and prolonged nasal discharge (>10 days) and less commonly of headache, facial pain, and halitosis.
- Complications include facial cellulitis/abscess, orbital cellulitis, osteomyelitis of the skull (Pott's puffy tumor), cavernous sinus thrombosis, epidural abscess, subdural empyema, meningitis, and brain abscess.
- Imaging with computerized tomography is most useful when infectious extension is suspected.
- The causal organisms and treatment are the same as in acute otitis media. The treatment should extend for a minimum of 10 to 14 days, or longer depending on clinical response.

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QUESTIONS

1. You are seeing a 6-month old infant with history of fever and prolonged nasal discharge. You suspect possible sinusitis. Which of the following sinuses does this baby have?
 - A. No sinuses
 - B. Frontal sinuses
 - C. Maxillary and Ethmoid sinuses
 - D. Sphenoid sinuses
 - E. B, C, and D
2. A 13 year-old male presents with acute otalgia. The patient denies swimming, rhinorrhea, or fever. On examination, the patient has pain when his tragus is touched and purulent drainage in the external ear canal. There is no external ear swelling, and the tympanic membrane cannot be visualized. What is the best management for this condition?
 - A. Oral antibiotics
 - B. Intravenous antibiotics
 - C. Computerized tomography of temporal bones
 - D. Topical antibiotic-steroid preparation
 - E. Myringotomy
3. A 2-year-old male presents with a complaint of fever to 102 and right ear pain. On examination, there is a bulging poorly mobile tympanic membrane. Which of the following decreases the incidence of this condition?
 - A. Exposure to tobacco smoke
 - B. Breastfeeding
 - C. Daycare attendance
 - D. Upper respiratory infection
 - E. Allergic rhinitis
4. A 7-year-old patient presents with spontaneous epistaxis. By history, there have been recent recurrent episodes. On examination, there is no active bleeding. You also note scattered ecchymoses on the trunk and extremities. What is the appropriate management?
 - A. Nasal saline moisture
 - B. Cautery
 - C. Anterior nasal packing
 - D. Complete blood count
 - E. Cutting fingernails short
5. A 5-year-old patient has placed a small bean in her ear. Which of the following is correct regarding the appropriate management of this patient?
 - A. Curettage or suctioning may attempted if the patient is cooperative.
 - B. Irrigation is the method of choice for removal of this foreign body.
 - C. An emergent referral to an otolaryngologist is indicated if unsuccessful removal.

- D. Child abuse and neglect should be considered and a report made.
- E. Oral antibiotics should be administered .
6. You are seeing a febrile 10-month-old infant with fever, rhinorrhea, and an erythematous nonmobile tympanic membrane. The patient has no drug allergies. There is no tympanic membrane perforation. What is the most appropriate treatment?
- A. Amoxicillin/clavulanate
- B. A macrolide
- C. High-dose amoxicillin
- D. A topical antibiotic preparation
- E. 48-hour observation without antibiotics
7. A toddler has put a bead in their left nostril. There are no signs of purulent drainage. The parents are concerned that the child will be uncooperative for a removal procedure but would like to avoid procedural sedation. What is the best initial option for removal?
- A. Caregiver giving mouth-to-mouth breath while occluding clear nostril
- B. Intramuscular ketamine and curettage.
- C. Papoose and suction.
- D. Immediate consultation of otolaryngologist.
- E. Nasal irrigation.
8. You are seeing a 3-year-old male with recurrent episodes of otitis media. The patient again has fever and otalgia. On examination, you note that the patient has some tenderness behind the ear as well as displacement of the pinna. Which of the following is true regarding the management of this patient?
- A. Prescribe an oral antibiotic appropriate for resistant *Streptococcus pneumoniae*.
- B. Admit the patient and initiate broad-spectrum IV antibiotics
- C. Skull films should be ordered to visualize the temporal bones.
- D. Urgent follow up is indicated for tympanostomy tube placement.
- E. An incision and drainage should be done using procedural sedation in the ED.
9. You are seeing a febrile and fussy 8-month-old child. The child is completely immunized. On examination, both tympanic membranes appear red and bulging. What is the best next step?
- A. Oral antibiotics
- B. Topical antibiotics
- C. Pneumatic otoscopy
- D. 48-hour observation period
- E. Intravenous antibiotics
10. A toddler has placed a small button battery in her right nostril. After multiple techniques and attempts, you are unable to remove it. What is the best next step?
- A. Immediate consultation of otolaryngology.
- B. Close outpatient follow-up without antibiotics.
- C. Close outpatient follow-up with oral antibiotics.
- D. Nasal irrigation.
- E. Topical antibiotic drops.

ANSWERS

1. C. Maxillary and ethmoid sinuses are present at birth, while sphenoid sinuses appear at around age 5, and frontal sinuses appear around age 7.
2. D. This patient has signs of acute otitis externa. A history of swimming is not always required. The lack of external swelling and pinna deviation make mastoiditis unlikely. Acute otitis media would not have tenderness of the tragus.
3. B. Breastfeeding has been shown to decrease the incidence of acute otitis media. Common conditions predisposing to otitis media include upper respiratory infections, allergic rhinitis, supine bottle-feeding, daycare attendance, and exposure to tobacco smoke.
4. D. The recurrent nature of the bleeding as well as the bruising elevates concern for a bleeding disorder. A CBC is a good screen for disorders such as Idiopathic Thrombocytopenic Purpura and Leukemia. There is no active bleeding, so the patient does not require cautery or packing at the present time. Although a dry environment and nose picking may contribute to many cases of epistaxis, this patient requires more of an evaluation for a hematologic problem.
5. A. Options for removal of this foreign body include alligator forceps, curettage, and suctioning if the patient is cooperative and can be help still during the procedure. Irrigation should be avoided in wood or vegetable matter because the object may swell, complicating removal. Foreign bodies are common in the nose and ears of children. Unremoved foreign bodies should be referred to an otolaryngologist but not an emergently.
6. C. This patient should receive high-dose amoxicillin, which is appropriate empiric therapy for resistant *Streptococcus pneumoniae*. Amoxicillin/clavulanate is second-line therapy, and macrolides are for those who are penicillin allergic. A topical preparation would be appropriate if the patient had eardrum rupture. With a certain diagnosis of otitis media, this patient is too young for the “watchful

waiting” observation period. Those patients should be older than two years or have an uncertain diagnosis without severe symptoms.

7. A. A positive pressure breath is a relatively easy and painless method for solid objects such as a bead. The child may respond best to the caregiver attempting this removal technique. Use of a papoose and patient distraction may be a reasonable second-line option to assist with another method such as suction, curettage, or forceps. Immediate consultation of a specialist is rarely needed in uncomplicated nasal foreign bodies.
8. B. This patient has signs and symptoms of mastoiditis. The patient should be admitted on broad-spectrum IV antibiotics. A CT should be performed to determine extent of bone involvement and to rule out intracranial extension.
9. C. This patient has symptoms of possible acute otitis media; however, the presence of crying confounds the examination of erythematous tympanic membranes. The next step would be to accurately test for membrane mobility with pneumatic otoscopy.
10. A. Button batteries can cause tissue necrosis and septal perforation in a short period of time. They should be removed immediately. Instilling drops or fluids of any kind is contraindicated.

91 EMERGENCIES OF THE ORAL CAVITY AND NECK

Erica Katz

Gregory Garra

DENTOALVEOLAR TRAUMA

- Falls are the most common cause of injury in young children. Injuries in teenagers are more commonly from MVCs, sports, and altercations. The maxillary central incisor is the most commonly injured dental structure. Dental trauma may be a marker for child abuse.
- Trauma to the tooth can result in loosening, avulsion, or fracture. Luxations result from damage to the supporting structures of the teeth (periodontal ligament (PDL) and alveolar bone). Dental fractures may involve the tooth and/or supporting structures.
- Analgesia is indicated for all dentoalveolar injuries. NSAIDs have been shown to be as effective as narcotics for the treatment of dental pain. All but the most minor cases should be referred to a dentist for evaluation and radiographic documentation.

- Trauma to the primary teeth may result in infection of the tooth, pulp necrosis with subsequent discoloration, displacement, premature loss, problems with root resorption, or abnormal permanent tooth development.
- Primary tooth luxation injuries generally heal without any treatment. Remove severely loosened teeth if there is a high risk of aspiration. Reimplantation of an avulsed primary tooth is not recommended as it may damage permanent teeth.
- Injuries to permanent teeth can result in pulp necrosis or abscess formation. Permanent tooth luxation injuries threaten the integrity of the PDL. Concussion injuries require no treatment, but all others should be repositioned, splinted, and referred to dental.
- Avulsion injuries of the permanent teeth require immediate reimplantation or placement in a nutritive storage media (milk, saliva, Hanks solution) and prompt dental referral. Contaminated avulsions require penicillin prophylaxis.

OROPHARYNGEAL TRAUMA

- Soft palate or lateral pharyngeal wall trauma may present as avulsions, lacerations, or impalements. The major concern with oropharyngeal trauma is injury to the carotid artery resulting in thrombosis and neurologic sequelae.
- Injuries to the oropharyngeal soft tissues may present with bleeding, erythema, swelling, visible breaches in mucosal integrity, or presence of foreign bodies. Most soft tissue oropharyngeal injuries heal without complications. However, severe complications of seemingly innocuous injuries have been reported.
- There is no definitive diagnostic protocol. Carotid angiography is considered the gold standard for diagnosis of carotid injury but is invasive. Carotid ultrasound, CT, and MRA are other imaging modalities that can be considered.
- Carotid artery injury may result in an intimal tear with subsequent carotid and cerebrovascular thrombosis. The appearance of neurologic signs can be delayed for up to 60 hours and there are no reliable clinical factors to identify patients at increased risk.
- Lacerations greater than 1–2 cm or contaminated should receive antibiotics. Large gaping wounds should be sutured.
- Evidence of neurovascular injury requires hospitalization and imaging. The treatment of diagnosed carotid injuries requires neurosurgery and/or vascular surgery expertise. Patients with unreliable social situations should be hospitalized and observed.

DENTOALVEOLAR INFECTIONS

- Dentoalveolar infections are infections of the teeth and/or supporting structures (periodontium, bone). These infections include dentoalveolar abscesses and periodontal infections (gingivitis, periodontitis, and pericoronitis).
- Endodontic (periapical) abscesses are the most common dental abscesses in children. They are located at the apex of the tooth and typically originate from dental caries. Periodontic abscesses are more common in adults. They involve supportive tooth structures (PDL or alveolar bone) and form following infection of the periodontal tissues. Pericoronitis is an acute, localized infection of the opercula of partially erupted or impacted teeth. Wisdom teeth are commonly involved, but any tooth may be affected.
- Reversible pulpitis presents as transient pain in response to thermal stimulation. Irreversible pulpitis manifests as persistent, throbbing pain exacerbated by heat and relieved by cold.
- Extension of the infection to the periapical tissues is suggested by exquisite pain on chewing, touch, or percussion of the teeth. Fever, lymphadenopathy, tooth mobility, or edema of the soft tissues suggests the presence of a periapical abscess.
- Dental radiography may demonstrate radiolucencies where abscesses are located. In patients with local extension, a CT scan with IV contrast may differentiate deep space cellulitis from abscess.
- Treatment of dental infections is generally analgesia and outpatient dental referral except in cases of life-threatening infections of the deep fascial planes. Most complications result from direct spread of infection to deep spaces.
- Antibiotics are indicated where local or systemic spread of infection is suspected. Penicillin and amoxicillin are effective against the majority of infections. Infections present for greater than 3 days have a higher incidence of anaerobic agents. Clindamycin or penicillin plus metronidazole are appropriate regimens.
- Ludwig's angina is an extensive, life-threatening cellulitis of the submandibular, sublingual, and submental spaces. Patients present with erythema and a brawny, board-like swelling of the anterior neck, as well as pain, edema, and elevation of the tongue. These patients often appear toxic and dehydrated due to inability to swallow.
- Systemically ill patients with infections of deep facial planes require admission to the hospital for airway monitoring and IV antibiotics. Surgical drainage may be indicated. Involvement of anesthesia and otolaryngologic specialists is mandatory in patients with spread to the floor of the mouth.

INFECTIONS OF THE ORAL SOFT TISSUE

- Infections and ulcerative lesions of the oral mucosa are common. Recurrent aphthous stomatitis (RAS) is the most common mucosal disease in North America. Oral mucosal infections may be a sign of systemic disease. Oral candidiasis and angular cheilitis are the most common oral manifestation of children with HIV infection.
- Primary herpetic gingivostomatitis is caused by HSV I and manifests with high fever and swollen, red, friable gums with diffuse oropharyngeal mucosal lesions that may become confluent.
- Herpangina causes small vesicular lesions and punched-out ulcers in the posterior pharynx. Buccal and lingual vesicles and maculo-vesicular lesions on the hands and feet are present in hand-foot-mouth disease. Both are caused by Coxsackie virus.
- Candida stomatitis produces curdy or velvety white plaques on the tongue and/or oral mucosa and usually responds to oral nystatin.
- Laboratory evaluation is generally not indicated unless systemic illness is suspected. Treatment of soft tissue infections of the oral mucosa is largely symptomatic.
- Children with stomatitis, ulcers, or severe sore throat may benefit from gargling or from oral administration of a combination of kapectate or Maalox, diphenhydramine, and viscous lidocaine ("magic mouthwash"). Acyclovir is recommended for the treatment of primary herpetic gingivostomatitis in immunocompromised hosts at a dose of 15–30 mg/kg/day in three divided doses for 7 to 14 days.

PHARYNGITIS

- Pharyngitis is inflammation of the mucous membranes and underlying structures of the throat. 10% of children seek medical care for pharyngitis annually. The peak incidence is in children aged 4–7 years.
- 40% to 60% of cases are viral in origin. Group A beta-hemolytic streptococcus (*Streptococcus pyogenes*, GAS) is the most common bacterial cause of pharyngitis in children. Noninfectious causes include post-nasal drip, sinusitis, respiratory irritants such as tobacco smoke, or caustic ingestions.
- Acute GAS pharyngitis commonly presents as sore throat, fever, andodynophagia, with tonsillopharyngeal erythema and may or may not have exudate on exam. Headache, vomiting, abdominal pain, and scarlatiniform rash are often present.

- Respiratory symptoms such as clear rhinorrhea, cough, hoarseness, or mucosal ulcers suggest a viral etiology. Epstein-Barr virus (EBV) and cytomegalovirus (CMV) often have associated pharyngeal inflammation, diffuse lymphadenopathy, and hepatosplenomegaly.
- In sexually active adolescents, *Neisseria gonorrhoeae*, *Chlamydia trachomatis*, and syphilis should be considered. Pharyngitis accompanied by rash, joint pain, and urethral or vaginal discharge may indicate gonorrhea. In a young child it is a marker for sexual abuse and must be reported.
- Low-grade fever, follicular conjunctivitis, sore throat, and cervical lymphadenopathy characterize pharyngoconjunctival fever. Diphtheria presents as an adherent, grayish pharyngeal membrane with bull neck and toxic appearance.
- Rapid streptococcus detection is useful if positive, but culture confirmation of negative tests is recommended. A CBC, EBV titer (Monospot), syphilis screening tests, and/or cultures for *N gonorrhoeae* are indicated for unusual presentations or recurrent disease.
- Treatment should be initiated in cases with high clinical suspicion is high or confirmed diagnosis. Penicillin is the first line agent. In penicillin-allergic patients, use advanced macrolide antibiotics such as azithromycin.
- Suppuration can spread to contiguous tissues causing peritonsillar abscess (PTA), life-threatening Lemiere's postanginal sepsis (bacteremia from septic thrombophlebitis of the tonsillar vein), and Ludwig's angina.
- Nonsuppurative syndromes due to GAS include scarlet fever, acute rheumatic fever (ARF), and post-streptococcal glomerulonephritis (PSGN). ARF typically begins 1–5 weeks after GAS infection and is suggested by the Jones criteria (Table 91-1). Treating

acute GAS pharyngitis within 10 days of the illness can prevent ARF. Management of ARF involves confirmation of the diagnosis and limiting the sequelae of chronic valvular pathology.

- PSGN symptoms (dark urine, edema) appear 1–3 weeks after pharyngitis, or 3–6 weeks after scarlet fever or impetigo. The clinical course of PSGN is usually benign. Management of edema and hypertension with fluid restriction and furosemide is recommended. There is no evidence that treatment of GAS pharyngitis prevents PSGN.

PERITONSILLAR ABSCESS

- PTA is the most common deep infection of the head and neck and is typically seen in children >12 years of age.
- PTA usually begins as an episode of acute tonsillitis. Most PTAs are polymicrobial infections. Group A streptococci predominate. Other normal mouth flora, including anaerobes, may also be detected.
- Patients with PTA usually present with fever, gradually increasing pharyngeal discomfort, ipsilateral otalgia, trismus, and dysarthria. Drooling is not unusual. The voice has a muffled “hot potato” quality, and patients often appear toxic. Inferomedial displacement of the tonsil, contralateral shifting of the uvula and ipsilateral cervical adenopathy are typical.
- Most complications of PTA result from direct spread and invasion of adjacent tissue. Septicemia, parapharyngeal abscess, glottic edema, and airway obstruction are reported. The lateral pharyngeal recess provides a natural communication to the anterior chest and can result in mediastinitis, lung abscess, thrombophlebitis, or necrotizing fasciitis.
- The WBC count may be elevated and the throat culture will often grow strep. Needle aspiration of purulent material is the diagnostic gold standard and useful for directing antibiotic therapy. CT of the neck with contrast can differentiate PTA from peritonsillar cellulitis and delineate extension to adjacent structures.
- All patients with PTA should receive IV hydration and parenteral antibiotics. Penicillin is the drug of choice. Clindamycin or a second- or third-generation cephalosporin is used for beta-lactamase organisms.
- Cure rates are similar for I&D or three-point percutaneous needle aspiration. Needle aspiration and antibiotic therapy are effective in 85–90% of cases. Needle aspiration can be performed safely in cooperative patients, provides immediate relief of symptoms, and confirms the diagnosis with minimal trauma. I&D more rapidly relieves symptoms and may prevent recurrence of the disease.

TABLE 91-1 Jones Criteria

Major manifestations	Carditis Chorea Erythema marginatum Polyarthritits Subcutaneous nodules
Minor manifestations	Clinical Arthralgia Fever Laboratory Elevated acute-phase reactants (ESR, leukocyte count) ECG Prolonged PR interval
Evidence of antecedent GAS infection	Elevated or rising streptococcal antibody titers Positive throat culture or rapid antigen test Recent scarlet fever

RETROPHARYNGEAL ABSCESS

- Retropharyngeal abscess (RPA) is a local infection with accumulation of pus in the prevertebral soft tissue of the neck. Most cases occur in children < 6 years of age. *S aureus* and group A hemolytic streptococci are the most common pathogens.
- RPA frequently originates from infection of the nose, paranasal sinuses, or nasopharynx with subsequent spread to retropharyngeal lymph nodes. RPA can also result from direct inoculation of the space from penetrating oropharyngeal trauma or spread from contiguous spaces (parapharyngeal and submandibular).
- There is usually a prodrome of nasopharyngitis progressing to the abrupt onset of high fever, dysphagia, refusal of feeding, severe throat pain, drooling, stridor, “hot potato voice,” and neck pain or torticollis. Meningismus may result from irritation of the paravertebral ligaments.
- An elevated WBC count with a left shift is common. Soft tissue lateral neck radiography will usually demonstrate the retropharyngeal mass in stable patients. CT of the neck with IV contrast is very sensitive and will provide information on local structures. Gram stain and culture of purulent material obtained from I&D is essential.
- The most serious acute complications are airway obstruction and aspiration. Airway obstruction by RPA may mimic epiglottitis, croup, PTA, and mono. A standard approach to airway maintenance is vital since airway obstruction and aspiration can occur at any time.
- The abscess may rupture into the esophagus, mediastinum, or lungs resulting in pneumonia, mediastinitis, and empyema. Spread to adjacent spaces and structures may result in osteomyelitis and jugular vein thrombosis. Blood vessels may be eroded resulting in severe hemorrhage.
- Patients require hospitalization for hydration, IV antibiotics, analgesia, and surgical drainage. Emergent surgical intervention and drainage may be necessary with particular attention to the airway and ventilation.
- A penicillinase resistant penicillin with addition of clindamycin, metronidazole, and/or a third-generation cephalosporin is recommended. Duration of treatment is controversial.

ORAL PIERCINGS

- Piercing of the tongue, lips, and cheeks is popular amongst adolescents. The tongue is the most frequently pierced oral structure.
- Acute complications usually arise within 24 hours and include severe bleeding, infection, pain and

swelling of the tongue, changes in speech, galvanic current generation between the jewelry and resident metallic fillings, and allergy to metal.

- Chronic complications include traumatic injury to the teeth, localized tissue overgrowth, various mucogingival defects, and aspiration of the jewelry fragments.
- Life-threatening complications such as Ludwig’s angina have been described. Transmission of hepatitis B and HIV has also been reported.
- Tongue inflammation should be treated with removal of the piercing, local debridement, antibiotic therapy, and chlorhexidine mouthwashes.
- Dental injuries and infections of the fascial planes and spaces of the oral cavity and neck (eg Ludwig’s angina) should be treated as previously described.

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QUESTIONS

1. A 3-year-old girl presents to the emergency department minutes after a fall at home. On examination she is well appearing and playful, blood is noted on her upper lip, and she is missing a superior central incisor. The tooth socket is clean and there are no other obvious injuries and the parents have the tooth with them. What next most appropriate management?
 - A. Reimplant the tooth immediately
 - B. Place the tooth in Hanks Balanced Salt Solution and refer the patient to dental

- C. Call Child Protective Services
 D. Reassurance and outpatient dental follow-up
 E. Discharge the patient on penicillin
2. A 5-year-old boy is brought in by ambulance 1 hour after sustaining an injury to his oral cavity. According to mom, he fell while running around the apartment with a spoon in his mouth. He appears in no distress. Physical exam reveals a 5 mm laceration to the right lateral soft palate with mild swelling, with no active bleeding or airway compromise. His exam is otherwise unremarkable. On further history, it is unclear who was caring for the child at the time of injury. What is the appropriate management?
- A. Discharge the patient home with monitoring instructions
 B. Discharge the patient home on penicillin and monitoring instructions
 C. Suture the laceration and discharge the patient home on penicillin with monitoring instructions
 D. Perform carotid angiography and admit the patient
 E. Admit the patient for observation
3. A 16-year-old male presents with 1 week history of dental pain and 3 days of right lower jaw pain and swelling. On exam you see an ill-appearing patient in no respiratory distress with erythema, induration, and swelling to the right jaw and anterior neck. He has obvious dental caries and slight elevation of the tongue. What is the most appropriate management of this patient?
- A. Discharge the patient with analgesia and dental referral.
 B. Discharge the patient with analgesia, clindamycin, and dental referral.
 C. Admit the patient to the floor for IV antibiotics.
 D. Admit the patient to the pediatric ICU for IV antibiotics and airway monitoring.
 E. Admit the patient to the pediatric ICU for IV antibiotics and airway monitoring and obtain immediate ENT consultation.
4. An 11-month-old girl is brought to the pediatric emergency department by her mother with a history of 2 days of fever and 1 day of rash around her mouth and refusal to eat. The patient has vesicles around her mouth and in the posterior pharynx and on further exam you notice maculo-vesicular lesions on her hands and feet. Which organism is likely responsible for this patient's illness?
- A. Herpes simplex virus type 1
 B. *Candida albicans*
 C. Group A beta-hemolytic strep
 D. Coxsackie virus type A
 E. *Neisseria meningitidis*
5. An 8-year-old girl with no medical problems or allergies presents with a 3 day history of fever, sore throat, and abdominal pain. She has no rhinorrhea or cough and her 5-year-old sister was treated yesterday with similar symptoms. On examination you note well-appearing child with bilateral tonsillar erythema and exudates, tender cervical lymphadenopathy, and mild diffuse abdominal tenderness. What is the most appropriate management?
- A. Treat the patient with penicillin VK 25–50 mg/kg/day PO div Q6-8h × 10 days.
 B. Treat the patient with azithromycin 12 mg/kg PO daily × 5 days.
 C. Perform a rapid strep test and treat with penicillin if positive.
 D. Perform a throat culture and treat the patient with azithromycin if positive.
 E. Send a CBC, monospot, throat culture, syphilis screening and gonococcal culture and treat any identified pathogens.
6. A 7-year-old boy with a recent history of strep pharyngitis is brought to the ED by his mother for 2 days of decreased urination, dark urine and extremity swelling. On exam you note a well-appearing boy with a blood pressure of 130/90 and generalized body edema. His urinalysis is positive for protein and blood and his creatinine is 1.3. What is the most appropriate management?
- A. Discharge the patient on penicillin × 10 days.
 B. Discharge the patient with furosemide, fluid restriction, and PMD follow-up.
 C. Discharge the patient with furosemide, fluid restriction, and nephrology follow-up.
 D. Admit the patient to the floor on furosemide, fluid restriction, and bed rest.
 E. Admit the patient to the pediatric ICU for IV nifedipine and furosemide.
7. A 14-year-old boy presents with a sore throat and fever for 5 days. On exam you note an ill-appearing boy who has a muffled speaking voice, difficulty opening his mouth, left tonsillar swelling with contralateral uvular deviation, and left cervical adenopathy. What is the most likely diagnosis?
- A. Group A strep pharyngitis
 B. Retropharyngeal abscess
 C. Peritonsillar cellulitis
 D. Peritonsillar abscess
 E. Epiglottitis
8. A 2-year-old girl is brought into the emergency department in respiratory distress. She has a fever of 104, stiff neck, stridor, muffled voice, and is drooling. Her parents state she has had a sore throat for 3 days that has been increasingly severe and she has

been refusing to eat all day. On exam she is in obvious distress with tachypnea and tachycardia. What is the best next step?

- A. Order a CT neck with IV contrast to evaluate for abscess
 - B. Administer a 20 cc/kg fluid bolus and begin IV nafcillin and clindamycin
 - C. Place the patient on oxygen, alert anesthesia, and prepare for intubation
 - D. Call the pediatric ICU attending to admit the patient
 - E. Obtain a throat culture, CBC, chemistry and lateral neck X-ray
9. A 17-year-old girl presents with 1 week of tongue pain and difficulty speaking. On exam you note a tongue ring with surrounding erythema and swelling, with no signs of infection in the rest of the mouth or neck. The patient states she's had the piercing for 9 months. What is the appropriate management?
- A. Saline mouthwashes and cold beverages as needed
 - B. Saline mouthwashes and removal of the piercing
 - C. Antibiotics and chlorhexidine mouthwashes
 - D. Antibiotics, chlorhexidine mouthwashes, and removal of the piercing
 - E. Chlorhexidine mouthwashes, removal of the piercing, and admission for IV antibiotics

ANSWERS

1. D. Avulsion injuries of primary teeth require limited intervention and should not be reimplanted. Avulsed secondary (permanent) dentition should be placed in a physiologic solution and reimplanted as soon as possible. Although tooth fractures can be a sign of child abuse and should be reported to child protective services when suspected, falls are the most common cause of primary tooth avulsions in young children and abuse would not be suspected in this happy child with no other injuries. Antibiotics are indicated for contaminated tooth avulsions. Consider aspiration in avulsion injuries where the tooth is not found.
2. E. Admit the patient for observation. Patients with oropharyngeal injuries are at risk for carotid artery thrombosis, which although rare, can result in serious neurological sequelae. In this asymptomatic patient with a potentially unreliable social situation, hospitalization and observation are warranted as neurological symptoms can be delayed as much as 60 hours. Answer (A) would be appropriate in this patient if he had a reliable social situation. Patients with contaminated or large lacerations (>1–2 cm) should receive antibiotic prophylaxis. Large or gapping lacerations should be sutured. Patients with evidence of neurovascular injury should undergo carotid angiography.
3. E. This patient needs PICU admission, IV antibiotics, airway monitoring, and ENT consultation. This patient has Ludwig's angina, a life-threatening cellulitis of the submandibular, sublingual, and submental spaces. Admission to the ICU for IV antibiotics and airway monitoring with ENT consultation for incision and drainage is indicated. Choices (A) and (B) are appropriate for the treatment of simple caries and periodontic infections, respectfully. The need for close airway monitoring makes floor admission inappropriate.
4. D. Coxsackie virus type A. This patient has Hand-Foot-Mouth disease, which typically has a prodrome of fever followed by a vesicular rash in/around the mouth and on the hands and feet. The vesicles are painful, often causing the child to refuse to eat. The pain can be treated with analgesics and with "magic mouthwash" (kaopectate/Maalox, viscous lidocaine, and Benadryl). Coxsackie also causes herpangina, small punched out vesicles in the posterior pharynx. HSV1 causes primary herpetic gingivostomatitis, characterized by high fever and swollen, red, friable gums with diffuse oropharyngeal mucosal lesions. Candida stomatitis is characterized by curdy/velvety white plaques on the tongue and oral mucosa. Group A beta-hemolytic strep causes pharyngitis often with exudates. Infection with *Neisseria meningitidis* typically results in a toxic appearing child and may manifest with an exanthem.
5. A. This patient likely has Group A Strep pharyngitis based upon history and physical exam. Other symptoms that support the diagnosis of GAS pharyngitis are the abdominal pain and absence of respiratory symptoms. Patients may also present with a headache, vomiting, or scarlatiniform rash. Treatment based upon clinical grounds is appropriate. Penicillin, either oral or IM, is the first-line treatment for Group A Strep pharyngitis. Azithromycin is the correct treatment for a penicillin-allergic patient. Strep testing is appropriate in cases of diagnostic uncertainty. A negative rapid strep test should be followed by a culture. Choice (E) is unnecessary in this patient with an acute pharyngitis, but would be indicated in a patient with recurrent disease or unusual presentation.
6. D. Admit the patient to the floor on furosemide, fluid restriction, and bedrest. This patient with a recent strep infection has post-streptococcal glomerulonephritis (PSGN), the most common symptoms of which are dark urine and edema. Hypertension is very common. As this patient has moderate hypertension, generalized edema, oliguria, and an elevated creatinine, he should be admitted to the hospital

for treatment. Those without hypertension can be managed as outpatients with furosemide and fluid restriction and close PMD follow-up. Nephrology consultation is only necessary in severe, prolonged, or recurrent cases. ICU admission would be required for severe hypertension or encephalopathy. Choice (A) is the treatment for acute Group A strep pharyngitis and does not treat or prevent PSGN.

7. D. This patient has a peritonsillar abscess. Peritonsillar abscess is a polymicrobial infection that is usually preceded by acute pharyngitis. Trismus, muffled voice, unilateral tonsillar swelling with ipsilateral lymphadenopathy and contralateral uvular deviation are classic presentations of peritonsillar abscess. It should be treated with drainage and IV antibiotics. Group A strep pharyngitis is typically less severe, with bilateral tonsillar erythema with or without exudate, bilateral lymphadenopathy, and no trismus or uvular deviation. Retropharyngeal abscess is usually found in children <6 year old, and often presents with high fever, stiff neck, drooling, dysphagia, and stridor. Peritonsillar cellulitis is characterized by diffuse tonsillar erythema and swelling. Epiglottitis is

associated with fever, hoarse voice, stridor, and drooling, without tonsillar swelling and uvular deviation.

8. C. Place the patient on oxygen, alert anesthesia, and prepare for intubation. This patient likely has a retropharyngeal abscess (RPA) resulting in high fever, sore throat, muffled voice, stridor, and stiff neck. Her examination is consistent with respiratory distress and a potentially difficult airway management. RPAs can cause airway obstruction or aspiration, intubation may be difficult and anesthesia backup may be necessary. CT of the neck may be useful in defining the extent of the infection, but is unsafe in this patient with respiratory distress. Choices (B), (D), and (E) are all appropriate, but should come after airway management.
9. D. Antibiotics, chlorhexidine mouthwashes, and removal of the piercing are indicated in the treatment of this patient. Infection is a common complication of oral piercing and should be treated with antibiotics to prevent the spread of the infection. Life-threatening complications such as Ludwig's angina require admission for IV antibiotics. Removal of the piercing is necessary to aid recovery.

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92 EYE TRAUMA

Jeremiah J. Johnson

Stephen A. Colucciello

EPIDEMIOLOGY

- Ocular trauma is the leading cause of noncongenital blindness in individuals younger than 20 years
- 840,000 children annually injure an eye with an estimated cost of more than \$88 million
- Motor vehicle crashes and recreational injuries account for the majority of injuries
- Firearms and projectiles remain a significant source of severe injuries

HISTORY

- Caustic or chemical exposure requires immediate irrigation
- Mechanism of injury: have a high index of suspicion for an intraocular foreign body if the child hit metal striking metal
- Diplopia—ascertain if monocular or binocular
- Ask about pre-existing eye abnormalities to include corrective lenses
- Important aspects of the past medical history include
 - Coagulopathy, factor deficiencies, and sickle cell disease increase risk of intraocular hemorrhage
 - Osteogenesis imperfecta—increased risk of globe rupture

PHYSICAL EXAMINATION

- Visual acuity
 - Vital sign of the eye, document on every verbal and conscious patient.
 - Best predictor of outcome in traumatic eye injuries.
 - Topical anesthetic decreases pain and blephorospasm, and aids in the evaluation and diagnosis (Fig. 92-1).
 - Check vision with child's glasses, if worn. If glasses are lost or not available, correct refractive error by having the child look through a pinhole in a piece of paper.
- Visual fields tested in usual manner for older children, toy or light brought in from peripheral vision with younger children
- Adenexae
 - Examine peripherally to centrally
 - Avoid pressure on the eye, use eyelid retractors
 - Crepitus of periorbital tissues indicates orbital or sinus fracture
 - Lid lacerations involving medial third of eyelid often involve canalicular system—refer to ophthalmologist
 - Lid lacerations of tarsal edge should be referred to the ophthalmologist
- Conjunctiva and sclera
 - Examine for conjunctival hemorrhages. Circumferential subconjunctival hemorrhage and chemosis is seen with scleral rupture.
 - Perilimbal redness (ciliary flush) is seen with anterior chamber inflammation.

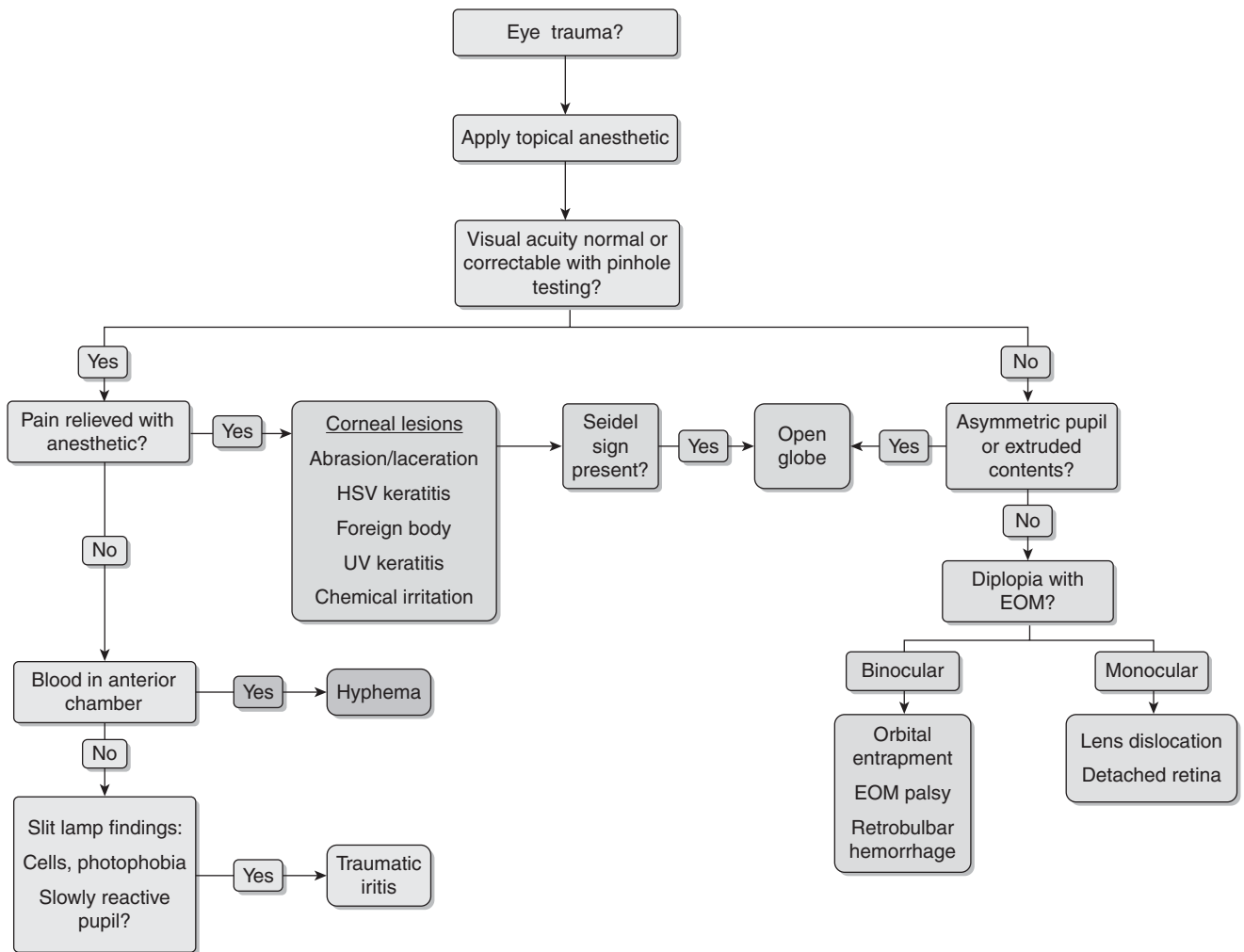


FIG. 92-1. Diagnostic algorithm for ocular trauma

- Pupils, iris, lens, and anterior chamber
 - Evaluate for asymmetry, reactivity, irregularity, a pointing pupil indicated iris detachment, and possible extrusion (Fig. 92-2).
 - Dilated, sluggish pupil seen in post-traumatic iritis and increased ocular pressures, third nerve compression due to herniation.
 - Evaluate for afferent pupillary defect (APD) (Marcus Gunn Pupil) seen in optic nerve lesions using the swinging flashlight test assessing.
 - Consensual pupillary constriction
 - Absence of direct constriction
 - Evaluate anterior chamber depth and clarity with slit lamp
- Funduscopic examination and intraocular pressure
 - Evaluate for clear vitreous and retinal detachment
 - Do not attempt to measure pressures if globe rupture suspected or in penetrating injuries



FIG. 92-2. Ruptured globe with extravasation of ocular contents. (Reproduced with permission from Knoop KJ, et al. *The Atlas of Emergency Medicine*, 3rd ed. McGraw-Hill, 2002)

MEDICATIONS

- Topical anesthetic (0.5% tetracaine or proparacaine) useful adjunct to physical exam
 - Onset within 1 minute, lasts 15–20 minutes
 - Pain relief indicated pathology of cornea or conjunctiva
 - Never prescribe for home use due to corneal toxicity and loss of protective reflexes
- Cycloplegics (homatropine, cyclopentolate)
 - Dilate pupil, treat pain of ciliary spasm
 - Aids in funduscopy exam
 - Contraindicated in angle closure glaucoma
- Ocular steroids
 - Useful in inflammatory conditions
 - Consult ophthalmology prior to use
 - May lead to glaucoma, cataract formation, acceleration of fungal, and herpetic infections
- Evaluate need for tetanus prophylaxis

SPECIFIC INJURIES

- Lid lacerations
 - Superficial lacerations not involving tarsal plate or lid margins are usually repairable by emergency physicians
 - Lacerations to margins, tarsal plate, area of medial canthus require referral
 - Lacerations containing fat are concerning for globe injury, consider imaging
 - Lacerations involving levator palpebrae require repair of the muscle to prevent posttraumatic ptosis, consider referral to ophthalmology or plastic surgery
- Subconjunctival hemorrhage—generally benign lesion, but may be a marker of more serious injury
 - Signs of significant ocular injury include decreased vision, severe pain, photophobia, and circumferential hemorrhage/chemosis
 - Exclude scleral rupture, open globe injury
 - Assess for associated orbital fracture
 - Benign causes include minor trauma, valsalva, coughing and will resolve in 2 weeks
- Corneal abrasion
 - Present as foreign body sensation, red eye, blepharospasm in older children, infants may only cry
 - Best seen with fluorescein staining and examination with ultraviolet or cobalt blue light
 - Exclude foreign body and corneal penetration. Evaluate for corneal ulcers and herpetic lesions which require ophthalmologic consultation
 - Treat abrasions with inexpensive ophthalmic antibiotics to prevent infection every 4–6 hours for 3–5 days. Mydriatic drops in the ED will decrease pain from ciliary spasm

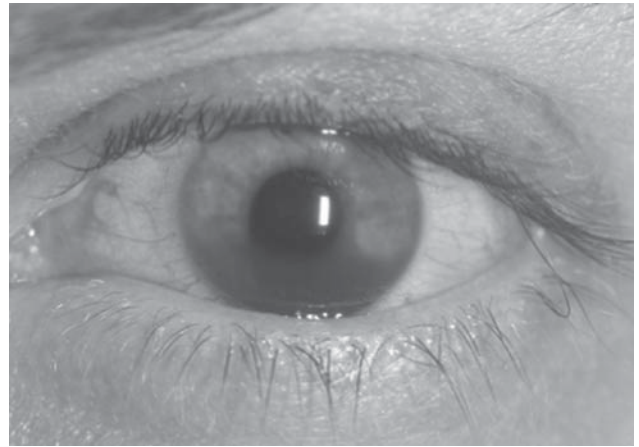


FIG. 92-3. Hyphema. (Reproduced with permission from Knoop KJ, et al. *The Atlas of Emergency Medicine*, 3rd ed. McGraw-Hill, 2002)

- Eye patching is not recommended
- Follow up in 24–48 hours if not resolved
- Traumatic iritis
 - Present 1–2 days after eye injury with photophobia, pain, and tearing decreased vision
 - Pain with accommodation. Slit lamp examination will reveal cells in the anterior chamber
 - Treat with cycloplegic drops, oral anti-inflammatory medications, and or topical steroids in consultation with ophthalmologist
- Hyphema
 - Blood in anterior chamber (see Fig. 92-3), usually due to blunt trauma
 - Described as percentage of chamber that fills with blood
 - Associated injuries: lens dislocation, vitreous hemorrhage, retinal damage
 - Can lead to loss of visual acuity due to corneal staining, deprivational amblyopia
 - Complications:
 - Rebleeding in up to 25%. Sickle cell disease/trait increase risk of rebleeding
 - Increased intraocular pressure due to aqueous outflow obstruction
 - Treatment
 - Bed rest with head of bed at 30°
 - Shield affected eye
 - Correct any underlying coagulopathy or bleeding disorder
 - Consultation with ophthalmology—treatment is directed at preventing rebleeding and treating increased ocular pressure
 - Avoid acetazolamide or osmotic agents in sickle cell patients as it increases risk of rebleeding

- Retinal injury
 - Injuries include concussion (commotio retinae), pre-retinal, superficial and deep hemorrhages, and retinal tears or detachments
 - Older children complain of light flashes or curtain
 - Refer retinal injuries to ophthalmologist
- Retrobulbar hemorrhage
 - Bleeding behind the eye can lead to orbital compartment syndrome, optic nerve injury
 - Findings: proptosis, decreased extraocular motion, decreased vision, APD, increased intraocular pressure
 - Treatment: surgical release (lateral canthotomy)
- Conjunctival and scleral lacerations
 - Assess depth by slit lamp
 - Scleral rupture often occurs at insertion of extraocular muscles and at the limbus
 - Clues to scleral rupture (open globe)
 - Decreased visual acuity
 - Abnormal anterior chamber
 - Positive Seidel's test—fluorescein drop placed on eye over area of interest has swirling dilution due to leaking aqueous.
 - Treatment
 - Conjunctival lacerations: topical antibiotics
 - Scleral laceration: eye shield, systemic antibiotics, ophthalmology consultation
- Corneal and scleral foreign bodies (FB)
 - Sensation of FB is a FB until proven otherwise
 - Evert lids, evaluate for retained foreign body in conjunctiva
 - Superficial FB can be removed with a moistened cotton tip applicator
 - FB adherent to cornea or sclera can be removed with ocular spud or a 25 gauge tuberculin needle under a slit lamp in a cooperative patient
 - Ferrous material will result in a rust ring, refer to ophthalmology
 - For scleral or corneal stroma penetration, consult ophthalmology in the ED (Fig. 92-4)
- Intraocular foreign body
 - Presentation may be subtle, high index of suspicion
 - Metal on metal or grinding, can result in the greatest risk for projectiles penetrating the globe
 - Imaging with CT or ultrasound for any concern (avoid MRI for metallic FB)
 - Treatment: eye shield, bed rest, broad spectrum antibiotics such as vancomycin and ceftazidime, ophthalmology consult
 - No topical antibiotics
 - For intubation, avoid succinylcholine and ketamine as they increase ocular pressures
 - For FB protruding from eye, cover affected eye with a cup or other protective shield, patch unaffected eye to prevent extraocular movements



FIG. 92-4. Intraocular foreign body. Note foreign body at arrow. Also note perforation of globe with asymmetric pupil and leaking vitreous. (Reproduced with permission from Shah, BR, Lucchesi, M. *Atlas of Pediatric Emergency Medicine*. McGraw-Hill, 2006)

- Chemical injuries to eye
 - Treatment (irrigation with normal saline) must occur prior to examination or vision testing
 - Extent of injury dependent on pH and duration of exposure, alkali exposures cause more damage than acids due to liquefaction necrosis
 - Irrigation is beneficial up to 24 hours after alkali exposure
 - Topical anesthetic +/- sedation may be necessary to effectively irrigate
 - Irrigate for at least 20 minutes with 2 L of normal saline, continue until pH is between 7.4 and 7.6. Recheck pH 10 minutes after stopping irrigation to ensure stable pH
- Ultraviolet keratitis
 - Eye pain, redness, photophobia occur 8–12 hours postexposure
 - Punctate shallow lesions seen with fluorescein staining of cornea
 - Sources: snow blindness, welding torches, tanning booths
 - Treatment: cycloplegia and oral analgesia
 - Self limited, resolves in 24–48 hours

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QUESTIONS

- A 3-year old presents with crying, red eyes and appears to be in pain after getting drain cleaner in the eye. Initial course of action should be:
 - Obtain visual acuity
 - Consult social work
 - Irrigate the eyes with normal saline
 - Neutralize eye pH with 0.1% acetic acid
 - Check pH of eye with litmus paper
- A 12-year-old boy presents after being struck in the right eye with a baseball. He complains of decreased vision and mild discomfort. Examination reveals visual acuity of 20/100 and a 30% hyphema. Which of the following medical conditions increases his risk of complications from rebleeding?
 - Diabetes
 - High blood pressure
 - Juvenile rheumatoid arthritis
 - Seizure disorder
 - Sickle cell trait
- A 14-year-old Boy Scout presents with eye pain, decreased vision and photophobia after smashing coins on an anvil with a sledgehammer. He was not using eye protection. Which of the following conditions is the most likely cause of his discomfort?
 - Infectious conjunctivitis
 - Exposure to caustic chemicals
 - Intraocular foreign body
 - Orbital fracture with entrapment
 - Ultraviolet keratitis
- A 7-year-old girl is complaining of a laceration to the eyelid after colliding with the family dog, a 140 pound Labrador retriever. Examination of the eye is normal with the exception of a small vertical laceration between the nasal bridge and medial canthus. Which important anatomical structure is at risk?
 - Lacrimal gland
 - Cannicular system
 - A branch of the trigeminal nerve
 - Frontal sinus
 - A branch of the facial nerve
- A 9-year-old male presents after his cousin “shot his eye out” with a BB gun. Examination reveals a pointing pupil and extrusion of iris. Which of the following is appropriate?
 - Placing a shield over the affected eye
 - Gentle irrigation with normal saline
 - Topical anesthetics for pain control and to assist with further examination
 - Measure intraocular pressures
 - Ketamine sedation for pain control and immediate ophthalmology consultation
- A 7-year-old female was involved in an MVA and sustained severe trauma to the face and right orbit. On examination, the right eye is proptotic; there is an afferent pupillary defect. CT of the orbits demonstrates a large amount of bleeding behind the globe. Which procedure is most indicated?
 - Posterior nasal packing
 - External carotid artery ligation
 - Morgan lens placement
 - Lateral canthotomy
 - Dilated funduscopic evaluation
- A 10-year-old male presents 24 hours after a fist fight in which he was hit once in the left eye. Initially, he was asymptomatic. He is now complaining of blurry vision, photophobia and eye pain the just started 1 hour ago. Examination reveals periorbital ecchymosis, intact extraocular movements, pain with accommodation, perilimbal injection and cells in the anterior chamber. Which of the following is most consistent with his presentation?
 - Retinal detachment
 - Subconjunctival hemorrhage
 - Traumatic iritis
 - Corneal abrasion
 - Lens dislocation

ANSWERS

1. C. The most important action is irrigating in cases of chemical or caustic exposure. Never try to neutralize acids with alkali or vice versa as it generates heat and can worsen the condition. All of the other answers are part of the patient's care but not the immediate action.
2. E. Diabetes, high blood pressure and JRA all have ocular complications, but are not associated with an increased risk of rebleeding from hyphema. Sickle cell disease and sickle cell trait have been associated with an increased risk of rebleeding of hyphema and warrant more conservative treatment.
3. C. History of metal striking metal is classic for intraocular foreign body and best explains the patient's symptoms. Infectious conjunctivitis typically presents with redness and discharge, but no significant decrease in visual acuity. There is no history of caustic exposure. Orbital fracture is a blunt trauma injury. UV keratitis is seen from UV exposure from snow, water, white sand, welding torches, etc.
4. B. The lacrimal gland is located in the superior temporal area of the orbit and produces tears. Tears are drained medially through the superior and inferior puncta into the canalicular system to the nasal lacrimal ducts. Laceration in the medial canthal area may involve this drainage system. The other answers are not consistent with the patient presentation.
5. A. Protecting the eye from further injury is of the highest priority. Avoid any maneuvers that might put pressure on the eye and further extrude intraocular contents. Nothing should go in the eye if an open globe is suspected. Ketamine will increase intraocular pressure and is not an ideal drug for sedation in this case.
6. D. This case describes a retrobulbar hemorrhage stretching the optic nerve. The treatment is lateral canthotomy to release retrobulbar pressures. The other maneuvers are either not indicated or contraindicated.
7. C. Post-traumatic iritis presents 12–24 hours after injury. Pain with accommodation, photophobia (direct and consensual) and decreased vision are seen. Retinal detachment is typically painless. A lens dislocation is possible in this scenario but the patient would have had immediate symptoms. Corneal abrasions present with a foreign body sensation. Subconjunctival hemorrhages are generally asymptomatic.

93 EYE EMERGENCIES IN CHILDHOOD

Lauren P. Ortega

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Ghazala Q. Sharieff

INTRODUCTION

- Children with eye disorders often come to the emergency department (ED). A thorough and complete history must be taken. A general physical examination should be performed and rapport should be built with the child.
- The eye should be touched and dilated only after a thorough systemic examination, and only if indicated. If the possibility of a globe perforation exists, manipulation of the eye should not be performed. Physicians must know when to stop and when to consult an ophthalmologist.
- Common errors to avoid when evaluating and treating eye emergencies include
 - Forgetting to examine the unaffected eye
 - Not thoroughly examining the injured eye
 - Failing to recognize globe perforation and/or using eye drops or ointment when perforation exists
 - Prescribing topical anesthetics and steroids
 - Failure to ensure proper follow-up

PHYSICAL EXAMINATION OF THE EYE AND DIFFERENTIAL CONSIDERATIONS

VISION

- Exam: Newborns/infants—ability to fix +/- follow an object
- Older children- Snellen letters or Allen figures
- Normal acuity:
 - 3 years—20/40
 - 4 years—20/30
 - Age 5 and up—20/20
- Differential diagnoses (DDX)—See sections below. Also consider cortical blindness due to head trauma, hypoglycemia, cerebrovascular accidents, and anesthetic accidents

LIDS AND ORBIT

- Note any erythema, edema, lacerations, or ecchymosis
- Lacerations involving the medial canthus may result in a lacrimal duct injury and should be repaired by an ophthalmologist

- The upper lid must be everted to rule out the presence of a foreign body
- Palpate for defects in the orbital bony structure and subcutaneous emphysema
- DDX- periorbital cellulitis, foreign body, lacerations, orbital fracture, and sinus fracture

ANTERIOR SEGMENT

- Inspect the sclera and conjunctiva for swelling, erythema, foreign bodies, hemorrhage, or discharge. Perform fluorescein examination for suspected corneal abrasion. Examine aqueous humor, iris, and lens using slit lamp and direct ophthalmoscope
- DDX- infection, trauma, iritis, cataracts, aniridia, and lens displacement

PUPILS AND EXTRAOCULAR MOVEMENTS

- Pupils should be black, round, symmetric, and equally reactive to light. Assess extraocular movements in all visual fields
- DDX- ruptured globe, intracranial process (elevated ICP), orbital fracture with entrapment of inferior rectus muscle, and coloboma

POSTERIOR SEGMENT

- Use direct ophthalmoscope to examine the vitreous, retina, and optic nerve for papilledema, hemorrhages, retinal detachment, and intraocular foreign bodies
- DDX—uveitis, endophthalmitis, trauma, diabetes, hypertension, sickle cell disease, leukemia, retinopathy of prematurity, retinal dysplasia, congenital toxoplasmosis, retinal tears, retinal detachment, central retinal vein occlusion, retinal artery occlusion, tumor, retinal phlebitis, and optic neuritis

INTRAOCULAR PRESSURE

- Evaluate papillary response and visual acuity. Measure intraocular pressure
 - Normal range in children—10–22 mm Hg
- DDX—glaucoma

COMMON EYE COMPLAINT

- The Red Eye
 - History is extremely important in differentiating the etiology of the red eye

TABLE 93-1 Differential Diagnosis of the Red Eye

Conjunctivitis
Bacterial
Viral herpes simplex
Other viral etiology
Chemical
Allergic (including vernal)
Neonatal ophthalmia
Corneal abrasion and corneal ulcer
Foreign bodies
Glaucoma
Hordeolum or chalazion
Iritis
Keratitis, episcleritis, scleritis
Periorbital or orbital cellulitis
Systemic disorders
Ataxia-telangiectasia
Collagen vascular disease
Infectious disease—mumps, measles, rubella, otitis media, influenza, EBV, CMV, coxsackie, Perinaud oculoglandular syndrome
Inflammatory bowel disease
Juvenile rheumatoid arthritis
Kawasaki disease
Lyme disease
Leukemia
Stevens–Johnson syndrome
Trauma
Chemical burns and thermal burns
Ruptured globe
Subconjunctival hemorrhage
Hyphema

- Although conjunctivitis is common in childhood, other etiologies must be thoroughly considered prior to arriving at the diagnosis (see Table 93-1)
- Eye Pain
 - DDX- foreign bodies, corneal abrasions, conjunctivitis, episcleritis, acute dacryocystitis, congenital glaucoma, uveitis, optic neuritis, hordeolum, keratitis, and trauma
- Excessive Tearing
 - In infants often due to nasolacrimal obstruction (dacryostenosis), corneal abrasion or glaucoma. May also be secondary to bacterial, viral, or allergic conjunctivitis
- Eye Discharge
 - Purulent eye discharge—most often associated with bacterial conjunctivitis
 - Mucoïd discharge—viral and allergic conjunctivitis
 - Crusting in addition to discharge—blepharitis

NONTRAUMATIC EYE DISORDERS

- **Eyelid infections (blepharitis):** Often involve glands of Zeis (hordeolum or “stye”) or Meibomian glands (chalazion)

- Hordeolum: Painful swelling and erythema of the eyelid. Treatment- warm compresses, eyelid hygiene, anti-staphylococcal antibiotic ointment or drops
- Chalazion: Painless, hard nodule often located in the mid-portion of the tarsus. Treatment similar to that of hordeolum but antibiotic treatment should be continued for several days after rupture of the chalazion. Lack of response to medical treatment requires surgical incision and drainage
- Impetigo contagiosa: Presents with vesicles then develops a yellowish crust. Treatment- remove crusts and apply topical antistaphylococcal /antistreptococcal antibiotics
- Herpetic blepharitis: Presents with vesicles which break down and form a yellowish-crusted surface. Treatment- trifluorothymidine topical drops

CELLULITIS OF THE PERIORBITAL AND ORBITAL REGION (SEE FIG. 93-1)

- Class I: periorbital or preseptal cellulitis
 - Periorbital edema, erythema, pain+/- chemosis, conjunctivitis, fever, and leukocytosis. Vision and function remain normal
 - Treatment: mild cases—oral antibiotics such as amoxicillin, amoxicillin-clavulanate or cefuroxime. In patients requiring hospitalization consider laboratory tests, cultures, and LP prior to initiating broad-spectrum antimicrobial therapy (ceftriaxone, with the addition of vancomycin or clindamycin in severe cases)
- Class II: orbital cellulitis
 - Similar to periorbital cellulitis but also with proptosis, impaired or painful eye movements, and chemosis +/- impairment of vision and septicemia
 - Hospitalization, workup and parenteral antibiotic treatment as above (periorbital cellulitis) + ophthalmologic and otolaryngology consult. Surgical indications- diminishing visual acuity, lack of improvement or suggestion of development of orbital abscess or cavernous venous thrombosis
- Class III: subperiosteal abscess
 - Edema, chemosis, and tenderness with ocular movement +/- globe displaced inferiorly or laterally, vision loss and proptosis
 - Treatment similar to that of orbital cellulitis +/- surgery
- Class IV: orbital abscess
 - Exophthalmos, chemosis, ophthalmoplegia, pain and visual impairment are generally severe; systemic toxicity may be impressive
- Treatment- similar to that of orbital cellulitis +/- surgery
- Class V: Cavernous sinus thrombosis
 - Marked lid edema, cranial nerve palsies, nausea, vomiting, headache, fever, pupillary dilation +/- systemic signs
 - Treatment similar to that of orbital cellulitis +/- surgery

SCLERITIS AND EPISCLERITIS

- Scleritis: severe pain and inflammation. Uncommon but can be associated with juvenile rheumatoid arthritis or various infectious processes (herpes simplex, varicella zoster, mumps, syphilis, and tuberculosis). Management consists of treating the underlying disease and some combination of oral non-steroidal anti-inflammatory agents, topical corticosteroids, and cycloplegics
- Episcleritis: inflammation (often a distinct area of injected episclera) and irritation (but not the severe pain associated with scleritis). Associated with a variety of diseases (varicella zoster, syphilis, Henoch-Schonlein purpura, erythema multiforme, and penicillin sensitivity). Management—similar to that of scleritis

OPHTHALMIA NEONATORUM

- Conjunctivitis in newborn infants (first 28 days of life)
 - Diagnosis—conjunctival scrapings for Gram's stain, Giemsa's stain, and viral/bacterial cultures (including Neisseria), and a rapid antigen test for Chlamydia
 - Differential diagnosis
- Chemical conjunctivitis: caused by silver nitrate drops. No treatment needed
- Bacterial conjunctivitis: *Chlamydia trachomatis* has a typical incubation period of 1–2 weeks. Unilateral, purulent involvement is characteristic and the infant may have an otitis media or afebrile pneumonia. Management—is with oral erythromycin for 2–3 weeks
- *Neisseria gonorrhoea*: Mean incubation period 6.5 days (range 1–31 days). Presents as a purulent, bilateral conjunctivitis +/- conjunctival hyperemia, chemosis, eyelid edema, and erythema. Neonates with suspected gonococcal infection should have a sepsis evaluation, including a lumbar puncture and gram stain/culture of eye discharge. Management—parenteral ceftriaxone or cefotaxime for 7 days (no meningitis) or

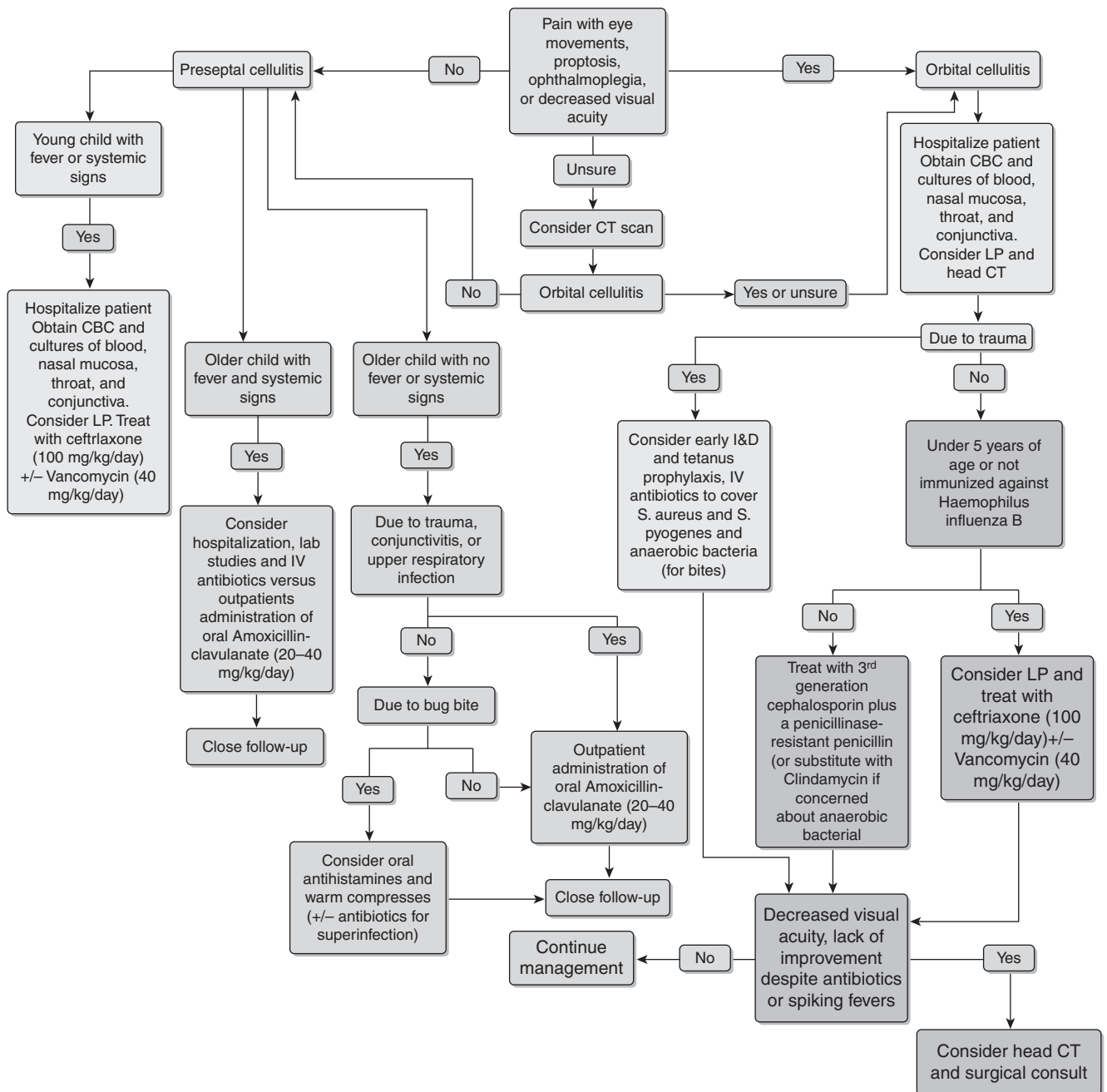


FIG. 93-1. Algorithm delineating the inpatient and outpatient diagnosis and management of preseptal and orbital cellulitis. (Algorithm courtesy of Dr. Lauren P. Ortega, Medical City Hospital, Dallas, TX.)

- 10–14 days (with meningitis) + frequent saline irrigation of the eyes
- Viral conjunctivitis: herpes simplex has an incubation of 2–14 days after birth. Characteristics not clinically distinctive; however, unilateral or bilateral epithelial dendrites are virtually diagnostic. A fluorescent antibody test and viral culture should be obtained. Treatment—IV acyclovir for 10 days and topical trifluorothymidine. Patients have a high risk of recurrence

- of keratitis later in life and may need ophthalmologic follow up. There are a few other viral etiologies.
- Obstructed nasolacrimal duct (dacryostenosis)
 - Failure to canalize a membrane (valve of Hasner). Causes pooling of tears onto the lower lid and cheeks and maceration of the eyelids
 - Treatment—massaging the lacrimal sac, warm compresses, +/- suppressive topical antimicrobials
 - Other— corneal abrasions, glaucoma, etc.

CONJUNCTIVITIS BEYOND THE NEONATAL PERIOD

- Characterized by normal vision, a gritty sensation in the eye, diffuse injection, and exudates (but not with visual changes, photophobia or lacrimation)
- Bacterial Conjunctivitis is associated with marked exudates, severe injection, and lid matting. Treatment is empiric with topical antimicrobial ointments or ophthalmic drops
- *N gonorrhoea* infections may be suspected in sexually active patients. Treat with ceftriaxone and saline irrigation
- Chlamydia trachomatis should be treated with oral erythromycin for 2–3 weeks or single oral dose of azithromycin (age \geq 1 year)
- Viral conjunctivitis has preauricular adenopathy is often associated with viral infections. Adenoviruses are the most common cause of viral conjunctivitis in children. Patients may also have URI symptoms, fever, rash, aseptic meningitis, myalgias, and headache. Treatment is symptomatic with cool compresses and removal from school as needed to prevent outbreaks
- Herpes simplex and varicella-zoster has unilateral, follicular conjunctivitis with vesicles localized to the eyelids and preauricular lymphadenopathy. Treat with trifluorothymidine drops +/- corneal debridement (rare). 25% of all children will have recurrences- ophthalmology f/u needed
- Allergic conjunctivitis is characterized by itching associated with mildly inflamed conjunctiva +/- edema. Treat with topical vasoconstrictors, antihistamines, mast cell stabilizers, and/or systemic antihistamines. Ophthalmologists may use steroids
- Vernal conjunctivitis has an itching, foreign-body sensation, clear mucoid discharge, photophobia, injection, and large “cobblestone” papillae on upper tarsal conjunctiva. Treat with topical mast cell stabilizers and antihistamines. Resistant disease requires ophthalmologic consult and may include systemic antihistamines, anti-inflammatory drops, steroid drops, and/or cyclosporine drops
- Special forms of conjunctivitis
 - Stevens–Johnson syndrome
 - Kawasaki disease
 - Chronic blepharoconjunctivitis from *Phthirus pubis*
 - Molluscum contagiosum
 - Parinaud oculoglandular syndrome (rare manifestation of cat scratch disease)
 - Infections with rubella, influenza, mumps, measles, infectious mononucleosis, cytomegalovirus,

papillomavirus, *Francisella tularensis*, *Sporothrix schenckii*

- *Mycobacterium tuberculosis*, *Treponema pallidum*, or *Borrelia burgdorferi*

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QUESTIONS

1. A 16-year-old male comes in after sustaining a direct blow to his left eye from a paintball. The patient is in severe pain and cursory exam reveals extensive conjunctival hemorrhage and scleral buckling. Which of the following is contraindicated at this time?
 - A. Visual acuity assessment
 - B. Palpation of the rim
 - C. Intraocular pressure assessment
 - D. Undilated slit lamp exam
 - E. Immediate ophthalmology consult
2. The patient in question #1 has also sustained several lacerations in the periorbital region. Which procedures should be deferred until subspecialty consultation can be obtained?
 - A. Repair of a laceration involving the medial canthus
 - B. Repair of a laceration of the involved eyelid
 - C. Eversion of the upper lid to rule out the presence of a foreign body
 - D. Choices B and C may be performed safely by an ED provider at this point
 - E. None of the procedures should be attempted at this time

3. A 5-year-old female is referred to the ED by her primary physician for evaluation of conjunctivitis, which has worsened despite treatment with appropriate topical antibiotics. In addition to a red eye, the child is also complaining of “seeing rainbows” and nausea. Exam reveals a sluggish pupil in the affected eye. Confirmation of your suspected diagnosis can be achieved by:
 - A. Obtaining corneal scrapings for gram’s stain and culture
 - B. Intraocular pressure measurement
 - C. Fluorescein examination
 - D. CT scan of the orbits
 - E. Rapid PCR testing for the suspected organism
4. A mother brings her 1-week-old infant into the ED for evaluation of her right eye. The mother states that she has noticed pooling of tears and small amounts of whitish secretions in the child’s right eye for the last few days. The infant seems comfortable and is consolable on exam. There is no crusting noted despite the lower lid appearing mildly macerated. The rest of the evaluation is within normal limits. What is the most likely diagnosis?
 - A. Dacryostenosis
 - B. Corneal abrasion
 - C. Congenital glaucoma
 - D. Bacterial conjunctivitis
 - E. Normal infant
5. What treatments may be considered for the patient in question #4?
 - A. Evaluation by an ophthalmologist for possible duct probing
 - B. Massage of the lacrimal sac
 - C. Oral antibiotics
 - D. Suppressing topical antibiotics
 - E. Both B and D
6. An 11-year-old male presents to the ED for evaluation of a painless nodule in the mid-portion of the tarsus of his left eyelid. The lesion has been present for 4 months and has not responded to warm compresses, lid hygiene and several rounds of topical antibiotics. At this point this patient requires:
 - A. Oncologic consultation to evaluate for eyelid rhabdomyosarcoma
 - B. Inpatient admission for IV antibiotic therapy
 - C. Trifluorothymidine drops
 - D. Ophthalmologic consultation for possible incision and drainage
 - E. Oral antibiotics
7. A 3-year-old female presents to the ED with right-sided periorbital edema, erythema, and pain. Exam reveals injected conjunctiva and fever. The patient is not cooperative, but you are concerned that she is not moving the affected eye as well as she should. At this point you should consider:
 - A. Oral antibiotics with close primary care follow-up
 - B. Oral antibiotics with ophthalmologic follow-up
 - C. Inpatient admission for further work-up, consultation, and IV antibiotic therapy
 - D. Immediate transfer to the OR for emergency surgical intervention
 - E. Outpatient treatment with warm compresses and an oral antihistamine
8. A 12-year-old male has been ill with sinusitis and periorbital edema, erythema, and pain for several days despite treatment with amoxicillin-clavulanate. He is brought via EMS to the ED from his primary care doctor’s office because he is now noted to have marked chemosis, high fevers, nausea, vomiting, and headache. He appears rather toxic and is not as responsive as he was earlier in the day. Exam reveals lateral gaze palsy and ptosis on the affected side. The most likely diagnosis is:
 - A. Periorbital cellulitis
 - B. Orbital cellulitis
 - C. Subperiosteal abscess
 - D. Orbital abscess
 - E. Cavernous sinus thrombosis
9. A 3-week-old infant of a mother with poor prenatal care presents to the ED for evaluation of unilateral purulent discharge from her right eye for the last 2 weeks. The affected eye is mildly swollen and irritated. The patient’s mother reports that her child has also developed a cough over the last 2 days despite remaining energetic and afebrile. Had this child presented earlier and ONLY with the eye findings she currently has, appropriate treatment AT THAT TIME would have been:
 - A. Oral erythromycin for 2–3 weeks
 - B. Inpatient admission for IV azithromycin
 - C. Inpatient admission for IV ceftriaxone
 - D. Inpatient admission for IV acyclovir
 - E. Topical erythromycin ointment
10. A 12-year-old female presents to the ED with a 2 day history of low-grade fever, myalgias, runny nose, congestion, and conjunctivitis. Her conjunctivae are injected bilaterally and she states that she has had mild discomfort as well as tearing and clear-yellow thin eye discharge. Exam reveals bilateral preauricular, cervical, and submandibular

shotty lymphadenopathy. Treatment of this patient requires:

- A. Topical antimicrobials
- B. Oral erythromycin for 2–3 weeks
- C. Trifluorothymidine drops
- D. Topical steroids
- E. None of the above

ANSWERS

1. C. The nature of the injury and the findings on exam should raise the suspicion for globe rupture. It is important to assess visual acuity in both the affected and unaffected eye in nearly every case of pediatric eye injury. Palpation of the orbital rim can detect orbital rim fractures or sinus fractures, which are possible in high-impact injuries. Care must be taken not to palpate the globe or apply pressure to it. The non-invasive nature of an undilated slit lamp exam makes this an acceptable means of assessing the patient's injuries. Serious eye injuries with the potential for globe rupture are ophthalmologic emergencies and warrant immediate subspecialty care. Until globe perforation has definitively been ruled out, the globe should not be manipulated in any fashion, including checking intraocular pressure.
2. E. A laceration of the medial canthus may have resulted in lacrimal duct injury and should be repaired by an ophthalmologist. In many cases, the repair of an eyelid laceration or eversion of the eyelid can be safely performed by a competent ED provider. In this case, however, where perforation of the globe is still a possibility, ALL procedures that may result in pressure being applied to the globe are contraindicated until an ophthalmologist has evaluated the patient.
3. B. One should suspect elevated intraocular pressures (glaucoma) in any child with a red eye. Although much less common than conjunctivitis, glaucoma must not be missed. In addition to presenting with a red eye, the child with acute glaucoma may also present with severe eye pain, complaints of blurred vision or "rainbow halos," nausea, and vomiting. Exam may reveal a cloudy cornea, and/or poorly reactive pupil. The definitive test for glaucoma is measurement of intraocular pressure. Other tests may be performed as part of a thorough workup of a red eye, depending on what etiologies are suspected.
4. A. Dacryostenosis (nasolacrimal duct stenosis) is due to failure of the valve of Hasner to canalize. Symptoms include pooling of tears and accumulation of non-purulent discharge in the affected eye. Lid maceration can result from the pooling of tears. A child with dacryostenosis is at higher risk for bacterial conjunctivitis due to poor drainage of eye secretions. Yellow or green purulent discharge, lid matting and erythema often mark these infections. A corneal abrasion may cause excessive tearing in an infant but is usually painful and unlikely to result in lid maceration due to its acuity. Congenital glaucoma must be considered in any child with excessive tearing but is much less common than dacryostenosis and is often accompanied by corneal clouding, eye redness, and light sensitivity. One may also see enlargement of the affected eye in congenital glaucoma. Although common, dacryostenosis is not considered normal.
5. E. One may consider treating dacryostenosis with suppressive topical antibiotics and warm compresses. Parents can be taught to massage the lacrimal sac with clean fingers at each feeding to potentially speed resolution. Watchful waiting is also a valid approach. A vast majority of children will experience spontaneous resolution by 1 year of age. Ophthalmologic evaluation in an otherwise uncomplicated case of dacryostenosis is not warranted at this time. Systemic oral antibiotics play no role in the treatment of this child who does not appear to have an infection.
6. D. From the description, this patient most likely has a chalazion that has not responded to conservative treatment. At this point incision and drainage must be considered. Further topical antibiotics as well and oral or IV antibiotics are unlikely to be helpful and this patient certainly does not meet requirements for inpatient admission. Trifluorothymidine drops are used to treat herpetic infections. The differential diagnosis for a chalazion does include rhabdomyosarcoma; however, ophthalmologic evaluation is a more logical first step in the treatment of this patient.
7. C. One must consider orbital cellulitis in this patient. Periorbital (preseptal) cellulitis often presents with edema, erythema, and pain. One may also see chemosis, conjunctivitis, fever, and leukocytosis. Children with mild cases may be successfully treated as outpatients with oral antibiotics, as long as close primary care or ophthalmologic follow-up is assured. In the presence of proptosis, impaired or painful eye movements, or impaired vision, one must suspect orbital cellulitis. Treatment includes hospitalization for further work-up, possible consultation, and treatment with

IV antibiotics. Surgical intervention may also be required; however, immediately transferring this patient to an OR is premature as she does not demonstrate absolute indications for surgery at this time. Indications for surgery include diminishing visual acuity, lack of improvement despite proper treatment, or suggestion of development of orbital abscess or cavernous venous thrombosis. Warm compresses and oral antihistamines may be appropriate for mild cases of periorbital irritation due to an allergy or bug bite, but are not appropriate in this case.

8. E. All of the listed conditions can be marked by periorbital edema, erythema, and pain. Fever and chemosis can also be seen in each of these cases. Nausea and vomiting are non-specific but are associated with more severe disease. Headache (especially in the distribution of the ophthalmic and maxillary branches of the fifth cranial nerve) leads to a higher suspicion for cavernous sinus thrombosis. Cranial nerve compromise, as manifested by ptosis and gaze palsy, is a hallmark of cavernous sinus thrombosis. One may also see significant toxicity, meningeal signs, and CNS depression with cavernous sinus thrombosis.
9. A. Based on the history, this patient most likely has a Chlamydia infection. Conjunctivitis with Chlamydia trachomatis is characterized by unilateral purulent involvement. The incubation period is typically 1–2 weeks after birth, but may appear later. If left untreated the infant may develop otitis media

or afebrile pneumonia. Chlamydia pneumonia is marked by a dry staccato cough and rales on exam. The treatment of Chlamydia pneumonia in the setting of possible poor compliance is beyond the scope of this text. Treatment of simple conjunctivitis due to Chlamydia trachomatis is achieved with 2–3 weeks of oral erythromycin therapy. Treatment does not require IV administration of a macrolide. IV ceftriaxone is appropriate treatment for many infections, including gonococcal conjunctivitis, but is not effective against Chlamydia infections. IV acyclovir is appropriate treatment for certain herpetic infections. Topical erythromycin is not effective treatment for Chlamydia conjunctivitis.

10. E. This patient appears to have a viral illness causing her conjunctivitis. Adenoviruses are the most common cause of viral conjunctivitis in children. Topical antibiotics are ineffective treatment for viral conjunctivitis without bacterial super-infection. Some practitioners may use topical antibiotics if there is no improvement in 24 to 48 hours. A child with viral conjunctivitis can be symptomatically treated with cool compresses alone. Oral erythromycin is appropriate treatment for conjunctivitis due to Chlamydia trachomatis. Trifluorothymidine drops are used in cases of confirmed or suspected Herpes simplex or Varicella-Zoster conjunctivitis. Topical steroids are not used routinely for viral conjunctivitis and in most cases an ophthalmologist should be the one to prescribe steroids in order to prevent possible complications.

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94 THE ADOLESCENT PREGNANT PATIENT

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INTRODUCTION

Despite the decrease in overall teenage pregnancies, there are over 850,000 teen pregnancies a year. Pregnant teens are less likely to receive prenatal care, are more likely to participate in high risk behaviors, and often use the emergency department for medical treatment. Thus, it is important for emergency physicians who routinely care for the adolescent patient to have basic knowledge and skills to recognize, stabilize, and treat complications of pregnancy.

PREECLAMPSIA

Also known as toxemia of pregnancy, preeclampsia is characterized by hypertension (BP of 140/90 mm Hg or greater), proteinuria, and/or edema during pregnancy of greater than 20 weeks gestation.

- Mild preeclampsia: the patient is asymptomatic, and the BP is less than 160/110 mm Hg. Mental status, deep tendon reflexes, abdominal exam, liver function tests and coagulation studies are all normal.
- Severe preeclampsia: the diastolic BP is greater than 110 mm Hg. The patient may have systemic problems like blurred vision, headache, abdominal pain, coagulopathy, disseminated intravascular coagulation (DIC), hyperreflexia, or vaginal bleeding (Table 94-1)
- Diagnostic studies: to assess maternal organ injury obtain
 - Complete blood count (CBC) with platelet count
 - Liver function transaminases

- Creatinine level
- Consider a cranial CT or MRI if there are mental status changes, seizures, or lateralizing neurologic signs
- Treatment: the most effective treatment is to deliver the infant
 - Reduces risk of maternal seizures and permanent maternal end organ damage
 - Reduces risks of complications to the infant
 - Seizure treatment includes magnesium sulfate and controlling blood pressure with hydralazine or labetalol (Table 94-2)
 - Immediate obstetrical consultation

ECLAMPSIA

Defined as the occurrence of seizures attributed to being a pregnant woman with preeclampsia.

- Complications of eclampsia include
 - Abruptio placentae
 - DIC
 - Pulmonary edema
 - Acute renal failure
 - Aspiration pneumonia
 - Cardiopulmonary arrest
 - Perinatal death rate ranges from 5.6% to 11.8%
- Diagnostic studies and treatment: same as in preeclampsia

HELLP SYNDROME

Hemolysis (microangiopathic hemolytic anemia), **E**levated Liver enzymes, **L**ow Platelets (<100,000/mm³)

- Part of the spectrum of eclampsia
- Hypertension may be absent in 10–15% of cases
- Lower platelets are associated with higher maternal and fetal morbidity and mortality
- Diagnostic studies and treatment: same as in preeclampsia

TABLE 94-1 Mild and Severe Preeclampsia Diagnostic Criteria

Mild preeclampsia
Systolic blood pressure ≥ 140 mm Hg or diastolic blood pressure ≥ 90 mm Hg (in women with previously normal blood pressure)
Proteinuria: ≥ 300 mg/L in 24 h or $\geq 1+$ urine dipstick
Severe preeclampsia
Systolic blood pressure ≥ 160 mm Hg or diastolic blood pressure ≥ 110 mm Hg
Proteinuria: urinary excretion of 2 g or more on 24-h urine collection or 3+ protein on urine dipstick on two random urine samples 4 h apart
Serum Cr ≥ 1.2 mg/dL (in patients with otherwise normal renal function)
Oliguria < 500 cc of urine in 24 h
Cerebral/visual disturbance
Pulmonary edema
Hemolysis
Epigastric or RUQ pain
Elevation of liver transaminases
Thrombocytopenia (platelets $< 100,000$)
Fetal growth restriction
Headache

TABLE 94-2 Treatment For Severe Preeclampsia

Systolic BP ≥ 160 mm Hg or diastolic BP ≥ 110 mm Hg:
Hydralazine 5–10 mg doses at 15–20 min intervals OR
Labetolol 10 mg and reevaluate in 10 min, increase to 20 mg in 10 min, then 40 mg, followed by another 40 mg, then another 80 mg, if not effective. Total dosage not to exceed 220 mg
Seizure prophylaxis: Magnesium sulfate IV load with 4–6 g over 15–20 min, then 2 g/h for desired levels of serum Mg of 4.8–8.4 mg/dL
Fluid resuscitation for oliguria (urine output < 30 cc/h)
500 cc of crystalloid IV bolus
Provide maximum of 3×500 cc crystalloid IV boluses to attain urine output of 100–125 cc/h
Delivery of infant

VAGINAL BLEEDING IN PREGNANCY

Common causes of vaginal bleeding in the first trimester include ectopic pregnancy, threatened abortion, spontaneous abortion, sexually transmitted infection, and trauma. Common causes in the later trimesters include placenta previa and placental abruption.

- Placenta previa: proximity of the implantation of the placenta in relation to the cervical os. The placenta is classified as complete, incomplete, marginal or low lying.
 - Primary cause of painless third trimester bleeding
 - Risk factors include maternal age greater than 35 years, ethnic minority, multiparity, cocaine use, previous placenta previa, prior cesarean section
 - Classic presentation: painless bright red blood from the vagina. Contractions may occur in 20% of patients

- Diagnostic studies
 - Digital examination may further exacerbate hemorrhaging and is contraindicated
 - Transvaginal ultrasonography is controversial but yields more information than transabdominal ultrasonography
 - Ultrasonography is necessary to make the diagnosis
 - MRI may occasionally be used to determine invasion into the myometrium (placenta accreta)
- Treatment
 - Patients with nonbleeding placenta previa: pelvic rest, limiting long distance travel, maintaining a safe hemoglobin level
 - Patients with hemorrhage: stabilization, insertion of two large bore intravenous (IV) catheters, crystalloid fluid resuscitation, packed red blood cell (PRBC) transfusions, and if needed, cesarean section and possibly hysterectomy.
- Transfer
 - Obtain early obstetrical consultation
 - Initiate early transfer to a facility capable of managing both the mother and newly born
- Placental abruption: characterized by the premature separation (partial or total) of the normally implanted placenta
 - May be concealed or present as vaginal bleeding
 - Accounts for 30% of bleeding during the latter half of pregnancy
 - Maternal risk factors include advanced maternal age, hypertension, advanced parity, tobacco use, poor nutrition, cocaine use, premature rupture of membranes, diabetes, preeclampsia, trauma, polyhydramnios, chorioamnionitis
 - Classic presentation includes dark red blood from the vagina, abdominal pain, uterine tenderness, and contractions
 - Amount of bleeding does not correlate with severity of abruption
 - Severe cases present with severe hemorrhage, uterine tetany and tenderness, maternal hypotension, coagulopathy, and fetal distress
 - Diagnostic studies
 - Ultrasonography is insensitive and unreliable in diagnosis of placental abruption
 - Diagnosis is often made postpartum upon inspection of placenta
 - Laboratory studies
 - CBC with platelet count
 - Coagulation studies
 - Fibrinogen, fibrin degradation products
 - Prothrombin (PT) and partial thromboplastin time (aPTT)
 - Type and cross match for maternal blood

- Management of patient with suspected placental abruption
 - Stabilize the airway, give 100% oxygen
 - Two large bore IV catheters
 - Fluid resuscitation
 - Monitor both mother and fetus
- Complications include preterm delivery, low birth weight, intrauterine growth restriction, still birth, and perinatal death
- Transfer
 - Obtain early obstetrical consultation
 - Initiate early transfer to a facility capable of managing both the mother and newly born
- Resuscitate with crystalloid and PRBC transfusions
- Obtain early obstetrical consultation for surgical management and transfer
- Give Rh immunoprophylaxis (50 µg anti-D immunoglobulin) intramuscularly (IM)
- Hemodynamically stable patient
 - Methotrexate (folic acid antagonist) can be used for medical management of an early ectopic pregnancy
 - Compared to laparoscopy, there is no difference in tubal preservation, but there are reduced hospitalization and costs
 - Contraindications to methotrexate treatment include unstable vital signs, active bleeding, tubal rupture, or contraindication to methotrexate

ECTOPIC PREGNANCY/THREATENED ABORTION

Ectopic pregnancy is the implantation of the blastocyst outside the endometrial lining of the uterine cavity.

- Mortality rate is highest in the 15–19 year old
 - Due to delay in seeking medical care
- 95% ectopic pregnancies are in the fallopian tube
- Risk factors are prior ectopic pregnancy, previous salpingitis, pelvic inflammatory disease (PID), and intrauterine device (IUD)
- Clinical presentation usually presents with first trimester abdominal pain and abnormal vaginal bleeding
 - Pelvic or abdominal pain is associated with exquisite tenderness
 - Vaginal bleeding may be scant dark colored blood, and may be confused with menses
 - Hemorrhage may be intra-abdominal and associated with hypovolemia, dizziness, or shock
 - Rarely a patient will present with the classic triad of abdominal tenderness, shock and adnexal mass
 - More commonly, the abdominal exam may be unremarkable or may be associated with adnexal or cervical motion tenderness
 - Physical findings are often unreliable
 - Diagnostic studies
 - Positive serum beta-HCG
 - Transvaginal ultrasound revealing an empty uterus
 - Serum progesterone less than 5 ng/mL (ectopic pregnancy or dead fetus)
 - Serum progesterone greater than 25 ng/mL (not an ectopic pregnancy)
 - CBC may reveal anemia and leukocytosis
- Treatment
 - Unstable patient
 - Stabilize the patient
 - Assess the airway, breathing, and circulation, apply 100% oxygen
 - Place two large bore IV catheters

DEEP VEIN THROMBOSIS AND PULMONARY EMBOLISM IN PREGNANCY AND POSTPARTUM

- Risk of developing a deep vein thrombosis (DVT) and or subsequent pulmonary embolus (PE) is five times greater during pregnancy and postpartum
- PE causes up to 15% maternal death during pregnancy or puerperium
- Risk factors for thrombotic events include the following: venous stasis, blood vessel trauma, hypercoagulability (increased circulating clotting factors, decreasing protein C), advanced maternal age, obesity, infection, prolonged bed rest (Table 94-3).
- Clinical presentation
 - DVT: leg pain or discomfort, especially of the left leg, swelling, tenderness, increased warmth, edema, and/or lower abdominal pain
 - PE: variable, but most commonly include dyspnea and tachypnea (90% cases), and to a lesser degree, chest pain, cough, tachycardia, and hemoptysis. Having a high index of suspicion is necessary.
- Diagnostic studies
 - DVT: diagnosis can be made using compression ultrasonography with or without color Doppler

TABLE 94-3 Risk Factors for Thrombotic Events in Pregnancy

Cesarean section delivery
Operative vaginal delivery
Maternal age (advancing age yields increasing risk)
Thrombophilias
Antiphospholipid antibodies
Obesity
Trauma
Infection
Prolonged bed rest

TABLE 94-4 Treatment of Acute PE

Intravenous heparin bolus 5000 units
Continuous infusion of 30 000 or more units of heparin over 24 h to prolong the PTT to 1.5–2.5 times control
Continue anticoagulation for 5–7 d
Therapeutic heparinization with subcutaneous dosing for ≥ 3 mo
May use low molecular weight heparin or Coumadin (if postpartum) after initial intravenous dosing

or when not definitive, with magnetic resonance venography (MRV).

- PE: diagnosis is often difficult.
 - Arterial blood gas (ABG), chest radiograph and electrocardiographic changes may be helpful, but not diagnostic
 - Ventilation/perfusion lung scan has low specificity
 - Spiral chest CT is currently the first line diagnostic test
 - Serum D-dimers is not recommended for the diagnosis of PE in the pregnant patient
 - ◻ Often increased during pregnancy, especially if there is preeclampsia
- Treatment
 - When DVT or PE is suspected clinically, begin treatment with unfractionated heparin or low-molecular-weight heparin until the diagnosis is excluded (Table 94-4)
 - DVT: elevate the affected leg, apply thromboembolic deterrent stockings
 - PE: give oxygen and analgesia as needed for pleuritic chest pain
- Transfer
 - Obtain early obstetrical consultation
 - Initiate early interfacility transfer to a facility capable of managing both the mother and newly born

HYDATIDIFORM MOLE

Defined as a proliferative abnormality of trophoblastic tissue or abnormality of chorionic villi that consists of trophoblastic proliferation and edema of villous stroma. Hydatidiform moles are classified as complete or partial molar pregnancies.

- Risk factors include maternal age younger than 15 years or older than 45 years, and previous molar pregnancies
- Clinical presentation and signs and symptoms
 - Abnormal uterine bleeding
 - Large uterus size
 - No fetal activity
 - Gestational hypertension
 - Preeclampsia prior to 24 weeks

- Hyperemesis
- Thyrotoxicosis
- Embolization
- Diagnostic studies
 - Serum beta-HCG level higher than expected for estimated gestation
 - Molar pregnancy on ultrasonography
- Treatment
 - Immediate uterine dilation and curettage

EMERGENCY CONTRACEPTION

Defined as a drug or device used to prevent pregnancy after sexual intercourse or after contraception fails. There are two common methods that can be used for up to 5 days post-sexual intercourse.

- Levonogestrel 1.5 mg (Plan B®)
 - Available without prescription
 - Most effective if used within the first 48 hours
 - Side effects include nausea, vomiting, irregular menstrual bleeding, dizziness, fatigue, breast tenderness, headache, and abdominal pain
 - If vomiting occurs within 1–2 hours post ingestion, repeat the dose
- Copper IUD containing 380 mm of copper (Paragard®)
 - No reduced efficacy if placed between 48 and 120 hours
 - Complications are the same as that when used for conventional contraception and include heavier menses, infection, uterine perforation, and expulsion

TRAUMA IN PREGNANCY

Trauma is most commonly due to motor vehicle crashes, assault, falls, and intimate partner violence (IPV)

- Injury to the fetus increases from 10% to 15% in the first trimester to 50% in the third trimester. This is due to increased uterine and fetal size.
- Teens are at the highest risk of IPV
- It is important to know the anatomic and physiologic changes that occur in pregnancy. These changes make the diagnosis of shock difficult.
 - Increased respiratory rate, heart rate, and cardiac output
 - Relative hypotension
 - Increased blood volume (50%)
 - Relative decrease in hematocrit
- Management of the traumatized pregnant patient is similar to all trauma victims
 - Follow ATLS guidelines
 - Pregnant women are at higher risk of aspiration
 - Protect the airway early

- Optimize oxygenation
- When needed, place tube thoracostomy 1–2 intercostal spaces higher than usual
- Place two large bore IV catheters
- Deflect the uterus to the left side once the airway is secured
 - Limits aortocaval compression
 - Improves venous return
- Monitor fetal heart tones in addition to mother’s vital signs
 - Fetal distress may be the first sign of maternal hemodynamic instability
 - If vasopressor support is indicated, ephedrine and mephentermine are preferred
 - Consult obstetric and surgical services early
- Diagnostic studies
 - Laboratory studies include
 - CBC
 - Type and cross match
 - Urinalysis
 - Blood gas analysis
 - Fibrinogen levels
 - Diagnostic imaging
 - Plain radiographs of the cervical spine, chest, and pelvis while shielding the uterus
 - Cranial and chest CT are generally safe
 - Avoid abdominal CTs early in pregnancy and consider alternative imaging modalities such as ultrasonography
 - Ultrasonography can be helpful to:
 - ◻ Locate free intraperitoneal fluid for DPL
 - ◻ Establishes gestational age
 - ◻ Placenta localization
 - ◻ Aids in diagnosis of placental abruption
 - ◻ Estimates amniotic fluid volume
 - Disposition
 - Dependent upon severity of injury and gestational age (GA)
 - Patients greater than 20 weeks GA are typically monitored for 4–6 hours
 - Maternal cardiopulmonary arrest or death may require emergent cesarean section within the first 4 minutes to preserve the unborn child

Rh sensitization: When an Rh negative mother is exposed to Rh positive fetal blood, sensitization occurs.

- Risks for Rh sensitization
 - Following trauma
 - Spontaneous or therapeutic abortion
 - Ectopic pregnancy
- Treatment
 - Anti-D immune globulin (RhoGAM)
 - Less than 12 weeks gestational age: 50 µg IM
 - Greater than 12 weeks gestational age: 300 µg IM

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QUESTIONS

1. A 16-year-old female presents to the ED with a severe headache and diffuse abdominal pain. She thinks her last menstrual period was “many” months ago but has not yet seen her doctor for this condition. At triage, she is noted to have a blood pressure of 175/130 mm Hg, swelling of the hands and feet, and an obvious gravid uterus. These findings are most worrisome for which of the following?
 - A. Mild preeclampsia
 - B. Severe preeclampsia
 - C. Eclampsia
 - D. Hydatidiform mole
 - E. Placental abruption
2. During the examination, the 16-year-old pregnant and hypertensive patient complains of increasing headache and has changes in mental status. She then has a generalized tonic clonic seizure. What would be the initial treatment of choice to control the seizures?
 - A. Valium
 - B. Lorazepam
 - C. Magnesium sulfate
 - D. Nifedipine
 - E. Terbutaline

3. A 14-year-old pregnant female with no previous medical problems presents to the ED with vaginal bleeding. The estimated gestational age is 30 weeks. The bleeding is painless and described as bright red in color. What would be the safest way to diagnose the cause of vaginal bleeding?
- Transvaginal ultrasonography
 - Digital examination of the vagina and cervix
 - Helical CT
 - Transabdominal ultrasonography
 - Postpartum upon inspection of the placenta
4. A 17-year-old female presents to the ED with abdominal pain and vaginal bleeding. The vaginal bleeding is described as scant in volume and dark in color. She reports that her last menstrual period was 2 months ago. Physical examination reveals a mildly tender abdomen and cervical motion tenderness on pelvic examination. The uterus is barely palpable and of normal size. The most likely diagnosis of this pregnant adolescent teen is which of the following?
- Ectopic pregnancy
 - Placental abruption
 - Placenta previa
 - Hydatidiform mole
 - Preclampsia
5. A 19-year-old female with a history of multiple sexual partners and one prior hospitalization for pelvic inflammatory disease, presents with abdominal and pelvic pain, nausea, and vomiting. Her last menstrual period (LMP) was 4 weeks ago. Vital signs reveal a HR 120, RR of 50, BP is 120/90 mm Hg, and oxygen saturations of 99%.
- The most helpful diagnostic tests that will aid in the diagnosis include which of the following?
- CBC with platelet count
 - Urinalysis
 - Serum progesterone > 25 ng/mL
 - Positive serum beta-HCG and a transvaginal ultrasound
 - Fibrinogen levels
6. An 18-year-old female presents to the ED with shortness of breath and chest pain with deep inspiration. The pain does not radiate, is primarily located on the right side of her chest. There is a strong family history of asthma and an aunt who now lives with her has recently immigrated from Mexico. The patient also reports having a cesarean section 2 weeks ago. Her newborn infant is doing well. What test would be diagnostic in determining the cause of shortness of breath and chest pain in this postpartum patient?
- Serum D-dimer
 - Arterial blood gas
 - Chest radiograph
 - ECG
 - Spiral chest CT
7. While evaluating the above patient and monitoring for other complications, what treatment(s) should be instituted while the work up is in progress?
- Albuterol nebulized solution
 - Low-molecular-weight heparin infusion
 - Baby aspirin
 - Labetolol
 - Magnesium sulfate
8. An 18-year-old pregnant female is brought in by paramedics after crashing her car into the freeway median. She was placed in a cervical collar and onto a back board for precaution. With pregnancy and the physiologic changes that occur during this time, making the diagnosis of shock can be difficult. What is one of the many normal physiologic changes found in a pregnant female?
- Decreased respiratory rate
 - Increased heart rate
 - Decreased cardiac output
 - Relative increase in hematocrit
 - Increased gastric emptying time
9. A 16-year-old female presents to the ED with epistaxis and scant vaginal bleeding for one day. She denies trauma other than tripping down the stairs. She is living with her boyfriend and her 2 year old daughter. She is hemodynamically stable with normal vital signs. Physical exam is remarkable for bruising around the left eye and cheek, bruising about the neck, dried blood in the nares, and circumferential bruising of the right arm. Abdominal exam reveals a nontender, approximately 20 week gravid uterus. There is scant dark blood at the vaginal opening. These findings are worrisome for a diagnosis of which of the following?
- Intimate partner violence
 - von Willebrand disease
 - Placenta previa
 - Leukemia
 - Threatened abortion

ANSWERS

- Severe preeclampsia or toxemia of pregnancy is characterized by hypertension with a diastolic blood pressure greater than 110 mm Hg, proteinuria, and/or edema and systemic symptoms such as headache blurred vision or abdominal pain. At this point, the patient does not have eclampsia because she has not had a seizure. However, prophylactic treatment with magnesium sulfate is indicated. Patients with mild preeclampsia are asymptomatic with a blood

pressure less than 160/110 mm Hg. Hydatidiform mole presents with abnormal uterine bleeding, large uterus, and no fetal activity, or preeclampsia prior to 24 weeks. Placental abruption presents with dark red blood from the vagina, abdominal pain, uterine tenderness, and in severe cases, maternal hypotension, coagulaopathy, and fetal distress.

2. C. Magnesium sulfate is the medication of choice for seizure prophylaxis and treatment in the patient with severe preeclampsia and/or eclampsia. Seizures usually terminate after the loading dose of magnesium sulfate. The loading dose is 4–6 g over 15–20 minutes, then 2 g/h for desired levels of serum Mg of 4.8–8.4 mg/dL. Once seizures terminate, the majority of patients will show improvement in blood pressure as well. Hypertension must be addressed after initiation of magnesium sulfate. The goal is to maintain a systolic blood pressure between 140 and 160 mm Hg and a diastolic blood pressure between 90 and 110 mm Hg. Hydralazine or labetalol are the preferred antihypertensive agents for the treatment of hypertension. Hydralazine is administered at 5–10 mg doses at 15–20 minutes intervals. Labetolol 10 mg is given initially. Reevaluate in 10 minutes, increase to 20 mg in 10 minutes, then 40 mg, followed by another 40 mg, then another 80 mg, if not effective. Total dosage should not exceed 220 mg. Valium and lorazepam are benzodiazepines used in the treatment of status epilepticus. Terbutaline is not indicated for the treatment of seizures.
3. D. The patient in the above vignette presents with the classic presentation of placenta previa. The patient is in the later trimester and has painless bright red bleeding from the vagina. Diagnosis of placenta previa requires caution since examination of the cervix may exacerbate hemorrhaging by further tearing the placenta. Digital examination is contraindicated. Transvaginal ultrasonography is controversial because of the risk of further tearing the placenta. Transabdominal ultrasonography is safe. Helical CT is not indicated.
4. A. Ectopic pregnancy is the leading cause of maternal morbidity in the first half of pregnancy in the United States, and should be suspected in any patient with vaginal bleeding and abdominal pain. Clinically, most cases of ectopic pregnancy present within the first 8 weeks of gestation with abdominal pain or abnormal vaginal bleeding. The vaginal bleeding may be confused with menses, and is usually scant and dark in color. Commonly the abdominal examination may be unremarkable or there may be adnexal or cervical motion tenderness. Placental abruption can be seen with vaginal bleeding, abdominal pain and uterine tenderness. Bleeding is also dark in color and the amount is variable. Typically the bleeding occurs in the latter half of pregnancy. Placenta previa presents with painless bright red bleeding from the vagina. The uterus is usually soft; however, contractions can occur in up to 20% of patients. Hydatidiform mole presents also with abnormal uterine bleeding, but the uterus is large in size.
5. D. The patient in the vignette has risk factors, symptoms, and physical exam findings worrisome for an ectopic pregnancy. Often, the history and physical examination are insufficient to detect an unruptured ectopic pregnancy, and thus other modalities to aid in the diagnosis must be used. The diagnosis of an ectopic pregnancy can be confirmed by a positive serum beta-HCG and a Transvaginal ultrasound showing an empty uterus with or without an adnexal mass. If the serum progesterone is greater than 25 ng/mL, an ectopic pregnancy is unlikely. If the serum progesterone is less than 5 ng/mL, an ectopic or dead fetus is likely. A CBC may reveal a leukocytosis but this finding is nonspecific. Fibrinogen levels are measured when there are concerns for DIC and vaginal bleeding.
6. E. The diagnosis of PE can be difficult and a high index of suspicion is necessary. The current first line diagnostic test is a spiral or helical chest CT. Spiral CT allows visualization of the entire lung with a single breath and has high specificity and sensitivity. The negative predictive value is 99%. Arterial blood gas, chest radiograph and ECG changes will be seen only variably. The most common ECG abnormalities in the setting of PE are tachycardia and nonspecific ST-T wave abnormalities. These findings are seen only in 20% of the cases. Ventilation/perfusion (V/Q) lung scan is safe during pregnancy; however, it also has low specificity. D-dimer levels are naturally increased during pregnancy and are thus not helpful in the diagnosis of PE.
7. B. It is recommended that when a DVT or PE is suspected clinically, treatment with unfractionated heparin or low-molecular-weight heparin should be started until the diagnosis is excluded, unless anticoagulation is contraindicated.
8. B. The approach to the traumatized pregnant patient requires understanding of the physiologic changes that occur during pregnancy. Changes include an increase in respiratory rate, heart rate and cardiac output. There is also a relative hypotension, an increase in blood volume by 50% and a relative decrease in hematocrit. All of these changes

make the diagnosis of shock difficult. There is an increased risk for aspiration from delayed gastric emptying during pregnancy.

9. A. Pregnant adolescents are at the highest risk of intimate partner violence and are at an increased risk of vaginal bleeding, poor weight gain, alcohol or drug abuse, and death. This vignette describes an at risk pregnant teen with unexplained bruises of her face, neck, and arm. Her physical findings are worrisome for intimate partner violence. She has evidence of vaginal bleeding and thus, potential trauma to the uterus and fetus must be considered. In the evaluation of a pregnant adolescent with vaginal bleeding, other causes such as leukemia and clotting disorders, threatened abortion, infection, placenta previa, and placental abruption must be considered.

95 GYNECOLOGIC DISORDERS OF INFANCY, CHILDHOOD, AND ADOLESCENCE

Maria Stephan

INTRODUCTION

The pathologic processes in infants and young children differ significantly for the adolescent female patient. It is important to have knowledge of basic anatomic and physiologic variations among age groups as well as be

able to provide compassionate and quality care to young female patients.

EVALUATION OF PREMENARCHAL PATIENTS

INFANTS AND TODDLERS

- Labia majora appear full. Labia minora appear thickened and enlarged up to 8 weeks from maternal estrogens.
- Hymen is circumferential, surrounding the vaginal orifice. Its tissues are thick, redundant, and elastic (Fig 95-1).

YOUNGER SCHOOL-AGED CHILDREN (3–6 YEARS)

- Lowest estrogen levels between ages 3 and 9 years.
- Clitoris is less prominent. Labia appear full.
- Hymen is thin, translucent “crescentic” appearance as tissue recedes from anterior vaginal orifice. Great degree of variability exists in appearance.
- Alkaline vaginal pH.

OLDER SCHOOL-AGED CHILDREN (7–12 YEARS)

- Hymen and vaginal mucosa thicken.
- Vaginal pH becomes acidotic.
- Physiologic leucorrhea may be present. This is seen as a thin white vaginal discharge.

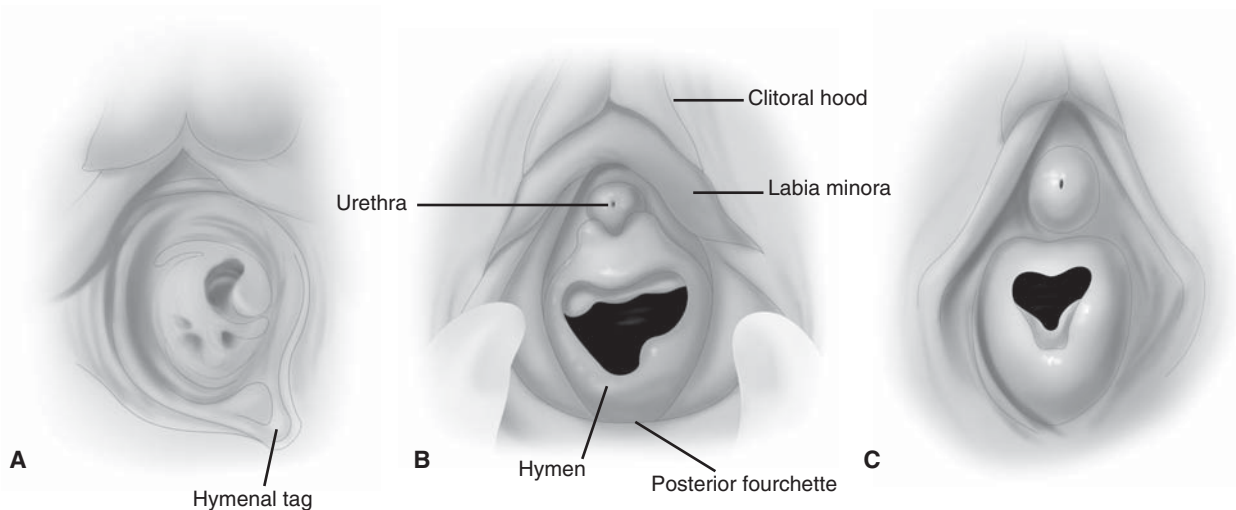


FIG. 95-1. Developmental changes in genital morphology. (A) Infant: hymen circumferential and redundant. (B) Preschool through school age: rudimentary labia minora, thinner hymen. (C) Early puberty: labia minora develop and hymen thickens.

TABLE 95-1 Stages of Sexual Development

SEXUAL MATURITY RATING (SMR)	BREAST DEVELOPMENT	PUBIC HAIR DEVELOPMENT
1	Elevation of papilla	No pubic hair
2	Breast buds appear, areola appears as small mound	Straight hair extends along labia
3	Breast enlargement with protrusion of papilla or nipple	Darker and increased in quantity on pubis and remains in triangle
4	Breast enlargement with projection of areola and nipple as secondary mound	Darker, more coarse and curly, adult distribution but not as abundant
5	Adult configuration with no projection of areola as secondary mound	Adult pattern with extension of hair into medial aspect of thigh

APPROACH TO THE PHYSICAL EXAMINATION

- A successful examination requires sensitivity to the child and family, addressing their concerns and fears.
- Sexual development stages are summarized in Table 95-1.
- An internal speculum exam is indicated only for patients who are sexually active or those with bleeding from trauma or suspected foreign bodies. Sometimes sedation or general anesthesia may be required for an adequate exam.
- On prepubertal girls, direct visualization is usually all that is required.
- Best positions for examination are: frog leg or the knee chest position. Use of labial traction will allow better visualization of the vaginal introital area.
- Specimen collection for gram stain, culture and wet mount is indicated for any vaginal discharge. Samples can be collected from the vaginal introital area using saline-moistened swabs.

DISORDERS OF INFANCY AND CHILDHOOD

NEONATAL PHYSIOLOGIC VARIANTS

- Breast buds, milky nipple discharge, and transient vaginal secretions or bleeding may all be seen in newborn girls secondary to circulation of maternal estrogens. This should subside within 2 weeks.

CONGENITAL VAGINAL OBSTRUCTION

- *Etiology*: occurs secondary to imperforate hymen (a remnant of urogenital membrane) or vaginal atresia, also known as transverse vaginal septum (failure of canalization of the vaginal plate at various levels). Other causes include: cloacal malformation and common urogenital sinus.

- *Clinical presentation*: may present as abdominal mass or bulging mass at the introitus. An imperforate hymen undetected until puberty may present as primary amenorrhea with cyclic intermittent monthly abdominal pain. Physical examination reveals a bluish bulge at the introitus, known as hematocolpos, hydrocolpos (fluid distension of the vagina), or hydrometrocolpos (fluid distension of the uterus and vagina).
- Congenital renal anomalies are associated with vaginal atresia.
- *Management*: surgical correction is required.

LABIAL ADHESIONS

- *Etiology*: also known as labial agglutination, begins posteriorly and extends toward the clitoris. Etiology unknown, but thought to be secondary to prepubertal estrogen deficiency and inflammation resulting in thinning of the superficial mucosal layers.
- Occurs ages 1–6, peaking at 1–2 years. Prevalence 1–3%.
- *Clinical presentation*: parent notices vaginal area “closing.” Distinguished from ambiguous genitalia by the raphe or vertical line connecting the adhesion.
- *Management*: most resolve without intervention. First line therapy is estrogen cream applied BID \times 1–2 weeks if patient is symptomatic with pain or UTIs. If adhesions and symptoms persist, surgical separation may be required. Manual separation is not recommended.

PUBERTAL VAGINAL BLEEDING

VAGINAL FOREIGN BODY

- *Etiology*: commonly from small pieces of toilet paper. Causes of prepubertal vaginal bleeding are listed in Table 95-2.
- *Clinical presentation*: vaginal foreign bodies are common and present with brown or bloody discharge, dysuria, or abdominal pain.

TABLE 95-2 Differential Diagnosis of Prepubertal Vaginal Bleeding*Visible lesion or mass present*

Traumatic injury including sexual assault, straddle injury
 Urethral prolapse
 Lichen sclerosis
 Genital warts or ulcers
 Neoplasm
 Hemangioma

No visible lesion or mass

Vaginal foreign body
 Infectious vaginitis
 Traumatic injury including sexual assault, straddle injury
 Rectal bleeding
 Premature menarche
 Exogenous hormone withdrawal
 Hematuria
 Coagulopathy
 Neoplasm

- **Management:** removal of the foreign body is necessary. Saline irrigation to flush out small foreign bodies is useful. Large objects may require sedation or general anesthesia for exam and removal. X-rays may aid in determination of size of object if radio-opaque.

URETHRAL PROLAPSE

- **Etiology:** urethral prolapse is a protrusion of urethral mucosa through the urethral meatus, felt to be secondary to increased intra-abdominal pressure. Increased prevalence in prepubescent African American girls.
- **Clinical presentation:** painless bleeding. Examination reveals ring of congested red/purple edematous tissue at the introitus, often obscuring the vagina.
- **Management:** topical estrogen cream daily \times 1–2 weeks; sitz baths, treatment for constipation. Surgical excision may be required for failed cases.

PRECOCIOUS PUBERTY

- **Etiology:** Definition: onset of sexual characteristics at an age that is >2.5 standard deviations below the mean age of puberty for the population, usually defined as age 8. More common in African American girls. Majority of cases are idiopathic.

However, two defined etiologies exist: gonadotropin-dependent precocious puberty (GDPP) secondary to hypothalamic-pituitary-gonadal axis dysfunction; or gonadotropin-independent precocious puberty (GIPP), from steroid production not related to gonadotropin secretion (Fig. 95-2).

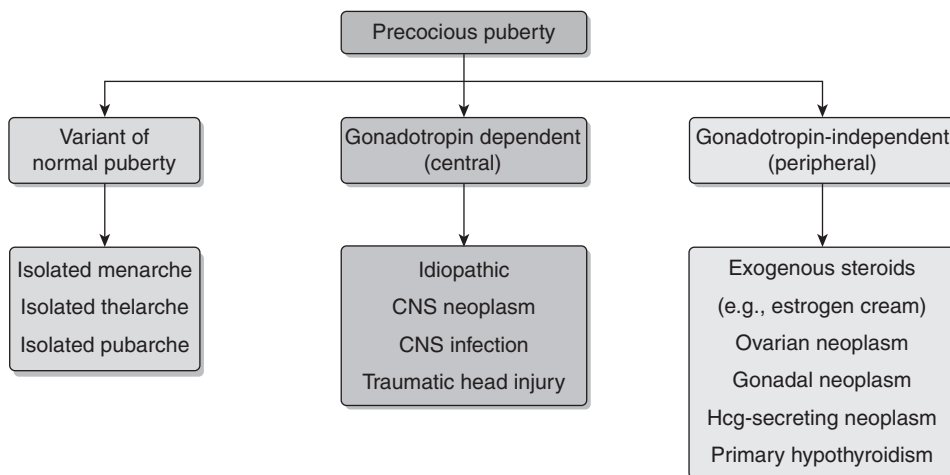
- **Clinical presentation:** GDPP: vaginal bleeding, breast and pubic hair development, headaches, seizures. GIPP: present with thyroid or abdominal uterine masses.
- **Management:** referral to pediatrician or pediatric endocrinologist.

DISORDERS OF ADOLESCENCE**ADOLESCENT HISTORY AND PHYSICAL EXAM**

- Obtain a thorough history including sexual and menstrual.
- Have the parent step outside if needed during the history to maintain patient confidentiality.
- Speculum and bimanual examination may be necessary to assess gynecologic disorders in adolescence.

OVARIAN TORSION

- **Etiology:** the ovary twists on its pedicle, composed of lymphatic and vascular structures, resulting in

**FIG. 95-2.** Algorithm of precocious puberty.

infarction when arterial supply is compromised. Incidence: 15% occurs during childhood.

- *Clinical presentation:* abdominal pain, nausea, vomiting and fever. More common on right secondary to protective effect of sigmoid colon on left.
- *Management:* early diagnosis important for ovarian salvage. Diagnosis made by ultrasound with or without Doppler flow studies. Normal blood flow does not exclude the diagnosis. Surgical laparoscopy or laparotomy should be performed for ovarian salvage.

BARTHOLIN ABSCESS

- *Etiology:* infection of a Bartholin gland cyst. These glands are located at the 4-o'clock and 8-o'clock position on the vaginal vestibule. Bartholin gland cysts are usually asymptomatic.
- *Clinical presentation:* pain and mass will be noted in the labia minora.
- *Management:* incision and drainage often results in recurrence. Marsupialization of the abscess or placement of a Word balloon catheter \times 4–6 weeks is required.

TABLE 95-3 Differential Diagnosis of Pelvic Pain in the Adolescent Female Patient

Ectopic pregnancy
Ovarian torsion
Mittelschmerz
Imperforate hymen
Endometriosis
Pelvic inflammatory disease, tubo-ovarian abscess
Urinary tract infection, urolithiasis
Dysmenorrhea
Appendicitis

ACUTE PELVIC PAIN

- *Etiology:* causes are listed in Table 95-3.
- *Management:* pregnancy test is required in any post-menarchal female with abdominal pain. Other laboratory studies (UA, CBC, basic metabolic profile, LFTs) are obtained as needed for evaluation of each cause. Pelvic examination to identify STDs, PID or masses may be required. Imaging studies such as CT and ultrasound will aid in differentiating the diagnoses of appendicitis, ovarian torsion, tubo-ovarian abscess, or urolithiasis (Fig. 95-3).

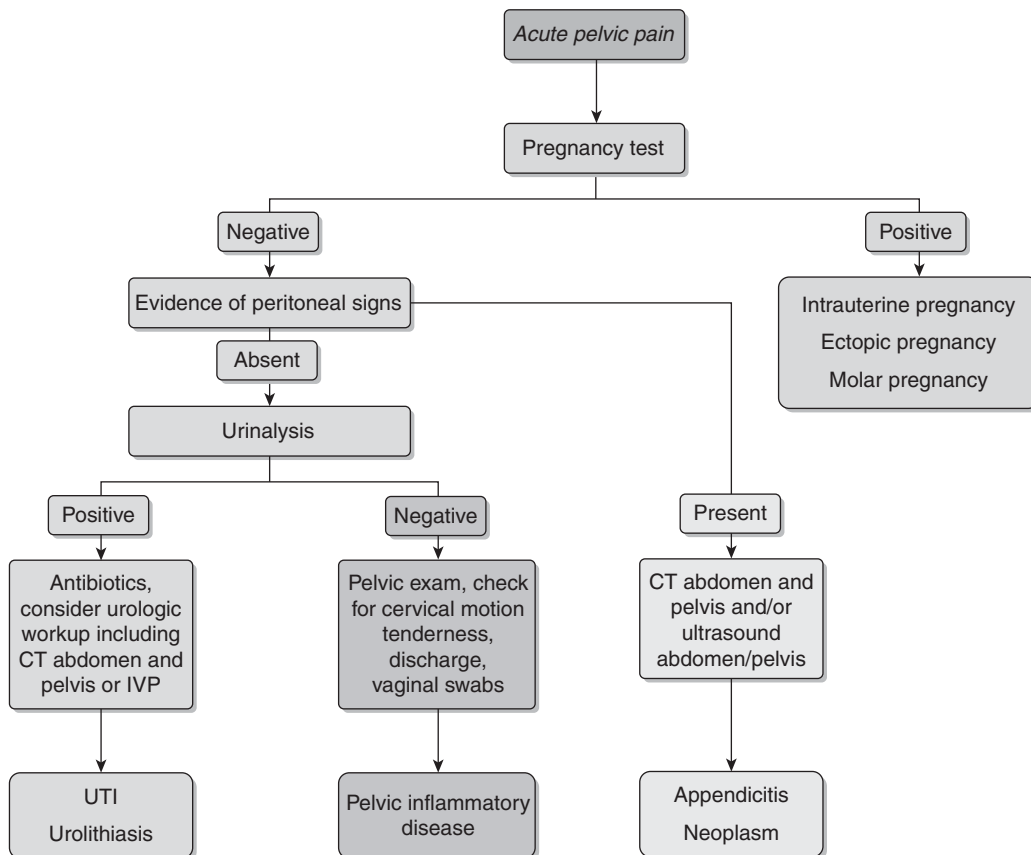


FIG. 95-3. Algorithm of acute pelvic pain.

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QUESTIONS

1. A 10 year-old female is brought by her mother to the ED after noting staining on her underwear for 2 days. There are no complaints of fever, vomiting, dysuria, hematuria or abdominal pain. She has been eating and acting normally. She is nonmenarchal. Both child and mother deny possibility of sexual abuse or insertion of foreign bodies. On genital exam, the external genitalia are normal without evidence of trauma, hymen is intact and normal in appearance. There is a scant thin white vaginal discharge present. No foreign bodies are visualized. Which of the following is the most appropriate next step?
 - A. Ask for social service consult
 - B. Diagnose physiologic leucorrhea
 - C. Perform a speculum exam under sedation
 - D. Refer to Child Abuse Team evaluation
 - E. Begin antibiotics
2. A 6-day-old female is brought to the ED by her parents after they noted blood in her diaper for 1 day. She is the result of a 40 week gestational uncomplicated pregnancy. There are no complaints of fever, vomiting, trauma, lethargy or irritability. She has been breast feeding well. Her exam is unremarkable with the exception of developed breast buds and on genital exam a small amount of blood is noted from the vagina. Which of the following best describes the etiology of this presentation?
 - A. A sign of abuse
 - B. Consistent with hemorrhagic disease of the newborn
 - C. A Normal physiologic variant
 - D. Concerning for Von Willebrands disease
 - E. Requires vitamin K administration
3. A 14 year-old female presents to the ED with complaints of abdominal pain. She has been to 2 outside physicians additionally for this complaint. Mother states that the pain is recurrent, cramping, occasionally causing the patient to vomit. There is no fever, diarrhea, weight loss, dysuria, hematuria, or vaginal discharge. She denies sexual activity and is nonmenarchal. Mother returns today stating that her pain is increasing in severity and now she has a fever to 100.3°F. On exam, HR 110, the patient is a healthy appearing normally developed female in mild distress, but non toxic. Abdominal exam is significant for lower pelvic tenderness and distension. Your evaluation includes: β -HCG=negative, Hgb =15 g%. The genital exam reveals a large mass protruding from the vagina. By inspection, you determine this to be which of the following?
 - A. An imminent delivery
 - B. Foreign Body
 - C. Hematocolpos
 - D. Hydatidiform mole
 - E. Bartholin abscess
4. A 6-month-old female infant is brought to the ED with fever. She is up to date on immunizations. She is eating and acting well otherwise. Nursing informs you that they noted the patient to have labial adhesions when attempting to catheterize the patient for urine culture. Which of the following is the best management option in this patient?
 - A. Manually separate the labia
 - B. Prescribe estrogen cream
 - C. Apply antifungal cream
 - D. Consult pediatric surgery
 - E. Perform chromosomal testing
5. A 14-year-old nonsexually active teenager presents with severe abdominal pain, vomiting and low grade fever. On exam the patient has pain on palpation of her right lower quadrant. Pelvic exam reveals a tender adnexal mass, no cervical motion tenderness or discharge. Which of the following tests will most likely give you the diagnosis?
 - A. CBC
 - B. Urinalysis
 - C. Doppler ultrasound of the ovaries
 - D. DNA probe for sexually transmitted diseases
 - E. CRP

ANSWERS

1. B. Between the ages of 7 and 12, the labia develop, the hymen thickens and vagina elongates. The vaginal mucosa thickens and the vaginal pH becomes acidic. A thin white vaginal discharge known as physiologic leucorrhea may be noted.

2. C. The vaginal discharge is a result of effect of circulating maternal estrogens. Breast buds, sometimes with a milky discharge from the nipple may be seen. Maternal estrogens stimulate mucoid secretions in the neonate and these may appear as a whitish, clear mucous or bloody discharge. The discharge subsides in 2 weeks as maternal estrogen levels fall.
3. C. Hematocolpos results secondary to imperforate hymen. An imperforate hymen may not be diagnosed until puberty when primary amenorrhea is present. On physical exam, a bluish bulge, known as hematocolpos may be seen at the introitus. Treatment is by surgical correction.
4. B. Labial adhesions are believed caused by an estrogen deficiency in the prepubertal period and inflammation that result in thinning of the superficial mucosal layers. Estrogen cream is recommended if the patient is symptomatic with UTI, pain, or inflammation. Surgical separation is recommended only if the adhesions and symptoms persist.
5. C. Fifteen percent of ovarian torsions occur during childhood. The ovary twists on its pedicle made of lymphatic and vascular structures. Pregnancy appears to predispose women to adnexal torsion. Ovarian tumors are discovered in 50% to 60% of women with adnexal torsion, but a normal ovary or adnexum is also possible and occurs more frequently in children. Patients with torsion present with acute, severe, unilateral, lower abdominal and pelvic pain, which may be recurrent. A unilateral, extremely tender adnexal mass is found in more than 90% of patients. 60% have nausea and vomiting. These associated gastrointestinal symptoms which often lead to a diagnosis of acute appendicitis or small intestinal obstruction. Doppler ultrasound has been used to evaluate ovarian arterial blood flow. Abnormal color Doppler flow is highly predictive of ovarian torsion. Normal blood flow through the ovary does not exclude the diagnosis. Patients often require laparoscopic surgery for diagnosis and ovarian salvage.

96 VAGINITIS

Maria Stephan

INTRODUCTION

- Vulvovaginitis is the most common gynecologic disorder of childhood. Inflammation of vulvar and vaginal tissues is caused by physical, chemical, or

TABLE 96-1 Causes of Nonspecific Vulvovaginitis in Children

Poor hygiene, including inadequate front-to-back wiping
Small labia minora, with a short distance from anus to vagina
Vulvovaginal epithelium that is thin and not well estrogenized, making the area more prone to irritation
Foreign body including toilet paper, small toys, pieces of cloth
Chemical irritants including soap, shampoo, bath oils, deodorant soaps, bubble baths
Eczema and seborrhea
Chronic disease or immunodeficiency
Sexual abuse

infectious agents. This chapter will review nonsexually transmitted causes of vaginitis (see Chapter 97 for a review of STDs).

- History should include the following: hygienic practices, medical conditions, potential for sexual abuse, and presence of pruritis, odor, character and amount of discharge, menstrual and sexual history.

NONSPECIFIC VULVOVAGINITIS

- *Etiology:* accounts for 60% of cases of vulvovaginitis. Often find alteration of vaginal flora with fecal bacteria overgrowth. See Table 96-1 for a review of causes.
- *Clinical presentation:* erythematous dermatitis of vulva and perianal tissues in addition to vaginal discharge.
- *Management:* proper hygiene measures (Table 96-2). Culture of discharge should be obtained.
 - Cases of bacterial superinfection treated with antibiotics such as amoxicillin, amoxicillin/clavulinate acid, or a cephalosporin.

VAGINITIS DUE TO UPPER RESPIRATORY INFECTION

- *Etiology:* commonly group A β -hemolytic *Streptococcus* (GAS) or *Hemophilus influenzae* via self inoculation.

TABLE 96-2 Treatment of Nonspecific Vulvovaginitis

Improve local hygiene, including using front-to-back wiping after a bowel movement, and keeping the vulvar area clean and dry
Sitz bath (lukewarm bathwater with 2 tbsp baking soda)
Discontinue bubble baths, deodorant soaps, lotions and use mild bath soap instead
Use unscented toilet paper
Wash hands before and after using toilet
Remove wet bathing suit soon after leaving a swimming area

- **Clinical presentation:** erythematous dermatitis of vulva and perianal tissues in addition to vaginal discharge.
- **Management:** obtain culture of discharge. Initiate antibiotic treatment with penicillin or amoxicillin after cultures are obtained.
 - For recurrent cases, use clindamycin or penicillin with rifampin.

CANDIDAL VULVOVAGINITIS

- **Etiology:** caused by *Candida albicans*. Occurs more commonly after puberty when estrogen promotes fungal growth.
 - Uncommon in prepubertal girls except in the presence of diabetes mellitus, immunodeficiency, or antibiotic use.
- **Clinical presentation:** inflammation of vulva/perianal regions with a thick white discharge. Satellite lesions may be present.
- **Management:** diagnosis by wet mount with potassium hydroxide or specific fungal cultures. Treatment with topical antifungal preparations. Oral fluconazole may be used in complicated cases or in immunosuppressed children.

SHIGELLA VAGINITIS

- **Etiology:** infection by *Shigella flexneri*.
- **Clinical presentation:** a mucopurulent bloody discharge sometimes seen after an episode of diarrhea.
- **Management:** diagnosis made by culture of discharge. Empiric treatment with trimethoprim-sulfamethoxazole.

PARASITIC VAGINITIS

- **Etiology:** caused by *Enterobius vermicularis* (pinworms). 1 cm long thin white worms travel from the anus to the vulvar area.
- **Clinical presentation:** persistent genital itching, erythema, or pinworms visualized by caregiver.
- **Management:** diagnosis by identification of anal pinworms by sight or by microscopic visualization of eggs obtained after application of transparent adhesive tape to anal region in morning. Treatment with mebendazole 100 mg orally once.

GARDNERELLA VAGINITIS

- **Etiology:** also known as bacterial vaginosis. Secondary to overgrowth of mixed vaginal flora when vaginal pH is above 4.5. Consider potential for sexual abuse if present in prepubertal child.

- **Clinical presentation:** grayish white malodorous discharge is seen. A fishy, amine-like odor is released when potassium hydroxide solution is added to a sample of discharge.
- **Management:** diagnosis is by wet mount looking for clue cells. Treatment is with oral metronidazole for 7–10 days.

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QUESTIONS

1. A 6-year-old girl is brought to the ED by her mother with concerns of vaginal redness, and some bloody discharge. Both child and mother deny trauma or sexual abuse. On exam, the vulva and perianal tissues are red and blood tinged with scant vaginal discharge seen. The hymen is normal appearing with no evidence of trauma. The MOST LIKELY source of symptoms would be which of the following?
 - A. *N. gonorrhoea*
 - B. Group A β -hemolytic *Streptococcus*
 - C. Physical or chemical irritant
 - D. Foreign body
 - E. *Shigella*
2. A 12-year-old female is brought by her mother to the ED after noting discharge on her underpants for over 1 week. The child and mother deny dysuria, fever, abdominal pain, or sexual abuse. Menarche occurred at age 9. On exam, a whitish vaginal discharge is noted. Clue cells are seen on saline wet mount. Which of the following is the best treatment option in this patient?
 - A. Reassure the patient and mother that the discharge is physiologic leucorrhea.
 - B. Prescribe fluconazole
 - C. Investigate for sexual abuse
 - D. Exam the patient for a vaginal foreign body
 - E. Test for *E. vermicularis*

ANSWERS

1. C. Nearly two thirds of non-specific vaginitis in prepubertal girls is due to physical or chemical irritants (see Table 96-1). All of the above causes should be considered, but the most likely cause is an irritant.
2. C. Bacterial vaginosis, also known as Gardnerella vaginitis, occurs primarily in sexually active women; therefore, investigating for potential sexual abuse is warranted in this patient.

97 SEXUALLY TRANSMITTED DISEASES

Maria Stephan

INTRODUCTION

Many sexually transmitted diseases (STD) occur concurrently; when one is suspected, screen and treat appropriately for others. HIV screening is recommended for all patients seeking care for an STD. Adolescent HIV testing and counseling can be done without parental consent in most states. Always remember to assure that sexual partner(s) of a patient with an STD gets appropriately treated.

CHLAMYDIA

- **Etiology:** infection with *Chlamydia trachomatis*
 - Most common treatable STD in the US.
 - Up to 45% are co-infected with gonorrhea.
 - Second most common STD after human papilloma virus.
- **Clinical presentation:** most men and women are asymptomatic.
 - **In women:** Symptoms: vaginal discharge, mild abdominal pain, dysuria, urinary frequency, post-coital/intermenstrual bleeding. Physical exam: pyuria without bacteria, cervical edema, mucopurulent discharge.
 - **In men:** Symptoms: dysuria, urethral itching, clear to white urethral discharge. Physical exam: meatal edema, whitish-clear discharge, pyuria.
- **Diagnosis:** DNA probes from swabs collected from cervical or male urethral sites. Chlamydia is detected by chemiluminescence of chlamydial rRNA from hybridization of the DNA probe. It is highly specific.
 - **Culture** of Chlamydia from cervical/vaginal or male urethral swabs remain the standard for

diagnosis in children for cases of medico-legal implications (i.e. sexual assault).

- Other available testing includes: PCR (polymerase chain reaction-nucleic acid amplification, LCR (ligase chain reaction), TMA (transcription-mediated amplification).
- All these may be performed on cervical, urethral or urine specimens.
- **Treatment:** see Table 97-1 for treatment guidelines.

GENITAL WARTS

- **Etiology:** infection caused by DNA-containing human papilloma virus (HPV) types 6, 11, 16, 18.
 - More common than genital herpes.
 - 10–20% of sexually active women are infected annually.
 - Most common in 15–30 year olds.
- **Clinical presentation:** usually asymptomatic.
 - **In women:** lesions seen on external genitalia, cervix, and vagina.
 - **In men:** lesions noted in coronal sulcus, penile shaft, and urethra.
 - Intra-anal warts seen in both sexes associated with receptive anal sex.
 - Lesions may also occur on conjunctiva, nose, mouth, or larynx.
 - Condyloma lata associated with syphilis may be confused with HPV. Distinction can be made by aspirate of lesion demonstrating spirochetes on dark-field microscopy or serologic testing for syphilis.
 - Other similar appearing lesions: molluscum contagiosum, Bowenoids papules, and pearly penile papules.
- **Diagnosis:** by visual inspection. Include diagnostic testing for other STDs, including serologic testing for syphilis (VDRL, RPR).
- **Treatment:** see Table 97-1.
 - Podofilox is a keratolytic and antimitotic applied only to external warts, not mucous membrane warts.
 - Imiquimod is an immunomodulator, which induces cytokine and interferon. Its use is indicated for patients over 12 years and only external warts.
- Investigate for sexual abuse when genital warts are seen in children.

GONORRHEA (GC)

- **Etiology:** infection by gram negative diplococci, *Neisseria gonorrhoeae*

TABLE 97-1 Summary table of sexually transmitted diseases

DISEASE	ETIOLOGY	CLINICAL MANIFESTATIONS	DIAGNOSIS	THERAPY	TREATMENT ADMINISTRATION
Chlamydia	<i>Chlamydia trachomatis</i>	Vaginal discharge, dysuria, abdominal or testicular pain, or asymptomatic	NAAT, culture	po	Azithromycin 1 g
				po	or Doxycycline 100 mg bid × 7 d
				po	or EES base 500 mg qid × 7 d
				po	or EES ethylsuccinate 800 mg qid × 7 d
				po	or Ofloxacin 300 mg bid × 7 d
				po	or Levofloxacin 500 mg qid × 7 d
				po	Children <45 kg: EES (base or ethylsuccinate) 50/kg/dose × 14 d Children >45 kg + <8 y: Azithromycin 1 g × 1 Children >45 kg + >8 y: Adult dosing
Gonorrhea	<i>Neisseria gonorrhoeae</i>	Asymptomatic, dysuria, penile or vaginal discharge, abdominal pain, rash	NAAT, culture, DNA probe	IM	Ceftriaxone 125 mg × 1
				po	or Cefixime 400 mg × 1
				po	or Ciprofloxacin 500 mg × 1
				po	or Ofloxacin 400 mg × 1
				po	or Levofloxacin 250 mg × 1
				IM	Children >45 kg: adult dosing used on above medications Children <45 kg: Ceftriaxone 125 mg × 1 Add treatment for chlamydia if not ruled out
Trichomoniasis	<i>Trichomonas vaginalis</i> (protozoan)	Asymptomatic (men & women) or frothy, odorous vaginal discharge, dyspareunia, urgency	Saline wet prep, culture, rapid antigen testing	po	Metronidazole 2 g × 1
				po	or Tinidazole 2 g × 1
Genital warts	Human papilloma virus (HPV), types 6,11, 16,18	Asymptomatic or pain, itching, bleeding, or obstructive effects from wart size	Visual exam, biopsy,	Topical	Podofilox 0.5% gel/solution bid × 3 d or Imiquimod 5% cream qhs 3 times/wk × 16 wk
				Specialist administered	Cryotherapy, intralesional interferon, podophyllin, laser & surgical removal
Genital Ulcers Syphilis	<i>Treponema pallidum</i>	Chancre, genital ulceration Secondary: lymphadenopathy rash, condyloma lata Latent Syphilis: no evidence of disease Tertiary: neuropathy, dementia, tabes dorsalis, aortitis, gumma of skin	Dark-field DAT serologic testing (treponemal & nontreponemal)	IM	Benzathine penicillin G 2.4 million units × 1
				IM	Children: Benzathine penicillin G 50,000 U/kg up to adult dose × 1
				po	Alternatives: doxycycline 100 mg bid × 14 d
				po	Tetracycline 500 mg qid × 14 d. Not recommended for children <8 y.
				IM	Ceftriaxone 1 g qid × 8–10 d Secondary syphilis treatment: same as primary
				IM	Late Latent disease: Benzathine penicillin G 2.4 million units q wk × 3 wk Children: Benzathine penicillin G 50,000 U/kg up to adult dose q wk × 3 wk

(continued)

TABLE 97-1 Summary table of sexually transmitted diseases (Continued)

DISEASE	ETIOLOGY	CLINICAL MANIFESTATIONS	DIAGNOSIS	THERAPY	TREATMENT ADMINISTRATION
Herpes Simplex	Herpes simplex virus type 2	Painful genital ulcers, dysuria, dyspareunia	Viral culture-specific antibody assays	po	<i>First clinical episode</i> Acyclovir 400 mg tid 7–10 d or
				po	Acyclovir 200 mg 5 × d × 7–10 d or
				po	Famciclovir 250 mg tid × 7–10 d or
				po	Valacyclovir 1 g bid × 7–10 d
					Episodic treatment for recurrent infections
				po	Acyclovir 400 mg tid × 5 d or
				po	Acyclovir 800 mg bid × 5 d or
				po	Acyclovir 800 mg tid × 2 d or
				po	Famciclovir 125 mg bid × 5 d or
				po	Famciclovir 1000 mg bid × 1 d or
po	Valacyclovir 500 mg tid × 3 d or				
po	Valacyclovir 1 g qid × 5 d				

Adapted from CDC guidelines, MMWR Recomm Rep. 2006;55(RR-11):1–95.

- *Clinical presentation:*
 - **In women:** 80% are asymptomatic. Vaginal discharge, dysuria, abdominal pain, red friable cervix with mucopurulent discharge seen.
 - **In men:** urethral discharge, urgency, penile discharge, proctitis with rectal discharge. Rectal and oropharyngeal infection may be asymptomatic.
 - Symptoms usually present 2 weeks after exposure.
 - Disseminated GC may appear as pustular lesions on hands/feet, polyarthritis, tenosynovitis or endocarditis.
- Investigate for sexual abuse in pediatric patients presenting with gonococcal infections.
- *Diagnosis:* see Table 97-1.
 - **In children always culture**, using Thayer-Martin media, as legal implications are present.
- *Treatment:* see Table 97-1.
 - Single therapy with ceftriaxone or cefixime is recommended.
 - Pediatric dosing of cefixime is 8mg/kg orally (maximum 400 mg orally as a single dose).
 - Antibiotic resistance to quinolones is increasing in the US.
 - **Disseminated infection dosing:** Pediatric: Ceftriaxone 25–50 mg/kg/dose every 12 hours × 7 days. **Adult:** Ceftriaxone 1 gram every 24 hours

till improvement, then switch to cefixime 400 mg every 12 hours × 7 days.

TRICHOMONIASIS

- *Etiology:* infection caused by flagellated protozoan, *Trichomonas vaginalis*.
- *Clinical presentation:*
 - **In women:** malodorous discharge and strawberry cervix may be noted on exam.
 - **In men:** often asymptomatic or urethritis.
- *Diagnosis:* flagellated motile trichomonads seen on saline prep and pH of discharge may be elevated.
- *Treatment:* single dose metronidazole is curative in 95%.

SYPHILIS

- *Etiology:* infection caused by spirochete, *Treponema pallidum*.
- *Clinical presentation:*
 - **Primary syphilis:** chancre develops within 3 weeks after exposure with spirochetes present on ulcer surface.
 - **Secondary syphilis:** manifestations variable. May see rash, lymphadenopathy, condyloma lata months after resolution of primary lesion.

- **Latent syphilis** is detected by serologic testing without clinical manifestations.
- **Diagnosis:** in first or second stage, detected by dark-field and direct fluorescent antibody (DAT).
 - Serologic testing via: non treponemal VDRL, RPR and treponemal FTA-ABS. Treponemal test stays positive for life, VDRL and RPR become negative 1 year after treatment.
- **Treatment:** for various stages, see Table 97-1.

HERPES SIMPLEX

- **Etiology:** 90% caused by Herpes simplex virus (HSV) type 2.
- **Clinical presentation:** painful genital ulcers and vesicles. See Table 97-1.
 - 75% of infections are asymptomatic
- **Diagnosis:** specific antibody assays (HSV glycoprotein G-1 and G-2) available.
 - Diagnosis should be confirmed by **culture in children**.
- **Treatment:** see Table 97-1.

HUMAN IMMUNODEFICIENCY VIRUS (HIV) AND ACQUIRED IMMUNODEFICIENCY DISEASE SYNDROME (AIDS)

- **Etiology:** caused by human immunodeficiency virus (primarily HIV-1 in the US)
 - Modes of transmission: homosexuality, heterosexuality, bisexuality, needle sticks, intravenous drug use, maternal neonatal transmission, blood transfusions before 1985.
- **Clinical presentation:** most asymptomatic for 10 years. Wide spectrum of symptoms: fever, rash, lymphadenopathy, failure to thrive, opportunistic infections, and malignancies.
- **Diagnosis:** rapid HIV-1 enzyme immunoassay (EIA),
 - Western blot or immunofluorescence assay (IFA) available.
 - Perform testing for other STDs at same time.
- **Treatment:** retroviral medications include: zidovudine, didanosine, stavudine, indinavir, and ritonavir. Refer to specialist for treatment and counseling.

URETHRITIS, EPIDIDYMITIS, AND CERVICITIS

URETHRITIS

- Inflammation of the urethra, most commonly caused by *N gonorrhoeae* and *C trachomatis*.

- Noninfectious causes: irritation, topically applied agents, and systemic diseases.
- Hematuria is uncommon.
- STD testing and treatment guidelines should be followed.

EPIDIDYMITIS

- **Etiology:** inflammation of the epididymis caused by STDs in post-pubertal males and *Escherichia coli* or *Hemophilus influenzae* in prepubertal boys.
 - Noninfectious causes include trauma and vigorous physical activity.
- **Clinical presentation:** scrotal pain, swelling, edema, reactive hydrocoele, and fever. Must investigate for testicular torsion if indicated.
- **Diagnosis:** evaluate as urethritis in men with STD testing, positive leucoesterase on first void urine.
- **Treatment:** follow STD guidelines (Table 97-1)
 - Rest, analgesia, scrotal elevation.

CERVICITIS

Inflammation of the cervix with a mucopurulent discharge most commonly caused by STDs. Evaluate and treat as per STD guidelines. See Table 97-1.

PELVIC INFLAMMATORY DISEASE (PID)

- **Etiology:** inflammation of female genital tract involving endometrium, uterus, fallopian tubes and ovaries. Often leading to pelvic abscesses, infertility and subsequent ectopic pregnancies.
 - PID is commonly caused by *C trachomatis* and *N gonorrhoeae*; plus other anaerobic bacteria.
- **Clinical presentation:** Classic findings are: fever, pelvic pain, mucopurulent endocervical discharge, adnexal and cervical motion tenderness.
- **Diagnosis:** Diagnostic criteria: fever $\geq 38.3^{\circ}\text{C}$ (101°F), abnormal cervical or vaginal discharge, elevated ESR and CRP, confirmed GC or Chlamydia infection.
 - Ultrasound may reveal tubo-ovarian abscess.
 - Ectopic pregnancy must be excluded.
- **Treatment:**
 - **Outpatient:** single dose of ceftriaxone (250 mg IM) or ceftioxin (2 grams IM + probenecid 1 g PO) plus oral doxycycline (100 mg BID) plus oral metronidazole to complete a 14 day course.
 - **Inpatient: Admission criteria:** high fever, ill-appearing, failed outpatient management, pregnancy, or tubo-ovarian abscesses. **Parenteral**

management: cefotetan or cefoxitin plus oral or IV doxycycline. Alternative parenteral treatment: clindamycin plus gentamycin. Continue parenteral therapy until clinical improvement for 24 hours, and then switch to oral outpatient regimen.

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QUESTIONS

1. A 15-year-old female presents with a complaint of vaginal discharge. On pelvic exam you note a frothy white foul-smelling vaginal discharge with a “strawberry cervix.” Which of the following is true regarding this patient?
 - A. The wet prep will demonstrate “clue cells”
 - B. DNA probe will be positive
 - C. The wet prep will demonstrate a flagellated organism
 - D. RPR will be positive
 - E. The pH of the vaginal discharge will be 3
2. A 17-year-old sexually active female presents with low grade fever, vaginal discharge, and abdominal pain. You suspect pelvic inflammatory disease. Which of the following would be a reason for inpatient care with intravenous antibiotics?
 - A. Abnormal copious purulent vaginal discharge
 - B. A fever of 38.4°C
 - C. Absence of tubo-ovarian abscess on ultrasound exam
 - D. Positive pregnancy test
 - E. Elevated ESR and CRP
3. A 4-year-old female is brought to the ED by her mother because she noted bumps in her private areas for the past 3 months. On exam, you note genital warts surrounding her vaginal and perianal areas. Which of the following is true regarding genital warts in children?
 - A. 100% of condyloma accuminata are the result of sexual abuse in children
 - B. Warts in the genital area can be acquired without sexual abuse
 - C. The most common HPV serotypes, 6 and 11 predispose to cervical cancer
 - D. Diagnosis is by VDRL
 - E. Treatment includes ceftriaxone
4. A 15-year-old male presents with a rash noted on his palms and soles which has been present for 3 months. He has no complaints of itching, penile discharge, fever, arthritis, tick bites, or other recent known exposures. He denies history of STDs. His exam is otherwise unremarkable except for rose-colored macular lesions on palms and soles and also on his back. Which of the following would be the most appropriate management of this patient?
 - A. Obtain treponemal FTA-ABS
 - B. Treat symptomatically with emollient ointment
 - C. Apply hydrocortisone cream
 - D. Obtain blood culture
 - E. Empirically treat with single dose Ceftriaxone
5. The vaginal discharge cultures for a 3-year-old 15 kg female seen by you several days ago are now positive for *N gonorrhoeae*. In addition to reporting the results to Child Protective Services, the most appropriate treatment includes which of the following?
 - A. Ceftriaxone 75 mg/kg × 1 IM + Azithromycin 1 g PO
 - B. Ciprofloxacin 500 mg PO + Azithromycin 1 g PO
 - C. Benzathine penicillin G 50,000 units/kg IM + Erythromycin 50 mg/kg/day divided every 6 hours × 14 days PO
 - D. Ceftriaxone 125mg IM + Erythromycin 50 mg/kg × 14 days PO
 - E. Benzathine penicillin G 50,000 units/kg IM + Metronidazole 1 g PO

ANSWERS

1. C. Trichomoniasis is caused by the flagellated protozoan, *Trichomonas vaginalis*. 50% of women are asymptomatic, but classically presents with white foul-smelling vaginal discharge, with a “strawberry cervix.” Diagnosis is made by viewing flagellated organisms on microscopy of the saline preparations of the vaginal discharge or urine with a sensitivity of 70%. Additionally, the pH of the vaginal discharge is elevated. A single dose metronidazole 2 g orally, is curative in 95% of cases.
2. D. A positive pregnancy test is a clear indication for inpatient care for PID. Other indications are an ill appearing patient, and presence of tubo-ovarian abscess. Low grade fever and elevated ESR and CRP are commonly seen with PID. Copious vaginal discharge is not a reason for hospitalization.
3. B. It has been estimated that at least 50% of the cases of condylomata acuminata in children are the result of sexual abuse. Warts in the genital area can be acquired without sexual abuse. A child with warts on the hands can transfer the warts to the mouth, genitals, and anal area. A mother with hand warts can transfer warts to the child. The incubation period for warts are often many months; making it difficult to associate past events. More than 80 genotypes have been identified; HPV-6, HPV-11, and HPV-16 are most commonly associated with genital warts and HPV-6 and HPV-11 are rarely associated with cervical cancer. Treatment options include patient administered podofilox, imiquimod and provider administered cryotherapy, podophyllin, interferon, laser therapy.
4. A. This patient has a rash consistent with secondary syphilis. Secondary syphilis may occur months after the primary lesion resolves. Clinical symptoms include: rash, usually rose-colored, macules presenting on palms of hands and soles of feet, fever, malaise, generalized lymphadenopathy, condyloma lata, or pharyngitis. These also resolve after 3–5 months. Diagnosis is made with serologic testing via nontreponemal VDRL and RPR or treponemal FTA-ABS. Treatment consists of Benzathine penicillin 2.4 million units IM and physician follow-up.
5. D. In children < 45 kg, ceftriaxone 125 mg intramuscularly once is effective for gonorrhea. Treatment for Chlamydia should additionally be administered. The Center for Disease Control (CDC) recommendations for pediatric Chlamydia treatment is: erythromycin 50 mg/kg/day divided every 6 hours × 14 days for children <45 kg. Azithromycin for Chlamydia is recommended for children <8 years of age, but greater than 45 kg. Fluoroquinolones have not been recommended for persons < 18 year of age.

98 DYSMENORRHEA AND DYSFUNCTIONAL UTERINE BLEEDING

Pamela J. Okada
Mercedes Uribe

DYSMENORRHEA

- Dysmenorrhea is defined as cyclic cramps and pain associated with menstruation
 - Primary: menstrual pain without pelvic pathology
 - Secondary: menstrual pain with pelvic pathology (most often seen in older women)
- Incidence: highest in older adolescent girls
 - The most common cause of school and work absenteeism
- Pathophysiology of primary dysmenorrhea
 - Not completely clear
 - Associated with prostaglandin F₂ release in endometrium
 - Sloughing of endometrial cells release prostaglandins
 - Prostaglandins cause myometrial contractions and vasoconstriction
 - Results in pain and cramping
 - Role of vasopressin is unclear
 - Found to be increased in menstrual fluids
 - Similar role as prostaglandins
- Pathophysiology of secondary dysmenorrhea
 - Involves prostaglandin activity
 - Influenced by underlying pelvic pathology
- Etiology/risk factors
 - Primary dysmenorrhea
 - Affects all races
 - Risk factors are numerous (Table 98-1)

TABLE 98-1 Risk Factors for Dysmenorrhea

Smoking or passive smoking
Early menarche, age <11 y
Nulliparity
Prior abortions
Nonoral-contraceptive use
Poor mental health
Severe distress
Single or younger age of marriage
Sexual violence
Alcohol use
Anxiety
Depression
Obesity
Infertility
Heavy menstrual flow
History of sterilization
Intrauterine device

- Secondary dysmenorrhea
 - Risk factors
 - History of sterilization
 - Sexual assault
 - Intrauterine device
 - Presence of pelvic pathology
 - Clinical presentation
 - Onset of primary dysmenorrhea: first 6 months after menarche or when ovulatory cycles are established
 - Symptoms: Pain in pelvis, lower back, anterior upper legs occurs at onset of menses, lasts 8–72 hours
 - Associated symptoms:
 - Primary: Headache, diarrhea, nausea, fatigue, dizziness, vomiting
 - Secondary: pain after established painless menstrual cycles and infertility, dyspareunia, itchiness vaginal discharge, irregular or heavy bleeding, or dysuria during times other than menses
 - Diagnostic evaluation
 - No laboratory tests or imaging studies are necessary for the evaluation of primary dysmenorrhea.
 - The evaluation of secondary dysmenorrhea may include
 - Complete blood count
 - Urinalysis
 - Pregnancy test
 - Gonococcal and chlamydial cervical cultures
 - Erythrocyte sedimentation rate
 - Abdominal and transvaginal ultrasonography
 - Other diagnostic modalities may be indicated depending on underlying pathology. Table 98-2
 - Management
 - Pharmacologic treatment
 - Nonsteroidal anti-inflammatory drugs (NSAIDs) are the mainstay of treatment for dysmenorrhea
 - Reduces prostaglandin production by inhibiting cyclo-oxygenase 1 and 2 enzymes and decreases menstrual flow
 - Side effects are minor and include gastritis, indigestion, and drowsiness
 - Newer NSAID class: cyclooxygenase-2 (COX-2) inhibitors
 - Inhibits cyclooxygenase-2 enzyme, which is responsible for the metabolism of arachidonic acid to prostaglandin
 - Side effects include insomnia, headache, flatulence, stomach ulcers, bleeding, and perforation of the stomach and intestines. They may also increase the risk of heart attacks and stroke.
 - Oral contraceptive pills (OCPs)
 - Inhibits ovulation and reduces endometrial lining of the uterus
 - Decreases menstrual fluid volume and prostaglandin production
 - Side effects include nausea, headache, breast tenderness, weight gain, breakthrough bleeding. They may increase the risk of hypertension, blood clots, and stroke.
 - Experimental treatments that require prospective clinical trials to evaluate efficacy and safety before routine use include
 - Oral nifedipine
 - Intravenous terbutaline
 - Nonpharmacologic treatment
 - Herbal and dietary supplements reduce pain
 - Magnesium, vitamin B₆, vitamin B₁
 - Omega-3 fatty acids, fish oil
 - Japanese herbals
 - Behavior therapy may improve symptoms
 - Pain management training
 - Relaxation
 - Imagery
 - Biofeedback
 - Chiropractic-spinal manipulation and acupuncture
 - Help anecdotally
 - Little research published to establish efficacy
- Treatment of secondary dysmenorrhea
 - Dependent upon underlying diagnosis and cause of pain
 - Nonacute pain can be managed as in primary dysmenorrhea
 - Refractory pain may require surgical interventions including
 - Laparoscopy
 - Presacral neurectomy
 - Laparoscopic uterosacral nerve ablation
 - Hysterectomy

DYSFUNCTIONAL UTERINE BLEEDING

- Definition: DUB is defined as any disturbance in regularity, frequency, duration, or volume of menstrual flow when organic or structural causes have been ruled out
 - Patterns of menstrual bleeding: Table 98-3
 - Menstruation and the normal menstrual cycle (Fig. 98-1)
 - Occurs every 21 to 35 days

TABLE 98-2 Diagnostic Modalities for Secondary Dysmenorrhea

Laparoscopy: Endometriosis, pelvic adhesion, pelvic inflammatory disease

Hysteroscopy and saline sonohysterography: Endometrial polyps and submucosal leiomyomas, obstructing müllerian malformation

Sonography: Pelvic mass, ovarian cyst

MRI: Adenomyosis

TABLE 98-3 Patterns of Menstrual Bleeding

Menorrhagia	Excessive bleeding (>80 mL) at regular intervals
Metrorrhagia	Prolonged bleeding (>7 d) and irregular intervals
Menometrorrhagia	Excessive, prolonged, and irregular bleeding (>80 mL for >7 d)
Oligomenorrhagia	Interval of 35 d to months
Polymenorrhea	Interval less than 21 d
Hypermenorrhea	Regular menses for more than 7 d duration
Amenorrhea	No uterine bleeding for 6 mo or longer
Intermenstrual bleeding	Spotting, variable amounts between regular menstrual periods

- Lasts from 2 to 7 days
- Produces an average of 35 to 80 mL of blood loss a cycle
- Incidence: Accounts for 20% of all gynecologic visits
 - 20% are adolescents
 - 50% are aged 40 to 50 years

- Pathophysiology

- Anovulatory cycle
 - Accounts for 90% of DUB
 - Adolescents: Results from immaturity of the hypothalamic—ovarian axis
 - ◻ LH surge is unable to respond to rising estradiol levels
 - ◻ Ovulation does not occur
 - ◻ Corpus luteum does not form
 - ◻ Progesterone levels remain low or absent
 - ◻ Endometrium proliferates under the effects of unopposed estrogen levels
 - Endometrium outgrows its blood supply resulting in excessive prolonged and irregular bleeding (menometrorrhagia)
 - Perimenopausal women: results from short follicular phase
 - ◻ Ovarian follicles secrete less estradiol leading to insufficient endometrial proliferation

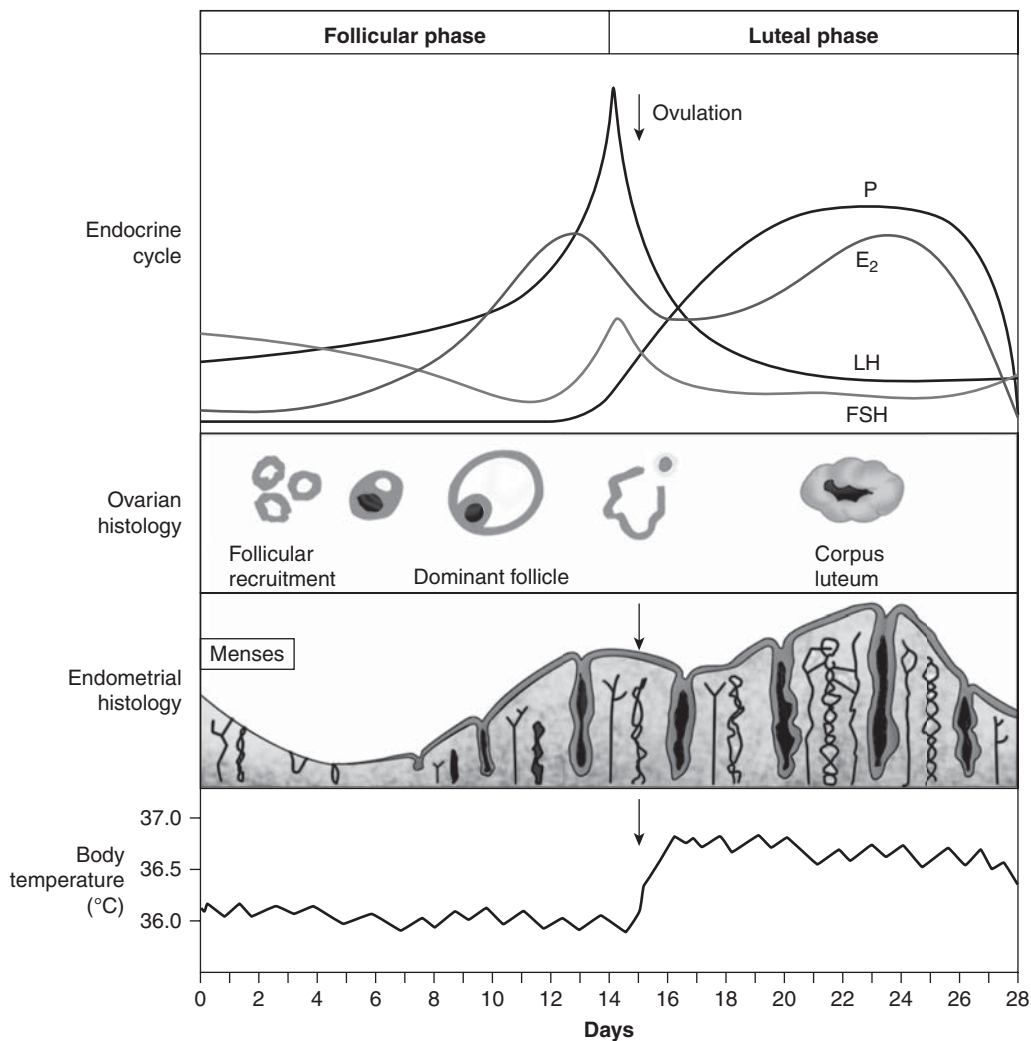
**FIG. 98-1.** Normal menstrual cycle

TABLE 98-4 Pathologic Causes of Dysfunctional Uterine Bleeding**Systemic**

- Coagulopathies:
 - Thrombocytopenia, von Willebrand disease, leukemia
- Liver disease
- Sepsis
- Endocrinopathies:
 - Diabetes mellitus, polycystic ovarian syndrome, Addison's disease, hypothyroidism, congenital adrenal hyperplasia, Cushing's disease, Turner's syndrome, hyperprolactinemia
- Others:
 - Hypertension, cystic fibrosis, inflammatory bowel disease

Miscellaneous

- Stress
- Excessive exercise
- Excessive weight gain
- Poor nutritional status

Iatrogenic

- Intrauterine contraceptive device (IUCD)
 - Medications: Oral contraceptive, steroids, tamoxifen anticoagulants, antihistamines, metoclopramide, methyl dopa, and phenothiazine
- Trauma to cervix, vulva, or vagina
- Foreign body
- Sexual abuse

Reproductive Tract

- Myomas:
 - Adenomyosis, endometriosis, polyps, fibroids, ovarian cyst
- Carcinoma of the vagina, cervix, uterus, and ovaries
- Condyloma, cervicitis, vaginitis, endometritis, oophoritis
- Pregnancy related: Threatened, incomplete, missed abortion, molar or ectopic pregnancy, placenta previa, and abruption

- Leads to irregular and light menstrual bleeding
 - Ovulatory cycle
 - Accounts for 10% of DUB in women with normal ovulatory cycles
 - Menses are regular but are characterized by heavy menstrual loss
 - Pituitary-ovarian axis and hormone profile are normal
- Etiology/risk factors
 - Anovulatory cycle is the most common cause of abnormal bleeding
 - Risks include stress, eating disorders, hypothyroidism, hyperthyroidism, diabetes mellitus, and excessive exercise
 - Polycystic ovary syndrome (PCOS) is associated with heavy bleeding
 - PCOS is associated with type 2 diabetes mellitus, hirsutism, acanthosis nigricans, overweight, insulin resistance
 - May be a family history of PCOS
 - Pathologic causes of DUB include systemic diseases, iatrogenic causes, and disorders of the reproductive tract Table 98-4
 - 20% of the time DUB may be associated with systemic disease
 - Coagulopathies such as thrombocytopenia, von Willebrand disease, or leukemia account for half of the systemic diseases causing DUB
- Clinical presentation
 - History—obtain a detailed menstrual and sexual history
 - Age of menarche and timing of last menstrual period
 - Frequency, amount, duration of menses
 - Associated pain
 - Impact on quality of life
 - Number of sexual partners
 - History of sexually transmitted diseases
 - Reproductive and family history
- Review of systems, signs, and symptoms
 - Pallor
 - Bleeding—epistaxis, easy bruising, excessive bleeding during surgery or dental procedures
 - Evaluate for signs of hypovolemia, tachycardia, diaphoresis
 - Examine for signs of PCOS such as acne, hirsutism, acanthosis nigricans
 - Ask about weight loss or gain, cold or heat intolerance, hair loss seen in thyroid disease

TABLE 98-5 Treatment of Dysfunctional Uterine Bleeding**Oral contraceptive pills (OCPs)***Mild bleeding*

- 30 µg ethinyl estradiol plus 0.3 mg norgestrel one pill per day
- 30 µg ethinyl estradiol plus 0.15 mg levonorgestrel one pill per day
- 20–35 µg ethinyl estradiol plus a progestin one pill daily

Moderate bleeding

- 35 µg ethinyl estradiol plus progestin twice to every 6 h for 5–7 d followed by one pill to completion of 28 d pack; then one pack per month for 3–6 mo.

Severe bleeding

- 35 µg ethinyl estradiol plus progestin one pill every 6 h for 4 d, one pill every 8 h for 4 d, one pill every 12 h for 4 d, one pill to completion of 28 d-pack; then one pack per month for 3–6 mo

Intravenous conjugated estrogens

Hemorrhagic shock (Premarin): 25 mg IV every 4–6 h for 24 h

Progestins

- Medroxyprogesterone acetate 10 mg per day for 12–14 d per month
- Norethindrone acetate 5 mg per day for 12–14 d per month
- Micronized progesterone 200 mg per day for 12 d per month

Elemental iron 60 mg three times per day for 8 wk**Nonsteroidal anti-inflammatory drugs**

- Ibuprofen 400–600 mg bid–qid
- Mefenamic acid 250 mg qd–qid
- Naproxen 250–500 mg bid

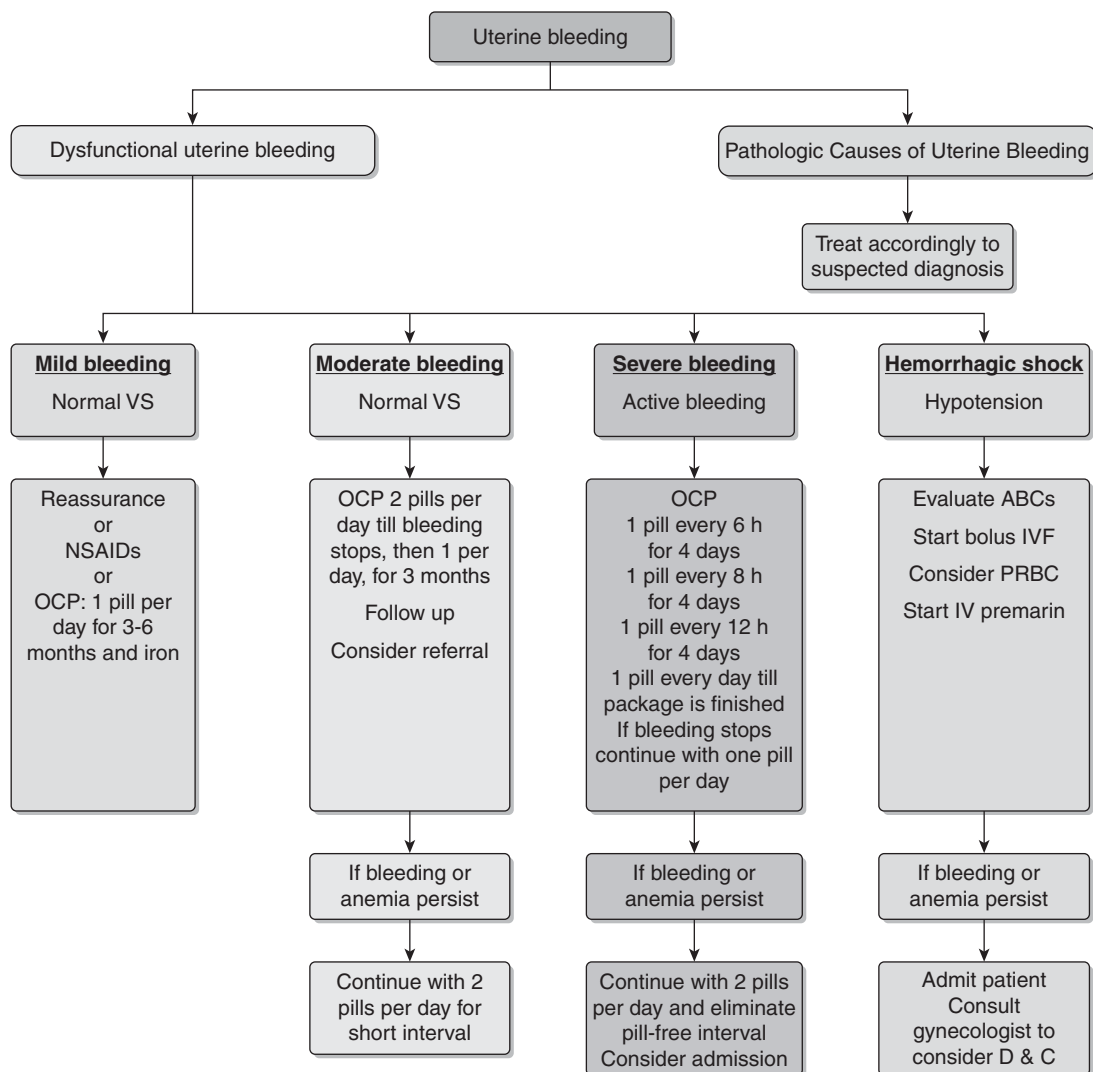


FIG. 98-2. Treatment of dysfunctional bleeding. VS (vital signs), NSAIDs (nonsteroidal anti-inflammatory drugs). Hgb (hemoglobin), ABCs (airway, breathing, circulation), PRBC (packed red blood cells), OCP (oral contraceptive pill), IVF (intravenous fluids), D&C (dilation and curettage).

- Physical examination
 - Assess for hemodynamic stability by evaluating vital signs, patency of airway, and establishing that breathing and circulation are adequate
 - Pelvic examination
- Diagnostic evaluation
 - Consider laboratory studies as clinically indicated
 - Complete blood count
 - Prothrombin time (PT)
 - Activated thromboplastin time (aPTT)
 - Type and cross-match for packed red blood cells
 - Pregnancy test
 - Cervical cultures or urine NAATs for *Chlamydia trachomatis* and *Neisseria gonorrhoea*
 - Imaging studies
 - Platelet function analysis (PFA-100) is the study of choice to determine platelet function
 - ◻ 96% sensitivity and 98% specificity for measuring platelet function
 - Ristocetin cofactor assay of von Willebrand factor (vWF), vW antigen, and factor 8 (FVIII) level
 - Consider thyroid function tests to rule out thyroid dysfunction
 - Consider FSH, LH, testosterone, and DHEAS levels to rule out PCOS
 - Not indicated in patients with DUB
 - Ultrasonography can rule out structural or anatomical pathology

- Management
 - Treatment of patients with nonovulatory DUB: Ensure hemodynamic stability (Table 98-5 and Fig. 98-2)
 - Severe anemia (<7 g/dL), hemorrhagic shock, persistent heavy bleeding
 - ◻ This patient is unstable
 - ◻ Address priorities of resuscitation according to PALS guidelines
 - ◻ Rapid infusion of isotonic crystalloid which may be repeated multiple times
 - ◻ Packed red blood cell transfusion
 - ◻ Intravenous estrogen (Premarin) 25 mg IV q 4 to 6 hours until bleeding ceases for 24 hours
 - ◻ Once bleeding has stopped, begin OCPs containing high progestin
 - ◻ If bleeding does not stop, admit and obtain a gynecology consultation to consider dilation and curettage.
 - Hemodynamically stable patients
 - ◻ Begin oral contraception and NSAIDs
 - ◻ Refer to gynecologist
 - ◻ Mild bleeding
 - Reassurance, NSAIDs
 - Monophasic OCP
 - Iron supplements
 - ◻ Moderate and heavy bleeding
 - Start OCPs at a higher dose
 - Treatment of stable patients with ovulatory DUB
 - NSAIDs
 - Levonorgestrel-releasing intrauterine system
 - OCPs
 - Androgens, antifibrinolytics
 - Desmopressin and surgery

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QUESTIONS

1. A 14-year-old female presents to the ED with complaints of pain in the pelvis and lower back every time her menses starts. Menarche began at age 11 and has been regular for about one year. First line treatment for primary dysmenorrhea includes which of the following?
 - A. Oral contraceptive pills (OCPs)
 - B. Danazol
 - C. Nonsteroidal anti-inflammatory drugs (NSAIDs)
 - D. Leuprolide acetate
 - E. Oral nifedipine
2. A 13-year-old female presents to the emergency department with severe cramping pelvic and low back pain associated with her menses. She reports starting her menses at age 10 ½ years, denies sexual activity, alcohol and drug use, and is doing well in school. Diagnostic evaluation for this child should include which of the following?
 - A. CBC with differential
 - B. Urinalysis
 - C. Erythrocyte sedimentation rate
 - D. Transvaginal ultrasound
 - E. A thorough history and physical exam only
3. A once active and outgoing 17-year-old teenager is brought in to the emergency department because she is tired, appears pale, and seems apathetic about school. Parents are concerned that their teenager may be pregnant because she is also gaining weight. Although, the teen denies sexual activity and cold or heat intolerance, she does report that her menstrual periods last 12 days, come irregularly, and seem to be heavy such that she must change her pads frequently. What should be the first priorities of her evaluation?
 - A. Perform a complete pelvic examination
 - B. Evaluate airway, breathing, and circulation to assess for hemodynamic stability
 - C. Examine the abdomen to determine pregnancy status

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- D. Evaluate for signs of a bleeding disorder or leukemia
 - E. Place an intravenous catheter and obtain peripheral blood samples for type and cross-matched blood
4. In the above patient, vital signs are obtained: HR = 150 bpm, RR = 20 bpm, and supine blood pressure = 120/85 mm Hg. Oxygen saturation is 99% on room air. Weight = 90 kg.

Physical exam reveals a teenager with facial acne, facial hair and acanthosis. Her neck is full but without a palpable mass or lymphadenopathy. Heart sounds are tachycardic with a soft flow murmur, but are regular and without gallop or rub. Lung examination is normal. Abdominal exam reveals normoactive bowel sounds, soft without tenderness and guarding, and there are no palpable masses. The parents refuse a speculum examination, but upon visualization, blood clots are present at the vaginal opening. Skin examination is negative for bruises, rashes, and petechiae. A CBC and type and cross are obtained. What laboratory study(s) may be diagnostic in this patient?

- A. Pregnancy test
 - B. Stool guaiac test for blood
 - C. Thyroid function tests
 - D. FSH, LH, testosterone and DHEAS levels
 - E. PT/PTT and bleeding time
5. During the visit, the above patient continues to experience heavy vaginal bleeding with passage of blood clots. Despite crystalloid fluid resuscitation, she remains tachycardic and with a blood pressure of 110/60 mm Hg. Her hemoglobin is reported as 5 g/dL. Packed red blood cells have been ordered. What initial treatment option for the ongoing vaginal bleeding would be recommended at this time?
- A. Oral contraceptive pills: 1 pill by mouth per day for 3–6 months
 - B. Oral contraceptive pills: 2 pills by mouth per day until bleeding stops
 - C. Platelet transfusion
 - D. Dilation and curettage
 - E. Intravenous estrogen 25 mg every 4 to 6 hours until bleeding stops
6. The teenager described in the above vignette has excessive, prolonged, and irregular bleeding. What pattern of menstrual bleeding is described?
- A. Hypermenorrhea
 - B. Metrorrhagia
 - C. Menometrorrhagia
 - D. Menorrhagia
 - E. Polymenorrhea

7. A healthy 17-year-old with a negative medical, surgical, and family history presents to the ED with the complaint of recent onset of irregular and heavy menstrual bleeding. She denies pain, dizziness, pallor, shortness of breath, or lethargy. She has normal vital signs and appears well. What tests, if any, would be recommended to obtain in this patient first?
- A. CBC with platelet count
 - B. Ristocetin cofactor assay of von Willebrand factor
 - C. Urine analysis
 - D. Pregnancy test
 - E. No tests are necessary

ANSWERS

1. C. Nonsteroidal anti-inflammatory drugs (NSAIDs) are the first line of treatment for primary dysmenorrhea. NSAIDs reduce prostaglandin production via cyclo-oxygenase inhibition and decreases menstrual flow. OCPs are an off label method of treatment for dysmenorrhea and is frequently used after or in conjunction with NSAIDs. Danazol (an androgen) and Leuprolide acetate (a gonadotropin releasing hormone) may be considered in refractory cases. Oral nifedipine has in experimental models shown to decrease myometrial contractions and thus pain, but prospective clinical trials still need to be done.
2. E. Nothing but an excellent history and thorough physical exam. The patient in the above vignette is a girl with classic symptoms of primary dysmenorrhea. Primary dysmenorrhea is usually a benign condition that requires little diagnostic evaluation other than a careful history and physical examination. On the other hand, secondary dysmenorrhea requires a more detailed assessment to determine the underlying pathology. Evaluation may include a complete blood count (CBC), urinalysis, pregnancy test, cultures for sexually transmitted diseases such as *Neisseria gonorrhoeae* and *Chlamydia trachomatis*, and an erythrocyte sedimentation rate. Transvaginal ultrasonography is recommended in the evaluation of secondary dysmenorrhea to identify anatomic abnormalities.
3. B. Evaluate the airway, breathing, and circulation to assess for hemodynamic stability

The first step in the management of a patient with bleeding is to assure hemodynamic stability and evaluate the airway, breathing, and circulation of the patient according to established Pediatric Advanced Life Support (PALS) guidelines. Once this is assessed, obtain vascular access and begin infusion of isotonic crystalloid followed by transfusion of packed red blood cells as indicated. A more

thorough and complete examination can be done while the resuscitation is ongoing, and a pregnancy test and other laboratory tests can be sent to aid in the diagnosis of vaginal bleeding.

4. D. FSH, LH, testosterone, and DHEAS levels. The patient in the above vignette exhibits signs and symptoms of polycystic ovarian syndrome (PCOS) as the cause of her dysfunctional uterine bleeding. The child is overweight, hypertensive and has findings of hirsutism, acne, and acanthosis nigricans. Although the results of the FSH, LH, testosterone and DHEAS levels will not be known immediately, these tests will be helpful and diagnostic in this patient. Although, we recommend a pregnancy test in all females with irregular vaginal bleeding, this test would not explain all of her other physical findings. Stool for blood would not be helpful. There are no history or physical findings consistent with a coagulopathy in this child. Although, patients with hypothyroidism have symptoms of weight gain, apathy, depression, and hair and skin changes, thyroid disease would not completely explain her physical findings and thyroid function tests would not be diagnostic.
5. E. Intravenous estrogen 25 mg every 4 to 6 hours until bleeding stops is the management of choice. Refer to the flow diagram in Fig. 98-2 and Table 98-5. This patient exhibits signs of hemodynamic instability with severe anemia (hemoglobin <7 g/dL), tachycardia, and decreased blood pressure. Despite fluid resuscitation, she remains mildly hypotensive.

In addition to PRBCs, it is recommended to administer intravenous estrogen (Premarin) at a dose of 25 mg every 4 to 6 hours until the bleeding stops for more than 24 hours. Once bleeding is controlled, OCP with a strong progestin can be started. If the bleeding is not controlled despite the above therapy, gynecology can be consulted for dilation and curettage. Unless there is documented thrombocytopenia, platelet transfusion is not indicated. Oral contraceptive pills are recommended as first line therapy in patients who are hemodynamically stable and present with moderate to severe vaginal bleeding.

6. C. Menometrorrhagia is defined as excessive, prolonged and irregular bleeding (>80 mL for > 7 days). See Table 98-3 for the definitions of the other patterns of menstrual bleeding.
7. D. A pregnancy test is indicated first in this patient. In healthy female teenagers, who present with unexplained heavy bleeding, and previously regular cycles, pregnancy must be ruled out. Intrauterine or ectopic pregnancy, threatened or spontaneous abortion all can present with vaginal bleeding. Further testing for other causes of dysfunctional uterine bleeding can be obtained as indicated. This child has no history of bleeding with dental procedures, a negative family history, and is otherwise healthy. She does not exhibit signs of anemia or hypovolemia, systemic or iatrogenic causes of bleeding, and there is nothing in the history or physical examination that concerns us for cancers of the reproductive tract.

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Section 17

HEMATOLOGIC AND ONCOLOGIC EMERGENCIES

99 ANEMIA

Audra L. McCreight

INTRODUCTION

- Anemia is a common incidental finding in the emergency management of infants and children presenting for other reasons. A complete blood count is obtained and reveals an unexpected low hemoglobin level. Table 99-1 lists important basic screening tests which can help to define the source of anemia, and Table 99-2 describes red blood cell morphology used to interpret a cause for anemia.

MICROCYTIC ANEMIA

- Most common cause in children is iron deficiency
- Thalassemia, anemia of inflammation, hemoglobin C disease, hemoglobin E disease, and sideroblastic anemia may also lead to microcytic anemia
- Table 99-3 reviews a method for sorting out the common causes of microcytic anemia

IRON DEFICIENCY

- Risk factors include
 - age between 6 months and 2 years
 - increased duration of breast-feeding
 - lack of use of iron-fortified formulas
 - early introduction of whole cow's milk into the diet
 - low socioeconomic status.
- Whole cow's milk produces deficiency by
 - inherent lack of bioavailable iron
 - inherent high caloric content; reducing appetite and intake of iron-rich foods

- leading to occult gastrointestinal bleeding from the effect of unmodified cow's milk proteins on gastrointestinal mucosa
- Premature infants are at a greater risk
 - most total body iron is absorbed during the last trimester
- Develops slowly; patients present compensated and hemodynamically stable
 - Diagnosis is made on the basis of the history
- Laboratory findings
 - CBC with anemia and significant microcytosis
 - low or normal reticulocyte count
 - reduced serum iron, elevated total iron binding capacity (TIBC), reduced ferritin level
 - ferritin, an acute-phase reactant, may be elevated despite the presence of iron deficiency anemia
- Trial of iron therapy is both diagnostic and therapeutic.
 - ferrous sulfate administered to provide 4 to 6 mg/kg of elemental iron per day in two divided doses for 3 to 4 months
 - multivitamins with iron do not provide adequate amounts of iron
- Hemoglobin level increases in 1 to 2 weeks
- Treatment involves the identification and elimination of the cause of the iron deficiency

THALASSEMIA

- Inherited defects resulting in the absence or decreased production of normal hemoglobin
- Most common in people of Mediterranean, Southeast Asian, and African ancestry
- Disease is classified by the number of abnormal globin genes
 - heterozygous: thalassemia trait
 - produces marked microcytosis out of proportion to the degree of anemia

TABLE 99-1 Anemia Screening Tests For Cause

Mean corpuscular volume (MCV)

- Microcytic?
- Normocytic?
- Macrocytic?

Reticulocyte count

- Low—Decreased production
- High—Increased destruction or blood loss

Red cell distribution width (RDW)—iron deficiency (high) vs. thalassemia (low)

Peripheral smear examination (see Table 99-2)

- typically a high total RBC count and narrow RDW (helps differentiate thalassemia trait from iron deficiency anemia)
- the reticulocyte count should be normal or slightly elevated
- α -Thalassemia results from a decreased production of α -globin
 - due to a deletion or mutation in one or more of the four α -globin genes
 - normal hemoglobin electrophoresis outside of the newborn period
 - silent carrier state: defect in a single gene
 - ◻ no anemia and normal-appearing red cells
 - α -thalassemia trait: defect in two genes
 - ◻ mild microcytic anemia
 - hemoglobin H disease: only one normal α -globin gene
 - ◻ moderate anemia in the 8 to 10 g/dL range
 - ◻ may have increased hemolysis with stress or infection
 - α -thalassemia major: defect in all four α -globin genes
 - ◻ severe fetal complications
- β -Thalassemia: caused by decreased production of β -globin
 - a mutation or deletion in one or more of the two β -globin genes

TABLE 99-2 Peripheral Smear Interpretation

Fragmented RBCs, schistocytes, burr cells

- Hemolytic uremic syndrome
- Thrombotic thrombocytopenic purpura
- Disseminated intravascular coagulopathy

Spherocytes

- Hereditary spherocytosis
- ABO incompatibility

Eliptocytes: Hereditary eliptycotosis

Target cells

- Thalassemia
- Hemoglobinopathies

Sickle cells: Sickle hemoglobinopathies

Basophilic stippling: Lead poisoning

Variation in size and shape: See interpretation of MVC and RDW

- hemoglobin concentration is 2 to 3 g/dL below normal values
- β -thalassemia intermedia: patients maintain a hemoglobin of 6 to 8 g/dL and do not require chronic transfusion
- β -thalassemia major produces severe hemolytic anemia with marked microcytosis and reticulocytosis
 - presents within the first year of life
 - require lifelong transfusion therapy: the use of uncrossmatched blood is avoided, except in the most dire circumstances
 - major side effect of long-term transfusion therapy is iron overload

LEAD POISONING

- Anemia is due to iron deficiency which:
 - leads to pica and the ingestion of lead
 - increases absorption of lead from the gastrointestinal tract
- Must be considered in the child with microcytic anemia
- Basophilic stippling of the red blood cells is noted on peripheral blood smear

TABLE 99-3 Differentiating Microcytic Anemia

PARAMETER	IRON DEFICIENCY	THALASSEMIA	ANEMIA OF CHRONIC DISEASE
History	Prematurity or high milk intake	None or family history	Chronic inflammation or disease
Reticulocyte count	Low	Normal	Low
MCV	Low	Very low	Normal to low
RDW	High	Low	Normal to high
Serum ferritin	Low	Normal	High
Serum iron	Low	Normal or elevated	Low
TIBC	High	Normal	Low

MCV, mean corpuscular volume; RDW, red cell distribution width; TIBC, total iron binding capacity.

TABLE 99-4 Differential Diagnosis for Normocytic Anemia*Blood loss (high reticulocyte count)**Hemolytic anemia (high reticulocyte count)*

Immune

- Autoimmune hemolytic anemia
- Neonatal–maternal blood group incompatibility

Nonimmune

Microangiopathic

- Disseminated intravascular coagulation (DIC)
- Hemolytic uremic syndrome (HUS)

Macroangiopathic

- Artificial cardiac valve
- Membrane abnormalities
- Spherocytosis
- Elliptocytosis

Metabolic abnormalities

- G6PD deficiency
- Pyruvate kinase deficiency

Hemoglobinopathies

Nonhemolytic anemia (low or normal reticulocyte count)

Abnormality isolated to red cell line

- Chronic hemolytic anemia with concurrent aplastic crisis
- Transient erythroblastopenia of childhood (TEC)

Chronic disease

- Renal insufficiency
- Diamond–Blackfan anemia

Abnormality affecting other cell lines

- Bone marrow infiltration
- Leukemia
- Lymphoma
- Tumor metastasis
- Acquired aplastic anemia

NORMOCYTIC ANEMIA

- Differential diagnosis in childhood is extensive. See Table 99-4.
- Important to determine whether the anemia is due to decreased production, increased destruction, or blood loss
- Diagnosis is based on history, reticulocyte count, and review of RBC morphology

NORMOCYTIC ANEMIA WITH HIGH RETICULOCYTE COUNT

- Most common cause is blood loss
- No evidence of blood loss, a hemolytic anemia is likely
- Evaluation includes a Coombs or direct anti-globulin test (DAT)
 - determines whether the hemolytic anemia is immunologic in nature

- Immune hemolytic anemia
 - may be the result of a drug reaction, infection, collagen vascular disorder, malignancy, or undetermined
 - presentation is acute with severe anemia, pallor, jaundice, and hemoglobinuria
 - transfusions may be necessary
 - difficult due to the circulating antibodies causing hemolysis
 - immunosuppression with corticosteroid is initial therapy
 - severe cases may require plasmapheresis or IVIG
- Nonimmune hemolytic anemia
 - micro and macroangiopathic destruction, membrane disorders, metabolic abnormalities, and hemoglobinopathies
 - microangiopathic RBC destruction occurs with disseminated intravascular coagulation (DIC), thrombotic thrombocytopenic purpura (TTP), and hemolytic uremic syndrome
 - peripheral smear will demonstrate schistocytes, burr cells, and other RBC fragments
 - hereditary spherocytosis (HS) and elliptocytosis (HE) result from mutations in the red cell membrane
 - hemolytic anemia occurs due to splenic destruction of abnormally shaped RBCs
 - often presents as jaundice and anemia in infancy
 - laboratory studies reveal anemia, reticulocytosis, and hyperbilirubinemia
 - major hematologic crisis is aplastic anemia, usually secondary to a parvovirus infection
 - ◻ may also have an increased rate of hemolysis with stress or infection
 - splenectomy is curative and is considered in patients with severe hemolysis leading to frequent transfusion or hospitalization
 - inherited metabolic disorders, such as pyruvate kinase and glucose-6-phosphate dehydrogenase (G6PD) deficiencies cause chronic hemolysis
 - ◻ Episodes of acute hemolysis can occur with many variants of G6PD deficiency, including the A variant found in 10% of African American boys
 - symptoms can include pallor, malaise, fever, scleral icterus, abdominal pain, and dark urine
 - most common cause of acute hemolysis in American children is exposure naphthalene containing mothballs
 - ◻ Treatment involves blood transfusion with severe anemia and counseling regarding avoidance of oxidant stressors

NORMOCYTIC ANEMIA WITH LOW RETICULOCYTE COUNTS

- Indicates bone marrow underproduction
- Abnormality isolated to the RBC line; primary considerations include transient erythroblastopenia of childhood (TEC) and an aplastic crisis complicating an underlying hemolytic anemia
- TEC is an acquired pure RBC aplasia; WBC and platelet counts are normal
 - affects children between 1 and 4 years of age
 - often an associated history of preceding viral illness
 - no causative viral agent has been identified
 - supportive therapy is usually sufficient as patients recover spontaneously over several weeks; steroids have not been shown to speed recovery
 - transfusion may be necessary in symptomatic patients
- Other causes include anemia of chronic disease, inflammatory processes, and decreased erythropoietin from renal insufficiency
 - anemia of chronic disease (ACOD) occurs in patients with acute or chronic immune activation, resulting in disorder iron hemostasis and blunted erythropoietin response
 - ACOD is usually a mild normocytic, normochromic anemia; but may be microcytic in long-standing cases, making it difficult to differentiate from iron deficiency
- Thrombocytopenia or WBC abnormalities with normocytic anemia and poor reticulocyte response suggests marrow infiltration or acquired aplastic anemia
 - marrow infiltration is most commonly due to leukemia
 - acquired aplastic anemia has been associated with drugs and infections
 - often no etiology is determined
 - prognosis is poor often requiring bone marrow transplantation

MACROCYTIC ANEMIA

- Relatively uncommon in pediatric patients
- Folate and vitamin B₁₂ deficiencies
 - rare in otherwise healthy children
 - consider in
 - children with underlying gastrointestinal pathology
 - infants fed only Goat's milk (contains very little folate)
 - children receiving some drugs, notably AZT

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QUESTIONS

1. A 17-month-old male is referred to the Emergency Department (ED) for pallor. The patient was delivered full term and has an uncomplicated past medical history. Parents report no recent travel, viral infections, or new medications. On physical exam, vital signs are within normal limits. The infant is awake, alert and playful. Physical exam is notable only for generalized pallor. CBC reveals a hemoglobin level of 3 g/dL, a low MCV, normal WBC and platelet counts. The reticulocyte count is also noted to be low. Peripheral blood smear reveals pale red blood cells but is otherwise unremarkable. What is the most common etiology of the patient's anemia?
 - A. Iron deficiency anemia
 - B. Thalassemia
 - C. Lead poisoning
 - D. Acute blood loss
 - E. Aplastic anemia
2. What is the most common cause of the anemia described in Question 1?
 - A. Excess ingestion of goat's milk
 - B. Excess ingestion of cow's milk
 - C. Prematurity
 - D. Marrow infiltration by leukemic cells
 - E. Red cell membrane abnormality
3. A 2-year-old African-American male presents to the emergency department for evaluation of yellow eyes, getting worse over the past 24 hours. They also report that the patient has had decreased energy and was complaining of abdominal pain. Of note, they say his urine is extremely dark. The only thing they can think of out of the ordinary the past week is they found him eating a moth ball at grandmother's house 3 days ago. CBC reveals a hemoglobin level of 4 g/dL and an increased reticulocyte count.

What underlying disorder is causing this patient's anemia?

- A. Iron deficiency anemia
 - B. Thalassemia
 - C. Glucose-6-phosphate dehydrogenase deficiency (G6PD)
 - D. Acute blood loss
 - E. Anemia of chronic disease
4. A 2-year-old female presents to the ED as a referral from her primary physician for evaluation of anemia. The patient was seen for a follow-up visit in the office today following resolution of a viral infection 2 weeks prior. Physical exam reveals a stable female toddler with normal vital signs and is otherwise unremarkable. Complete blood count reveals a hemoglobin of 5 g/dL, low reticulocyte count and a normal MCV. White blood cell (WBC) and platelet counts are noted to be normal. What is the most common etiology of the patient's anemia?
- A. Marrow infiltration by leukemic cells
 - B. Excess ingestion of cow's milk
 - C. Hereditary spherocytosis
 - D. Transient erythroblastopenia of childhood (TEC)
 - E. Aplastic anemia
5. A 3-year-old female presents to the emergency department with new onset rash. Her parents report that the rash began 2 hours prior and has now gotten worse. Vitals: T: 40C, HR: 165, RR: 30, BP: 75/30. On physical exam the patient is lethargic with delayed capillary refill of 5 seconds. Bounding femoral pulses are palpated. Petechiae are noted to her lower extremities. You diagnose decompensated septic shock and initiate appropriate therapy. What type of anemia do you expect this patient to manifest?
- A. Iron deficiency anemia
 - B. Thalassemia
 - C. Immune hemolytic anemia
 - D. Non-immune hemolytic anemia
 - E. Anemia of chronic disease
6. A 1-year-old male presents to the ED as a referral from his primary physician for anemia. The family recently emigrated from Greece and was seen as a new patient visit today. On physical exam the patient is alert and active with mild pallor. Otherwise physical exam is unremarkable. Hemoglobin is measured at 8.7 g/dL with an MCV of 65. Iron deficiency anemia is ruled out with normal iron studies. What is the etiology of the patient's anemia?
- A. α -thalassemia trait
 - B. α -thalassemia silent carrier state
 - C. Hemoglobin H disease

- D. α -thalassemia major
- E. β -thalassemia major

ANSWERS

1. A. Iron deficiency anemia. The patient presents to the ED in no distress. He is pale with a microcytic anemia, yet is hemodynamically compensated with normal activity and vital signs. This indicates that the anemia is not acute, but has developed over a period of time. This eliminates acute blood loss as the etiology. Aplastic anemia would cause a decrease in WBC and platelet count which this patient does not have. Iron deficiency is the most common cause of a compensated microcytic anemia in children of this age group. It develops slowly and patients rarely present with acute symptoms. Even with drastically reduced hemoglobin levels, patients are usually well compensated and hemodynamically stable. Thalassemia could be the etiology of the patient's anemia, yet it is most often associated with a normal to high reticulocyte count. The reticulocyte count is low in this patient. Finally, no basophilic stippling is noted on peripheral blood smear eliminating lead poisoning as the cause of the patient's anemia.
2. B. Excess ingestion of cow's milk causes iron deficiency due to its inherent lack of iron, by reducing appetite and intake of iron rich foods, and by leading to occult gastrointestinal bleeding from the effect of unmodified cow's milk proteins on the gastrointestinal mucosa. Ferrous sulfate is administered to provide 4–6 mg/kg of elemental iron per day in two divided doses for 3–4 months. Over the counter supplements, such as poly-vi-sol, do not provide adequate amounts of iron to correct the deficiency. Hemoglobin level corrects in 1–2 weeks following the initiation of iron therapy. Elimination of excess cow's milk in the child's diet is necessary. Excess goat's milk in the diet will cause a macrocytic anemia due to the low amount of folate. Premature infants often suffer from iron deficiency because the majority of fetal iron is absorbed in the last trimester. However, this would present earlier and by history, the patient in this scenario was born full term. Marrow infiltration by leukemic cells would produce anemia as well as abnormalities in the WBC and platelets. These patients present ill with abnormalities on physical exam such as petechiae and hepatosplenomegaly. Finally, abnormalities in red cell membranes, such as hereditary spherocytosis and elliptocytosis, would produce a normocytic anemia. Most cases are inherited as autosomal dominant, making family history

extremely important. These patients have an elevated bilirubin due to destruction of abnormal red cells in the spleen, which is often enlarged at presentation. Splenectomy is curative.

3. C. Inherited metabolic disorders, such as pyruvate kinase and glucose-6-phosphate dehydrogenase (G6PD) deficiencies, can cause acute and chronic hemolysis. There are multiple variants of G6PD deficiency, with varying degrees of severity. The A-variant is seen in approximately 10% of African American males and becomes symptomatic only after a significant challenge from a drug or infection. The most common cause of acute hemolysis in American children is exposure to naphthalene containing mothballs. The acute hemolysis seen following oxidant exposure in G6PD deficiency presents 1 to 3 days later. Symptoms can include pallor, malaise, fever, scleral icterus, abdominal pain, and dark urine. Treatment involves blood transfusion with severe anemia and counseling regarding avoidance of oxidant stressors. Iron deficiency anemia, thalassemia, and anemia of chronic disease are not hemolytic in etiology. None present with scleral icterus or hemoglobinuria, as did this patient. The family does not report a history of acute blood loss.
4. D. TEC is an acquired pure RBC aplasia resulting in a normocytic anemia with a low reticulocyte count. WBC and platelet counts are normal. It typically affects children between 1 and 4 years of age. There is seasonal clustering and an associated history of preceding viral illness, but no causative viral agent has been identified. Supportive therapy is usually sufficient as patients are typically hemodynamically stable and recover spontaneously over several weeks. Transfusion may be necessary in symptomatic patients. Steroids have not been shown to speed recovery. Marrow infiltration by leukemic cells would cause a normocytic anemia as well as abnormalities in the WBC and platelets. These patients often present ill with abnormalities on physical exam such as petechiae and hepatosplenomegaly. Excess cow's milk ingestion would produce a microcytic anemia due to iron deficiency. Hereditary spherocytosis would result in a normocytic anemia with high reticulocyte count and associated hyperbilirubinemia and hepatosplenomegaly.
5. D. The patient described is manifesting symptoms of disseminated intravascular coagulation (DIC) which causes a normocytic microangiopathic hemolytic anemia. This type of hemolytic anemia is often seen in the face of overwhelming infection, as with

this patient, and is not immunogenic in nature. The peripheral smear will demonstrate schistocytes, burr cells, and other RBC fragments. Immune hemolytic anemia may be the result of a drug reaction, infection, collagen vascular disorder, or malignancy, but commonly no etiology is determined. Patients often present acutely with severe anemia, pallor, jaundice, and hemoglobinuria. Transfusions may be necessary with severe symptomatic anemia, but may be difficult due to the circulating antibody causing "incompatibility" in vitro and the rapid destruction of transfused cells. Iron deficiency anemia and thalassemias are both microcytic anemia. Anemia of chronic disease (ACOD) occurs in patients with acute or chronic immune activation, resulting in disorder of iron hemostasis and blunted erythropoietin response. ACOD is usually a mild normocytic, normochromic anemia; but may be microcytic in long-standing cases, making it difficult to differentiate from iron deficiency (see Table 99-1). However, as the name describes, ACOD is a chronic disorder and does not present with an acute onset as with this patient.

6. C. Thalassemias are inherited defects resulting in the absence or decreased production of normal hemoglobin, leading to a microcytic anemia. The condition is most common in people of Mediterranean, Southeast Asian, and African ancestry and is the most common single gene disease worldwide. In general, the disease is classified by the number of abnormal globin genes. The heterozygous form of thalassemia is often referred to as thalassemia trait. Thalassemia trait produces marked microcytosis out of proportion to the degree of anemia. α -Thalassemia results from a decreased production of α -globin because of a deletion or mutation in one or more of the four α -globin genes. The silent carrier state results from a defect in a single gene and patients have no anemia and normal-appearing red cells. α -Thalassemia trait refers to a defect in two genes, resulting in mild microcytic anemia. Hemoglobin H disease occurs when there is only one normal α -globin gene. These patients have moderate anemia in the 8 to 10 g/dL range, but may have increased hemolysis with stress or infection. A defect in all four α -globin genes results in α -thalassemia major, a condition leading to severe fetal complications. β -Thalassemia results from a decreased production of β -globin because of a mutation or deletion in one or more of the two β -globin genes. In β -thalassemia trait, the hemoglobin concentration is often 2 to 3 g/dL below normal values. In β -thalassemia intermedia, patients maintain a hemoglobin level of

6 to 8 g/dL and do not require chronic transfusion. β -Thalassemia major produces severe hemolytic anemia with marked microcytosis and reticulocytosis. It usually presents within the first year of life. Pallor, jaundice, and hepatosplenomegaly are often present.

100 SICKLE CELL DISEASE

Audra L. McCreight

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INTRODUCTION

- Hemoglobin S (Hgb S): results from a single nucleotide mutation in the β -globin gene leading to the substitution of valine for the normal glutamic acid
- Sickle cell trait: patients have a single abnormal gene for HbS
 - concentration of HbS is typically 40%
 - large percentage of normal hemoglobin allows patients to remain asymptomatic except under the most severe hypoxic stress
 - sickle trait should be considered a benign condition
- Sickle cell disease (SCD): homozygous for Hgb S or is a compound heterozygote for Hgb S and another interacting β -globin variant
 - Hgb SS: sickle cell anemia
 - Hgb SC: hemoglobin SC disease
 - Hgb S- β thalassemia (either β^0 or β^+)
 - Hgb SD
- Patients with double heterozygous states such as Hgb SC, Hgb S β^+ thalassemia, and Hgb SD are typically less seriously affected than those with Hgb SS or Hgb S β^0 (no hemoglobin A production)
- Approximately 8% of the African-American population are sickle trait carriers
 - 1 in 600 have sickle cell anemia
 - also occurs in people of Mediterranean, Indian, Central and South American, and Middle Eastern descent
- Vasoocclusion occurs via a combination of sickle cell interactions with endothelial cells and obstruction from nondeformable sickled cells
- Dactylitis or hand-foot syndrome
 - vasoocclusion in marrow of the metacarpal or metatarsal bones
 - often the earliest presentation of SCD
 - usually occurs between 6 and 18 months of life
 - infants present with hand and foot swelling and tenderness
 - may lead to refusal to walk and irritability
 - dactylitis declines with age as hematopoiesis shifts to the long bones
- Older patients experience vasoocclusive events in the long bones, back, joints, and abdomen
- Events may be precipitated by dehydration, hypoxia, cold exposure, or infection
 - often no instigating factor is identified
- Diagnosis is made on history alone
- Formation of gallstones due to chronic hemolysis may lead to cholecystitis or pancreatitis and abdominal pain
- Laboratory evaluation
 - complete blood count (CBC) and reticulocyte count
 - patients typically remain at baseline levels of Hgb during a painful event
- Treatment
 - hydration at maintenance to correct and prevent dehydration
 - overhydration may lead to acute chest syndrome (ACS) and should be avoided
 - oxygen has not been shown to be beneficial in the management of pain crises unless hypoxemia is present
 - pain relief
 - assessed at least every 30 minutes using standardized pain scales
 - medication choice depends on the severity of the crisis and what the patient has required in past crisis
 - mild to moderate pain: scheduled oral agents such as acetaminophen, nonsteroidal anti-inflammatory drugs (NSAIDs), and codeine used separately or in combination
 - severe pain: parenteral opioids, preferably morphine or hydromorphone
- Patients who receive scheduled oral opioid and NSAID and require parenteral opioid analgesics are observed for 3 or 4 hours
 - if adequate pain relief is not achieved, the patient is admitted for further parenteral analgesia
- Analgesia should be provided at frequent regular time intervals to avoid breakthrough pain
 - recurring pain is more difficult to control

VASOOCCLUSIVE CRISIS

- Most common complication of SCD and the most frequent cause of ED visits
- Result from the obstruction of blood flow in the microcirculation leading to tissue ischemia and microinfarction

- Hypoventilation as a result of opiate use may increase the risk of developing ACS
 - encourage incentive spirometry

ACUTE CHEST SYNDROME (ACS)

- Presence of a new lobar or segmental pulmonary infiltrate in the presence of fever, respiratory symptoms, and/or chest pain
- Contributing causes may include infection, pulmonary infarction due to vasoocclusion, and fat emboli from marrow infarction
- May cause splinting and hypoventilation
- Often difficult to differentiate vasoocclusion from pneumonia
 - both etiologies cause similar manifestations
- Infectious organisms associated with ACS:
 - *S pneumoniae* in younger children
 - *Mycoplasma* or *Chlamydia* in adolescents
- May present with or rapidly progress to respiratory failure
- All patients are admitted to the hospital
- Laboratory evaluation:
 - CBC, reticulocyte count
 - blood culture
 - type and crossmatch should be considered
 - chest radiograph
- Treatment
 - supplemental oxygen via face mask or nasal cannula in hypoxemic patients
 - antibiotic therapy directed at *S pneumoniae* and atypical organisms
 - a third-generation cephalosporin and a macrolide
 - analgesia for chest pain
 - manage carefully to prevent hypoventilation
 - hydration limited to 1 to 1.25 times maintenance (po + IV) to avoid fluid overload
 - a hemoglobin >2 g/dL below baseline, hypoxia, or a rapidly progressing process will require blood transfusion
 - either simple or by exchange depending on the severity of symptoms and level of anemia
 - bronchodilators in patients with a history of reactive airway disease

INFECTION

- High risk primarily due to functional asplenia
- Major risk from encapsulated bacteria, especially *S pneumoniae*
- Despite prophylactic penicillin and vaccines for *S pneumoniae* and *H influenzae* type B, overwhelming

pneumococcal sepsis remains a significant cause of death

- Laboratory evaluation:
 - CBC and reticulocyte count
 - blood culture
 - lumbar puncture in the absence of meningeal signs is not indicated
- Treatment
 - parenteral antibiotics effective against *S pneumoniae* and, if unimmunized, *H influenzae* B
- Hospitalization
 - required for any toxic-appearing child
 - considered in patients presenting with a temperature >40°C, a WBC count >30,000 per mm³, thrombocytopenia, hemoglobin significantly below baseline value, or in a situation where follow-up is uncertain or unlikely
- Children with SCD are also susceptible to pneumonia, meningitis, and osteomyelitis
 - etiology is most frequently encapsulated organisms
 - most common organism identified as the cause of osteomyelitis in patients with SCD is *Salmonella*

STROKE (CEREBROVASCULAR ACCIDENTS)

- Occurs in approximately 10% of children with sickle cell anemia before 18 years of age
 - rare in children with Hgb SC and Hgb Sβ⁺
- Most strokes in children are ischemic events, involving large arteries
- Hemorrhagic events are more common in adults, but may be seen in adolescents
- Common presenting signs and symptoms
 - hemiparesis, refusal to use an arm or leg, aphasia, dysphasia, seizures, cranial nerve palsy, or coma
- Initial management
 - a careful history, focusing on any previous neurologic events and results of previous neuroimaging
 - laboratory evaluation:
 - CBC, reticulocyte count, and type and crossmatch
 - MRI and MRA with diffusion-weighted imaging should be obtained as soon as possible
 - noncontrast enhanced CT is only necessary if there will be a delay in obtaining MRI
 - results of neuroimaging may be normal early in the event and the diagnosis of stroke may be made clinically
- Treatment
 - exchange or simple transfusion to reduce Hgb S to less than 30% is the management of choice
 - admission to the intensive care unit for further monitoring

ACUTE SPLENIC SEQUESTRATION CRISIS (ASSC)

- Occurs when red cells become trapped in the spleen
 - results in a rapidly enlarging spleen
 - sudden drop in Hgb
 - risk of hypovolemic shock
- Occurs most often in patients with Hgb SS between 3 months and 5 years of age
- Repeated infarctions of the spleen in patients with Hgb SS leads to autosplenectomy by 3 to 5 years of age
 - decreases the risk of ASSC
- Less common in patients with Hgb SC and S β^+
 - persistent splenomegaly may occur in adolescence or adulthood
- May present with the sudden onset of weakness, pallor, tachycardia, tachypnea, or abdominal fullness.
- Parents of children with SCD are taught to palpate the spleen regularly at home
 - present to the ED for a newly palpable spleen or enlargement of a chronically enlarged spleen
- Laboratory evaluation
 - CBC with reticulocyte count
 - may demonstrate marked anemia, an elevated reticulocyte count, and often thrombocytopenia
 - consider type and crossmatch
- Treatment:
 - mainstay of therapy is simple blood transfusion
 - in the unstable patient:
 - judicious use of fluid expansion in the initial stages of resuscitation while awaiting blood
 - volume overload and congestive heart failure can result
- Consider blood transfusion with a decline in hemoglobin of 2 g/dL or more below baseline, hemoglobin below 5 g/dL, or in any patient with signs of cardiovascular compromise
 - goal of transfusion is hemoglobin of 9 g/dL
- Sequestration frequently recurs
 - up to 50% of children having a second ASSC
 - splenectomy may be necessary

APLASTIC CRISIS

- Infections with Parvovirus B19 may cause transient red cell aplasia
- RBC lifespan in normal children: 120 days
 - infection will not lead to a significant drop in Hgb
- RBC lifespan in children with SCD : 10–25 days
 - marrow suppression may lead to a significant drop in hemoglobin
- Patients have fever, upper respiratory symptoms, nausea and vomiting, arthralgias, or myalgias, but may also have minimal symptoms

- Aplastic crisis may present with fatigue, pallor, tachycardia, and tachypnea
- Laboratory evaluation:
 - CBC with reticulocyte count
 - reveals a drop in the hemoglobin from baseline and a decreased reticulocyte count (usually <1%)
 - recovery phase may have a high reticulocyte count and numerous nucleated RBCs noted on the blood smear
- Mildly anemic and asymptomatic children:
 - managed with supportive care and close outpatient observation pending marrow recovery
- Simple transfusion of RBCs is necessary in patients with hemoglobin below 5 g/dL or with cardiovascular compromise
- Hgb SC or S β^+ rarely require transfusion with aplastic crisis
 - these patients have higher baseline hemoglobin and longer RBC lifespan
- Patient is infectious and proper isolation procedures must be followed
- Parents should be made aware of the risk to siblings, other family members with SCD, or pregnant females

PRIAPISM

- Occurs in up to 50% of boys with SCD before 21 years of age
- May occur in stuttering episodes that last less than 2 hours but with frequent recurrence, or as a sudden event lasting for hours
- Prolonged episodes may lead to impotence
- No specific therapy is necessary for a single episode of stuttering priapism
- Hydration, warm showers or baths, opioid pain medication, or frequent urination may end an episode
- Oral adrenergic agents such as pseudoephedrine, may treat brief episodes
 - often used prophylactically in patients with frequent events
- Events lasting > 2 hours require immediate medical management and evaluation by an experienced urologist

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QUESTIONS

1. A 3-year-old African-American male with Hgb SS disease presents to the emergency department (ED) with a chief complaint of weakness. His parents report decreased energy and abdominal pain. Vital signs are within normal limits except for mild tachycardia (heart rate: 130). On exam the patient is pale. He is noted to have an enlarged abdomen with a palpable spleen measured 4 cm below the left costal margin. What is the etiology of this patient's symptoms?
 - A. Stroke
 - B. Intussusception
 - C. Acute splenic sequestration crisis
 - D. Dactylitis
 - E. Acute chest syndrome (ACS)
2. A 2-year-old female with Hgb SS presents to the ED with fever. Her parents report a temperature of 103°F at home. Vital signs are significant for tachycardia and borderline hypotension. On exam, the patient is pale and lethargic. You are concerned about bacteremia and possible sepsis. Considering the patient's underlying medical condition, what would be the best antibiotic of choice?
 - A. Oral cephalixin
 - B. Oral azithromycin
 - C. Parenteral ceftriaxone
 - D. Parenteral gentamycin
 - E. Oral trimethoprim-sulfamethoxazole
3. A 7-year-old African-American male with Hgb SS disease presents to the emergency department with the inability to move his right hand or foot. The patient experienced the acute onset of symptoms one hour prior to arrival. Vital signs are stable. Physical exam is significant for right hemiparesis. What is the most likely etiology of the patient's symptoms?
 - A. Atypical migraine headache
 - B. Hemorrhagic cerebrovascular accident
 - C. Embolic cerebrovascular accident
 - D. Conversion disorder
 - E. Ischemic cerebrovascular accident
4. A 15-year-old African-American female with Hgb SS disease presents to the emergency department with new onset of chest pain, cough, and fever of 38.5°C. Chest x-ray reveals a right middle lobe infiltrate. Complete blood count reveals anemia with a hemoglobin level of 3g/dL below the patient's baseline level. You are concerned about ACS. What is the appropriate initial workup and management of this patient?
 - A. Single dose of parenteral third-generation cephalosporin and discharge home
 - B. Initiation of parenteral third-generation cephalosporin, oral macrolide antibiotic, and hospital admission
 - C. Initiation of oral macrolide antibiotic and discharge home
 - D. Hospital admission with initiation of antibiotic only if blood culture is positive
 - E. Discharge home to follow up with hematologist in the morning
5. A 16-year-old African-American male presents to the emergency department after 3 hours of priapism. The patient has tried over the counter pseudoephedrine, warm showers and frequent urination without resolution. What is the next step in the appropriate management of this patient's condition?
 - A. Obtain CBC with reticulocyte count
 - B. Administer a second dose of oral pseudoephedrine
 - C. Administer additional intravenous hydration to enhance urine output
 - D. Consultation with urology for management
 - E. Administration of oral analgesics to control pain
6. A 4-year-old female with Hgb SC disease presents to the emergency department with bilateral leg pain. This is the typical location for her pain during a crisis. Her mother has initiated oral NSAIDs and acetaminophen with codeine at home without relief. Appropriate laboratory studies have been sent. What is the most appropriate management of this patient?
 - A. Continue oral NSAIDs and acetaminophen with codeine alone
 - B. Simple blood transfusion
 - C. Provide parenteral opioid alone
 - D. Provide parenteral opioid and continue oral NSAIDs and acetaminophen with codeine
 - E. Evaluate for other sources of pain as Hgb SC should not cause acute vaso-occlusive events.
7. A 3-year-old female with Hgb SS disease presents to the emergency department from her primary pediatrician's office for evaluation of pallor. She had been in her doctor's office for a 1 week recheck following a mild upper respiratory infection. Vital signs are stable and physical exam is normal other than pale mucus membranes. Complete blood count reveals

hemoglobin 2 g/d below baseline with a low reticulocyte count. What is the explanation for this patient's drop in hemoglobin?

- A. Infection with Parvovirus B19
- B. Acute blood loss
- C. Acute splenic sequestration crisis
- D. Anemia from vasoocclusive crisis
- E. Marrow infiltration with leukemia

ANSWERS

1. C. Acute splenic sequestration crisis (ASSC) occurs when red cells become trapped in the spleen, resulting in a rapidly enlarging spleen, a sudden drop in hemoglobin, and the potential for shock. ASSC occurs most often in patients with Hgb SS between 3 months and 5 years of age. ASSC may present with the sudden onset of weakness, pallor, tachycardia, tachypnea, or abdominal fullness. Parents of children with SCD are taught to palpate the spleen regularly at home and present to the ED for a newly palpable spleen or enlargement of a chronically enlarged spleen. Laboratory studies demonstrate marked anemia, an elevated reticulocyte count, and often thrombocytopenia. Intussusception is possible in this patient with abdominal pain but the enlarging spleen makes ASSC the most likely etiology. The patient is not experiencing any focal deficits that may indicate a stroke. Dactylitis is unlikely in the absence of hand/foot swelling and pain. In addition, by age 3, hematopoiesis has moved to the long bones of the body. Acute chest syndrome does affect this patient population; however the absence of chest pain and respiratory symptoms makes this diagnosis unlikely.
2. C. Patients with SCD are at high risk for infection primarily due to functional asplenia. The major risk comes from encapsulated bacteria, especially *S pneumoniae*. Although prophylactic penicillin and vaccines for pneumococci and *Haemophilus influenzae* type B have reduced the incidence of sepsis in this vulnerable population, overwhelming pneumococcal sepsis remains a significant cause of death in children with SCD. Children younger than 3 years are particularly susceptible to bacteremia, which can occur as commonly as nine bacteremic events per 100 patient-years. The fatality rate is high, even though many of these children appear well at initial presentation. Children with SCD who present to the ED with fever ($>38.5^{\circ}\text{C}$) are at highest risk for bacteremia. After obtaining a CBC and blood culture, all persons with SCD should be promptly treated with parenteral antibiotics effective against *S pneumoniae* and, if unimmunized, *H influenzae* B. Of the parenteral antibiotic choices provided, ceftriaxone is the best choice as it is long acting and provides good coverage against *S pneumoniae*.
3. E. Stroke occurs in approximately 10% of children with sickle cell anemia before 18 years of age, but is rare in children with Hgb SC and Hgb S β^+ . Most strokes in children are ischemic events, involving large arteries. Hemorrhagic events are more common in adults, but may be seen in adolescents. Common presenting signs and symptoms of ischemic stroke include hemiparesis, refusal to use an arm or leg, aphasia, dysphasia, seizures, cranial nerve palsy, or coma. Initial management should include a careful history, focusing on any previous neurologic events and results of previous neuroimaging. A CBC, reticulocyte count, and type and crossmatch should be drawn. An MRI and MRA with diffusion-weighted imaging should be obtained as soon as possible. A non contrast enhanced CT is only necessary if there will be a delay in obtaining MRI. Results of neuroimaging may be normal early in the event and the diagnosis of stroke may be made clinically. While there is no treatment proven to change the acute outcome, exchange or simple transfusion to reduce Hgb S to less than 30% is the management of choice. Care must be taken to maintain the patient's hemoglobin below 11 g/dL until Hgb S is known to be below 30%. Chronic transfusion to maintain Hgb S below 30% has been shown to reduce recurrent stroke events. Hemorrhagic and embolic cerebrovascular accidents are much less likely in this patient. While a migraine headache and conversion disorder are possible in this patient population, one must evaluate the most likely and most potentially devastating etiology first.
4. B. Acute chest syndrome (ACS) is the presence of a new lobar or segmental pulmonary infiltrate in the presence of fever, respiratory symptoms, and/or chest pain. Various causes may contribute to ACS including infection, pulmonary infarction due to vasoocclusion, and fat emboli from marrow infarction. Chest pain from vasoocclusion may cause splinting and hypoventilation, leading to the development of ACS in a patient who initially presents with a painful episode. Infectious organisms associated with ACS include *S pneumoniae* in younger children and *Mycoplasma* or *Chlamydia* in adolescents. ACS may present with or rapidly progress to respiratory failure requiring mechanical ventilation. All patients with ACS should be admitted to the hospital. Laboratory evaluation should include a CBC, reticulocyte count, and blood culture. Antibiotic therapy directed at *S pneumoniae* and atypical organisms such as a third-generation cephalosporin and a macrolide should be initiated. A type and crossmatch should be considered as red cell transfusion may be necessary. Room air oxygen saturation should be checked and supplemental oxygen initiated via face mask or nasal

cannula in hypoxemic patients. Analgesia for chest pain should be provided, but managed carefully to prevent hypoventilation. Hydration should be limited to 1 to 1.25 times maintenance fluids in order to avoid fluid overload. Patients with hemoglobin >2 g below baseline, hypoxia, or a rapidly progressing process will require blood transfusion. Transfusion may be either simple or by exchange depending on the severity of symptoms and level of anemia compared to the patient's baseline hemoglobin. Transfusion will decrease the percentage of Hgb S and increase the blood's oxygen-carrying capacity.

5. D. This patient is experiencing a prolonged episode of priapism. Priapism, a prolonged painful erection of the penis, may occur in up to 50% of boys with SCD before 21 years of age. Priapism may occur in stuttering episodes that last less than 2 hours but with frequent recurrence, or may occur as a sudden event lasting for hours. Maneuvers such as hydration, warm showers or baths, opioid pain medication, or frequent urination may end an episode. Oral adrenergic agents such as pseudoephedrine may treat brief episodes, and are often used prophylactically in patients with frequent events. Events lasting longer than 2 hours require immediate medical management and evaluation by an experienced urologist. All choices other than choice D may be performed during the patient's stay in the emergency department. Due to the fact that prolonged episodes of priapism may lead to impotence, consultation to urology is the most appropriate next step in this patient's management.
6. D. Acute vasoocclusive events, or painful "crisis," are the most common complication of SCD and are the most frequent cause of ED visits. Pain episodes result from the obstruction of blood flow in the microcirculation leading to tissue ischemia and microinfarction. Vasoocclusion occurs via a combination of sickle cell interactions with endothelial cells and obstruction from nondeformable sickled cells. On average, patients with SCD experience 0.8 hospitalizations per patient-year, but 5% of patients have frequent pain crises and account for approximately one-third of all medical contacts for painful crisis. Pain should be assessed at least every 30 minutes using standardized pain scales. Pain relief is achieved with a variety of analgesics, depending on the severity of the crisis and what the patient has required to manage past crises. Oral agents such as acetaminophen, nonsteroidal anti-inflammatory drugs (NSAIDs), and codeine used separately or in combination are the mainstays of treatment for mild to moderate pain. Parenteral opioid administration is given in conjunction with oral pain relievers for severe pain. If adequate pain relief is not

achieved, the patient is admitted for further pain control. Analgesia should be provided at frequent regular time intervals to avoid breakthrough pain. As needed (PRN) pain medications should be avoided in a crisis.

7. A. This patient is suffering from acute infection with Parvovirus B19. Infections with this virus cause transient red cell aplasia. In normal children whose RBC lifespan is 120 days, brief marrow suppression will not lead to a significant drop in Hgb. However, the shortened lifespan of the RBC in sickle cell anemia (10–25 days) may lead to a significant drop in hemoglobin with even a short period of red cell aplasia. Patients may present with fever, upper respiratory symptoms, nausea and vomiting, arthralgias, or myalgias, but may also have minimal symptoms. Fatigue, pallor, tachycardia, and tachypnea may be present with severe anemia. A CBC will reveal a drop in the hemoglobin from baseline and a decreased reticulocyte count (usually $<1\%$), compared to the normally elevated reticulocyte count seen in sickle cell anemia at baseline. Patients presenting in the recovery phase may have a high reticulocyte count and numerous nucleated RBCs noted on the blood smear. Mildly anemic and asymptomatic children can be managed with supportive care and close outpatient observation pending marrow recovery, symptomatic patients may require simple blood transfusion. Acute blood loss is not supported by the history in this patient. Although ASSC and VOC are both possible etiologies for increased anemia in patients with SCD, neither is supported by history, physical exam, or laboratory findings. Finally, marrow infiltration with leukemia is not supported by the laboratory findings.

101 BLEEDING DISORDERS

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HEMOPHILIA

- X-linked recessive disorder.
- Deficiency of factor VIII (hemophilia A) or factor IX (hemophilia B).
- Percentage of factor present determines the severity of disease.
 - Five to 25% denotes mild disease:
 - no tendency for spontaneous hemorrhage
 - bleeding occurs usually only with surgery or severe trauma.

- Two to 5% denotes moderate disease
 - bleeding with mild trauma.
- Less than 1% denotes severe disease
 - proclivity to spontaneous hemorrhage.
- Two-thirds of male patients with hemophilia have severe disease.
- In both hemophilia A and B
 - prothrombin time (PT) is normal
 - partial thromboplastin time (PTT) is prolonged.
- Same types of bleeding occur in factors VIII and IX deficiency
 - bruising, hemarthroses, and intramuscular hematomas predominate
 - intracranial hemorrhage is less common but can be devastating.

ACUTE HEMARTHROSIS

- Knees, elbows, ankles, hips, and shoulders are the most commonly affected joints.
- Older patients may report pain prior to onset of swelling.
- Younger patients may present with new onset limp or limited range of motion.
- Even if joint bleeding cannot be confirmed, treatment is indicated based on the potentially crippling sequelae of hemarthrosis.
 - Intra-articular bleeding causes synovial inflammation, erosion of the cartilage, synovial hypertrophy, and friability.
 - Muscle atrophy around the joint leads to instability
 - increases the likelihood of more frequent hemarthroses.
 - Repeated bleeding into a “target joint” can lead to complete cartilaginous destruction
 - causes secondary osteoarthritis.
- Persistent joint swelling associated with fever may indicate a septic joint
 - aspiration preceded by appropriate factor replacement may be necessary
 - joint aspiration is not recommended for most cases of bleeding.
- Treatment of hemarthroses.
 - Splinting, ice, immobilization, elastic bandages, and analgesia with acetaminophen with or without codeine.
 - A single factor infusion to raise levels to 30–50%
 - A joint that has bled repeatedly may require several doses of factor.
 - Begin range of motion and physical therapy as soon as possible.
- Bleeding into the hip is especially worrisome.
 - Pressure within the joint can lead to aseptic necrosis of the femoral head.
 - Factor replacement to 80–100% levels with subsequent daily replacement to 50% may be necessary.

INTRAMUSCULAR BLEEDING

- Affects the large weight-bearing muscles
 - Iliopsoas, calf, gluteal, and forearm muscles, but can affect any muscle of the body.
- Bleeding occurs slowly over extended periods of time prior to onset of symptoms.
- Often present as a large hematoma.
- Treatment requires factor replacement to a level of 30–50%.
- Forearm, calf, and hand bleeding can result in a compartment syndrome.
- Classic triad of iliopsoas hemorrhage (which can be massive)
 - Flexion of the thigh
 - Groin, or iliac fossa pain
 - Paresthesias along the anterior thigh
 - secondary to femoral nerve compression by the swollen iliopsoas muscle as it passes under the anterior ligament;
 - Ultrasound or computed tomography (CT) will confirm the diagnosis.
- Compartment syndromes and psoas hemorrhages are treated with correction to achieve factor levels of 80–100%
 - Require admission for observation and continued factor replacement.

INTRACRANIAL HEMORRHAGE

- May be traumatic or spontaneous
 - Minor trauma may present with neurologic changes days after the event.
- Symptoms may include headache, lethargy, loss of consciousness, vomiting, and seizures
- Forceful blows to the head, regardless of symptoms, are empirically treated with factor replacement.
- If intracranial hemorrhage is suspected, immediate factor replacement to a 100% level is necessary
 - Factor infusion should not be delayed for imaging studies.

OTHER BLEEDING MANIFESTATIONS

- Subcutaneous hemorrhage, abrasions, and lacerations that do not require sutures do not require factor replacement.

- Factor replacement is necessary prior to laceration repair, lumbar puncture, surgery, and dental extractions.
- Males with hemophilia can present with painless, gross hematuria
 - an anatomic source is often not found;
 - treatment with factor may or may not be necessary.
- Intramuscular injections, aspirin, and jugular and femoral venipuncture are avoided in this patient population.
- Simple peripheral venipuncture is followed by at least 5 minutes of pressure to the site.

MANAGEMENT ISSUES

- Factor replacement is accomplished by transfusion of factor concentrates.
- Products are made from either pooled donor plasma or recombinant proteins
 - Recombinant products may contain human albumin and are not necessarily superior.
- The amount of factor to be delivered depends on the nature and severity of the bleeding
 - minor bleeding: target factor level is 30–40%
 - major bleeding or prior to surgery a minimum target factor level of 50% is required.
 - life- or limb-threatening bleeds: target factor level of 80–100% is needed
 - Treatment with factor replacement is required every 12 hours or by continuous infusion until healing occurs.
- Patients often present with their home supply of factor and this should be utilized
 - Always give an entire vial of factor even if it results in a higher than calculated dose per weight.
- Approximately 20% of patients with severe factor VIII deficiency develop an inhibitory IgG antibody against factor VIII
 - Treatment can be problematic, as the infused factor VIII is neutralized by the circulating antibody
 - Treatment of bleeding in these children depends on inhibitor titer and the severity of the bleeding
 - Low titers and serious hemorrhage may respond to large doses (up to 100–200 U/kg) of factor VIII.
- Alternatives for treating patients with high titers of inhibitor
 - Prothrombin complexes
 - Bypasses the need for factor VIII through the presence of factors II, VII, and X
 - Recombinant factor VIIa
 - Very short half-life and must be given every 2 hours.
- Purified factor VIII concentrate prepared from pooled plasma donations transmitted hepatitis virus and human immunodeficiency virus (HIV) to 95% of hemophilia patients in the United States in the 1970s and early 1980s
 - A combination of mandatory donor screening and viral attenuation techniques have greatly reduced, but not completely eliminated, viral transmission
 - No cases of HIV-1 transmission from clotting factor concentrates have been documented since 1986.
- Adjuncts to therapy in hemophilia are available in certain situations
 - Corticosteroids: management of hematuria or recurrent joint bleeds
 - Epsilon aminocaproic acid (Amicar) and tranexamic acid (Cyklokapron): clot stabilizers used for the prevention or treatment of oral hemorrhage
 - Desmopressin (DDAVP): increases factor VIII levels in patients with mild hemophilia.

VON WILLEBRAND DISEASE

- Von Willebrand factor (vWf)
 - the carrier protein in plasma for factor VIII
 - acts as a bridge between platelets and subendothelial collagen fibers.
- Von Willebrand disease exists when there are decreased levels of or defective vWf proteins.
- Transmitted as an autosomal dominant trait showing variable expression and penetrance.
- Most patients present as young adults.
- Clinical manifestations include epistaxis, easy bruising, menorrhagia, prolonged oozing from superficial cuts, bleeding after dental extraction, and posttraumatic or postsurgical hemorrhage.
- Hemarthroses are uncommon.
- Many exhibit no clinical problems with bleeding in spite of biochemical abnormalities.
- Laboratory findings: a normal PT and platelet count, a prolonged bleeding time, PTT may be normal or prolonged
 - Measurement of antigenic vWf (vWf:Ag) and ristocetin cofactor (vWf R:Co) activity are decreased in most patients.
- Approximately 80% of patients have Type I von Willebrand disease
 - Often amenable to DDAVP therapy
 - DDAVP stimulates the endogenous release of vWf
 - dose of 0.3 µg/kg intravenously infused over 20–30 minutes
 - can be repeated every 4–6 hours for continued bleeding

- *stimate* (1.5 mg/mL): a concentrated intranasal preparation of desmopressin that has demonstrated effectiveness (dose in teens and adults: 150 µg/nostril, with a total dose of 300 µg; dose in children <50 kg: total dose is 150 µg).

ACQUIRED COAGULOPATHIES

- Include Vitamin K deficiency, liver disease, disseminated intravascular coagulation (DIC), thrombocytopenia, and platelet dysfunctions.
- Vitamin K deficiency decreases the levels of vitamin K-dependent factors (II, VII, IX, and X) resulting in prolongation of the PT
 - Can be seen in malabsorption syndromes, biliary obstruction, or prolonged diarrhea.
 - Children with poor nutrition who receive broad-spectrum antibiotics are also at risk.
 - Drugs may cause vitamin K deficiency as a side effect
 - diphenylhydantoin, phenobarbital, isoniazid, and Coumadin
 - Can lead to hemorrhagic disease of the newborn unless supplementation is provided routinely at delivery
 - Administration of vitamin K is safest by the subcutaneous route
 - rare but severe anaphylactoid reactions are described with intravenous infusion
 - dose of 10 mg of vitamin K given subcutaneously will correct the PT within 24 hours.

DISSEMINATED INTRAVASCULAR COAGULATION (DIC)

- Characterized by simultaneous activation of coagulation and fibrinolysis within the microvasculature
 - microthrombi form in small blood vessels, leading to vessel occlusion, tissue ischemia, and end organ damage
 - excessive bleeding occurs due to thrombocytopenia, consumption of clotting factors, and fibrinolysis.
- Leading cause in pediatric patients is overwhelming infection.
- Conditions that can precipitate DIC
 - Tissue injuries: burns, multiple trauma, crush injuries, and severe head trauma;
 - Abruption placenta and eclampsia
 - Tumors
 - Hemolytic transfusion reactions
 - Myocardial infarctions
 - Giant hemangiomas

- Respiratory distress syndrome
- Snake bites
- Heat stroke or hypothermia.
- Common ischemic complications include hemorrhagic necrosis of the skin, renal failure, seizures, coma, hypoxemia, and pulmonary infarcts.
- Laboratory abnormalities include the following
 - Hemolytic anemia with schistocytes
 - Thrombocytopenia
 - Prolonged PT and PTT
 - Decreased levels of factor V, factor VIII, and fibrinogen
 - Increased fibrin split products
 - Marked decrease in protein C, protein S, and anti-thrombin III.
- Therapeutic options to correct coagulation abnormalities
 - factor replacement with fresh frozen plasma (10–20 mL/kg) to keep the PT in the normal range
 - cryoprecipitate increases the concentration of fibrinogen, factor VIII, and vWf
 - platelet transfusion to keep platelet counts >50,000.

PLATELET DISORDERS

- Platelet activation, adherence, recruitment, and aggregation and binding of fibrinogen result in the cellular clot
 - Responsible for primary hemostasis following a disruption of a vessel wall.
- Deficit in number or function can lead to excessive bleeding.
- Congenital platelet dysfunction can affect a variety of platelet functions
 - receptors, platelet–vessel wall adhesion, and platelet–platelet interactions
 - typically present with severe bleeding diatheses early in life
 - even minor platelet dysfunction can result in easy bruisability and significant bleeding from mucosal membranes.
- Acquired platelet dysfunction is most commonly caused by aspirin
 - Inhibits production of thromboxane A₂ causing decreased platelet aggregation and vessel constriction.
- Deficits in platelet number are more common in pediatric patients.
- Thrombocytopenia is defined as a platelet count less than 150,000/µL
 - rare to develop any abnormal bleeding with counts greater than 50,000/µL

- counts below 20,000/ μ L indicate severe thrombocytopenia
 - increased risk for life-threatening hemorrhage and intracranial bleeding.
- Thrombocytopenia is often an unexpected finding on a complete blood count obtained for unrelated reasons.
- Symptomatic patients may present as well-appearing children with a petechial or purpuric rash.
- Extensive ecchymoses in the absence of a history of significant trauma can wrongly suggest child abuse.
- Physical examination should focus on evidence of systemic disorders such as recent weight loss, hypothyroidism, lymphadenopathy, and hepatosplenomegaly.
- Single most common cause of thrombocytopenia in the well-appearing child is immune thrombocytopenic purpura (ITP).
- Other causes include
 - autoimmune diseases, eg, systemic lupus erythematosus
 - anemia and lymphopenia are usually seen
 - secondary immune destruction of platelets from infectious agents
 - hepatitis B and Epstein–Barr viruses
 - sepsis
 - bone marrow infiltration from leukemia, lymphoma, and other malignancies
 - often have associated hepatosplenomegaly, anemia, and abnormalities of the white blood cells
 - cancer chemotherapy agents cause suppression of all cell lines
 - aplastic anemia
 - idiosyncratic immune reactions following administration of various agents
 - commonly valproic acid, phenytoin, and trimethoprim/sulfamethoxazole.

HEMOLYTIC UREMIC SYNDROME

- Presents with a triad of acute renal failure, microangiopathic hemolytic anemia, and thrombocytopenia.
- Thrombocytopenia is usually mild to moderate.
- Typical presentation: a pale, lethargic young child with a prodromal history of a gastrointestinal infection
 - abdominal pain, vomiting, and bloody diarrhea are common
 - acute renal failure and neurologic manifestations may be present.
- Laboratory examination reveals anemia with schistocytes, thrombocytopenia, electrolyte and acid–base disturbances, and elevated serum creatinine.

- Management consists of early dialysis to treat the effects of renal failure
 - also reduce the fluid overload and hyperkalemia associated with the required frequent blood transfusions.

IMMUNE THROMBOCYTOPENIC PURPURA (ITP)

- Most common cause of thrombocytopenia in a well-appearing young child.
- Peak age of diagnosis is 2–4 years.
- Female to male ratio 1:1.
- Children typically have a history of a preceding viral illness.
- Platelet surface is covered with increased amounts of IgG
 - the spleen removes the affected platelets from the circulation
 - platelet production is increased in the bone marrow, but not enough to offset the rapid destruction.
- Patients present with the acute onset of bruising, petechiae, and purpura
 - physical examination is normal other than for skin findings
 - mucosal or gastrointestinal bleeding can occur.
- Most serious complication is intracranial hemorrhage
 - occurs in >0.1–0.5% of patients.
- Diagnosis is likely with a complete blood count revealing thrombocytopenia with normal red and white blood cell numbers and morphology.
- Patients usually do not require hospitalization and can be followed as outpatients.
- 85% of children make a full recovery within 6 months
 - of the 15% with persistently low platelets, bleeding symptoms are rare and splenectomy is rarely needed.
- Treatment is controversial; consultation with a pediatric hematologist is recommended.
- Therapeutic options include corticosteroids, intravenous immune globulin (IVIG), and anti-Rh(D) immunoglobulin (WinRho-SD)
 - corticosteroids or IVIG may promptly increase the platelet count in patients with profound thrombocytopenia
 - both modalities are presumed to block reticuloendothelial destruction of platelets
 - No evidence that treatment diminishes the risk of major bleeding
 - Infusion of anti-Rh(D) immunoglobulin in Rh-positive individuals results in immune clearance

of the antibody-coated red cells and coincident prolonged survival of autoantibody-coated platelets

- Anti-D may only be administered in Rh-positive patients who have a normal hemoglobin level.
- Transfused platelets will be rapidly destroyed due to the immune response
 - not indicated except in cases of life-threatening hemorrhage
 - platelet transfusion along with intravenous gamma globulin and high-dose intravenous steroids are administered for life-threatening bleeding.
 - emergency splenectomy may be required with life-threatening bleeding.

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QUESTIONS

1. A 14-year-old male with severe factor VIII deficiency presents to the emergency department after being hit by a car while he was skateboarding. The car was traveling 20 MPH on a residential street and the patient was wearing a helmet. The patient reports no loss of consciousness, but is complaining of headache and right elbow pain. Vital signs are normal except for mild tachycardia. Primary and secondary surveys reveal mild posterior scalp swelling and significant swelling of the right elbow with limitation in range of motion. What is the next step in management of this patient?
 - A. Obtain head computed tomography to evaluate for intracranial hemorrhage
 - B. Obtain right elbow films to evaluate for elbow fracture
 - C. Consult the hematologist on call for management suggestions
 - D. Immediate factor replacement to achieve 100%
 - E. Obtain laboratory values such as CBC, coagulation studies, and liver function enzymes as part of the trauma evaluation.
2. A 5-year-old male presents to the emergency department with pain and swelling in his right knee after falling down three steps at home. The child is well known in the emergency department as a patient with severe factor VIII deficiency who often presents with the same symptoms localized to the right knee. His family has brought their own supply of factor. What is the next step in management of this patient?
 - A. Factor replacement using family's supply of factor
 - B. Obtain x-rays of the knee to evaluate for fracture
 - C. Consult orthopedics for possible knee aspiration
 - D. Splinting and immobilization of the knee with orthopedic referral in 1 week
 - E. Consultation with the institutions child abuse team
3. A 14-year-old female presents to the emergency department with a chief complaint of prolonged and heavy menstrual bleeding. The patient reports the onset of menarche 2 month prior to presentation. She is currently using 15 pads per day, which she describes as "soaked through," for 10 days. Vital signs reveal mild tachycardia and mild hypotension. Exam is significant for pallor, but otherwise normal. Mother reports that she often experiences heavy menstruation as well. A complete blood count reveals a hemoglobin of 7 g/dL. You institute appropriate therapy with volume resuscitation and blood transfusion. What is the most likely underlying cause of the patient's bleeding?
 - A. Factor VIII deficiency
 - B. Factor IX deficiency
 - C. Disseminated intravascular coagulation (DIC)
 - D. Vitamin K deficiency
 - E. Von Willebrand disease
4. A 3-year-old female presents to the emergency room after being involved in a low-speed motor vehicle collision. Airbags did not deploy. She was restrained properly in a backseat, front facing three point harness car seat. The only damage to the car was to the bumper. There is a significant seat belt mark across her lower abdomen, out of proportion to the severity of the accident. Her vital signs are stable and physical exam otherwise normal. Complete blood count obtained for trauma workup is abnormal only for a platelet count of 65,000/ μ . What is the most likely etiology of the patient's thrombocytopenia?
 - A. Hemolytic uremic syndrome
 - B. Immune thrombocytopenic purpura (ITP)
 - C. Marrow infiltration by leukemic cells
 - D. Aplastic anemia
 - E. Acute blood loss

ANSWERS

1. D. This patient has severe factor VIII deficiency characterized by <1% factor activity. Intracranial bleeding in this population is rare but can be devastating. Intracranial hemorrhage may be traumatic, as with this patient, or spontaneous. Symptoms can include headache, lethargy, loss of consciousness, vomiting, and seizures. Forceful blows to the head, regardless of symptoms, are empirically treated with factor replacement. If intracranial hemorrhage is suspected, immediate factor replacement to a 100% level is necessary. Factor infusion should not be delayed for imaging studies of any type.
2. A. This patient is presenting with acute hemarthrosis in a “target joint”; his right knee. Knees, elbows, ankles, hips, and shoulders are the most commonly affected joints. It is generally agreed that even if joint bleeding cannot be confirmed, treatment is indicated. This philosophy is based on the potentially crippling sequelae of hemarthrosis. Intra-articular bleeding provokes a strong synovial inflammatory reaction causing erosion of the cartilage, synovial hypertrophy, and friability. Muscle atrophy around the joint leads to instability, which increases the likelihood of more frequent hemarthroses. Unless treated early and adequately, repeated bleeding into a “target joint” can lead to complete cartilaginous destruction causing secondary osteoarthritis. Radiographic evaluation of the joint can be performed following factor replacement. Joint swelling that is persistent and associated with fever may indicate a septic joint. However, this patient has no fever and a known history of trauma to the area. Joint aspiration is not recommended for most cases of bleeding. Symptomatic treatment of hemarthroses consists of splinting, ice, immobilization, elastic bandages, and analgesia with acetaminophen with or without codeine. A single factor infusion to raise levels to 30–50% is usually sufficient to terminate bleeding. A joint that has bled repeatedly may require several doses of factor. Range of motion and physical therapy are instituted as soon as possible. Bleeding into the hip is especially worrisome because pressure within the joint can lead to aseptic necrosis of the femoral head. Factor replacement to 80–100% levels with subsequent daily replacement to 50% may be necessary. This injury, given the child’s underlying diagnosis and history of trauma, does not warrant and evaluation by the institution’s Child Abuse team.
3. E. Von Willebrand disease exists when there are decreased levels of or defective vWf proteins. vWf is the carrier protein in plasma for factor VIII and it also acts as a bridge between platelets and subendothelial

collagen fibers. Unlike the sex-linked hemophilias, von Willebrand disease is typically transmitted as an autosomal dominant trait showing variable expression and penetrance; hence, the mother having similar symptoms, but to a lesser degree. Most patients present as young adults with clinical manifestations including epistaxis, easy bruising, menorrhagia, prolonged oozing from superficial cuts, and bleeding after dental extraction. Many people exhibit no clinical problems with bleeding in spite of biochemical abnormalities. Hemophilia is an X-linked recessive disorder of coagulation caused by deficiency of factor VIII (hemophilia A) or factor IX (hemophilia B). The majority of affected patients are male and present in infancy.

DIC is an acquired syndrome characterized by simultaneous activation of coagulation and fibrinolysis within the microvasculature. Excessive bleeding occurs due to thrombocytopenia, consumption of clotting factors, and fibrinolysis. In pediatric patients, the leading cause of DIC is overwhelming infection. Vitamin K deficiency is not the etiology of the patient’s bleeding as in general, she is an otherwise healthy teen. She is not malnourished and does not suffer from a chronic malabsorption syndrome which would inhibit her ability to obtain adequate vitamin K from her diet.

4. B. Immune thrombocytopenic purpura (ITP) is the most common cause of thrombocytopenia in a well-appearing young child. The peak age of diagnosis is 2–4 years, occurring equally in female and male patients. Children typically have a history of a preceding viral illness, although the link to the development of antiplatelet antibodies is not clear. Patients present with the acute onset of bruising, petechiae, and purpura. They have normal physical examinations other than skin findings. The diagnosis of ITP is likely when the complete blood count reveals thrombocytopenia in association with normal red and white blood cell numbers and morphology. The natural history of the condition is that 85% of children make a full recovery within 6 months. Of the 15% with persistently low platelets, bleeding symptoms are rare and splenectomy is rarely needed. Hemolytic uremic syndrome presents with a triad of acute renal failure, microangiopathic hemolytic anemia, and thrombocytopenia. The typical presentation is that of a pale, somewhat lethargic young child with a prodromal history of a gastrointestinal infection. Marrow infiltration and aplastic anemia would both cause abnormalities in other cell lines. Acute blood loss is possible in the setting of trauma; however, in this patient it was noted that her complete blood count was normal except for thrombocytopenia.

102 BLOOD COMPONENT THERAPY

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INTRODUCTION

Transfusion of blood and blood components is often necessary in the emergency department (ED). Whole blood, packed red blood cells (PRBCs), platelets, granulocytes, fresh frozen plasma (FFP), cryoprecipitate, specific clotting factors, albumin, and immunoglobulins each have specific indications and risks associated with their use. As blood for transfusion is a scarce commodity, the component that will specifically address the patient's need is generally transfused.

BLOOD AND BLOOD COMPONENTS

WHOLE BLOOD

- Transfusion is rarely performed.
- May be indicated for prompt restoration of red cells and volume after trauma or surgery.
- Activity of certain components such as platelets, granulocytes, and clotting factors V and VIII is diminished following 24 hours of storage.
- High risk of transfusion reactions due to the large volume of foreign proteins and antibodies contained within whole blood.

PACKED RED BLOOD CELLS

- Contain ~30–50 mL of plasma and have a hematocrit ranging from 55% to 80%, depending on the preservative used for storage.
- Units are stored for up to 42 days.
- Contains no functional platelets or granulocytes.
- Filtered, leukocyte-poor red cells are recommended for patients with a previous history of febrile reactions to transfusions or to avoid contamination by cytomegalovirus (CMV) transmission.
- Leukocyte depleted preparations contain $<10^7$ leukocytes per unit.

PLATELET CONCENTRATE

- Preparations contain $\sim 5.5 \times 10^{10}$ platelets in 50 mL of plasma.

- Crossmatching is not necessary.
- 0.1–0.2 U/kg of donor platelets are required to raise the platelet count by 50,000–100,000/ μL .
- Indicated for patients with thrombocytopenia or platelet dysfunction who are actively bleeding.
- Counts above 20,000/ μL rarely result in spontaneous bleeding; counts below 10,000/ μL , increase the risk of spontaneous bleeding.
- Immune thrombocytopenia or thrombotic thrombocytopenic purpura do not benefit from platelet transfusions except in cases of life-threatening hemorrhage; the ongoing antibody-mediated disease process quickly destroys the transfused platelets.

GRANULOCYTE CONCENTRATES

- Transfusion is indicated only in a severe, prolonged neutropenic patient with documented or strongly suspected antibiotic resistant sepsis.

FRESH FROZEN PLASMA

- Consists of pro-coagulant clotting factors such as V and VIII, the anticoagulants protein S, protein C, and antithrombin III.
- Crossmatching is not necessary.
- Table 102-1 reviews the indications for use of FFP.
- 10–20 mL/kg infused to gravity, not indicated for acute volume expansion.
- Risk of disease transmission is similar to that of whole blood transfusion.
- Allergic reactions are possible.

CRYOPRECIPITATE

- A purely pro-coagulant preparation rich in fibrinogen, factor VIII, vWf, and factor XIII.
- Does not require crossmatching.
- Used for treatment of hypo- or a-fibrinogenemia.
- One unit of cryoprecipitate is given per 5 kg of body weight.

TABLE 102-1 Indications for Use of Fresh Frozen Plasma

- Disseminated intravascular coagulopathies
- Acute blood loss (trauma)
- Unknown factor deficiencies
- Chronic liver disease
- Vitamin K deficiency
- Treatment of excessive warfarin or dicumarol therapy

FACTORS VIII AND IX

- Highly purified concentrates of factors VIII and IX produced by monoclonal antibody techniques and by recombinant DNA technology.
- Greatly diminishes the risk of infectious disease transmission.
- Used in the treatment of factor VIII and factor IX deficiencies.

ALBUMIN

- Most frequently used for blood volume expansion in shock, trauma, burns, and surgery.
- Available in 5% and 25%.
- The 5% solution is iso-osmotic with plasma.
- The 25% solution is never used to treat shock without other fluids.

IMMUNE GLOBULINS

- Antibody-rich preparations used as postexposure disease prophylaxis to treat conditions such as rabies and tetanus.
- A mainstay of therapy in immune-mediated diseases (eg Kawasaki's disease).

INDICATIONS FOR MASSIVE TRANSFUSION

- Refers to the replacement of a patient's total blood volume in >24 hours, or as the acute replacement of more than half the patient's estimated blood volume in any 4-hour period.
- Result in rapid release of O-negative blood (universal donor) and blood products such as platelets and FFP.
- Patient-specific blood typing usually should be done in the hemodynamically stable patient.
 - Hgb >10 g/dL, transfusion is rarely indicated.
 - Hgb <5 g/dL, transfusion is usually necessary.
 - Hgb is between 5 and 10 g/dL; clinical status and risk of ongoing blood loss are helpful in determining transfusion requirements.

COMPLICATIONS OF TRANSFUSION

ACUTE HEMOLYTIC TRANSFUSION REACTIONS (AHTR)

- Patient's anti-A or anti-B antibodies bind to incompatible transfused red cells.

- Immediate reaction from lysis of the transfused cells releasing inflammatory mediators.
- Symptoms: fever, tachycardia, chills, back or flank pain, nausea and vomiting, dyspnea, flushing, abnormal bleeding, and hypotension with possible progression to disseminated intravascular coagulation, shock, renal failure, and death.
- Laboratory findings include hemoglobinemia, and/or hemoglobinuria, an increased serum bilirubin, and a positive direct antibody test.

DELAYED HEMOLYTIC TRANSFUSION REACTIONS (DHTR)

- Sensitization to non-ABO antigens from a previous transfusion.
- Signs and symptoms include unexplained anemia, jaundice, fever, back pain, and rarely, hemoglobine-mia and/or hemoglobinuria.
- Detected 3–14 days after transfusion.
- No treatment is usually required.

FEBRILE NONHEMOLYTIC TRANSFUSION REACTIONS (FNHTR)

- Account for the majority of transfusion reactions.
- Occur most commonly in the multiply transfused patient; patients often report a history of FNHTR.
- Symptoms include fever and chills that may be difficult to distinguish from AHTR.
- No laboratory tests available to predict or prevent these reactions.

ALLERGIC TRANSFUSION REACTION

- Three types of reactions are as follows:
 - Urticarial reactions: They involve allergens, cytokines, or histamine in stored blood products; can progress to anaphylaxis.
 - Anaphylactic reactions: These are severe urticarial reactions that commonly occur in patients with congenital IgA deficiency who have high-titer IgG anti-IgA antibodies.
 - Transfusion-related acute lung injury (TRALI): This occurs when the permeability of the pulmonary microvasculature is acutely increased, leading to massive pulmonary edema, usually within 6 hours of transfusion.
 - Thought to be related to the presence of granulocyte antibodies in either the donor product or the recipient.

TABLE 102-2 Complications of Massive Transfusion Therapy

- Dilutional thrombocytopenia
- Citrate-induced hypocalcemia
- Hyperkalemia
- Hypokalemia
- Acid/base disturbances
- Acute respiratory distress syndrome

COMPLICATIONS OF MASSIVE TRANSFUSIONS

Table 102-2 summarizes complications that may occur when large amounts of whole blood or PRBCs are given rapidly.

INFECTIOUS COMPLICATIONS

- Estimated risk of transmitting HIV through a blood transfusion is 1 in 2,135,000 units transfused; hepatitis B, 1 in 205,000 units transfused; and hepatitis C, 1 in 1,935,000 units transfused.
- Bacterial contamination of blood products can occur and accounts for other transfusion reactions and fatalities.
- Symptoms include fever, chills, rigor, vomiting, and hypotension present soon after the transfusion is begun.
- AHTR is in the differential if the patient is receiving red blood cells.

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QUESTIONS

1. A 2-year-old female is involved in a high-speed rollover motor vehicle collision. She presents to the emergency department (ED) with tachycardia and hypotension. Multiple deep lacerations are noted on her abdomen and lower extremities. Her hemoglobin measures 6 g/dl. What is the most appropriate blood product to transfuse?
 - A. Fresh frozen plasma (FFP)
 - B. Whole blood
 - C. Packed red blood cells (PRBCs)
 - D. Platelets
 - E. Albumin
2. A 7-year-old female is involved in a fall from a two-story window and presents to the emergency department unresponsive and in hemorrhagic shock. To appropriately treat this patient, the ED attending should do which of the following?
 - A. Rapidly transfuse whole blood
 - B. Rapidly transfuse PRBCs
 - C. Rapidly transfuse platelets
 - D. Rapidly transfuse FFP
 - E. Call for massive transfusion protocol to be initiated releasing O-negative PRBCs, platelets, and FFP for transfusion
3. A 7-year-old male with sickle-cell anemia is in the emergency department receiving a blood transfusion for splenic sequestration crisis. His measured hemoglobin is 6 g/dL. Five minutes after initiation of transfusion the patient develops a fever, tachycardia, hypotension, nausea, and vomiting. STAT laboratory values reveal a hemoglobin of 4 g/dl and elevated liver function enzymes. The cause of this reaction is which of the following?
 - A. Allergic reaction to the blood products
 - B. Delayed hemolytic transfusion reaction (DHTR)
 - C. Further complication of splenic sequestration crisis
 - D. Acute hemolytic transfusion reaction (AHTR)
 - E. Febrile nonhemolytic transfusion reaction (FNHTR)
4. A 3-year-old female develops a severe urticarial reaction, wheezing, and hypotension upon receiving a blood transfusion for aplastic anemia. The initial therapy indicated following cessation of transfusion is which of the following options?
 - A. Oral Benadryl
 - B. Nebulized albuterol
 - C. Intravenous infusion of 20 cc/kg isotonic fluid
 - D. Intramuscular injection of 0.01 mg/kg of 1:1000 epinephrine
 - E. Intravenous infusion of 1 mg/kg solumedrol

ANSWERS

1. C. PRBCs are the most appropriate therapy in the setting of the unstable trauma patient. Units of PRBCs contain a hematocrit of 55–80% and are effective in improving the hemoglobinemia due to acute hemorrhage. PRBCs are also effective volume expanders and will improve both the tachycardia and hypotension. FFP is used in situations of acute blood loss to replace lost clotting factors. While this patient will eventually need FFP, in the initial resuscitation, she requires volume expansion. FFP is not used for volume expansion and correction of hemoglobinemia due to acute hemorrhage. Whole Blood can be used in this situation; however, the risk of complications is high and better products are available. Platelet concentrates do nothing to correct the hemoglobinemia and are not used as volume expanders. Although this patient will require platelet transfusion, this is not the appropriate first line intervention. Albumin in the 5% concentration is an effective volume expander and will improve both the hypotension and tachycardia. However, it will not improve the measured hemoglobin and oxygen-carrying capacity.
2. E. Massive transfusion is defined as the replacement of a patient's total blood volume in >24 hours, or as the acute replacement of more than half the patient's estimated blood volume in any 4-hour period. The most common scenario in the emergency department requiring massive transfusion is the unstable trauma patient. These patients suffer from hemorrhagic shock requiring both volume support with PRBCs and coagulation support with platelets and FFP. Whole blood can be used in this situation as it does contain all blood components. However, these components have decreased activity after 24 hours of storage. There also exists a high risk of transfusion reactions with whole blood. Answers B–D are all correct but none alone address the entire needs of the patient.
3. D. This scenario describes an acute hemolytic transfusion reaction. Answer A is possible; however, allergic reactions do not cause a decline in hemoglobin and elevation of liver function enzymes. Answer B, a delayed hemolytic transfusion reaction, is caused by sensitization to non-ABO antigens from a previous transfusion. This reaction may cause the symptoms described; however, it is usually detected 3–14 days following transfusion. Answer C, further progression of splenic sequestration crisis can cause a decline in hemoglobin as well as the tachycardia and hypotension described. However, the acute onset of symptoms and the increase in liver function enzymes

make this less likely. Answer E, a febrile nonhemolytic transfusion reaction, can be difficult to distinguish from an acute hemolytic transfusion reaction in the initial presentation. However, FNHTR are not associated with hemolysis.

4. D. This patient is having an anaphylactic reaction to the infusion of blood products. While answers A, B, C, and E may be needed, the first line therapy is the administration of intramuscular injection of 1:1000 epinephrine.

103 ONCOLOGIC EMERGENCIES

Audra L. McCreight

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COMMON PEDIATRIC MALIGNANCIES

ACUTE LEUKEMIAS

- Leukemia is caused by uncontrolled, clonal proliferation of immature white blood cells within the bone marrow, with subsequent suppression of normal hematopoiesis.
- Acute leukemia is the most common childhood malignancy
 - acute lymphoblastic leukemia (ALL) accounts for approximately 75% of pediatric leukemia
 - peak incidence between the ages of 3 and 5 years
 - 75–80% survival >5 years beyond diagnosis
 - acute myelogenous leukemia (AML) accounts for 25%
 - incidence is relatively constant throughout childhood
 - 50% survival at 5 years from diagnosis
- Chronic leukemia: rare in pediatrics
 - chronic myelogenous leukemia (CML): <1% of all childhood cancers.
- Common presenting symptoms
 - pallor, fatigue, petechiae, purpura, and infection
 - result from defective hematopoiesis from marrow replacement
 - lymphadenopathy, hepatomegaly, splenomegaly, and mediastinal or testicular masses
 - may represent extramedullary involvement
 - bone pain
 - leukemic involvement of the periosteum and bone
 - patients may limp or even refuse to walk
 - cranial nerve deficits, headache, or changes in vision
 - presence of leukemic cells in the CNS.

- Nonspecific symptoms such as anorexia, fever, and irritability.
- The leukocyte count may be high or low.
- Patients should be considered immunocompromised at diagnosis regardless of the leukocyte count
 - white blood cells produced may not be functional.
- Most will be anemic and/or thrombocytopenic.
- Diagnosis confirmed by bone marrow aspiration.

HODGKIN DISEASE

- Malignancy of the lymph nodes.
- Reed–Sternberg cell is considered to be the malignant cell.
- Approximately 5% of pediatric malignancies
 - peak incidence in adolescents and young adults.
- Majority have painless supraclavicular or cervical lymphadenopathy
 - rubbery, matted, and do not decrease in size.
- Abdominal examination may reveal hepatomegaly or splenomegaly.
- Mediastinal involvement is common
 - obtain chest radiograph for any patient suspected of having Hodgkin disease.
- Systemic or “B” symptoms may occur in one-third of the patients
 - unexplained fever, weight loss, and night sweats.
- The differential diagnosis includes
 - infectious mononucleosis, mycobacterial infections, or other metastatic malignancies.
- Treatment regimens include multidrug chemotherapy and/or radiation.
- Five-year survival rates of >90%.

NON-HODGKIN LYMPHOMAS (NHL)

- A heterogeneous group of malignancies of lymphatic tissue.
- No peak age incidence
 - cases increase steadily with age.
- Rapidly proliferating.
- Most common types seen in children
 - Burkitt’s lymphoma
 - most often present with abdominal involvement causing pain, nausea, vomiting, distension, ascites, or bowel obstruction
 - right lower quadrant pain reflects distal ileal, appendiceal, or cecal involvement; may mimic appendicitis
 - may be a lead point for an intussusception
 - Burkitt’s-like lymphoma
 - large B cell lymphoma

- lymphoblastic lymphoma
 - often present with a mediastinal mass or supraclavicular adenopathy
 - symptoms may include cough, wheeze, chest pain, airway obstruction, or signs and symptoms of superior vena cava obstruction
- anaplastic large cell lymphoma (ALCL).
- Patients often require emergency management at presentation
 - due to the rapid doubling time and growth rate of the tumor (Burkitt’s lymphoma)
 - due to tumor mass encroachment on vital structures (lymphoblastic lymphoma).
- Laboratory evaluation
 - CBC to assess for marrow involvement
 - electrolytes, blood urea nitrogen (BUN), creatinine, calcium, phosphorous, and uric acid to evaluate for tumor lysis syndrome
 - chest radiograph may reveal a mediastinal mass
 - abdominal masses are evaluated with abdominal ultrasound or computed tomography (CT) scan.
- Up to 80% or higher long-term, disease-free survival depending on histologic type.

CENTRAL NERVOUS SYSTEM TUMORS

- Represent the second most common pediatric cancer diagnosis.
- Classification of CNS tumors is based on histologic type
 - supratentorial tumors include cerebral astrocytoma, optic glioma, and craniopharyngioma
 - more commonly occur in neonates and infants
 - may cause headache, seizures, or visual impairment
 - infratentorial tumors include cerebellar astrocytoma, medulloblastoma, ependymoma, and brain stem glioma
 - more commonly seen after 2 years of age
 - truncal ataxia or incoordination is a typical finding.
- Impingement of the brain stem may lead to cranial nerve palsies or Horner’s syndrome.
- Raised intracranial pressure (ICP) may manifest as vomiting, anorexia, irritability, developmental regression, impaired upward gaze (“sunsetting” sign), or a change in behavior or personality.
- Headaches that are recurrent, intense, associated with vomiting, that awaken patients from sleep should raise the suspicion of a malignancy.
- Back pain, bladder, or bowel dysfunction, or focal neurologic deficits may suggest spinal cord or cauda equina involvement.

- Differential diagnosis of raised ICP or neurologic deficits
 - brain abscess, chronic subdural hematoma, and vascular malformations.
- Can be diagnosed by CT
 - magnetic resonance imaging (MRI) is more sensitive.

WILMS' TUMOR

- The peak age of diagnosis is 2–3 years
 - most cases diagnosed before 5 years of age.
- Most appear well at diagnosis with a nontender abdominal mass
 - systemic symptoms are rare
 - hematuria is rare
 - if present is usually microscopic.
- Rare cases are associated with an underlying genetic predisposition syndrome
 - Beckwith–Wiedemann, Denys–Drash, or Wilms' tumor, aniridia, genitourinary anomalies, and mental retardation (WAGR).
- Bilateral at diagnosis in 5–10% of cases.
- Initial evaluation
 - CBC
 - urinalysis
 - imaging of the chest and abdomen
 - ultrasound or CT will often reveal a large, encapsulated mass arising from the kidney

NEUROBLASTOMA

- Neuroblastoma is a malignant tumor arising from neural crest cells
 - can originate anywhere along the sympathetic chain or the adrenal medulla.
- Most common extracranial solid tumor in childhood
 - almost all cases diagnosed before 5 years of age.
- Two-thirds arise in the abdomen and pelvis
 - may present as an abdominal mass.
- Tumor manifestations may include
 - impingement of renal vasculature leading to renin-mediated hypertension
 - Horner's syndrome with cervical or high thoracic tumors
 - unilateral ptosis, miosis, and anhidrosis
 - spinal cord or nerve root compression
 - paraspinal ganglia tumors growing around and through the intervertebral foramina
 - retrobulbar involvement can cause proptosis or periorbital ecchymosis

- massive hepatomegaly from liver involvement can cause respiratory compromise or liver failure
 - more common in infants.
- Opsoclonus-myoclonus
 - a paraneoplastic syndrome
 - characterized by myoclonic jerking and random eye movements
 - seen in a small percentage of patients at diagnosis
 - may or may not resolve following treatment.
- Imaging with abdominal ultrasound or CT scan may reveal a suprarenal mass.
- Catecholamine metabolites homovanillic acid and vanillylmandelic acid are elevated
 - detectable in the urine >90% of patients.

MUSCULOSKELETAL TUMORS

- Osteosarcoma, Ewing's sarcoma, and rhabdomyosarcoma.
- Rhabdomyosarcoma: a malignant solid tumor of striated muscle tissue
 - may arise anywhere
 - most often presents as a painless mass.
- Osteosarcoma: most common malignancy of bone
 - predilection for the metaphysis of long bones
 - distal femur and proximal tibia
 - presents with bone or joint pain, often after an injury
 - may also present with a palpable mass or pathologic fracture.
- Ewing's sarcoma occurs equally between long bones and flat bones
 - presents with bone or joint pain, often after an injury
 - may present with a pathologic fracture
 - may also present as a soft tissue mass without bone involvement.
- Evaluation with plain radiographs should be performed first.

RETINOBLASTOMA

- Most common intraocular tumor of childhood.
- Strongly linked to deletions of the RB1 gene on chromosome 13.
- 30% of cases are bilateral.
- Most present with leukocoria or strabismus.
- CT or MRI is needed to determine the presence of extraocular involvement.
- Unilateral disease is treated with enucleation.

COMMON COMPLICATIONS OF CHILDHOOD CANCER

METABOLIC EMERGENCIES

TUMOR LYSIS SYNDROME (TLS)

- Results from the death of tumor cells and release of intracellular contents into circulation
 - causes hyperuricemia, hyperphosphatemia, and hyperkalemia.
- Occurs most often with hematologic malignancies
 - particularly Burkitt's lymphoma and T-cell lymphoma or leukemia.
- Rare with nonlymphomatous solid tumors.
- Usually begins within the first few days following treatment initiation.
- Breakdown of released nucleic acids from tumor cells leads to hyperuricemia:
 - uric acid can crystallize within the renal tubules
 - leads to obstruction, oliguria, and renal failure
- Release of intracellular potassium leads to hyperkalemia
 - exacerbated by declining renal function
- Hyperphosphatemia can lead to secondary hypocalcemia
 - may cause tetany, seizures, and arrhythmias
 - precipitation of calcium phosphate crystals in the renal tubules leads to renal failure.
- All patients with possible TLS require the following studies
 - CBC
 - electrolytes, BUN, creatinine, and glucose
 - calcium, magnesium, and phosphate
 - lactate dehydrogenase and uric acid
 - an electrocardiogram if hyperkalemia is found
 - hydration is the most important initial intervention
 - intravenous fluid is administered at a minimum of twice the patient's maintenance rate with 5% dextrose in 1/4 normal saline with 40 mEq of sodium bicarbonate per liter
- goal is a urine pH of 7.0–7.5
 - urine alkalization increases uric acid solubility and excretion
 - over alkalization may lead to crystallization of calcium phosphate in the kidneys
- fluids should not contain potassium unless symptomatic hypokalemia exists.
- Allopurinol inhibits xanthine oxidase: the enzyme that promotes the degradation of purine to uric acid
 - used to prevent hyperuricemia.
- Intravenous recombinant urate oxidase (0.2 mg/kg)
 - converts uric acid to allantoin
 - may be indicated in the place of allopurinol in patients with an elevated uric acid for
 - tumors of a high proliferative rate such as Burkitt's lymphoma
 - large tumor burden
- Calcium supplementation for hypocalcemia
 - indicated only in patients who are severely symptomatic with a normal serum phosphate
 - additional calcium in the face of hyperphosphatemia may increase the precipitation of calcium phosphate.
- Hyperkalemia may be treated with calcium gluconate, sodium bicarbonate, and insulin along with dextrose (see Table 103-1)
 - sodium polystyrene sulfonate (Kayexalate) should not be an initial choice for hyperkalemia
 - slow onset of action
 - desire to avoid per rectum route of administration in neutropenic patients.
- Dialysis is indicated with persistent oliguria or electrolyte abnormalities that do not correct with medical management.

HYPERCALCEMIA

- May occur with ALL, NHL, neuroblastoma, and Ewing's sarcoma.
- Symptoms include constipation, weakness, polyuria, and drowsiness.

TABLE 103-1 Management of Hyperkalemia

PURPOSE	AGENT	DOSE	RATE OF ADMINISTRATION
Improve membrane stability Shift potassium into cells	Calcium gluconate	1 mL/kg of 10% solution	Intravenous over 10–15 min
	β-agonist	5–20 mg	Nebulized with 5 L oxygen
	Sodium bicarbonate	0.5 mg/kg per dose	Intravenous over 10–15 min
	Glucose	0.5 g/kg	Intravenous over 30 min
	Insulin	0.1 U/kg	
Enhanced elimination	Kayexalate	1–2 g/kg	Per rectum may be contraindicated in immunocompromised patients
	Dialysis		Done in coordination with nephrology

- Treatment begins with intravenous hydration with normal saline, followed by furosemide to promote calcium excretion.

HEMATOLOGIC COMPLICATIONS

ANEMIA

- Transfusion therapy with packed red blood cells (PRBC).
- Hemoglobin <6–8 g/dl: 10–12 mL/kg PRBC given over 3–4 hours.
- Hemoglobin <5 g/dL: require transfusion with multiple smaller aliquots (3–5 mL/kg) to avoid congestive heart failure.
- Patients with signs of fluid overload may be given furosemide.
- Blood products should be irradiated and leukoreduced
 - irradiation helps minimize the occurrence of post-transfusion graft-versus-host disease
 - leukoreduction decreases the occurrence of transfusion reactions as well as the transmission of CMV.

HEMORRHAGE

- May occur due to thrombocytopenia.
- Secondary to leukemia, chemotherapy, or DIC
 - petechiae, bruising, and mucosal bleeding—may be seen with platelet counts <20,000/mm³
 - significant spontaneous hemorrhage is more likely with platelet counts <10,000/mm³.
- Hemorrhage can also be secondary to disseminated intravascular coagulation (DIC)
 - causes a prolongation of the prothrombin time and partial thromboplastin time
 - reduced fibrinogen level
 - thrombocytopenia
 - elevated fibrin degradation products
 - initial management
 - includes treatment of the underlying condition
 - replacement of coagulation factors with fresh frozen plasma (10 mL/kg)
 - platelet and PRBC transfusions as needed.

HYPERLEUKOCYTOSIS

- May be seen with acute leukemia.
- WBCs are larger and not easily deformed, increasing blood viscosity.
- Leukemia cells tend to aggregate and impair tissue perfusion
 - symptoms may include dyspnea, confusion, agitation, or blurred vision

- physical examination may reveal plethora, cyanosis, papilledema, retinal hemorrhage, ataxia, priapism, or focal findings on neurologic examination
- hyperleukocytosis increases the risk for TLS
- thrombocytopenia is corrected to a platelet count of at least 20,000/mm³.
- Significant risk of intracranial hemorrhage with hyperleukocytosis.
- Leukopheresis prior to initiation of chemotherapy may be indicated in patients with symptomatic hyperleukocytosis.

INFECTIOUS COMPLICATIONS

- One of the most common complications in the treatment of children with cancer.
- A significant cause of morbidity and mortality.
- Single most important factor is the development of neutropenia
 - seen 8–16 days post-therapy.
- Best estimate of production of neutrophils is the absolute neutrophil count (ANC):
 - $ANC = \text{total WBC} \times (\% \text{ Bands} + \% \text{ PMNs})$.
- Neutropenic: defined as an ANC <500/mm³ or an ANC <1000/mm³ with predicted decline to <500/mm³.
- Fever: defined as a single oral temperature of $\geq 38.3^\circ\text{C}$ (101°F) or a temperature of $\geq 38.0^\circ\text{C}$ (100.4°F) for ≥ 1 hour.
- Common pathogens are listed in Table 103-2.
- Evaluation
 - careful history and physical examination
 - particular attention is paid to the oropharynx, axillae, groin, perineum, and sites of previous procedures, as well as along the tract of any IVAD
 - fever may be the only positive sign
 - due to the decreased number of neutrophils, the inflammatory response is blunted
 - other findings, such as exudates, adenopathy, fluctuance, warmth, and swelling, may be absent.

TABLE 103-2 Common Pathogens in Children with Cancer

BACTERIA			
GRAM-POSITIVE	GRAM-NEGATIVE	FUNGI	VIRUSES
• <i>Staphylococcus aureus</i>	• <i>Escherichia coli</i>	• <i>Candida</i> species	• HSV
• Coagulase-negative staphylococci	• <i>Klebsiella</i> species	• <i>Aspergillus</i> species	• Varicella
• Alpha-hemolytic streptococci	• <i>Pseudomonas aeruginosa</i>		• CMV
• <i>Enterococcus faecalis</i>			• EBV
			• RSV
			• Influenza

- Initial studies
 - creatinine, CBC, and blood cultures for bacterial and fungal culture
 - blood culture from IVAD if present
 - peripheral culture depending on institution guidelines
 - chest radiograph if respiratory symptoms are present
 - aspirate for Gram stain and culture is sent from any other areas suggestive of focal infection.
- Prompt initiation of empiric antibiotic therapy
 - infection can progress rapidly.
- Admission to the hospital for continuation of intravenous antibiotics
 - combination therapy to provide broad-spectrum antibiotic coverage
 - includes an aminoglycoside and an antipseudomonal beta lactam
 - vancomycin is warranted if there is evidence of intravenous catheter-related infection, methicillin-resistant *Staphylococcus aureus* colonization, severe chemotherapy-induced mucosal damage, fluoroquinolone prophylaxis, recent administration of high dose cytarabine, or septic shock.

FUNGAL INFECTIONS

- High risk for infection with *Candida* and *Aspergillus* species.
- Oral cavity is the most common site.
- May present asymptotically as punctate foci or diffuse erythematous mucosal plaques and ulcerations.
- Difficulty breathing, hoarseness, or stridor often indicates epiglottic or laryngeal candidiasis.
- Afebrile neutropenic patients who are able to tolerate oral medication may be treated with topical antifungal agents.
- Empiric intravenous antifungal therapy is not initially indicated for febrile patients
 - may be added after several days of fever in a persistently neutropenic patient.

VIRAL INFECTIONS

- Herpes simplex virus (HSV) infections tend to be localized
 - commonly involve the mouth, nares, esophagus, genitals, and perianal region
 - pain is the predominant presenting symptom
 - mild mucocutaneous disease may be treated with oral acyclovir
- Moderate or severe disease requires hospital admission for intravenous acyclovir therapy.
- Varicella zoster virus (VZV) infections

- associated with significant morbidity and mortality, high potential for dissemination
- diagnosis is based on the characteristic vesicular lesions and history of recent exposure
- chest radiograph is obtained to assess for pneumonia
- requires admission for intravenous acyclovir.

PARASITIC INFECTIONS

- *Pneumocystis carinii* pneumonia—the most common parasitic infection in immunocompromised patients.
- Children with hematologic malignancies are at the highest risk.
- Presents with fever, dry cough, tachypnea, and intercostal retractions without detectable rales.
- Hypoxia out of proportion to the degree of tachypnea.
- Chest radiograph may be normal in early disease, but progresses to bilateral alveolar infiltrates.
- Empiric therapy with TMP-SMX is initiated pending definitive diagnosis.
- Incidence of infection has been reduced with the use of prophylaxis
 - TMP-SMX, pentamidine, or dapsone.

FEVER IN THE NON-NEUTROPENIC ONCOLOGY PATIENT

- Evaluation is the same as that for the neutropenic patient.
- Non-neutropenic patients remain at risk for infection from an IVAD and therapy-related immune dysfunction outside of neutropenia.
- Hospitalization: warranted in children whose ANC is expected to decline below 500 or in the septic appearing patient.

SUPERIOR VENA CAVA SYNDROME AND SUPERIOR MEDIASTINAL SYNDROME

- Non-Hodgkin lymphoma, Hodgkin lymphoma, neuroblastoma, germ cell tumors, and acute lymphoblastic lymphoma may present with a mediastinal mass.
- Mediastinal compression may result in Superior Vena Cava Syndrome (SVCS) or Superior Mediastinal Syndrome (SMS)
 - SVCS refers to the signs and symptoms resulting from obstruction, compression, or thrombosis of the SVC
 - SMS occurs with compression of the narrow, compliant trachea in children.
- SMS and SVCS often occur together in pediatrics.

- Presenting symptoms may include
 - edema and plethora of the face, conjunctivae, neck, and upper torso
 - headache, papilledema, seizures, coma, cerebral hemorrhage, and engorgement of retinal veins
 - result of cerebral venous hypertension
 - compression of the tracheobronchial tree may cause tachypnea, wheezing, stridor, orthopnea, or cyanosis
- Chest radiography reveals superior mediastinal widening and occasionally a pleural or pericardial effusion.
- Laboratory evaluation
 - CBC with differential may show evidence of leukemia or lymphoma
 - Electrolytes may show evidence of tumor lysis.
- Airway management
 - challenging as complete obstruction may be precipitated
 - difficult if the obstruction is at or below the distal trachea
 - intubation (either tracheal or selective bronchial intubation) or emergent tracheostomy after airway collapse may not be possible.
 - if intubation is achieved, collapse of the airway below the level of the endotracheal tube may lead to inadequate ventilation and life-threatening ventilation perfusion mismatch
 - if intubation is necessary, it is recommended that paralytics not be used
 - computed tomography to determine tracheal cross-sectional obstruction has been suggested as means to judge anesthetic risk
 - patients with a critical mass may not tolerate the supine positioning needed for the CT scan, and should not be sedated.
- Significant respiratory compromise may require the initiation of therapy prior to definitive tissue diagnosis.
- Supportive therapy includes
 - elevation of the head of the bed to minimize cerebral hypertension
 - intravenous hydration
 - upper extremity phlebotomy should be avoided as these veins are under high pressure and may bleed excessively.
- Correction of electrolyte abnormalities and treatment of hyperuricemia should be initiated.

SPINAL CORD COMPRESSION

- May occur with extradural metastatic tumors
 - soft tissue sarcomas, neuroblastoma, germ cell tumors, and Hodgkin disease, or rarely with an intradural cord tumor.

- Pain is the most common initial presenting symptom
 - usually worse when supine
 - may be tender to palpation.
- Symmetric muscle weakness is a later finding.
- Sensory deficits are less common than weakness
 - present with ascending numbness and paraesthesias.
- Changes in bladder or bowel function may occur.
- Hydrocephalus may result from physical obstruction of cerebrospinal fluid flow.
- Plain spine radiographs will show an abnormality in some patients with spinal cord compression
 - an MRI provides a more definitive study
 - should be done immediately in patients with progressive neurologic deficit.
- Spinal cord compression is a true neurologic emergency
 - consultation with an oncologist and neurosurgeon should be obtained immediately.
- Treatment begins with dexamethasone to reduce tumor-related edema.

CENTRAL NERVOUS SYSTEM EMERGENCIES

- Electrolyte abnormalities, hypoxia, renal or hepatic failure, disseminated intravascular coagulation, hyperleukocytosis, and sepsis may lead to altered mental status.
- Primary CNS tumors and metastatic lesions may present with acute mental status changes.
- Cerebrovascular accidents may complicate acute leukemia as a result of thrombosis or hemorrhage
 - hemorrhage can occur at the site of intracerebral metastases
 - subdural and subarachnoid hemorrhage may occur due to thrombocytopenia or coagulopathy
 - thrombosis may occur after CNS irradiation or chemotherapy.
- Seizures may arise from electrolyte abnormalities, infection, metastatic disease, or as a complication of CNS therapy.
- The initial evaluation of a child with a neurologic emergency should include
 - a detailed history and neurologic examination
 - history of recent administration of chemotherapeutic agents, either intravenous, or intrathecal
 - methotrexate, cytarabine, corticosteroids, and ifosfamide may cause neurologic toxicity.
- Laboratory evaluation includes CBC, electrolytes, glucose, creatinine, phosphate, calcium, uric acid, magnesium, blood culture, and coagulation studies.

- A CT scan of the head without contrast should be done to quickly assess for tumor or intracranial bleeding
 - MRI may be performed when the child is stable.
- Treatment
 - support and protection of the airway and breathing
 - endotracheal intubation should utilize medications that do not increase intracranial pressure.
 - if elevated ICP or cerebral edema is suspected
 - Controlled ventilation to a P_{CO_2} of 30–35 mm Hg will cause cerebral vasoconstriction and thereby help reduce cerebral blood flow in an emergency.
 - corticosteroid administration.
 - hyperosmolar agents.
- Prompt neurosurgical consultation is recommended.
- If meningitis is suspected, lumbar puncture is deferred but antibiotics are initiated.
- Correction of thrombocytopenia and coagulopathy.

GASTROINTESTINAL EMERGENCIES

- Esophagitis, typhlitis, enterocolitis, and perirectal abscess may occur as a result of immunosuppression and infection.
- Typhlitis
 - a severe necrotizing colitis of the cecum in neutropenic patients
 - may mimic signs and symptoms of acute appendicitis
 - patients must be started on broad-spectrum antibiotics to cover both Gram-negative pathogens as well as gastrointestinal anaerobes
 - early surgical consultation is recommended
 - indications for laparotomy include evidence of perforation, persistent gastrointestinal hemorrhage despite correction of existing coagulopathies, and clinical deterioration.
- Gastrointestinal hemorrhage
 - increased risk due to the use of high-dose corticosteroids in the treatment of leukemia and lymphoma
 - may also result from thrombocytopenia, coagulopathy, mucosal ulceration, or abnormal tumor vessels.
- Gastrointestinal obstruction
 - caused by tumor mass at presentation, adhesions from previous resection of an abdominal tumor, or paralytic ileus from medications such as vincristine.
- Pancreatitis due to the use of asparaginase.

- Veno-occlusive disease
 - presents with tender hepatomegaly, ascites, weight gain, and hyperbilirubinemia
 - most often occurs during stem cell transplant, but may be a complication of some chemotherapy regimens.
- Common causes of an acute abdomen, such as appendicitis, must also be considered.
- Laboratory evaluation includes a CBC, blood and urine cultures, urinalysis, electrolytes, glucose, amylase, and lipase.
- Abdominal films may reveal bowel obstruction, perforation, or pneumatosis intestinalis
 - abdominal CT may be helpful if plain films are nondiagnostic.

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QUESTIONS

1. A 3-year-old female with B-cell leukemia presents to the emergency room with a fever of 40°C for 2 hours. A complete blood count and blood culture are obtained. Intravenous antibiotics are initiated. The CBC reveals an absolute blood count (ANC) of 345. What is the appropriate management of this patient?
 - A. Discharge home with 24-hour follow-up with oncology
 - B. Admission to the hospital for continuation of intravenous antibiotics
 - C. Continue to observe patient in the ED until afebrile, and then discharge home

- D. Discharge home on 7 days of oral antibiotics
E. Admission to the hospital for observation; antibiotics only if culture becomes positive
2. A 12-year-old male presents to the emergency department with the chief complaint of right lower-quadrant pain. Evaluation for appendicitis is negative; however, an abdominal mass consistent with Burkett's lymphoma is found on CT scan in the area of pain. Electrolytes are concerning for a potassium of 5.3, an elevated uric acid, an elevated phosphate, and a low calcium. What is the first step in the management of this patient?
- A. Obtain IV access and initiate hydration at twice maintenance and sodium bicarbonate
B. Consult to oncology for immediate initiation of chemotherapy
C. Consult to surgery for removal of mass
D. IV replacement of calcium
E. Initiate rectal Kayexalate
3. Repeat laboratory testing on the above patient 4 hours following initiation of therapy reveals increased potassium to 6.7. Emergent ECG reveals peaked T-waves. What is the correct initial intervention for the management of this patient's hyperkalemia?
- A. Increase intravenous fluids to 2.5 times maintenance
B. Administer rectal Kayexalate
C. Administer intravenous calcium gluconate
D. Administer glucose and insulin drip
E. Start a B-agonist nebulizer treatment
4. A 15-year-old female presents to the ED with a chief complaint of shortness of breath. The patient states the symptoms began 2 weeks ago and have progressed to the point where she is dyspneic at rest. She also states that she now must sleep on two pillows at night in order to breathe. Physical exam reveals normal vital signs except a respiratory rate of 30. Cardiac exam is unremarkable and there is no hepatosplenomegaly to suggest congestive heart disease. Chest X-ray is obtained and a mass is noted in the mediastinum. During your evaluation, the patient's respiratory rate and mental status declines, and she requires intubation. What are the most appropriate medications to use in this situation?
- A. Midazolam and Rocuronium
B. Fentanyl, Lidocaine, Ketamine, and Rocuronium
C. Fentanyl and Succinylcholine
D. Midazolam or Ketamine
E. Propofol and Succinylcholine
5. A 5-year-old female presents to the emergency room with new onset back pain and loss of bowel/bladder function. On examination, you note that the patient is more comfortable sitting on the bed, rather than lying down. Also, there is exquisite point tenderness to the lumbar spine on palpation. What is the next step in therapy for this patient?
- A. Referral for outpatient MRI to evaluate the area of pain
B. Obtain plain films of the lumbar spine
C. Initiate pain control and consult with orthopedics for back pain
D. Initiate intravenous dexamethasone
E. Discuss with oncology the need for further chemotherapy and admission
6. A 7-year-old male with B-cell leukemia presents to the ED with right lower-quadrant pain and fever. Per parents, he is undergoing chemotherapy, which includes high-dose corticosteroids. The last dose of chemotherapy was 10 days ago. Exam is significant for pain at McBurney's point, rebound tenderness, and positive psoas and obturator signs. What is the most likely diagnosis in this patient?
- A. Necrotizing colitis
B. Bowel obstruction
C. Diverticulitis
D. Gastro-esophageal reflux disease
E. Constipation

ANSWERS

1. B. The patient described is febrile with neutropenia. These patients are at significant risk of bacteremia or fungemia. Infection is one of the most common complications in the treatment of children with cancer and is a significant cause of morbidity and mortality. The single most important factor is the development of neutropenia due to replacement of healthy bone marrow by malignant cells or from myelosuppressive chemotherapy, which often produces granulocytopenia 8–16 days post-therapy. The best estimate of production of neutrophils is the absolute neutrophil count (ANC), calculated as the total WBC count multiplied by the sum of the percentages of band cells plus polymorphonuclear neutrophils:

$$\text{ANC} = \text{total WBC} \times (\% \text{ Bands} + \% \text{ PMNs})$$

Patients are defined as being neutropenic if their ANC is $<500/\text{mm}^3$ or if it is $<1000/\text{mm}^3$ with predicted decline to $<500/\text{mm}^3$. Fever is defined as a single oral temperature of $\geq 38.3^\circ\text{C}$ (101°F) or a temperature of $\geq 38.0^\circ\text{C}$ (100.4°F) for ≥ 1 hour. Prompt initiation of empiric antibiotic therapy in febrile neutropenic children is critical as infection may progress rapidly. All febrile neutropenic patients are admitted to the hospital for continuation of intravenous antibiotics while cultures are pending.

2. A. The patient is newly diagnosed with Burkett's lymphoma, which is a tumor with high cell turnover, resulting in tumor lysis syndrome. Tumor lysis syndrome (TLS) results from the death of tumor cells and the subsequent release of intracellular contents into the circulation. The result is hyperuricemia, hyperphosphatemia, and hyperkalemia. The breakdown of released nucleic acids from tumor cells leads to hyperuricemia. With hyperuricemia, uric acid may crystallize within the renal tubules leading to obstruction, oliguria, and renal failure. The release of intracellular potassium leads to hyperkalemia, which may be exacerbated by declining renal function. Hyperkalemia may lead to life-threatening arrhythmias. Hyperphosphatemia will lead to secondary hypocalcemia. Severe hypocalcemia may cause tetany, seizures, and arrhythmias. Hypocalcemia should only be corrected if symptomatic as excess calcium will combine with phosphorous. Precipitation of calcium phosphate crystals in the renal tubules leads to renal failure. Early recognition or anticipation of TLS is important. Hydration is the most important initial intervention, facilitating uric acid, and phosphate excretion. Intravenous fluid is administered at a minimum of twice the patient's maintenance rate with 5% dextrose in $\frac{1}{4}$ normal saline with 40 mEq of sodium bicarbonate per liter, aiming to produce a urine pH of 7.0–7.5. Alkalinization of the urine increases uric acid solubility and excretion.
3. C. The initial and most important intervention in the treatment of hyperkalemia involves stabilization of the cardiac cell membranes, to prevent further arrhythmias. This is accomplished with intravenous calcium gluconate at a dose of 1 mL/kg of a 10% solution given over 10–15 minutes. Next, interventions should be performed to shift potassium into cells. This can be accomplished by administering a β -agonist via nebulizer, administering intravenous sodium bicarbonate, or a glucose/insulin drip. These three interventions will relocate the potassium, but will not eliminate it from the body. This is accomplished by enhanced elimination with either rectal Kayexalate or dialysis.
4. D. Several pediatric malignancies, including non-Hodgkin lymphoma, Hodgkin lymphoma, neuroblastoma, germ cell tumors, and acute lymphoblastic lymphoma may present with a mediastinal mass. Mediastinal compression from the tumors may result in Superior Vena Cava Syndrome (SVCS) or Superior Mediastinal Syndrome (SMS). SVCS refers to the signs and symptoms resulting from obstruction, compression, or thrombosis of the SVC. SMS occurs with compression of the narrow, more compliant trachea in children. Management of these patients is challenging as complete obstruction of the airway may be precipitated. Securing the airway may be difficult, particularly, if the obstruction is at or below the distal trachea. Attempts at intubation (either tracheal or selective bronchial intubation) or emergent tracheostomy after airway collapse may not be possible. Even if intubation is achieved, collapse of the airway below the level of the endotracheal tube may lead to inadequate ventilation and life-threatening ventilation perfusion mismatch. Several cases of death due to airway collapse at induction of anesthesia have been reported. If intubation is required, it is recommended that the patient should be awake with spontaneous respiration. Only answer D contains a combination of medications that does not include a paralytic.
5. D. This patient has presented with spinal cord compression, which is a true neurologic emergency. Spinal cord compression may occur with extradural metastatic tumors such as soft tissue sarcomas, neuroblastoma, germ cell tumors, and Hodgkin disease, or rarely with an intradural cord tumor. Pain is the most common initial presenting symptom. The pain is usually worse when supine; there may be tenderness with palpation. Changes in bladder or bowel function may also occur. Treatment begins with dexamethasone to reduce tumor-related edema. Consultation with an oncologist and neurosurgeon should be obtained as soon as possible. MRI should be done immediately in those patients with progressive neurologic deficits. Patients should be promptly referred for possible surgery or radiation therapy.
6. A. This patient is suffering from typhlitis, which is a severe necrotizing colitis of the cecum in neutropenic patients; it may mimic signs and symptoms of acute appendicitis (the patient received chemotherapy 10 days prior to arrival, and thus should be considered neutropenic, as chemotherapy often produces granulocytopenia 8–16 days post therapy). Determining the etiology of the abdominal pain may be difficult in neutropenic or immunosuppressed patients. The inflammatory response may be reduced due to leukopenia and normally localized processes may be generalized. The abdominal examination begins with careful observation, gentle palpation, auscultation, and serial re-examination. Examination of the perineum is important in detecting pelvic and perirectal disease.

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Section 18

NONTRAUMATIC BONE AND JOINT DISORDERS

104 INFECTIOUS MUSCULOSKELETAL DISORDERS

Kemedy K. McQuillen

ACUTE SEPTIC ARTHRITIS

- 75% of cases are in children <5 years (peak incidence: 6–24 months).
- More than 90% of cases are monoarticular.
- Neonates and young infants are vulnerable to hip infection. In older infants and children, the knee is more commonly affected.
- Bacterial causes are listed in Table 104-1.

CLINICAL PRESENTATION

- Infants: fever, failure to feed, lethargy, pseudo-paralysis, and pain with diaper changes.
- Older infants and children: fever, malaise, poor appetite, irritability, localized pain, and limp.

PHYSICAL EXAMINATION

- Erythema, warmth, swelling, decreased range of motion, and painful passive joint movement.
- Hip involvement: held in flexion, abduction, and external rotation; joint dislocation in infants.
- Gonococcal arthritis: fever, chills, polyarthralgia, mono- or polyarticular arthritis, and rash.

DIAGNOSTIC EVALUATION

- Septic arthritis more likely than transient synovitis with fever, inability to bear weight, ESR > 40 mm/h, and WBC > 12,000/mm³.

- Elevated WBC, ESR, and CRP; blood cultures (positive <50% of cases).
- Joint fluid culture is positive in ~60% of patients (Table 104-2).
- *Neisseria gonorrhoeae*.
 - Culture joint fluid, blood, pharynx, rash, cervix, urethra, vagina, and rectum.
 - Nucleic amplification testing of urine, urethra, cervix, and vagina.

RADIOGRAPHIC IMAGING

- X-rays: most useful in ruling out other conditions.
- Ultrasound: visualization of joint and needle with joint aspiration (Fig. 104-1A and B).
- Advanced imaging: magnetic resonance imaging (MRI) or radionuclide scanning.

DIFFERENTIAL DIAGNOSIS

- Transient synovitis, cellulitis, traumatic hemarthrosis, osteomyelitis, inflammatory arthritis, fracture, Legg–Calve–Perthes disease, slipped capital femoral epiphysis, psoas abscess, obturator internus abscess, diskitis, nonbacterial infections.
- Reactive arthritis: >2 weeks after streptococcal infection, gastroenteritis, or viral hepatitis.

TREATMENT

- Admission, antibiotics after joint fluid is obtained and joint drainage (Table 104-1).
- Although mortality is <1%, morbidity remains significant.
- Sequelae include leg-length discrepancy, persistent pain, limited range of motion, and necrosis of the femoral head.

TABLE 104-1 Pathogens and Treatment of Septic Arthritis and Osteomyelitis^a

AGE OR COMORBIDITY	ORGANISMS	INITIAL ANTIBIOTICS
0 mo to 2 mo	Group B Streptococcus <i>S. aureus</i> Gram-negative rods <i>Candida</i> ^b <i>N. gonorrhoeae</i>	Nafcillin 50 mg/kg and Cefotaxime 50 mg/kg
2 mo to 5 y	<i>S. aureus</i> Group A Streptococcus <i>S. pneumoniae</i> <i>K. kingae</i> <i>Haemophilus influenzae type b</i> ^c	Nafcillin 50 mg/kg and Cefotaxime 50 mg/kg or Ceftriaxone 50 mg/kg or Cefuroxime 50 mg/kg
5 y to 12 y	<i>S. aureus</i> <i>S. pyogenes</i> <i>Haemophilus influenzae type b</i> ^c	Nafcillin 50 mg/kg or Cefazolin 25 mg/kg
>12 y	<i>N. gonorrhoeae</i> <i>S. aureus</i>	Ceftriaxone 50 mg/kg
Immunocompromised	Gram-negative enteric bacilli <i>S. aureus</i> <i>P. aeruginosa</i>	Ceftazidime 50 mg/kg and Vancomycin 10 mg/kg
Sickle cell disease	<i>Salmonella spp.</i> Gram-negative enteric bacilli <i>S. aureus</i>	Ceftriaxone 50 mg/kg and Nafcillin 50 mg/kg
Puncture wounds of the foot	<i>P. aeruginosa</i> <i>S. aureus</i>	Ceftazidime 50 mg/kg and Nafcillin 50 mg/kg

^aIf MRSA is a concern or local rates are >5% to 10%, use intravenous vancomycin; consider supplementing with gentamicin +/- rifampin for synergy if ICU admission.

^bNeonates with indwelling catheters.

^cIf unimmunized.

OSTEOMYELITIS

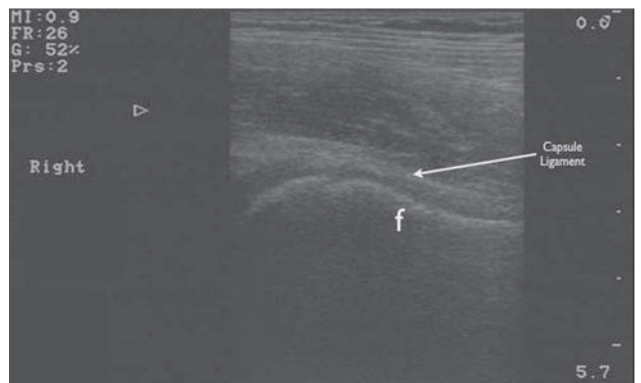
- Most commonly between 3 and 12 years (50% of cases occur in children <5 years old).
- Metaphyses of the long bones of the lower extremity are most commonly involved.
- Children <18 months have transphyseal vessels that spread infection to the joint space.

ETIOLOGY (Table 104-1)

- *Staphylococcus aureus*: 70–90% of cases of osteomyelitis.
- *Streptococcus pyogenes*: 10% of cases with peak incidence in preschool and early school-age children.
 - More pronounced fevers and higher WBC counts.
 - Recent history of varicella.



A



B

FIG. 104-1. (A) Ultrasound image showing hip effusion. (B) Ultrasound image of a normal hip for comparison.

TABLE 104-2 Analysis of Joint Fluid

	CHARACTER	WBC COUNT (/μL)	PMNS (%)	GLUCOSE (SYNOVIAL/BLOOD)	OTHER
Normal	Clear; yellow	<200	<10	>50	Good mucin clot
JRA	Turbid	250–50 000	50–70	>50	Fair to poor mucin clot 50% with low complement
Reactive arthritis	Clear or turbid	1000–150 000	50–70	>50	Fair to poor mucin clot Elevated complement
Lyme arthritis	Turbid	500–100 000	>50	>50	Poor mucin clot
Septic arthritis	Turbid; white-gray	10–250 000	>75	<50	Poor mucin clot High lactate

JRA, juvenile rheumatoid arthritis; PMN, polymorphonuclear leukocytes; WBC, white blood cell count.

- *Streptococcus pneumoniae*: younger children who are more likely to have joint involvement.

CLINICAL MANIFESTATIONS

- Neonates: irritability, fever, resistance to movement, +/- redness, swelling, and dissemination.
- Older infants and children: acute pain, limp, erythema, warmth, and swelling.

CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS

- It is characterized by recurrent bone pain and fever.
- Girls more commonly affected
- Associated with psoriasis and palmoplantar pustulosis
- Radiographs: often symmetric bone lesions of long bones and clavicles

DIAGNOSTIC TESTING

- Normal or elevated white blood cell count, elevated ESR and CRP.
- Blood and bone cultures are positive ~60% of the time.
- Radiographs: mottling and demineralization by days 7–10 with periosteal and lytic changes by days 10–21 (Fig. 104-2).
- Advanced imaging:
 - MRI (highest sensitivity and specificity)
 - CT
 - Bone scans
 - Ultrasound detects subperiosteal abscesses early in osteomyelitis and may be the only imaging test required in uncomplicated cases.

TREATMENT

- Admission and, after bone culture is obtained, empiric antibiotics (Table 104-1).

SEQUELAE

- They are rare although more frequent in neonates:
 - recurrence in 5% (risk factors: delayed diagnosis, abbreviated antibiotics, and young age);
 - limb-length discrepancies, arthritis, abnormal gait, and pathologic fractures;
 - chronic osteomyelitis occurs in <5%; most commonly *S aureus* and Gram (-) enterics
 - chronic swelling, pain, and drainage with exacerbations of fever, swelling, and redness;
 - treatment: surgical management augmented by a prolonged course of antibiotics.
 - Brodie abscess: subacute infection resulting in necrotic bone and pus in a fibrous capsule;
 - tends to occur in the long bones of adolescents and is treated with surgery and antibiotics.

INTERVERTEBRAL DISKITIS

- Acute infection of the vertebral disk usually seen in children younger than 5 years.
- The lumbar area is most commonly involved.



FIG. 104-2. Osteomyelitis of the clavicle.

- Pathogens: *S aureus* (less common: pneumococcus, Gram (–) organisms, mycobacterium (rare)).
- Diagnosis is often delayed: mean duration of symptoms of 33 days (range: 5 days to 3 months).

CLINICAL MANIFESTATIONS

- Irritability, refusal to sit or walk, back or leg pain, limp, and gastrointestinal symptoms.
- Physical examination: lordosis, torticollis, vertebral tenderness, and hip pain +/- fever.

DIAGNOSTIC TESTING

- Elevated ESR: positive blood cultures in ~40% of cases.
- Radiographs: disk space narrowing and erosion of vertebral end plates (Fig. 104-3).
- Advanced imaging with MRI and bone scintigraphy.

TREATMENT

- Outpatient oral antibiotic therapy directed against *S aureus*.

- Despite treatment, older children commonly develop spontaneous spinal fusion.

LYME DISEASE (For further discussion of Lyme disease see Chapter 65.)

- Most common tick-borne illness in North America and Europe.
- *Borrelia burgdorferi* infection (spirochete) from the bite of the *Ixodes scapularis* tick (deer tick).
- Initial presentation: erythema migrans (if untreated, 60% develop mono- or oligo-articular arthritis).

ACUTE SUPPURATIVE TENOSYNOVITIS

- Synovial sheath infection of the flexor tendons of the finger that can spread to deep palm spaces.
- Swollen finger held in flexion, pain with extension, and erythema/tenderness along flexor tendon.
- Pathogens: *S aureus* and group A *Streptococcus* (*N gonorrhoeae* if sexual contact).
- Management: antibiotics and surgical drainage.

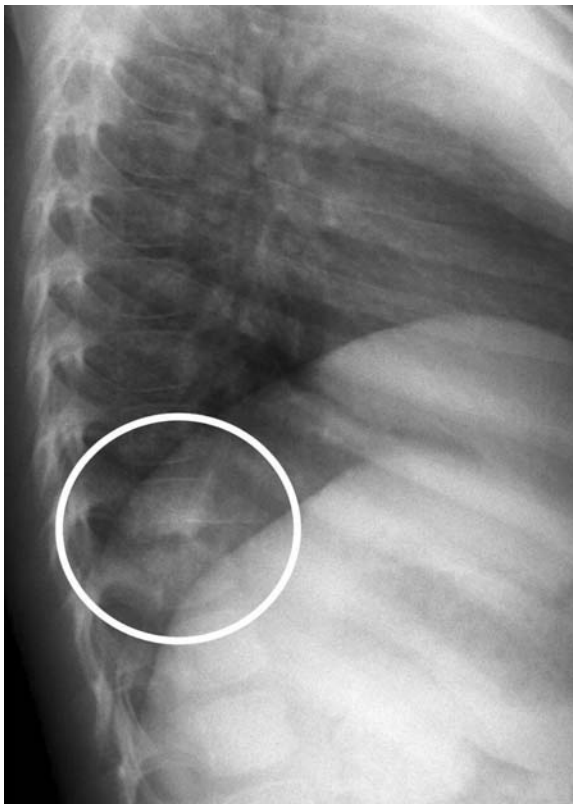


FIG. 104-3. Diskitis between T10 and T11 in a 3-year-old child.

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QUESTIONS

- A 2-year-old boy is brought in for evaluation of fever and fussiness. He has been much less active than usual and refuses to walk. He is tired but nontoxic appearing, febrile to 39°C, and is holding his left leg flexed, abducted, and externally rotated. Which of the following is true regarding his management?

 - Outpatient management is a viable option
 - The most common pathogen is *S aureus*
 - Single antibiotic treatment is the standard of care
 - Ultrasound can be diagnostic
 - Morbidity is negligible
- A 4-year-old girl is brought in with a red, swollen area at her upper left leg. She has had fever and leg pain for 3 days and is now refusing to walk. Which of the following is accurate?

 - This illness is more common in adolescence
 - Bony abnormalities are expected on X-ray within 24 hours of the onset of symptoms
 - Fever is uncommon
 - In a nontoxic appearing child, antibiotics can be held until a culture is obtained
 - This illness never involves adjacent structures
- After discharge, the child in the above question was lost to follow up. She returns with ongoing pain of her leg with intermittent fevers associated with worsening redness and swelling of the area. She likely has which of the following?

 - Chronic recurrent multifocal osteomyelitis
 - Septic arthritis
 - Chronic osteomyelitis
 - Cellulitis
 - Normal recovery
- A 14-year-old male presents with a week of pain to the left upper arm. He has a sense of chills but no documented fever at home. He has a palpable, tender mass of the proximal humerus. Which of the following is true regarding this patient?

 - Spontaneous resolution is expected
 - This illness is typically seen in adolescents
 - Requires advanced imaging modalities to detect
 - Follow up with his primary-care provider is sufficient
 - Onset of symptoms is usually acute.
- A 4-year-old girl presents with fever to 104°F for the past 3 days with redness, swelling, and tenderness to the left arm for the past 2 days. She got over chickenpox 1 week ago. She is fully immunized. Her WBC count is 22,000/mm³. What is the likely organism?

 - Haemophilus influenzae*
 - S pneumoniae*
 - S pyogenes*
 - Salmonella* spp.
 - Gram-negative enterics
- Antibiotics for the girl in Question 5 should include which of the following?

 - Nafcillin
 - Ceftriaxone
 - Penicillin
 - A and B
 - A and C
- Which of the following is true about the patient in Question 5?

 - Normal radiographs exclude the possibility of osteomyelitis
 - Antibiotics should be given as soon as osteomyelitis is suspected
 - Bone involvement precludes the chance of joint involvement
 - MRI or ultrasound may help confirm the diagnosis
 - She has no chance of complications from her illness
- A 2-year-old girl is brought in by her father because she has been fussy and refused to walk or sit. She had a fever to 101°F. She has no other symptoms. On examination, she is laying very still with her back arched and cries with movement of her hips and palpation of her mid-upper lumbar spine. Which of the following are true?

 - The most common organism is *S aureus*
 - Inpatient therapy is always required
 - Radiographs are not helpful in establishing the diagnosis
 - Laboratory tests are not helpful in establishing the diagnosis
 - Complications include limb-length discrepancy
- A 15-year-old presents with a target rash on her left arm. She removed a tick from that arm 4 days previously. She is afebrile and well appearing. You suspect Lyme disease. Which of the following is true?

 - Lyme disease is an infrequent illness in North America
 - The illness is due to *B burgdorferi*
 - The tick she removed is not related to the illness
 - No treatment is required for the rash; it will resolve without sequelae
 - The rash is called erythema marginatum
- A 16-year-old female presents with pain in her finger. She is unable to move it. She denies any recent injury. She also has a rash. On examination, she is febrile and has petechiae, papules, and pustular lesions with erythematous halos all over her body.

Her left finger is red, swollen, and held in flexion. She has pain with passive extension of her finger. Which of the following is true?

- A. This infection remains localized
- B. Her causative pathogen is likely mycoplasma
- C. A pelvic examination may help clarify the diagnosis
- D. Pathogens are invariably cultured from the skin lesions
- E. Surgical intervention is never required

ANSWERS

1. B. This child has septic arthritis. The most common pathogen is *S aureus*. Inpatient management is mandatory for septic arthritis. Empiric antibiotic coverage includes nafcillin and a cephalosporin. Vancomycin may also be used. Ultrasound can identify an effusion but cannot differentiate among purulent, nonpurulent, and hemorrhagic effusions. Morbidity is not negligible and includes leg-length discrepancy, persistent pain, limited range of motion, and necrosis of the femoral head.
2. D. In a nontoxic appearing child with osteomyelitis, antibiotics can be held until a culture is obtained. Osteomyelitis is most common in children between 3 and 12 years old. Although there may be soft tissue swelling early on in osteomyelitis, bony abnormalities do not appear until day 7–10 (with mottling and demineralization). Periosteal and lytic changes appear by day 10–21. Fever is common. Osteomyelitis can involve adjacent soft tissue and, especially in young infants, can include septic arthritis.
3. C. This child had a short course of antibiotics putting her at risk for developing chronic osteomyelitis. Chronic recurrent multifocal osteomyelitis is characterized by recurrent bone pain and fever. Her symptoms are not consistent with septic arthritis. Cellulitis is unlikely. With normal recovery she should be symptom free.
4. B. This child has a Brodie abscess which is a subacute infection resulting in necrotic bone and pus in a fibrous capsule. It is typically seen in adolescents. Treatment includes surgery and antibiotics. It does not resolve spontaneously. Brodie abscess can be seen on plain radiographs. This child requires treatment with surgery and antibiotics. Brodie abscess is a subacute process.
5. C. *S pyogenes* occurs in 10% of cases of osteomyelitis and is seen most commonly in preschool and early school-age children. It is associated with more pronounced fevers and higher WBC counts and is more commonly seen after a varicella infection. *H influenzae* can be seen in unimmunized children. *S pneumoniae* is more common in young children and is more likely to have joint involvement. *Salmonella* species cause osteomyelitis in children with sickle-cell disease. Gram-negative enterics cause osteomyelitis in immunocompromised children or children with sickle-cell disease.
6. D. Antibiotics for the girl in Question 5 should include nafcillin and a cephalosporin. Vancomycin may also be considered.
7. D. Ultrasound can detect osteomyelitis early in the course of disease if there is a subperiosteal abscess. MRI is also beneficial in detecting osteomyelitis early. Radiographs may be normal for the first 7–10 days of osteomyelitis. If possible, antibiotics should be held until a bone sample is obtained for culture. Osteomyelitis can be associated with septic arthritis especially in young infants. Osteomyelitis can be complicated by leg-length discrepancy, persistent pain, limited range of motion, and necrosis of the femoral head.
8. A. This child has diskitis. The most common organism in diskitis is *S aureus*. Outpatient therapy is an option. Radiographs can show narrowing of the disc space and erosion of vertebral end plates. Laboratory tests show an elevated ESR. Blood cultures are positive in ~40% of cases. Complications of diskitis include spinal fusion but not limb-length discrepancy.
9. B. This child has Lyme disease. It is due to *B burgdorferi*. Lyme disease is the most common tick-born illness in North America and Europe. The infection, caused by a spirochete is from the bite of the *I scapularis* tick (deer tick). If untreated, 60% or patients with erythema migrans will develop mono- or oligo-articular arthritis. The rash is called erythema migrans, not erythema marginatum.
10. C. This teenager has acute suppurative tenosynovitis caused by *N gonorrhoeae*. She may have vaginal discharge or cervical motion tenderness. Amplified assays or cultures will show the presence of *N gonorrhoeae*. This infection can extend into the deep palmar spaces. This infection, with rash and acute suppurative tenosynovitis, is usually caused by *S aureus* and group A *Streptococcus*, but *N gonorrhoeae* is the likely cause if there has been sexual contact. Skin lesions are usually sterile. Treatment is with antibiotics and surgical intervention.

105 INFLAMMATORY MUSCULOSKELETAL DISORDERS

Kemedy K. McQuillen

TRANSIENT SYNOVITIS (also known as toxic synovitis)

- Self-limited nonpyogenic inflammatory response of the synovium:
 - Most common cause of hip pain in childhood (can also affect the knee).
- Peak incidence: 3–6 years of age.
- Affects boys more commonly than girls and has a slight predilection for the right side.
- Etiology is unknown: possibly related to active or recent infection, trauma, or hypersensitivity.

CLINICAL PRESENTATION

- Hip or groin pain in most cases: thigh or knee pain in 10–30%.

PHYSICAL EXAMINATION

- Children are well appearing and may have a low-grade fever and malaise.
- The leg is held in flexion with slight abduction and external rotation.
- Passive movement is usually pain free: pain with extreme internal rotation or abduction.

DIAGNOSTIC EVALUATION

- Diagnostic evaluation of transient synovitis is one of exclusion.
- Normal or mildly elevated WBC count and ESR.
- In afebrile children with a normal WBC and ESR, septic arthritis is unlikely and the diagnosis of transient synovitis can be made without obtaining joint fluid.

RADIOGRAPHIC IMAGING

- It is helpful in excluding other diseases.
- AP and “frog-leg” lateral views of the pelvis.
- Radiographic findings: medial joint space widening, an accentuated pericapsular shadow, and Waldenström’s sign (lateral displacement of the femoral epiphysis with surface flattening secondary to effusion).

- Also seen in Legg–Calvé–Perthes (LCP) disease.
- If present, mandate close follow-up or further investigation with MRI.
- Ultrasound can diagnose effusion but cannot differentiate type of effusion.
- 60–70% of cases of transient synovitis have hip effusion.

DIFFERENTIAL DIAGNOSIS

- Septic arthritis, osteomyelitis, acute slipped capital femoral epiphysis (SCFE), LCP disease, rheumatoid and infectious arthritis, malignancy, and osteoid osteoma.

TREATMENT

- Rest via nonweight bearing or, in cases of extreme pain, bed rest.
- Reduction of synovitis with anti-inflammatory medications.
- Gradual return to activity with unrestricted activity when the hip is pain free.
- Repeat examination in 12–24 hours and again in 10–14 days if the symptoms persist.

PROGNOSIS

- It is excellent.
- 75% of patients have complete resolution within 2 weeks and 88% resolve within 4 weeks.
- Relapse is infrequent and usually occurs within 6 months.

SYSTEMIC LUPUS ERYTHEMATOSUS (SLE)

- Chronic, episodic multisystem autoimmune disease with protean manifestations.
- 15–20% of cases diagnosed in childhood (median age: 12.2 years): SLE rare <5 years old.
- Time from symptom onset to diagnosis: 1 month to 3.3 years with a median of 4 months.
- Female:Male ratio of 3:1 prepuberty and 9:1 after puberty.
- Higher incidence in Native Americans, Hispanics, Chinese, and Filipinos:
 - Greater disease severity in African Americans and Hispanics.
- Patients with SLE are more likely to have a relative with lupus or another autoimmune disease.

TABLE 105-1 Manifestations of Systemic Lupus Erythematosus (SLE)

MUCOCUTANEOUS	RENAL	NEUROPSYCHIATRIC
Malar rash (Fig. 105-1)	Microscopic hematuria	Headache
Photosensitivity	Proteinuria	Cerebral vein thrombosis
Nodules	Hypertension	Psychosis
Ulcerations	Azotemia	Altered mental status
Palmar/plantar erythema	Renal failure	Cognitive dysfunction
Alopecia	Pulmonary	Cerebrovascular disease
Discoid lupus	Pneumonitis	Seizures
Lupus profundus	Pleuritis	Movement disorders
Hyperemic mucosa	Pleural effusion	Neuropathy
Palatal petechiae	Pulmonary hemorrhage	Transverse myelitis
Painless oral ulcers	Pulmonary hypertension	Musculoskeletal
Hematologic	Shrinking lung syndrome	Arthritis
Anemia	Cardiac	Arthralgias
Hemolysis	Pericarditis	Prolonged morning stiffness
Thrombocytopenia	Pericardial effusion	Myalgia
Leukopenia	Myocarditis	Diffuse weakness
Gastrointestinal	Endocarditis	Tenosynovitis
Pancreatitis	Premature atherosclerosis	Periostitis
Mesenteric vasculitis	Constitutional	Myositis
Hepatitis	Fever	Avascular necrosis
Peritonitis (serositis)	Fatigue/malaise	Osteoporosis
Anorexia	Weight loss	Vertebral fractures

CLINICAL PRESENTATION OF PEDIATRIC SLE (pSLE) (Table 105-1)

- Most common presenting signs: rash, mucositis, and arthritis.
- Most common organ systems involved: skin, musculoskeletal, and renal:
 - Skin manifestations in 50–80% at diagnosis and in 85% over lifetime of patients:
 - The appearance of a malar rash often heralds a disease flare (Fig. 105-1);
 - Sun exposure can exacerbate the skin disease of lupus and cause a systemic flare;
 - Polyarticular arthritis: nonerosive, nondeforming, painful, symmetric, small/large joints
 - Pain and tenderness disproportionately greater than the degree of swelling;

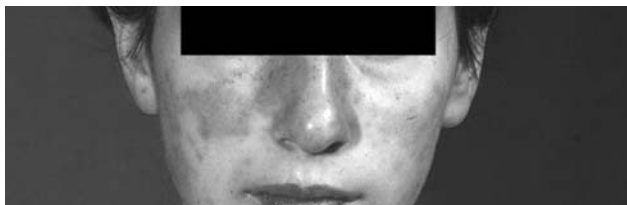


FIG. 105-1. The characteristic malar rash of systemic lupus erythematosus. Note the extension across the cheeks and the nasal bridge with sparing of the nasolabial folds.

- Renal involvement in 75%; usually within 2 years of disease onset –
 - Leading cause of serious morbidity and mortality in SLE;
 - Renal flares detected by proteinuria before constitutional symptoms appear.
- CNS disease: second leading cause of serious morbidity and mortality.

DIAGNOSTIC EVALUATION

- Complete blood count, prothrombin time (PT), international normalized ratio (INR), partial thromboplastin time (PTT), serum electrolytes, blood urea nitrogen, creatinine, erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP) are indicated.
- Urinalysis: microscopic hematuria and proteinuria.
- With signs of coagulopathy: lupus anticoagulant and antiphospholipid antibody.
- Antinuclear antibody (ANA) is positive in almost all patients who have active disease:
 - May also be positive in up to 33% of healthy patients.
- Double-stranded DNA (anti-ds DNA) more specific to SLE: in 60–70% of SLE patients.

DIFFERENTIAL DIAGNOSIS

- Differential diagnosis of pSLE is vast.

- Malignancies (especially leukemia), acute rheumatic fever, juvenile idiopathic arthritis (JIA), and infectious processes.
- Drug-induced lupus: anticonvulsants (phenytoin and carbamazepine), isoniazid, and minocycline.

TREATMENT

- NSAIDs, steroids, immunosuppressants, and biologic agents.

PROGNOSIS

- Prognosis of pSLE has improved over the past several years.
- 5-year survival rate is 92% and the 10-year survival rate is 85%.
- Primary causes of death: renal disease, infection, and CNS disease.
- Malar rash, neurologic features, and renal disease are more frequent in pSLE.
- Hematologic and renal involvement tends to be worse in pSLE than in adult SLE.

RHEUMATIC FEVER (ARF)

- Autoimmune inflammatory response to group A β -hemolytic *Streptococcus* (GA β HS) infection:
 - Occurs after GA β HS pharyngitis but is not associated with skin *streptococcal* infection;
 - Develops 2–4 weeks following the inciting infection (range: weeks to months);
 - One-third of children do not recall an antecedent sore throat.
- Incidence peaks between 5 and 15 years of age.
 - <5% of episodes arise in children younger than 5 years; almost unheard of <2 years old. It is most common during the winter and spring and is more common in girls.

CLINICAL FEATURES

- Modified Jones Criteria (Table 105-2) or the World Health Organization (WHO) criteria (Table 105-3).

JOINT

- Complaints are common: arthralgias to frank arthritis
- The arthralgias are especially intense at night and can wake children from sleep.
- Pain is out of proportion to the clinical findings.

TABLE 105-2 Diagnosis of Rheumatic Fever (Revised Jones Criteria)^{a,b}

Evidence of Antecedent GA β HS Infection

Positive throat culture or rapid antigen test for GA β HS
 Raised or rising streptococcal antibody titer (antistreptolysin O, anti-DNase B or antihyaluronidase)

Major Manifestation: J♥NES

J: Joints (Polyarthritis)

♥: Carditis

N: Nodules (subcutaneous nodules)

E: Erythema marginatum

S: Sydenham chorea

Minor Manifestation

Arthralgia

Fever

Elevated ESR or CRP concentrations

Prolonged PR interval on electrocardiogram

Recurrent Acute Rheumatic Fever

One major or several minor manifestations *plus* evidence of antecedent GA β HS infection

^aDiagnosis requires two major *or* one major and two minor manifestations *plus* evidence of antecedent GA β HS infection.

^bChorea and indolent carditis do not require evidence of antecedent GA β HS infection.

- Arthritis occurs in ~75% of cases:
 - Polyarticular, large joints (knees, ankles, wrists, and elbows), and migratory;
 - Monoarticular arthritis may occur with early use of anti-inflammatories;
 - Synovial fluid
 - 10,000–100,000 white blood cells/mm³ with a predominance of neutrophils;
 - protein of ≥ 4 g/dL and normal glucose.
- Responds dramatically to salicylates.
- Joint symptoms resolve within a month and leave no permanent damage.

TABLE 105-3 Diagnosis of Rheumatic Fever (Who Criteria)^a

First episode

As per Jones criteria

Recurrent episode

Patients without established rheumatic heart disease:

As per first episode

Patients with established rheumatic heart disease:

2 minor manifestations

plus

Evidence of antecedent GA β HS infection

Positive throat culture or rapid antigen test for GA β HS

Raised or rising streptococcal antibody titer

(antistreptolysin O, anti-DNase B or antihyaluronidase)

Recent scarlet fever

^aChorea and indolent carditis do not require evidence of antecedent GA β HS infection.

- Differentiate from poststreptococcal reactive arthritis (PSRA):
 - Develops 3–14 days after pharyngitis: anti-*Strep.* antibodies may not be present;
 - Nonmigratory, small, large joints, and axial joints, responds poorly to anti-inflammatories, and lasts from 1 week to 8 months (mean duration: 2 months);
 - Normal WBC count and elevated ESR, +/- positive throat culture or rapid antigen test;
 - 5% of patients with PSRA develop carditis;
 - Treatment of PSRA: antimicrobial prophylaxis with penicillin or erythromycin
 - For 1 year without carditis and for at least 5 years if carditis develops.

CARDITIS

- Occurs in 50–60% of cases and causes significant morbidity and mortality.
- Pancarditis: endocarditis (valvulitis) +/- myocarditis and pericarditis:
 - mitral valve is most commonly involved +/- aortic valve involvement.
- Typically presents with tachycardia and a valvular insufficiency murmur:
 - valvular stenosis appears years to decades after the acute illness.
- Echocardiography: pericardial effusion, decreased contractility, and valvular regurgitation.

NODULES

- Firm, painless, and subcutaneous nodules along the extensor surface near bony prominences.

ERYTHEMA MARGINATUM

- Nonpruritic, erythematous, serpiginous macule with a pale center (Fig. 105-2).
- On the trunk and extremities and tends to spare the face.
- Accentuated by warming the skin.

SYDENHAM CHOREA

- It is due to an autoimmune insult to the basal ganglia.
- Occurs in 10–15% of patients and may be the only manifestation of ARF.
- Involuntary unilateral or bilateral choreiform movements and facial grimacing:
 - Exacerbated by stress and disappears with sleep.



FIG. 105-2. Erythema marginatum: one of the major manifestations of acute rheumatic fever.

- Preceded by behavioral disturbances (emotional lability, personality changes, and anxiety)
- Patients may have “Sydenham speech” (bursts of dysarthric speech).
- Develops 1–6 months after the illness (streptococcal antibodies may be undetectable).
- Physical findings: irregular contractions of the hands when squeezing the examiner’s finger (milkmaid’s grip), spooning, and pronation of the hands when the arms are extended, and wormian movements of the tongue upon protrusion.
- Recurrence in 20–60% of patients and usually occurs within 2 years.
- Majority of patients also have cardiac involvement.

DIAGNOSTIC EVALUATION

- Throat culture and rapid antigen testing for GABHS.
- Titers for antistreptolysin O, anti-DNase B, and anti-hyaluronidase.
- ESR and CRP.
- Electrocardiogram and echocardiogram.
- Chest radiograph to exclude congestive heart failure (CHF).

DIFFERENTIAL DIAGNOSIS

- Septic arthritis, JIA, reactive arthritis, serum sickness, malignancies, SLE, Lyme disease, idiopathic juvenile arthritis, and sickle-cell disease.

TABLE 105-4 Characteristics of Spondyloarthropathies

TYPE	GENDER	AGE	TYPE OF INFLAMMATION	JOINTS	OTHER
Enthesitis-related arthritis (juvenile ankylosing spondylitis)	M >> F	Adolescence, adulthood	Oligoarthritis Enthesitis	Axial skeleton SI joints Legs > arms	HLA-B27(+) > 90%
Psoriatic arthritis	F >> M	Childhood	Oligoarthritis	Extremities	HLA-B27(-)
Inflammatory bowel disease arthropathies	M > F	Any age	Polyarthritis	Extremities	Reflects activity of GI inflammation
Inflammatory bowel disease arthropathies	M > F	Any age	Oligoarthritis	SI joints Extremities	HLA-B27(-) Independent of GI inflammation
Reactive arthritis	M > F	Any age	Oligoarthritis	Legs > arms	HLA-B27(+) ~30%–50%

TREATMENT

- It is initially inpatient.
- Antibiotics to eradicate GABHS infection.
- Arthritis without carditis: high-dose aspirin (100 mg/kg/d in four divided doses).
- Carditis and cardiomegaly or CHF: prednisone (2 mg/kg/d in four divided doses).
 - +/- digoxin, fluid and salt restriction, diuretics, and oxygen.
- Chorea: haloperidol, thorazine, valproic acid, or carbamazepine.
- Prophylactic antibiotics to prevent recurrent attacks that may cause or exacerbate carditis.

SPONDYLOARTHROPATHIES

- It includes enthesitis-related arthritis; formerly known as juvenile ankylosing spondylitis, psoriatic arthritis, inflammatory bowel disease-related arthritis, and reactive arthritis (Table 105-4).
- Inflammation of joints of the axial skeleton and limbs.
- Enthesitis: chronic inflammation at tendon, ligament, fascia, and capsule attachment sites
 - Causes calcification of ligaments and fusion of joints in adulthood.
- Absence of the rheumatoid factor.
- +/- tenosynovitis and periostitis.

ENTHESITIS-RELATED ARTHRITIS (JUVENILE ANKYLOSING SPONDYLITIS)

- Greater frequency of extra-axial symptoms than in adults.
- Involved joints: spine (Fig. 105-3), sacroiliac (SI) joints, and hips.

- Clinical findings:
 - hip, back, and thigh pain that is worse at night and improves with movement;
 - systemic symptoms include fatigue and low-grade fever.



FIG. 105-3. Radiograph of patient with enthesitis-related arthritis. Note the narrowed joint spaces between C6/C7 and C7/T1 with ankylosing of C6–T1 vertebral bodies.

- Physical examination: SI joint tenderness and loss of lumbar spine range of motion –
 - 25% of patients have acute, painful iridocyclitis;
 - +/- fever, aortic valve insufficiency, aortitis, weakness, or atlantoaxial subluxation.
- Sequelae: fusion of the spine and SI joints.

PSORIATIC ARTHRITIS

- Most commonly asymmetric oligoarthritis of the knees, ankles, and small joints.
- Psoriatic rash, nail pitting, dactylitis, onycholysis, and family history supports the diagnosis
- Chronic iridocyclitis in 15%.
- Antinuclear antibodies in 50% of affected patients.

INFLAMMATORY BOWEL DISEASE-RELATED ARTHROPATHIES

- These are characterized by a peripheral polyarthritis that is not a true spondyloarthropathy and is reflective of GI inflammation or an HLA-B27-associated spondyloarthropathy of the sacroiliac and peripheral joints that is independent of GI inflammation.
- Associations: erythema nodosum, pyoderma gangrenosum, fever, weight loss, and anorexia.

REACTIVE ARTHRITIS

- It is preceded by GI infection with *Salmonella*, *Yersinia*, *Campylobacter* or *Shigella*, or a GU infection with *Chlamydia trachomatis*.
- Symptoms starting 1–4 weeks after the inciting infection.
- Oligoarticular, sterile synovitis with pain, swelling, and erythema +/- enthesitis.
- Associated findings are listed in Table 105-5.
- Half have resolution within 6 months; the rest have a chronic, relapsing reactive arthritis.

DIAGNOSTIC EVALUATION

- Elevated inflammatory markers (ESR and CRP), WBC, and platelet counts.
- Rheumatoid factor: negative.
- Antinuclear antibodies: absent except in psoriatic arthritis.
- HLA-B27: present in >90% of patients with enthesitis-related arthritis –
 - found in the other spondyloarthropathies especially with sacroiliitis or anterior uveitis.

TABLE 105-5 Findings Associated with Reactive Arthritis

<i>Constitutional Symptoms</i>	<i>Musculoskeletal</i>
Fever	Arthritis
Malaise	Tenosynovitis
Weight loss	Periostitis
<i>Skin</i>	Enthesitis
Keratoderma blennorrhagica	Sacroiliitis
Balanitis circinata	Dactylitis
Oral ulcers	<i>Genitourinary</i>
Hyperkeratosis	Urethritis
Subungual keratosis	Cervicitis
Nail pits	Cystitis
Onycholysis	Hematuria
<i>Cardiac</i>	<i>Ocular</i>
Aortitis	Conjunctivitis
Aortic insufficiency	Acute anterior uveitis
Heart block	
Pericarditis	

- Radiographic findings: periarticular osteoporosis, enthesitis changes (loss of sharp cortical margins, erosions, and bony spurs), indistinct margins and erosions of the SI joints, sclerosis on the iliac side of the joint, and squaring of the corners of the vertebral bodies.

TREATMENT GOALS

- Control of inflammation, minimization of pain, and preservation of function.
- NSAIDs, sulfasalazine, and intra-articular steroid injections:
 - Etanercept is not approved for use in the pediatric spondyloarthropathies.
- Antibiotic treatment may be beneficial when *C trachomatis* is the inciting cause.

JUVENILE IDIOPATHIC ARTHRITIS

- Most common chronic rheumatic disease in childhood.
- Characterized by arthritis with onset <16 years of age, >6 weeks of symptoms, no identifiable cause.

ILAR classification system: based on the features in the first 6 months of illness (Table 105-6).

POLYARTICULAR

- Five or more joints.
- Swollen, warm joints with minimal, or no erythema with mild joint pain.
- Systemic involvement: fever, irritability, and hepatomegaly.

TABLE 105-6 Juvenile Idiopathic Arthritis (JIA): The ILAR Classification System

TYPE	FREQUENCY	AGE AT ONSET	GENDER	ARTHRITIS	JOINTS
Systemic arthritis	10%–20%	Childhood	F = M	Symmetrical Polyarticular	Large and small
Oligoarthritis	50%–60%	Early childhood (<6 y)	F >>> M	Asymmetric	Lower extremities
RF [+] polyarthritis	5%–10%	Late childhood/ adolescence	F >> M	Symmetric	Small joints (+/- large joints)
RF [-] polyarthritis	20%–30%	Childhood	F >> M	Asymmetric or symmetric	Large and small
Enthesitis-related arthritis	1%–7%	Late childhood/ adolescence	M >> F	Oligoarticular	Lower extremities Axial skeleton SI joints
Psoriatic arthritis	2%–15%	Childhood		Asymmetrical	Small and large
Undifferentiated arthritis	11%–21%			Oligoarticular	

ILAR, International League of Associations for Rheumatology; RF, rheumatoid factor.

RF[+] POLYARTHRITIS

- Positive IgM rheumatoid factor at least twice, >3 months apart.
- Same as adult RF[+] polyarthritis.
- May have involvement of the cervical spine (atlan-toaxial instability) or cricoarytenoid joint (hoarseness).
- Boutonniere deformities and swan-neck deformities of the fingers are common.
- Extra-articular manifestations: rheumatoid nodules and aortic regurgitation.
- Progressive, diffuse joint involvement with early radiographic changes.

RF[-] POLYARTHRITIS

- Biphasic onset: early peak between 2 and 4 years and late peak between 6 and 12 years.
- At least three distinct subsets:
 - Early onset asymmetric arthritis: positive ANA, iridocyclitis, and HLA association.
 - Polyarthritis: school-aged children, symmetric large and small joint disease, elevated ESR, and negative ANA;
 - “Dry synovitis”: negligible joint swelling, significant joint stiffness, flexion contractures, normal or minimally elevated ESR.
- Variable outcome depending on its subtype.

OLIGOARTICULAR ARTHRITIS

- Four or fewer joints during the first 6 months of disease.
- Knees are most commonly affected followed by the ankles.

- Normal or moderately increased acute phase reactants, positive ANA, and HLA association.
- 30% with asymptomatic iridocyclitis.
- Mild systemic manifestations.
- May develop extended oligoarthritis (more than four joints involved after the first 6 months).
- Remission rates: 12% with extended disease and 75% with persistent disease.

SYSTEMIC ONSET DISEASE

- It occurs throughout childhood.
- Diagnosis: arthritis accompanied or preceded by at least 2 weeks of quotidian fever plus typical evanescent rash (discrete, circumscribed, salmon-colored, 2- to 10-mm macules on the trunk, and proximal extremities that typically coincide with fever), hepatomegaly, splenomegaly, generalized lymphadenopathy, or serositis –
 - joint disease may be delayed obscuring the diagnosis.
- Ill-appearance, myalgias, and abdominal pain during episodes of fever.
- Leukocytosis with a preponderance of neutrophils, elevated liver enzymes, microcytic anemia, thrombocytosis, very high ESR and CRP, and negative ANA titer.
- Prognosis: 50% with relapsing-remitting course, 50% with unremitting, and severe arthritis.
- Complications: pericarditis, pericardial effusion, myocarditis, pleuritis, and amyloidosis:
 - Rarely patients develop macrophage activation syndrome (sustained fever, pancytopenia, hepatosplenomegaly, liver insufficiency, coagulopathy, and neurologic symptoms).

DIFFERENTIAL DIAGNOSIS

- ARF, SLE, bacterial arthritis, reactive arthritis, and neoplastic diseases.

DIAGNOSTIC EVALUATION

- Complete blood count, renal function studies, liver function tests, ANA, RF, ESR, and CRP.
- Rapid streptococcal screen.
- Electrocardiogram and echocardiogram with evidence of myocarditis or pericarditis.

TREATMENT

- Physical and occupational therapy, splints.
- NSAIDs, intra-articular steroids, glucocorticoids, cytotoxic drugs, or biological medications.
- Extended oligoarticular JIA: sulfasalazine.
- Severe pericarditis or myocarditis: prednisone.
- Macrophage activation syndrome: high-dose steroids and cyclosporine.
- Indicators of poor outcome include greater severity or extension of arthritis at onset, symmetrical disease, early wrist or hip involvement, presence of RF, persistent active disease, and early radiographic changes.

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QUESTIONS

1. A 4-year-old presents with a limp for the past 2 days. He has been otherwise well without any fever. On examination, he is lying on the examining table with his left hip flexed, abducted, and externally rotated. You are able to move his hip without much difficulty although he does resist extreme internal rotation. His WBC count, ESR, and X-ray are normal. Of the following, which is the most likely etiology of his limp?
 - A. Osteomyelitis
 - B. Septic arthritis
 - C. Transient synovitis
 - D. Leukemia
 - E. Slipped capital femoral epiphysis
2. Treatment for the above patient includes which of the following?
 - A. Steroids
 - B. Complete bedrest
 - C. Re-evaluation in 12–24 hours
 - D. Emergent orthopedic consultation
 - E. Antibiotics
3. A 15-year-old female comes in with concerns about a rash. She developed a rash across her nose and cheeks after spending the day at the beach. She has had a similar rash previously that has resolved spontaneously. On examination, she has a malar rash and mucositis. Which of the following are true of her illness?
 - A. Skin, musculoskeletal, and renal systems are commonly involved
 - B. CNS disease is the leading cause of morbidity and mortality
 - C. Diagnosis is confirmed with a positive ANA titer
 - D. The illness is usually mild and self-limited
 - E. Renal flares are heralded by fever
4. Treatments for the above patient may include which of the following options?
 - A. Steroids
 - B. NSAIDs
 - C. Immunosuppressants
 - D. Biologic agents
 - E. All of the above
5. A 7-year-old presents with complaints of knee and wrist pain. He had been complaining of elbow pain but that is now improved. He had a sore throat about a month ago that resolved spontaneously but

- has otherwise been well. He is afebrile with some swelling and redness of both knees and his left wrist. He has a mitral insufficiency murmur and tachycardia. His chest x-ray shows cardiomegaly. Which of the following is true?
- Antibiotics are not required
 - Joint symptoms resolve within a month and leave no permanent damage
 - Fever is required to meet diagnostic criteria
 - Arthritis is deforming
 - Evidence of an antecedent GAS infection is always required to make the diagnosis
6. Treatment of the patient above includes which of the following options?
- Antibiotics
 - Haldol
 - Prednisone
 - A and B
 - A and C
7. An 11-year-old presents 2 weeks after a short bout of diarrhea. He had been doing well until yesterday when he developed redness and swelling to his right knee, redness to his eyes, and pain with urination. He is able to walk but is complaining of pain. Which of the following are true?
- His current symptoms were triggered by a GI infection with Norwalk virus
 - Symptoms can start as late as 4 months after the inciting infection
 - The associated arthritis is an oligoarticular, sterile synovitis
 - Most patients have complete resolution of symptoms within 6 weeks
 - This illness is also associated with gonococcal infection
8. A 10-year-old presents to the ED with knee pain. His mother is frustrated because he just got over another illness that included a sore throat. He has also been complaining of diffuse joint aches that do not seem to improve with ibuprofen. You suspect that he has poststreptococcal reactive arthritis (PSRA). Which of the following is true regarding this illness?
- It develops 3–14 days after pharyngitis
 - Anti-Strep. antibodies will be positive
 - It is typically a migratory arthritis of large joints
 - There is no association with carditis
 - It is associated with an elevated WBC count and a normal ESR
9. A 6-year-old presents with several days of fever. He had been diagnosed with systemic onset Juvenile Idiopathic Arthritis (JIA) a year earlier. His typical fevers had been transient and predictable but this fever has lasted 2 days and he is more ill-appearing than usual. You are concerned that he has macrophage activation syndrome. Which of the following is true?
- Systemic onset JIA requires 1 week of quotidian fever and rash to be diagnosed
 - The rash of systemic onset JIA is typically petechial
 - Macrophage activating syndrome is associated with pancytopenia, neurologic symptoms, and coagulopathy
 - Children with systemic onset JIA have leukopenia and thrombocytopenia
 - Systemic onset JIA is easily diagnosed in the ED
10. Macrophage activation syndrome may be treated with which of the following?
- High-dose salicylates
 - High-dose steroids
 - Sulfasalazine
 - Cyclosporine
 - B and D

ANSWERS

- C. The child is well appearing with normal temperature, WBC count, ESR, and x-ray. Children with osteomyelitis would have fever and elevated ESR or WBC count. Children with septic arthritis have severe pain with joint movement, fever, and elevated WBC count and ESR. Patients with leukemia may also have anemia and thrombocytopenia. Patients with SCFE tend to be older and have abnormal x-rays.
- C. Re-evaluation in 12–24 hours and then in 10–14 days if symptoms persist. Steroids are not beneficial in toxic synovitis. Treatment is with rest via nonweight bearing; bedrest is required only for severe cases. Orthopedic evaluation is only necessary for atypical or prolonged courses. Antibiotics have no role in transient synovitis.
- A. In pediatric SLE, skin, musculoskeletal, and renal systems are most commonly involved. CNS disease is the second leading cause of morbidity and mortality; renal disease is the leading cause. ANA titers can be positive in up to 30% of well children. Anti-ds DNA is more specific to SLE and is present in 60–70% of SLE patients. SLE is progressive. Renal flares are heralded by proteinuria prior to the onset of systemic features.

4. E. All of the above. Treatment is determined by the organ involvement and severity of the disease.
5. B. Joint symptoms of acute rheumatic fever (ARF) resolve within a month and leave no permanent damage. Antibiotics are used to eradicate GABHS colonization and prevent re-infection. Fever is a minor JONES criteria and may or may not be present with ARF. Arthritis is nondeforming. Chorea and indolent carditis do not require evidence of antecedent GABHS infection.
6. E. In the presence of carditis, prednisone is added to the treatment of ARF. Haldol is used to treat Sydenham chorea.
7. C. The arthritis of reactive arthritis tends to be a sterile arthritis. Reactive arthritis is triggered by GI infections with *Salmonella*, *Yersinia*, *Campylobacter*, or *Shigella*. Symptoms of reactive arthritis begin 1–4 weeks after the inciting infection. Half of patients with reactive arthritis have resolution of symptoms within 6 months; the rest have a chronic, relapsing reactive arthritis. Reactive arthritis is triggered by a GU infection with *C trachomatis*.
8. A. Poststreptococcal reactive arthritis develops 3–14 days after pharyngitis. Due to the early onset of symptoms of PSRA after the inciting infection, antibodies may not be present. The arthritis of PRSA is a nonmigratory arthritis of small joints, large joints, and axial joints. It responds poorly to anti-inflammatories and lasts from 1 week to 8 months. 5% of patients with PSRA develop carditis. PSRA is associated with a normal WBC count and an elevated ESR.
9. C. Macrophage activating syndrome is associated with pancytopenia, neurologic symptoms, and coagulopathy. Systemic onset JIA requires 2 weeks of quotidian fever and rash to be diagnosed. The rash of systemic onset JIA is typically evanescent rash (not petechial) (discrete, circumscribed, salmon-colored, 2- to 10-mm macules on the trunk, and proximal extremities that typically coincide with fever). Children with systemic onset JIA have leukocytosis and thrombocytosis (not leukopenia and thrombocytopenia). Systemic onset JIA is NOT easily diagnosed in the ED. The fever may precede other findings often resulting in a delayed diagnosis.
10. E. Macrophage activation syndrome requires treatment with both high-dose steroids and cyclosporine. Treatment should be started in consultation with a rheumatologist.

106 NONMALIGNANT TUMORS OF BONE

Kemedy K. McQuillen

INTRODUCTION

- A number of histologically benign bone tumors present in childhood.
- They may be found as an incidental finding on radiographs or they may present with pain related to the tumor itself or from an associated pathologic fracture.

OSTEOID OSTEOMAS

- Presents between 5 and 20 years old and commonly involves the long bones of the lower extremities.
- Most common presentation: nocturnal pain that is exquisitely responsive to NSAIDs.
- Patients may have point tenderness, a swollen limb, a tender palpable mass, a painless limp, growth disturbance, angular deviations (if near a growth plate), scoliosis, torticollis, hyperlordosis, or kyphoscoliosis (if in the spine).
- Radiographs: small (<1 cm) radiolucent round or oval areas of osteolysis (nidus) surrounded by a regular ring of bony sclerosis (Fig. 106-1). Total size rarely exceeds 1.5 cm.
 - Cortical diaphyseal lesion: oblong thickening to one side of the shaft without obvious nidus;
 - Medullary lesion: osteosclerosis may be minimal or absent;
 - Spine lesions: involve the posterior elements and may be difficult to visualize.
- Computed tomography (CT) and bone scintigraphy preferred over MRI for advanced imaging.
- Osteoid osteomas tend to be nonprogressive and resolve over several years.
- Medical management with prolonged use of NSAIDs +/- surgical excision.

NONOSSIFYING FIBROMAS (NOF)

- Also known as fibrous cortical defects, metaphyseal fibrous defects, and cortical desmoids.
- Present in up to 40% of preadolescents and adolescents and may occur in multiple bones.
- Usually discovered incidentally but may present with pain from a pathologic fracture.
- Radiographs: metaphyseal cortical eccentric lucency with sharp margins and surrounding sclerosis
 - May be multilocular and expansile with extension into the medullary bone;



A



B

FIG. 106-1. Osteoid osteoma of the proximal tibia.

- Long axis of the fibroma parallels the long axis of the bone;
- Atypical appearance if imaged when filling in with normal bone during adolescence.
- Majority do not require treatment and recurrence is rare.

OSTEOCHONDROMAS (CARTILAGINOUS EXOSTOSES)

- Most common benign bone tumor in children and adolescents.
- Occur in the metaphysis of long bones (proximal tibia, proximal humerus, and distal femur).
- Cartilaginous in growing children, enlarge during skeletal growth, and ossify at skeletal maturity.
- Present as a single, bony, nonpainful mass in patients between 5 and 15 years of age.
- Radiographs: sessile or pedunculated projections arising from the surface of the bone usually directed away from the joint (Fig. 106-2)
 - Cartilage cap not seen on x-ray so it is smaller on radiographs than on physical examination.
- CT or MRI: cortex and marrow space of the bone are continuous with the lesion.



FIG. 106-2. Osteochondroma of the proximal humerus.



FIG. 106-3. Enchondroma of the pelvis.

- Negligible risk of malignant transformation unless associated with multiple hereditary exostoses (rare autosomal dominant disorder with multiple osteochondromas).
- Resection for cosmetics, pain relief, growth retardation, vascular obstruction, or pseudoaneurysm.

ENCHONDROMAS

- Benign lesion of hyaline cartilage occurring centrally in the bone primarily affecting the hands.
- Usually asymptomatic but may present as a mass or pathologic fracture.
- Radiographs: single, radiolucent, sharply marginated, medullary canal lesion with bone thinning and cortical bulge (Fig. 106-3). Punctate or stippled calcification may be seen in adults.
- Low risk for malignant transformation unless associated with Ollier disease (multiple enchondromas, bony dysplasia, short stature, limb length abnormalities, and joint deformity) (Fig. 106-4) or Maffucci syndrome (enchondromas and soft tissue angiomas).
- Curettage and bone grafting for large or symptomatic lesions.

UNICAMERAL BONE CYSTS (SIMPLE OR MULTILOCULATED BONE CYSTS)

- Expansile, fluid-containing defects of tubular and flat bones (proximal humerus/proximal femur).
- Mean age at discovery: 9 years (range: 3–14 years) with males affected more frequently.
- Start near the epiphyseal plate and extend toward the diaphysis during growth.
- Active in children <10 years: active cysts have a higher recurrence rate than inactive cysts.



FIG. 106-4. Pelvis radiograph in a patient with Ollier's disease. Note the stippled calcification.

- Found incidentally or present with pain, limp, failure to use the extremity or pathologic fracture.
- Radiographs:
 - Long bones—centrally located lytic lesions in the metaphyseal medullary cavity with expansion in all directions and an expanded, thinned but unpenetrated cortex (Fig. 106-5);



FIG. 106-5. Unicameral bone cyst of the distal tibia.



A



B

FIG. 106-6. Aneurysmal bone cyst of the proximal fibula. Note the faint trabeculations that give it a “soap bubble” appearance.

- Flat bones—centered between the inner and outer tables of the ilium and mid-portion of the superior pubic ramus;
- No periosteal reaction unless there is an associated fracture;
- “Fallen leaf” sign—pathognomonic of a multiloculated bone cyst and represents a broken piece of cortex that has fallen into the fluid-containing cavity of the cyst.
- Treatment goal is a functionally stable bone; usually requires excision or injection into the lesion.
- Faint trabeculations give the tumor a “soap bubble” appearance.
- ABCs are filled with blood and, on cross-sectional imaging, fluid–fluid levels may be seen.
- Treatment: curettage and bone grafting or cementation +/- preoperative embolization.
- Low-dose radiation therapy for incompletely resectable, aggressive, or recurrent ABCs.
- Recurrence in 10–50% usually within 2 years: more common in younger patients.

ANEURYSMAL BONE CYSTS (ABCS)

- Rare, rapidly growing, destructive bone tumor that presents in the first two decades of life.
- Commonly solitary with involvement of the humerus, femur, tibia, fibula, pelvis, or spine.
- Presents with pain, mass, neurologic deficit (with spine location), or pathologic fracture.
- Radiographs: eccentric, fusiform, aneurysmal dilation with a thin rim of cortex (Fig. 106-6):
 - Long bones—aggressive lytic bony destruction with laminated periosteal reaction;

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QUESTIONS

- Which of the following is true regarding osteoid osteomas?
 - Most commonly involve the long bones of the lower extremities
 - Are associated with nocturnal pain that is not responsive to NSAIDs
 - Show homogeneous bony sclerosis on x-ray
 - Are best visualized with MRI
 - Are treated surgically
- A 10-year-old presents with pain to the forearm and you obtain a radiograph to evaluate for bony injury. It shows a metaphyseal cortical eccentric lucency with sharp margins and surrounding sclerosis. This lesion is most likely which of the following?
 - Aneurysmal bone cyst
 - Unicameral bone cyst
 - Osteochondroma
 - Nonossifying fibroma
 - Osteoid osteoma
- A 16-year-old presents with arm pain and a mass. X-ray shows an eccentric, fusiform dilation of the humerus. The lesion has a thin rim of cortex and fine trabeculations. You tell the patient that this lesion requires which of the following management options?
 - Can be managed medically
 - Will not require orthopedic follow-up
 - Will never recur
 - May cause significant morbidity
 - Is never solitary
- A 10-year-old male presents with a lump on his arm. You palpate a hard, nonmobile mass in the upper arm. X-ray shows a pedunculated growth that is smaller than it seems on examination. Which of the following is true regarding this lesion?
 - Is the most common benign bone tumor in children and adolescents
 - Usually invades the joint space
 - Is entirely made of bone during childhood
 - Is not continuous with the bone
 - Has a high risk of malignant transformation
- A 9-year-old male presents with a limp and tenderness of the proximal femur. Your radiologist reports a bone tumor with a “fallen leaf sign.” This radiographic sign represents which of the following?
 - Is seen in multiple benign and malignant bone tumors
 - Represents fluid layering in the tumor
 - Is pathognomonic of a multiloculated bone cyst
 - Is usually discovered in late adolescence
 - Is part of a process that is always managed medically
- You are talking about the operative management of bone tumor with the parents of an 11-year-old who presented with upper arm pain. His x-ray shows a large, eccentric, fusiform tumor in the humerus with a thin rim of cortex. You tell them surgery may require preoperative embolization of the tumor. You have diagnosed their son with which of the following bony abnormalities?
 - Unicameral bone cyst
 - Aneurysmal bone cyst
 - Nonossifying fibroma
 - Enchondroma
 - Osteochondroma
- Which of the following are least likely to require surgical treatment?
 - Aneurysmal bone cyst
 - A large multiloculated bone cyst of the femur
 - An osteochondroma that is impinging on the femoral vein
 - A nonossifying fibroma that was found incidentally
 - An osteoid osteoma that is causing severe pain despite NSAIDs

ANSWERS

- A. Osteoid osteomas most commonly involve the long bones of the lower extremities. They are associated with nocturnal pain that is extremely responsive to NSAIDs. Osteoid osteomas are not homogeneously sclerotic. They show a small (<1 cm) radiolucent round or oval area of osteolysis (nidus) surrounded by a regular ring of bony sclerosis. They are best visualized with plain radiographs and are rarely treated surgically.
- D. This is a classic appearance for a nonossifying fibroma. Aneurysmal bone cysts are typically eccentric, fusiform, aneurysmal dilation with a thin rim of cortex and may have a periosteal reaction. Unicameral bone cysts of the long bones show concentrically located lytic lesions in the metaphyseal medullary cavity with expansion in all directions and an expanded, thinned but unpenetrated cortex. In flat

bones, they are centered between the inner and outer tables of the ilium and the midportion of the superior pubic ramus. Osteochondromas present as sessile or pedunculated projections arising from the surface of the bone usually directed away from the joint. Osteoid osteomas are a small (<1 cm) radiolucent round or oval area of osteolysis (nidus) surrounded by a regular ring of bony sclerosis. Total size rarely exceeds 1.5 cm.

3. D. This patient has an aneurysmal bone cyst (ABC). They can cause significant morbidity. ABCs are managed surgically. Biopsies are usually taken at that time to confirm the diagnosis and orthopedic follow-up will be required. The recurrence rate is 10–50% and recurrences tend to occur within 2 years. ABCs are commonly solitary.
4. A. This is an osteochondroma which is the most common benign bone tumor in children and adolescents. Osteochondromas usually are directed away from the joint space. Osteochondromas are made of cartilage during childhood. The cartilage does not appear on radiographs so the lesion is often larger on exam than on x-ray. The cortex and marrow space of the bone are continuous with the osteochondroma. Osteochondromas have a negligible risk of malignant transformation unless associated with multiple hereditary exostoses (rare autosomal dominant disorder with multiple osteochondromas).
5. C. The “fallen leaf” sign is pathognomonic of a multiloculated bone cyst and represents a broken piece of cortex that has fallen into the fluid-containing cavity of the cyst. Unicameral bone cysts have a mean age at discovery of 9 years (range: 3–14 years). The treatment goal for multiloculated, unicameral bone cysts is a functionally stable bone. Effective treatment usually requires excision or injection into the lesion.
6. B. Aneurysmal bone cysts are filled with blood and, on cross-sectional imaging, fluid–fluid levels may be seen. Surgical repair may be complicated by intraoperative bleeding and preoperative embolization is sometimes required. Unicameral bone cysts are not associated with excessive bleeding during surgical repair. Nonossifying fibromas generally do not require surgery. Enchondroma only require curettage and bone grafting for large or symptomatic lesions. Osteochondromas only require resection for cosmetic appearance, pain relief, growth retardation, vascular obstruction, or pseudoaneurysm. Resection is not complicated by bleeding.
7. D. Nonossifying fibromas rarely require resection. Aneurysmal bone cysts typically require surgical intervention. A large multiloculated bone cyst of the femur will require surgical intervention to allow for a functionally stable bone. Osteochondromas require resection for cosmetic appearance, pain relief, growth retardation, vascular obstruction, or pseudoaneurysm. Osteoid osteomas that are causing severe pain despite NSAIDs or osteoid osteomas in patients who are intolerant of NSAIDs require surgical resection.

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107 GENERAL APPROACH TO THE POISONED PEDIATRIC PATIENT

Timothy B. Erickson

EPIDEMIOLOGY

- Two-thirds of poisonings reported to the American Association of Poison Control Centers (AAPCC) children and adolescents. One-third are in children under 3 years of age and one-half occur in children younger than 6 years.
- Most exposures are accidental ingestions and result in minimal toxicity.
- Poisonings also result from inhalation, intravenous, dermal, ocular, and environmental exposure.

HISTORY

- History includes the toxin or medication, time of the exposure, what other medications were available, and amount taken. It is prudent to assume the worst-case scenario.

PHYSICAL EXAMINATION

- A comprehensive physical examination may provide valuable clues regarding the ingestion or exposure.
- Many agents have specific effects on the heart rate, temperature, blood pressure, and respiratory rate. Monitoring the vital signs may help in proper diagnosis (Table 107-1).
- The level of consciousness, pupillary size, and potential for coma or seizures may be directly affected in a dose-dependent fashion (Tables 107-2 and 107-3). Other diagnostic clues are obtained in

the skin exam and breath odor (Tables 107-4 and 107-5).

- Several toxins consistently present with recognizable patterns or signs. Recognizing these toxic syndromes or toxidromes may expedite not only the diagnosis of the toxic agent, but also its management (Table 107-6).

DIAGNOSTIC AIDS AND LABORATORY

- In children with significant poisonings, baseline laboratory studies include a complete blood cell count, renal functions, electrolytes, glucose, and arterial blood gases.
- In children with known ingestion demonstrating no overt signs of toxicity, a more selective approach to diagnostic studies is acceptable.
- If a venous or arterial blood gas value reveals a metabolic acidosis, calculating the anion gap can assist in formulating a differential diagnosis.
- A metabolic acidosis with an increased anion gap results from organically active acids and is characteristic of several toxins and various other disease states (Table 107-7). The anion gap can be calculated from serum electrolytes: anion gap calculation = $\text{Na} - (\text{Cl} + \text{HCO}_3)$. The normal anion gap ranges from 8 to 12 mEq/L.
- If ingestion of a toxic alcohol, such as methanol or ethylene glycol, is suspected, calculation of the osmolal gap is calculated (Table 107-8).

$$\text{Calculated osmolality} = 2(\text{Na}) + \text{glucose}/18 + \text{BUN}/2.8 \text{ ETOH}/4.6$$

$$\text{Osmolal gap} = \text{measured osmol} - \text{calculated osmol (normal } <10)$$

- When a particular drug or toxin is known or highly suspected, drug testing may confirm the ingestion and guide medical management.

TABLE 107-1 Diagnosing Toxicity from Vital Signs

Bradycardia (PACED) Propranolol (β -blockers), poppies (opiates), propoxyphene, physostigmine Anticholinesterase drugs, antiarrhythmics Clonidine, calcium channel blockers Ethanol or other alcohols Digoxin, digitalis	Hypotension (CRASH) Clonidine, calcium channel blockers Rodenticides (containing arsenic, cyanide) Antidepressants, aminophylline, antihypertensives Sedative-hypnotics Heroin or other opiates
Tachycardia (FAST) Free base or other forms of cocaine, Freon Anticholinergics, antihistamines, antipsychotics amphetamines, alcohol withdrawal Sympathomimetics (cocaine, caffeine, amphetamines, PCP), solvent abuse, strychnine Theophylline, TCAs, thyroid hormones	Hypertension (CT SCAN) Cocaine Thyroid supplements Sympathomimetics Caffeine Anticholinergics, amphetamines Nicotine
Hypothermia (COOLS) Carbon monoxide Opioids Oral hypoglycemics, insulin Liquor (alcohols) Sedative-hypnotics	Rapid respiration (PANT) PCP, paraquat, pneumonitis (chemical), phosgene ASA and other salicylates Noncardiogenic pulmonary edema, nerve agents Toxin-induced metabolic acidosis
Hyperthermia (NASA) Neuroleptic malignant syndrome, Nicotine Antihistamines, alcohol withdrawal Salicylates, sympathomimetics, serotonin syndrome Anticholinergics, antidepressants, antipsychotics	Slow respiration (SLOW) Sedative-hypnotics (barbiturates, benzos) Liquor (alcohols) Opioids Weed (marijuana)

- Toxicology screening can be helpful in diagnosing unknown ingestions but has limitations; blood toxicology screens may be negative if the drug has a short half-life and the specimen is not obtained immediately after the exposure.
- The urine toxicology screen is of greater value, since the drug's metabolites continue to be excreted in the urine for 48 to 72 hours following the ingestion.
- Toxicology panels typically screen for drugs of abuse such as opioids, amphetamines, cannabinoids, phencyclidine (PCP), and cocaine. However, most of these screens are qualitative; therefore, detection of a drug does not necessarily entail toxicity.
- Do not assume a child ingested nothing simply because the toxicology screen is reported as negative and the actual drug ingested has not been included in the screen.
- Radiological testing can help diagnose toxins that are radiopaque on abdominal studies or have symptoms of noncardiogenic pulmonary edema or chemical pneumonitis (Table 107-9).

TABLE 107-2 Agents that Affect Pupil Size

Miosis (COPS) Cholinergics, clonidine, carbamates Opiates, organophosphates, Phenothiazines (antipsychotics), pilocarpine, pontine hemorrhage Sedative-hypnotics	Mydriasis (SAW) Sympathomimetics Anticholinergics Withdrawal
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MANAGEMENT

STABILIZATION

- The cornerstone of management is supportive care, with particular attention to the airway, breathing, and circulation.
- Resuscitative measures are instituted prior to antidotal therapy or gastric decontamination (Fig. 107-1).

TABLE 107-3 Agents that Cause Coma or Seizures

COMA (LETHARGIC) L-Lead, lithium E-Ethanol, ethylene glycol, ethchlorvynol T-Tricyclic antidepressants, thallium, toluene H-Heroin, hemlock, hepatic encephalopathy, heavy metals, hydrogen sulfide, hypoglycemics A-Arsenic, antidepressants, anticonvulsants, antipsychotics, antihistamines R-Rohypnol (sedative hypnotics), risperidone G-GHB I-Isoniazid, insulin C-Carbon monoxide, cyanide, clonidine	SEIZURES (OTIS CAMPBELL^a) Organophosphates, oral hypoglycemics Tricyclic antidepressants Isoniazid, insulin Sympathomimetics, strychnine, salicylates Camphor, cocaine, carbon monoxide, cyanide, chlorinated hydrocarbons Amphetamines, anticholinergics Methylxanthines (theophylline, caffeine), methanol Phencyclidine (PCP), propranolol Benzodiazepine withdrawal, botanicals (water hemlock, nicotine), bupropion, GHB Ethanol withdrawal, ethylene glycol Lithium, lidocaine Lead, lindane
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^aFamous T. V. "town drunk" on the Andy Griffith Show.

TABLE 107-4 Agents that Cause Skin Signs

Diaphoretic skin (SOAP)	Flushed or red appearance
Sympathomimetics	Anticholinergics, niacin
Organophosphates	Boric acid
Acetylsalicylic acid or other salicylates	Carbon monoxide (rare)
Phencyclidine	Cyanide (rare)
Dry Skin	Cyanosis
Antihistamines, anticholinergics	Ergotamine
	Nitrates
	Nitrites
	Aniline dyes
	Phenazopyridine
	Dapsone
	Any agent causing hypoxemia, hypotension, or methemoglobinemia.
Bullae	Acneiform rash
Barbiturates and other sedative-hypnotics, Bites: Snakes and spiders	Bromides
	Chlorinated aromatic hydrocarbons

- When there is altered level of consciousness or hypoglycemia documented with a bedside glucose test, administer intravenous dextrose, or intramuscular glucagon.
- Naloxone is given to children or adolescents with lethargy or coma. Naloxone is a specific opiate antagonist with minimal side effects.
- Agitation and signs of withdrawal may develop in opiate-dependent adolescents or in neonates whose mothers are narcotic addicts or on methadone during pregnancy.
- Additional doses of naloxone are required for certain opiates, such as fentanyl, codeine, methadone, and propoxyphene, which have high potency and a prolonged half-life. If an intravenous line cannot be established, naloxone may be administered via the endotracheal tube, intramuscularly, intralingually, or subcutaneously.

TABLE 107-5 Odors that Suggest the Diagnosis

Odor	Possible Source
Bitter almonds	Cyanide
Carrots	Cicutoxin (water hemlock)
Fruity	Diabetic ketoacidosis, isopropanol
Garlic	Organophosphates, arsenic, dimethyl sulfoxide (DMSO), selenium
Gasoline	Petroleum distillates
Mothballs	Naphthalene, camphor
Pears	Chloral hydrate
Pungent aromatic	Ethchlorvynol
Oil of wintergreen	Methylsalicylate
Rotten eggs	Sulfur dioxide, hydrogen sulfide
Freshly mowed hay	Phosgene

TABLE 107-6 Common Toxidromes

Cholinergic	Opioid
Examples: organophosphates, carbamates, pilocarpine)	Examples: heroin, morphine, codeine, methadone, fentanyl, oxycodone, hydrocodone
(DUMBELLS)	Miosis
D iarrrhea, diaphoresis	Bradycardia
U rination	Hypotension
M iosis	Hypoventilation
B radycardia, bronchosecretions	Coma
E mesis	
L acrimation	
L ethargic	
S alivation	
Nicotinic	
M -Mydriasis	
T -Tachycardia	
W -Weakness	
T -Tremors	
F -Fasciculations	
S -Seizures	
S -Somnulent	
Anticholinergic	Withdrawal
Examples: antihistamines, cyclic antidepressants, atropine, benzotropine, phenothiazines, scopolamine	Diarrhea
Hyperthermia (HOT as a hare)	Mydriasis
Flushed (RED as a beet)	Goose flesh
Dry skin (DRY as a bone)	Tachycardia
Dilated pupils (BLIND as a bat)	Lacrimation
Delirium, hallucinations (MAD as a hatter)	Hypertension
Tachycardia	Yawning
Urinary urgency and retention	Cramps
	Hallucinations
	Seizures (with ETOH and benzodiazepine withdrawal)
Sympathomimetic	
Examples: cocaine, amphetamines, ephedrine, phencyclidine, pseudoephedrine	
Mydriasis	
Tachycardia	
Hypertension	
Hyperthermia	
Seizures	

TABLE 107-7 Agents Causing Metabolic Acidosis/Elevated Anion Gap

METAL
Methanol, metformin, massive overdoses
Ethylene glycol
Toluene
Alcoholic ketoacidosis,
Lactic acidosis
ACID
Acetaminophen (large ingestions)
Cyanide, carbon monoxide, colchicine
Isoniazid, iron, ibuprofen (large ingestions)
Diabetic ketoacidosis
GAP
Generalized seizure-producing toxins
Acetylsalicylic acid or other salicylates
Paraldehyde, phenformin

TABLE 107-8 Agents Increasing the Osmolar Gap

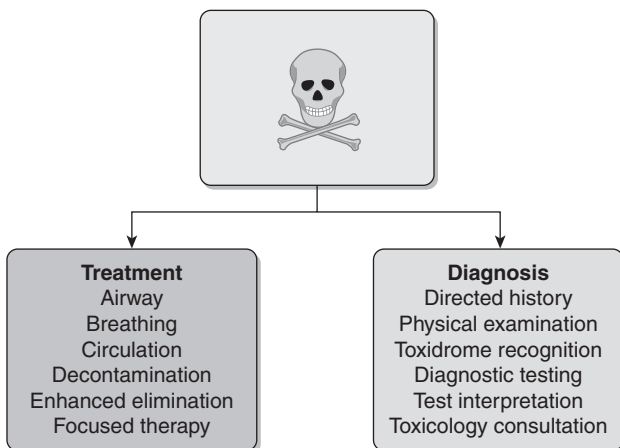
ME DIE
Methanol
Ethylene glycol
Diuretics (mannitol), diabetic ketoacidosis (acetone)
Isopropyl alcohol
Ethanol

GASTRIC DECONTAMINATION

- Gastric decontamination is not needed if the ingestion was nontoxic agent or a very small amount of a poison unlikely to cause toxicity.
- Gastric decontamination is recommended if the ingestion was recent and the child symptomatic or if the ingested toxin may cause delayed toxicity.
- Most treatment modalities have focused on stomach evacuation, while the ultimate goal should be to decontaminate the entire gastrointestinal tract.

INDUCTION OF EMESIS

- Historically, syrup of ipecac was the most commonly used gastric decontamination agent. Ipecac has now fallen out of favor in the emergency department (ED) and prehospital settings and is no longer advocated by the American Academy of Pediatrics or the American Association of Poison Centers for treatment of the acutely poisoned patient.
- Complications following ipecac include aspiration pneumonia, dehydration due to protracted vomiting, diaphragmatic rupture and death, Mallory–Weiss tears of the esophagus, and gastric rupture.

**FIG. 107-1.** General approach to the poisoned patient.**TABLE 107-9 Agents Visible on Abdominal Radiographs**

COINS
Chloral hydrate, cocaine packets, calcium
Opium packets
Iron, other heavy metals such as lead, arsenic, mercury
Neuroleptic agents
Sustained-released or enteric coated agents
DRUGS CAUSING ACUTE LUNG INJURY OR PULMONARY EDEMA
MOPS
Meprobamate, methadone
Opioids
Phenobarbital, propoxyphene, paraquat, phosgene
Salicylates

GASTRIC LAVAGE

- Gastric lavage mechanically removes toxins from the stomach through a large-bore orogastric tube, the opening of which is often too small to allow pill fragments to be removed. Gastric lavage does not remove toxic agents from the intestinal tract, where the majority of drug absorption occurs. No clinical trials evaluating the efficacy of gastric lavage in small children exist. Gastric lavage should not be routinely performed in overdosed pediatric patients.
- Consider gastric lavage if a patient has ingested a potentially life-threatening amount of a toxin and presents within one hour of ingestion. There may be some efficacy for lavage beyond 1 hour when the agent ingested slows gut motility, such as with anticholinergics or opioids, or when the toxin forms concretions, such as with iron and salicylates.
- Protect the airway protection with endotracheal intubation prior to lavage if there is a depressed level of consciousness to avoid aspiration pneumonitis. Weigh the risk of intubation against the potential benefits of gastric lavage in these cases.
- Gastric lavage should never be used as a punitive measure in cases of nontoxic overdoses or forced on pediatric patients who are combative or otherwise uncooperative.
- If the child has recently ingested an elixir or liquid, a simple nasogastric tube is adequate in order to avoid orogastric injury from traumatic insertion of a large bore tube.
- Gastric lavage is contraindicated in ingestions of most hydrocarbons, acids, alkalis, and sharp objects. Complications including aspiration, esophageal perforation, bleeding, electrolyte imbalance, and hypothermia have been described.

ACTIVATED CHARCOAL

- The majority of poisoned children who are not critically ill can be managed safely and effectively in the ED setting with charcoal alone.
- Activated charcoal is the most frequently used and most effective gastrointestinal decontamination agent. Evidence suggests that activated charcoal is more effective than induced emesis or gastric lavage for gastric decontamination.
- Activated charcoal can be administered rapidly and is most beneficial when administered within 1 hour after the ingestion. The absorptive properties of activated charcoal are effective beyond the gastric mucosa, absorbing drugs throughout the small intestine.
- While studies have demonstrated reduced drug absorption with activated charcoal use, it is important to note that there is no evidence that administration of activated charcoal ultimately improves patient outcome. Routine administration in nontoxic ingestions is not indicated.
- Although most sources recommend an activated charcoal dose of 1mg/kg, if the amount of drug ingested is known, a more accurate dose of activated charcoal can be calculated using a 10:1 ratio of charcoal to the ingested toxin.
- For some drugs, such as theophylline, phenobarbital, and carbamazepine, multiple dosing of activated charcoal may enhance elimination due primarily to enteroenteric circulation of the drug (Table 107-10).
- Repeated use of charcoal preparations premixed with cathartics such as sorbitol is to be avoided, since dehydration and electrolyte imbalance may result.
- Activated charcoal is the preferred mode of gastrointestinal decontamination when the history of the overdose or time of ingestion is unclear, since delayed administration may be beneficial, with minimal adverse side effects.

TABLE 107-10 Agents Responsive to Multiple Doses of Activated Charcoal

Substances adsorbable by activated charcoal (ABCD)

Antimalarials (quinine), Aminophylline (theophylline)

Barbiturates (phenobarbital)

Carbamazepine

Dapsone

Substances not adsorbable by activated charcoal (PHAILS)

Pesticides, potassium

Hydrocarbons

Acids, alkali, alcohols

Iron, insecticides

Lithium

Solvents

- Rare cases of vomiting, constipation, obstruction, and aspiration have been reported, but the incidence, particularly in pediatric cases, is extremely low.
- The administration of activated charcoal in the home or prehospital setting is gaining popularity as a superior substitute for syrup of ipecac.
- Activated charcoal is neither effective nor indicated in heavy metal poisonings, such as with iron, lithium or borates, ingestions of ethanol containing products, or following ingestion of acids or alkalis where gastric visualization or endoscopy may be required (Table 107-10).

CATHARTICS

- Cathartics are osmotically active agents that eliminate toxins from the gastrointestinal tract by inducing diarrhea.
- The most common agents are sorbitol, magnesium citrate, and magnesium sulfate. The efficacy of cathartic use in reducing the absorption or increasing the elimination of toxins has not been established. There is no published data demonstrating an improved outcome with cathartic use alone or combined with activated charcoal.
- In the pediatric population, cathartic agents have been reported to result in hypermagnesemia, severe dehydration, and electrolyte imbalances if used excessively or in repetitive doses. In young children, it is recommended that activated charcoal be administered with water only, without any cathartic agent.

WHOLE BOWEL IRRIGATION

- Whole bowel irrigation (WBI) is now used in the overdose setting to “flush” the toxin through the gastrointestinal tract and prevent further absorption.
- The solution used is nonabsorbable, isotonic polyethylene glycol electrolyte (PEG) solution that, unlike cathartic agents, does not appear to create fluid or electrolyte disturbances. The irrigation process is continued until the rectal effluent is clear, usually within 4 to 6 hours.
- There is currently no conclusive evidence that WBI improves clinical outcome of poisoned patients. Although WBI has been used in the pediatric population with minimal to no side effects.
- It has been effective in ingestions not well absorbed by activated charcoal such as iron tablets, lead paint chips, lithium, and sustained released calcium channel blockers. Use of WBI has been documented following cocaine and opiate packet ingestions.

TABLE 107-11 Antidotes and their Indications

Antidote	Indication (agent)
N-acetylcysteine	Acetaminophen
Ethanol/fomepizole (4-MP)	Methanol/ethylene glycol
Oxygen/hyperbarics	Carbon monoxide
Naloxone/Nalmefene	Opioids
Physostigmine	Anticholinergics
Atropine/pralidoxime (2-PAM)	Organophosphates
Methylene blue	Methemoglobinemia
Nitrites/Hydroxycobalamin	Cyanide
Deferoxamine	Iron
Dimercaprol (BAL)	Arsenic, lead
Succimer (DMSA)	Lead, mercury
CaEDTA	Lead
Fab fragments	Digoxin, Crotalids, Colchicine,
Glucagon	β -Blockers, Calcium channel blockers
Sodium bicarbonate	Tricyclic antidepressants
Calcium/insulin/dextrose	Calcium channel antagonists
Dextrose, glucagons, octreotide	Oral hypoglycemics

4-MP, 4-methylpyrazole; EDTA, calcium ethylenediamine tetraacetate; BAL, British antilewisite; DMSA, dimercaptosuccinic acid; 2-PAM, 2-pralidoxime.

- An abdominal radiograph demonstrating radioopacities can be followed in a serial manner to document the effectiveness of WBI.
- In agents well absorbed by charcoal, WBI is discouraged as it may actually decrease the efficacy of activated charcoal.

ANTIDOTES

- Although the majority of poisonings in the pediatric population respond to supportive care and gastric decontamination alone, there are a few toxins that require antidotes.
- The purpose of antidotal therapy is to reduce the agent's toxicity by inhibiting the toxin at the effector site or target organ, to reduce the toxin's concentration, or to enhance its excretion. The number of effective antidotes are limited (Table 107-11) and are not for indiscriminate use. Antidotal therapy should be used carefully and in clinical circumstances when specifically indicated.
- Newly approved antidotes include fomepizole for toxic alcohols, CroFab for pit viper envenomations, dextrose-insulin therapy for calcium channel blockers, and hydroxycobalamin administration for cyanide poisoning.

ENHANCED ELIMINATION

- Enhancing elimination is the process of removing a toxin from the body once absorption has already occurred.

TABLE 107-12 Toxins Accessible to Hemodialysis

UNSTABLE
Uremia
No response to conventional therapy
Salicylates
Theophylline
Alcohols (isopropanol, methanol)
Boric acid, barbiturates (phenobarbital)
Lithium
Ethylene glycol
Enhanced elimination by charcoal hemoperfusion
Theophylline
Barbiturates
Carbamazepine
Paraquat
Glutethamide

- Methods of enhanced elimination include multiple-dose activated charcoal, urinary alkalization, and extracorporeal elimination.
- Urinary alkalization involved the use of an intravenous sodium bicarbonate infusion and promotes urinary elimination of substances that are weak acids. It is important to maintain a normal potassium level when performing alkalization as appropriate alkalization cannot be achieved when hypokalemia is present.
- A common use of urinary alkalization is in the salicylate-poisoned patient. Another use may be in patients who overdose on phenobarbital.
- Urinary acidification is no longer recommended because of the high risk of myoglobinuria and rhabdomyolysis.

HEMODIALYSIS AND HEMOPERFUSION

- Drugs that may be adequately dialyzed include those with a low molecular weight, low volume of distribution, low protein binding, and high water solubility.
- Examples include isopropanol, salicylates, theophylline, uremia-causing agents, methanol, barbiturates, lithium, and ethylene glycol (Table 107-12).
- Theophylline is also responsive to charcoal hemoperfusion.
- In children with a severe overdose that may require dialysis early consultation with a nephrologist is critical.

DISPOSITION

- Admit unstable patients to the ICU.
- Observe nonsymptomatic patients in the ED for 6 to 8 hours as long as the ingested substances are known not to have delayed or prolonged action.

- If agents with delayed or prolonged actions were ingested, observe the patient for up to 24 hours. These agents include sustained release products, calcium channel antagonists, theophylline, lithium, methadone, lomotil, monoamine oxidase inhibitors, and oral hypoglycemic agents.
- Admit patients who have ingested a potentially dangerous toxin, are manifesting mild-to-moderate toxicity, require antidotal therapy, or have an unsafe home environment.
- Educate and counsel parents educated regarding proper poison prevention in the home.
- In adolescents with recreational drug abuse, encourage drug rehabilitation programs. If the adolescents are suicidal, obtain psychiatric consultation once they are stabilized.

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QUESTIONS

1. A 14-year-old male is brought in to the ED because he is suspected of having taken an unknown ingestion. He is noted to be cool to the touch and hypothermic when his temperature is taken. Which of the following toxic agents may produce hypothermia?
 - A. Salicylates
 - B. Cocaine
 - C. Amphetamines
 - D. Ethanol
 - E. Neuroleptics
2. A metabolic acidosis with an increased anion gap may be found with which of the following agents following a toxic exposure?
 - A. Mercury
 - B. Lead
 - C. Lithium
 - D. INH
 - E. Isopropanol
3. A 3-year-old child presents to the ED with altered mental status, agitation, tachycardia, fever, flushed skin, dilated pupils, and dry skin. Which of the following agents is the most likely cause of this toxidrome?
 - A. Methamphetamine
 - B. Ketamine
 - C. Diphenhydramine
 - D. Methadone
 - E. Phenobarbital
4. The majority of poisoned children who present following a recent ingestion and not critically ill can be managed safely and effectively in the ED setting with which of the following treatments?
 - A. Syrup of ipecac
 - B. Gastric lavage
 - C. Activated charcoal
 - D. WBI
 - E. Specific antidote therapy
5. Which of the following toxins is matched with its appropriate antidote?
 - A. Aspirin and *N*-acetylcysteine
 - B. Cyclic antidepressants and fab fragments
 - C. Cyanide and methylene blue
 - D. Organophosphates and naloxone
 - E. *B*-blockers and glucoagon

ANSWERS

1. D. Uncoupling of oxidative phosphorylation may lead to hyperthermia with severe salicylate intoxication. Ethanol, sedative hypnotics, and opioids all may lead to hypothermia. Cocaine and amphetamines are

- sympathomimetic drugs of abuse often producing hyperthermia. Neuroleptic agents may also produce hyperthermia secondary to neuroleptic malignant syndrome.
2. D. Agents that classically produce a metabolic acidosis with increased anion gap include iron, salicylates, cyanide, carbon monoxide, metformin, methanol, and ethylene glycol. (Isopropanol is a toxic alcohol that does not typically produce metabolic acidosis.) In addition, the classic triad of seizures, coma, and metabolic acidosis with an increased anion gap refractory to bicarbonate therapy and should alert the physician to the possibility of INH ingestion.
 3. C. This child is presenting with a classic anticholinergic toxidrome, which is most consistent with diphenhydramine (benadryl) ingestion. Other anticholinergic agents include cogentin, jimson weed, and cyclic antidepressants. Methamphetamines are sympathomimetics producing hyperthermia, hypertension, dilated pupils, and diaphoretic skin. Ketamine depresses the mental status, respiratory drive and classically causes drooling and nystagmus. Methadone and phenobarbital produce an opioid and sedative hypnotic syndrome, respectively.
 4. C. Syrup of ipecac is no longer recommended in the overdose setting. Single-dose activated charcoal is the most efficacious mode of gastric decontamination with the fewest side effects. WBI may be indicated following large iron, lead paint chip and drug packet ingestions. Gastric lavage may still be indicated following a potentially life-threatening overdose presenting within 1 hour of ingestion. The vast majority of poisonings in children resolve with observation and supportive care alone and specific antidote therapy is generally not necessary, save for a few specific agents.
 5. E. *N*-acetylcysteine is the antidote for acetaminophen poisoning. Cyclic antidepressants are effectively treated with sodium bicarbonate administration, while Fab fragments are indicated for digitalis toxicity. Cyanide is responsive to hydroxocobalamin and the cyanide antidote kit. Methylene blue is indicated for symptomatic methemoglobinemia. Organophosphates respond to atropine and praloxime (2-PAM), while naloxone is the antidote for the opioid overdose. Glucagon effectively reverses the clinical effects of *b*-blocker toxicity that includes bradycardia and hypotension.

Antipyretic Analgesics

108 ACETAMINOPHEN

Leon Gussow

- Death caused solely by acetaminophen ingestion is rare in the pediatric population. The small number of fatalities in this population can be explained by the availability of a very effective antidote, n-acetylcysteine (NAC); as well as the fact that young children are relatively resistant to acetaminophen-induced hepatotoxicity.

PATHOPHYSIOLOGY AND PHARMACOLOGY

- The therapeutic dose of acetaminophen in children is 15 mg/kg given every 4 to 6 hours, with a maximum recommended total daily dose of 75 mg/kg (or five doses). Therapeutic serum levels are 10 to 20 mcg/mL.
- Acetaminophen is eliminated primarily by hepatic pathways. After a therapeutic dose, 90% of the drug is metabolized to inactive sulfate and glucuronide conjugates. Less than 5% is excreted unchanged in the urine. The remaining 2% to 4% is metabolized by the cytochrome P450 mixed-function oxidase (MFO) system to the toxic intermediate NAPQI.
- In the presence of adequate hepatic stores of glutathione, NAPQI is rapidly converted to nontoxic conjugates. In overdose, the sulfate and glucuronide pathways become saturated, and increased amounts of acetaminophen are shunted through the MFO system. Glutathione becomes depleted and free NAPQI attacks hepatocytes, causing acute liver failure.
- The toxic dose of acetaminophen is generally considered to be 140 mg/kg.

CLINICAL PRESENTATION: THE FOUR STAGES OF ACETAMINOPHEN TOXICITY

STAGE I (0–24 HOURS AFTER INGESTION): GASTROINTESTINAL IRRITATION

- Patients in stage I may be asymptomatic, but young children frequently vomit after acetaminophen overdose.

STAGE II (24–48 HOURS AFTER INGESTION): LATENT PERIOD

- As nausea and vomiting resolve, patients appear to improve, but rising transaminase levels reveal evidence of hepatic necrosis.

STAGE III (72–96 HOURS AFTER INGESTION): HEPATIC FAILURE

- Severe hepatotoxicity presents with jaundice, hypoglycemia, renewed nausea and vomiting, right upper quadrant pain, coagulopathy, encephalopathy, hyperbilirubinemia, and markedly elevated transaminase levels.

STAGE IV (4–14 DAYS AFTER INGESTION): RECOVERY OR DEATH

- Patients who ultimately recover show improvement in laboratory parameters of hepatic function starting on about day 5, and go on to complete recovery. Less fortunate patients develop progressive encephalopathy, renal failure, coagulopathy, and hyperammonemia. The prognosis is poor for patients with these findings unless liver transplantation is performed.

LABORATORY STUDIES

- The Rumack–Matthew nomogram (Fig. 108-1) allows the clinician to predict the probability that hepatic toxicity will occur after single acute acetaminophen ingestion. A blood acetaminophen level is drawn 4 hours after the acute ingestion, or immediately if more than 4 hours have elapsed since the time of ingestion. This level is plotted on the nomogram against hours of post-ingestion.
- It is important to realize that this nomogram cannot be used in cases of chronic toxicity or when the time of ingestion is not well established.

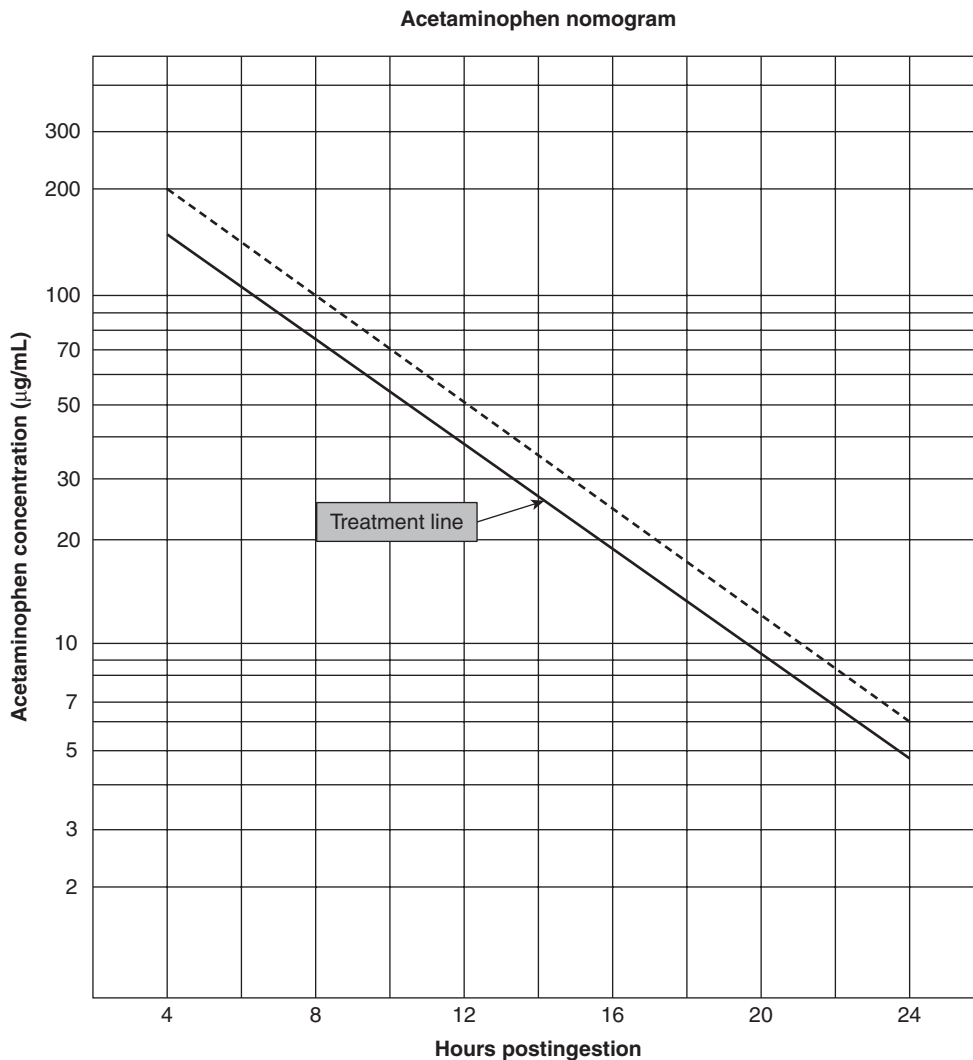


FIG. 108-1. The Rumack–Matthew nomogram predicts the probability that hepatic toxicity will occur after a single acute acetaminophen ingestion.

MANAGEMENT

GASTROINTESTINAL DECONTAMINATION

- Standard doses of activated charcoal can be given if the patient arrives within 1 hour of ingesting APAP alone, or if other toxic substances are also involved.

ANTIDOTE

- NAC helps restore the liver's ability to detoxify NAPQI and can prevent hepatonecrosis. It is most effective if started within 8 hours after an acute overdose.
- The Rumack–Matthew nomogram indicates which patients require treatment following a single,

acute APAP ingestion occurring at a known time (Fig. 108-1). It is current practice to treat any patient who has an APAP level that falls in the range of possible or probable hepatotoxicity with NAC.

- In cases where the patient has not taken a single acute overdose, but rather has taken multiple doses or overdoses of APAP, or has ingested APAP chronically, the nomogram cannot be used to predict hepatotoxicity or need for treatment with NAC.
- The oral NAC protocol long used in the United States for treating APAP toxicity consists of a loading dose of 140 mg/kg followed by 17 additional doses of 70 mg/kg given at 4-hour intervals.
- The commercial 20% solution (Mucomyst, Mead Johnson & Company) is unpalatable and should be diluted with three parts fruit juice or soda. If

vomiting occurs within 1 hour of treatment, the dose is repeated. Persistent vomiting that interferes with therapy can be suppressed with metoclopramide (0.25 mg/kg IV over 5 minutes) or ondansetron (0.15 mg/kg over 5 minutes).

- Some toxicologists believe that treatment with NAC should be continued, not for an arbitrary period of time or number of doses, but until a specific clinical end-point is reached. This end-point is often defined as: (1) acetaminophen level less than 10 mcg/mL, (2) alanine aminotransferase normal or clearly improving, and (3) patient is clinically well. This goal-oriented therapy can be used in all situations: acute overdose, repeated (chronic) supratherapeutic ingestion, and cases in which the time of ingestion and/or pattern of ingestion are unknown.
- Indications for the use of intravenous NAC (Acetadote, Cumberland Pharmaceuticals) include inability to tolerate oral NAC, intractable vomiting that does not respond to antiemetics, intestinal pathology (such as bowel obstruction or GI bleeding), encephalopathy, and neonatal acetaminophen toxicity secondary to maternal overdose. Some physicians prefer the IV preparation in almost all circumstances. When considering the use of IV NAC, it is good practice to seek toxicology consultation through the local poison center.

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QUESTIONS

1. A 14-year-old male presents after ingesting approximately twenty 500 mg acetaminophen tablets in a suicide attempt. In the management of this patient the toxic dose of acetaminophen is generally considered to be which of the following amounts?
 - A. 4 mg/kg
 - B. 14 mg/kg
 - C. 75 mg/kg
 - D. 140 mg/kg
 - E. 1 gm/kg
2. Which of the following statements about the stages of acute acetaminophen toxicity is true?
 - A. During stage I (0–24 hours postingestion), hypoglycemia may be a significant clinical problem.
 - B. During stage II (24–48 hours postingestion), patients may be asymptomatic.
 - C. During stage III (72–96 hours after ingestion), clinical improvement may be misleading.
 - D. During stage IV (4–14 days postingestion), continually rising transaminase levels are associated with progression to chronic liver disease.
 - E. None of the above.
3. The Rumack–Matthew nomogram can be used to determine need for treatment with NAC in which of the following situations?
 - A. 6-month-old who has received liquid acetaminophen hourly over the last 3 days for “fussiness.”
 - B. 4-year-old who may have ingested acetaminophen sometimes during the last 3 days while at her grandmother’s house.
 - C. 17-year-old who after breaking up with her boyfriend ingested a “handful” of acetaminophen tablets 5 hours before presenting to the emergency department.
 - D. 12-year-old with unexplained jaundice.
 - E. None of the above.
4. A 15-year-old female presents after ingesting several handfuls of 500 mg tablets of acetaminophen 6 hours prior to arrival. She is drowsy and has vomited multiple times. Her serum acetaminophen level is plotted on the Rumack–Matthew nomogram and found to be in the high-risk area. Which of the following is true regarding the management of this patient?
 - A. Standard doses of activated charcoal should be given.
 - B. NAC should be given in a single IV dose.
 - C. Oral NAC should not be given because the patient is vomiting and it will not be tolerated.

- D. Oral NAC should be given with a loading dose of 140 mg/kg followed by 17 doses of 70 mg/kg every 4 hours.
- E. NAC should always be given IV as oral NAC is unpalatable.

ANSWERS

1. D. The toxic dose of acetaminophen is generally considered to be 140 mg/kg.
2. B. During stage II of acute acetaminophen toxicity, patients seem to improve as the vomiting evident in stage I resolves. However, rising transaminase values reveal evidence of hepatic necrosis. Stage I manifests as gastrointestinal irritation, stage III as severe hepatotoxicity/hepatic failure and stage IV is either recovery or death.
3. C. The Rumack–Matthew nomogram can only be used in cases involving a single, acute acetaminophen overdose where the time of ingestion is known.
4. D. The oral NAC protocol used in the US for treating acetaminophen toxicity consists of a loading dose of 140 mg/kg followed by 17 additional doses of 70 mg/kg given at 4-hour intervals. If vomiting occurs within 1 hour of treatment, the dose is repeated. Persistent vomiting that interferes with therapy can be suppressed with antiemetics. Standard doses of activated charcoal can be given if the patient arrives within 1 hour of ingestion. The commercial NAC solution is unpalatable and should be diluted with three parts fruit juice or soda. Indications for the use of intravenous NAC include inability to tolerate oral NAC, intractable vomiting that does not respond to antiemetics, intestinal pathology (such as bowel obstruction or GI bleeding), encephalopathy, and neonatal acetaminophen toxicity secondary to maternal overdose.

109 ASPIRIN

Michele Zell-Kanter

PHARMACOKINETICS

- Aspirin's absorption can be delayed by the formation of concretions. Enteric-coated preparations will also have delayed absorption.
- Aspirin has a very narrow therapeutic range in higher doses because of altered pharmacokinetics.

- Ingestions of <150 mg/kg are generally nontoxic. Ingestions of 150 to 300 mg/kg can result in mild-to-moderate toxicity overdoses of >300 mg/kg can be lethal.

PATHOPHYSIOLOGY AND CLINICAL PRESENTATION

- Children have a quicker onset of toxicity from salicylate poisoning and exhibit more severe signs than adults.
- Patients may complain of tinnitus and impaired hearing.
- Monitor for tachypnea and anion gap metabolic acidosis.
- Fluid losses are especially severe in young children.
- Respiratory alkalosis may not occur.
- Agitation, delirium, seizures, and coma are signs of severe toxicity.
- Rhabdomyolysis can occur and can cause acute renal failure.
- Patients can develop noncardiogenic pulmonary edema
- Electrolyte abnormalities include hypo- or hypernatremia, hypokalemia, and hypocalcemia. Hypoglycemia is more common than hyperglycemia.
- Salicylate-induced ventricular dysrhythmias are infrequent, but indicative of a poor prognosis.

LABORATORY STUDIES

- Useful tests include complete blood count, serum electrolytes, creatinine, glucose, and arterial or venous blood gases.
- Plasma salicylate levels should be drawn upon presentation and repeated every 2 hours to ensure that the level is decreasing.
- Peak plasma levels following ingestion of enteric coated or extended release preparations will be delayed.
- Several products can be measured as salicylate (Table 109-1).
- Levels can continue to increase after they have leveled off, or decreased if a concretion is present.

TABLE 109-1 Products Measured as Salicylates

Acetylsalicylic acid (aspirin, ASA)
Bismuth subsalicylate
Choline salicylate
Magnesium choline salicylate (Trilisate)
Magnesium salicylate
Methyl salicylate
Salsalate (Disalcid)
Sodium salicylate

- Clinical findings are more predictive of toxicity than the plasma level in patients who chronically receive salicylate therapy. Generally, prognosis is worse with chronic toxicity.

MANAGEMENT

- Fluid replacement: use boluses of crystalloid at doses of 10 to 20 mL/kg until adequate perfusion is assured.
- Correction of metabolic disturbances: add potassium to the IV fluid as needed.
- Prevention of further absorption of the toxin: activated charcoal 1 g/kg po.
- Enhancement of elimination: systemic alkalization to achieve a urine pH of 7.5–8. Use sodium bicarbonate in an initial bolus of 1 to 2 mEq/kg followed by a bicarbonate drip titrated to the urine pH.
 - Obtain serial arterial or venous blood gases.
- Consider hemodialysis in extremely toxic patients and in patients with underlying disease states that will compromise the treatment.

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QUESTIONS

1. A 22-month-old male (15 kg) reportedly ingested an unknown amount of “childrens’ aspirin” about 4 hours ago. The child had appeared well until he started vomiting 30 minutes ago, which has since stopped. You draw blood to send off a salicylate level. The result is 9 mg/dL. What is the best option in managing this patient at this point?
 - A. As the child is no longer vomiting send him home because the level is nontoxic.
 - B. Observe two hours, if the child looks well and is no longer vomiting send him home.
 - C. Wait two more hours and repeat the salicylate level.
 - D. The clinical presentation should dictate the management and not the level therefore send the patient home.
 - E. There is no need to repeat the level, since serum salicylates should peak at 2 hours.
2. In the above stated case, a repeat salicylate level comes back at 18 mg/dL. This level is about 7 hours after ingestion. Which of the following is correct regarding this salicylate level?
 - A. This level represents a peak level since 7 hours is beyond the time it takes for aspirin to be absorbed.
 - B. The aspirin is still being absorbed possibly due to a concretion.
 - C. The aspirin is still being absorbed because of its anticholinergic effects.
 - D. Aspirin follows linear kinetics since the repeat level has doubled from the initial level.
 - E. The units are incorrect as the level should be 1.8 mcg/mL.
3. A 2-year-old boy is brought in by parents who found the child playing with an empty bottle of baby aspirin. The parents think there were about 10 tablets in the bottle. What is the significance of this information given by the parents?
 - A. This represents less than 150 mg/kg; therefore, this is a nontoxic ingestion.
 - B. If these are reliable parents, this is a nontoxic ingestion.
 - C. Aspirin tastes bad; therefore, this child would not have swallowed more than a few.
 - D. The history is notoriously incorrect.
 - E. The report is suspicious since you are wondering how the child could have removed the safety cap.
4. A 3-year-old male presents after ingesting multiple salicylate tablets and is initially awake and happy. During the ED course, the child appears increasingly somnolent. The initial salicylate level is 10 mg/dL. Which of the following statements is true regarding this patient?
 - A. Aspirin does not distribute into the brain until late in the presentation, so this is to be expected and does not affect management.
 - B. It is naptime, the somnolence has nothing to do with the ingestion.
 - C. You should wait for a repeat level before you decide what to do next.
 - D. You should begin a bicarbonate drip because aspirin causes a nonanion gap metabolic acidosis.

- E. You should begin a bicarbonate drip to increase the ionized fraction of salicylate and increase its elimination.
5. Which of the following is correct regarding aspirin toxicity?
- Aspirin causes an anion gap metabolic acidosis because of anaerobic metabolism.
 - Aspirin does not cause an anion gap acidosis.
 - Aspirin causes cardiogenic pulmonary edema.
 - Uncoupling of oxidative phosphorylation can result in hypothermia.
 - Aspirin causes no direct electrolyte abnormalities.
6. A 15-year-old female presents after ingesting a bottle of aspirin in a suicide attempt after getting into a fight with her boyfriend. Which laboratory studies will be helpful in the ED management of this patient?
- Electrolytes and LFT.
 - Electrolytes and INR.
 - Electrolytes, glucose, and LFT.
 - Electrolytes, glucose, and ABG or VBG.
 - Electrolytes, INR and LFT.
7. A 14-month-old male presents after ingesting approximately 15 baby aspirins after waking up from a nap. He presents within 45 minutes of the ingestion according to the baby sitter who brought him in to the ED. The major goal in the management of this patient is which of the following?
- Prevent further absorption with ipecac.
 - Prevent further absorption by gastric lavage.
 - Correct metabolic disturbances.
 - Prevent the ingestion from happening in the future.
 - Prevent further absorption using multiple dose activated charcoal.
8. Which of the following is most accurate in regards to the management of the patient from the previous question?
- A single dose of activated charcoal must be given to this child even if he is somnolent.
 - The goal in systemic alkalization is to increase the urine pH above 7.
 - A urine pH above 7 is easily obtainable irrespective of the serum potassium.
 - Potassium is exchanged for hydrogen in the tubular fluid.
 - Both B and D.
9. A 2-year-old presents with vomiting, agitation, and has a metabolic acidosis several hours after ingesting aspirin. A renal consult should be obtained for which of the following indications in this patient?
- If the patient has any underlying disease that may compromise salicylate elimination.
 - If the serum level is above 80 mg/dL.
 - If the patient develops noncardiogenic pulmonary edema.
 - If the patient develops congestive heart failure.
 - All of the above.
10. Which of the following is the most accurate statement regarding salicylate toxicity?
- In high doses, aspirin changes its pharmacokinetics to first order.
 - Tachypnea results from stimulation of the respiratory center.
 - Hemodialysis is equally effective to systemic alkalization in removing salicylate.
 - The only benefit of hemodialysis over alkalization is better fluid control.
 - Salicylate levels do not need to be repeated once you have two sequential levels that are trending down.

ANSWERS

- C. Salicylate levels can increase beyond 4 to 6 hours because a concretion may have formed or the aspirin product may have been enteric coated. Also, the time of ingestion may be incorrect. Choices A and B are incorrect since levels may still be increasing. Both levels and clinical presentation are important and should be consistent with each other. In non-overdose settings, salicylate absorption should peak within 4–6 hours.
- B. This level most likely indicates continued absorption from a concretion. A concretion or enteric-coated products will delay aspirin's absorption. Aspirin does not have anticholinergic effects. Aspirin follows first-order kinetics within the therapeutic range and zero-order kinetics in the toxic range. You must always check the units. Typically, salicylate levels are reported as mg/dL. Also, make sure that the clinical presentation is consistent with the reported level.
- D. Always be suspicious about the history, it may be intentionally or unintentionally incorrect. There may have been more than 10 tablets in the bottle. The parents may have had good intentions but were incorrect in estimating the amount of tablets in the bottle. Aspirin does taste bad but it is amazing what some kids will eat. The cap may not have been properly secured.

4. E. Clinically the child is worsening and this should not be ignored. Begin treatment and wait for a repeat level to return. Aspirin actually distributes more quickly to organs such as the brain, lung, and kidney in children than in adults. It is not possible to differentiate the etiology of the somnolence. Until proven otherwise, aspirin is the culprit. Aspirin causes an anion gap metabolic acidosis.
5. A. Uncoupling of the Krebs's cycle results in anaerobic metabolism. Therefore, aspirin does cause an anion gap metabolic acidosis. Aspirin causes noncardiogenic pulmonary edema, not cardiogenic. Uncoupling of oxidative phosphorylation results in hyperthermia, not hypothermia. Aspirin toxicity may cause hypo- or hypernatremia, hypokalemia, or hypocalcemia.
6. D. Electrolytes, glucose, and blood gases may be altered in aspirin toxicity. They will indicate the severity of toxicity and dictate management. Neither INR nor LFTs are altered in acute toxicity.
7. C. Treatment will not be effective unless electrolytes are appropriately replaced. Ipecac is not indicated in any exposure. Gastric lavage is not indicated in any exposure. Prevention is important but it is not the major goal in the acute treatment phase. Multiple dose activated charcoal has not consistently been shown to be effective in aspirin poisoning.
8. E. Salicylate is ionized at a urine pH of 7 and above 7 will be excreted. If the patient is hypokalemic, the kidney will hold on to potassium and exchange hydrogen for the potassium. It will not be possible to alkalinize the urine when hydrogen is being excreted. Potassium must be monitored and replaced. Activated charcoal should not be given to a somnolent child because of the risk of aspiration. There must be a normal serum potassium to achieve a urine pH above 7. Both B and D are correct.
9. E. Dialysis is indicated in all these circumstances. Systemic alkalinization is limited by the pulmonary edema/CHF. Hemodialysis is more effective at removing salicylate than alkalinization.
10. B. Aspirin's pharmacokinetics change to zero order when serum levels are above therapeutic. Hemodialysis is 3–5 times faster at removing salicylate compared to alkalinization. Electrolytes can be replaced and the pH can be monitored. Salicylate levels may rise even after you have two sequential levels that are trending down due to concretions or enteric-coated preparations.

110 NONSTEROIDAL ANTI-INFLAMMATORY DRUGS

Michele Zell-Kanter

CLINICAL PRESENTATION

- Patients who ingest NSAIDs typically exhibit only CNS or GI toxicity.
- Symptoms are generally seen within 4–6 hours of ingestion.
- CNS toxicity can include drowsiness, dizziness, and lethargy.
- Ponstel has a propensity to cause seizures. Other NSAIDs associated with seizures include piroxicam, naproxen, and ketoprofen.
- Headache is more likely to occur after ingestion of indomethacin than other NSAIDs.
- Aseptic meningitis has been reported with NSAIDs, most typically with ibuprofen.
- Symptoms of GI toxicity include nausea, vomiting, and epigastric pain, all of which can occur at therapeutic doses.
- Cardiovascular complications are generally limited to tachycardia and hypotension.
- Rare respiratory complications include hyperventilation and apnea.
- Renal toxicity is not associated with acute overdose.
- Long-term use of NSAIDs is associated with nephrotoxicity, including acute tubular necrosis, acute interstitial nephritis, and acute renal failure.
- Renal papillary necrosis has been reported in children being treated with NSAIDs for juvenile rheumatoid arthritis.
- Other long-term complications of NSAID use include hepatocellular injury and cholestatic jaundice.

LABORATORY STUDIES

- Useful laboratory tests include a complete blood count, electrolytes, glucose, creatinine, and coagulation profile.
- Overdose of NSAIDs has infrequently been associated with an anion-gap acidosis. For patients with severe clinical symptoms, an arterial blood gas is indicated.

MANAGEMENT

- Gastric decontamination with activated charcoal is indicated after the patient is stabilized if presentation is within 4 hours of ingestion.

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QUESTIONS

1. A 13 year-old female presents to the ED about 12 hours after ingesting an unknown quantity of ibuprofen as a suicide gesture. This is the second presentation after a similar ingestion. She is brought to the ED by her parents. The patient appears lethargic. Which of the following is the best option?
 - A. You should order an acetaminophen level because some patients cannot differentiate between the OTC analgesics.
 - B. You should order a salicylate level because some patients cannot differentiate between the OTC analgesics.
 - C. An ibuprofen level is of no value to you.
 - D. The patient's lethargy can be secondary to ibuprofen.
 - E. All of the above.
2. In the patient from the previous question you decide to administer activated charcoal. The patient begins vomiting soon after drinking the charcoal. Which of the following is the best option?
 - A. Wait 15 minutes before encouraging the patient to begin drinking the charcoal again.
 - B. Give the patient an antiemetic and then have the patient begin drinking the charcoal again.
 - C. Place a nasogastric tube to administer the charcoal.
 - D. Intubate the patient and then administer the charcoal.
 - E. Stop trying to administer the charcoal
3. A 15 year-old male presents after ingesting approximately fifty 200 mg ibuprofen tablets. What are the typical toxicities of acute NSAID overdose expected in this patient?
 - A. CNS or GI toxicity
 - B. Aseptic meningitis

- C. Seizures
 - D. Renal failure
 - E. Liver failure
4. Choose the most accurate statement regarding GI toxicity from NSAID ingestion.
 - A. Nausea, vomiting, and epigastric pain are only seen with large ingestions.
 - B. GI toxicity results from a direct irritant effect of NSAIDs on the GI mucosa.
 - C. GI toxicity results from inhibition of prostaglandin synthesis.
 - D. GI toxicity results from activation of the central vomiting center.
 - E. GI toxicity is specific for ibuprofen. Other NSAIDs do not cause GI toxicity.
 5. A 14 year-old female is taking daily ibuprofen for her juvenile rheumatoid arthritis. She is sent to the ED for abnormal renal function tests discovered during routine screening at her doctors office. Renal toxicity from chronic NSAID administration includes which of the following?
 - A. Acute tubular necrosis
 - B. Acute interstitial nephritis
 - C. Acute renal failure
 - D. Renal papillary necrosis
 - E. All of the above
 6. Complications from acute NSAID exposure include which of the following?
 - A. Hypertension
 - B. Bradycardia
 - C. Hypoventilation
 - D. Hypotension
 - E. Torsades des pointe
 7. Which of the following is a correct association of a specific NSAID with its toxic manifestation?
 - A. Tolmetin has a propensity for causing seizures
 - B. Indomethacin is associated with causing aseptic meningitis
 - C. Ibuprofen has a propensity for causing seizures
 - D. Ibuprofen is associated with causing aseptic meningitis
 - E. Mefenamic acid is associated with causing aseptic meningitis

ANSWERS

1. E. Some products contain a combination of OTC analgesics and some patients do not know the difference between the analgesics warranting levels of both salicylate and acetaminophen being drawn. CNS toxicity can include drowsiness, dizziness, and lethargy.

2. E. The interval between time of ingestion and treatment has been too long for charcoal to be effective. Charcoal will be of limited use since 12 hours have passed since the patient ingested the ibuprofen and it has already been cleared from the stomach. In addition, the patient presented lethargic and therefore may aspirate the charcoal. The morbidity associated with intubation is greater than the utility of the charcoal in this clinical setting.
3. A. Patients who ingest NSAIDs typically exhibit either CNS or GI toxicity. Aseptic meningitis has been reported but is a rare consequence. Seizures can occur but are rare. Renal failure is not associated with acute NSAID ingestion. It is associated with chronic ingestion. Hepatocellular injury and cholestatic jaundice have been associated with chronic NSAID use.
4. C. GI toxicity can be seen with therapeutic doses of NSAIDs. GI toxicity is thought to result from inhibition of prostaglandin synthesis. All NSAIDs can cause GI toxicity. Less GI toxicity is associated with celecoxib and meloxicam.
5. E. All of these are possible forms of renal toxicity due to NSAIDs.
6. D. Hypotension can occur secondary to volume depletion. Hypertension does not occur. Patients may develop tachycardia but not bradycardia; hyperventilation but not respiratory depression. NSAIDs are not associated with Torsades.
7. D. Ibuprofen is associated with aseptic meningitis. Mefenamic acid is known to cause seizures. Other agents associated with seizure activity are piroxicam, naproxen, and ketoprofen. Indomethacin is associated with headache.

Household Chemicals

111 TOXIC ALCOHOLS

Timothy B. Erickson

ETHANOL

- In addition to alcohol-containing beverages such as beer, wine, and hard liquors, children have access to over 700 ethanol containing medicinal preparations, colognes and perfumes and mouthwashes that can contain up to 75% ethanol.

PHARMACOKINETICS AND PATHOPHYSIOLOGY

- Ethanol undergoes hepatic metabolism via two metabolic pathways: Alcohol dehydrogenase and the microsomal ethanol-oxidizing system (MEOS). Alcohol dehydrogenase pathway is the major metabolic pathway and the rate-limiting step in converting ethanol to acetaldehyde.
- Nontolerant individuals metabolize ethanol at 10 to 25 mg/dL/h and alcoholics metabolize up to 30 mg/dL/h. Children can ingest large amounts of ethanol in relation to their body weight, resulting in rapid development of high blood alcohol concentrations. In children <5 years of age, the ability to metabolize ethanol is diminished due to immature hepatic dehydrogenase activity.

CLINICAL PRESENTATION

- Ethanol is a selective central nervous system (CNS) depressant at low concentrations, and a generalized depressant at high concentrations. Initially ethanol produces exhilaration and loss of inhibition, which progresses to lack of coordination, ataxia, slurred speech, gait disturbances, drowsiness, and, ultimately, stupor and coma.
- The intoxicated child may demonstrate a flushed face, dilated pupils, excessive sweating, vomiting and diarrhea, hypoventilation, hypothermia, and hypotension.

- Death from respiratory depression can occur at serum ethanol concentrations >500 mg/dL. Convulsions and death have been reported in children with acute ethanol intoxication due to alcohol-induced hypoglycemia. Hypoglycemia results from inhibition of hepatic gluconeogenesis and is most common in children <5 years of age.

LABORATORY

- In symptomatic pediatric patients who have suspected ethanol intoxication, the most critical laboratory tests are the serum ethanol and glucose concentrations.
- Although blood ethanol concentrations roughly correlate with clinical signs, treat the patient based on clinical manifestations. If the ethanol level does not correlate with the clinical picture, coingestants or other causes of altered mental status should be considered.

MANAGEMENT

- The majority of children with accidental acute ingestions of ethanol respond to supportive care. Attention is directed toward management of the patient's airway, circulation, and glucose status.
- Obtunded patients should receive intravenous dextrose after a specimen for blood glucose level is drawn. Alternatively, a bedside finger stick blood glucose determination can be immediately obtained and dextrose administered if hypoglycemia is documented. If children respond to glucose administration, serial glucose levels are followed to detect recurrent hypoglycemia.
- Unless children are comatose, or coingestion of another drug is suspected, gastric decontamination with a nasogastric tube is unnecessary. Activated charcoal is not efficacious in isolated ethanol ingestions.
- Hemodialysis increases ethanol clearance by three to four times, and may be considered in massive ethanol ingestions in which patients do not respond to conventional therapy.

DISPOSITION

- Any infant with significantly altered mental status following acute ethanol ingestion is admitted for observation of respiratory status, fluid resuscitation, and glucose monitoring. Asymptomatic patients may be discharged home with reliable caretakers.

- Adolescent patients should be referred for counseling if a pattern of ethanol abuse is suspected.

METHANOL

- Methanol is present in a variety of substances, including paint solvents, gasohol, gasoline additives, canned-heat products, windshield washer fluid, and duplicating chemicals.

PHARMACOKINETICS AND PATHOPHYSIOLOGY

- Methanol is rapidly absorbed following ingestion. Peak serum levels can be reached as early as 30 to 90 minutes post ingestion. Methanol is primarily metabolized by hepatic alcohol dehydrogenase.
- Methanol itself is harmless; however, its main metabolite, formic acid, is extremely toxic. Fatalities have been reported after ingestion of as little as 15 mL of a 40% methanol solution. Ingestion of only 10 mL can lead to blindness.

CLINICAL PRESENTATION

- Patients may have the classic triad consisting of visual complaints, abdominal pain, and metabolic acidosis.
- Eye signs and symptoms are generally delayed, and include blurring of vision, photophobia, constricted visual fields, snowfield vision, and hyperemia of the optic disk. Although the blindness is usually permanent, recovery has been reported.
- In small children, methanol toxicity should be suspected in the setting of altered mental status and metabolic acidosis of unclear etiology.
- Patients typically complain of nausea and vomiting, and can experience gastrointestinal bleeding and acute pancreatitis. Affected patients often lack the odor of ethanol on their breath, and typically have a clear sensorium.

LABORATORY

- Baseline laboratory data include a complete blood cell count, serum electrolytes and blood glucose, amylase, blood urea nitrogen (BUN) and serum creatinine, a urinalysis, and an arterial blood gas.
- Methanol-intoxicated patients develop an elevated anion gap metabolic acidosis, although this may not be present if the patient presents before a significant quantity of formic acid has been generated.

- The anion gap should be calculated using the equation: $(\text{Na}) - (\text{Cl} + \text{HCO}_3)$. The normal anion gap is 8 to 12 mEq/L.
- Another valuable clue in establishing the diagnosis is the presence of an elevated osmolal gap, which is the difference between measured and calculated serum osmolality. An elevated osmolal gap indicates that a highly osmotic compound not normally found in the serum is present in a significant quantity. The formula for calculating serum osmolality is: $2(\text{NA}) + \text{glucose}/18 + \text{BUN}/2.8 + \text{ETOH}/2.4$.
- Normally the difference between the measure serum osmolality and the calculated serum osmolality is less than 10 mOsm. Though the osmolal gap is a useful clue, cases of significant methanol overdose have been reported with normal osmolal gaps.
- Other toxicologic causes of elevated osmolal gaps include ethylene glycol, ethanol, and isopropanol poisoning, all of which are highly osmotically active compounds.
- Measurement of methanol and ethanol levels is critical in diagnosing these poisonings. Levels <20 mg/dL generally result in minimal effects. CNS effects appear with levels >20 mg/dL and peak levels >50 mg/dL indicate serious toxicity. Ocular effects occur at levels >100 mg/dL, and fatalities have been reported in untreated victims with levels >150 mg/dL.
- A problem with interpreting levels is the time lag between ingestion and the time of patient presentation. Patients with low serum methanol concentrations may still be significantly poisoned and acidotic when presenting late in their clinical course.

MANAGEMENT

- Gastrointestinal decontamination may be efficacious for patients presenting within 1 hour of ingestion.
- If a significant ingestion of methanol is likely, empiric treatment with the intravenous alcohol dehydrogenase inhibitor fomepizole is recommended, even if laboratory tests are unavailable.
- Fomepizole competitively binds hepatic alcohol dehydrogenase 500–1000 times more avidly than methanol, and prevents the formation of the toxic metabolite formic acid. Other indications for fomepizole therapy include serum methanol levels >20 mg/dL, or acidemia (pH <7.20). Unlike ethanol therapy, fomepizole lacks CNS-depressant and hypoglycemia effects.
- If fomepizole is unavailable, ethanol may be administered in an attempt to block alcohol dehydrogenase. To inhibit toxic metabolite formation, ethanol levels are maintained between 100 and 150 mg/dL with a 10% ethanol solution. Close monitoring of

the ethanol level every 1 to 2 hours is necessary in order to adjust the maintenance infusion rate for each individual patient.

- If IV ethanol preparations are unavailable, oral ethanol therapy can be instituted. Since hypoglycemia is a complication of toxic ethanol levels in young children, serum glucose levels are closely monitored. Continued therapy with fomepizole or ethanol is recommended until methanol levels fall below 20 mg/dL.
- Additional therapies for methanol poisonings may include bicarbonate if the serum pH falls below 7.20. Folate, the active form of folic acid, is a coenzyme in the metabolic step converting the toxic metabolite formate to CO₂ and H₂O, and is indicated in the methanol poisoned patient.
- Hemodialysis effectively removes methanol and formic acid. Indications for dialysis include visual impairment, metabolic acidosis not corrected with bicarbonate administration, renal failure, and methanol levels >50 mg/dL (with or without clinical signs or symptoms). Ethanol and fomepizol are readily dialyzed, so the rate of IV administration may have to be increased during dialysis.

DISPOSITION

- Any patients who are comatose and have abnormal vital signs, visual complaints, metabolic acidosis, or high methanol levels need admission to a pediatric intensive care unit.
- Asymptomatic patients without evidence of acidosis and with levels <20 mg/dL may be discharged from after close observation in the emergency department.

ETHYLENE GLYCOL

- Ethylene glycol is an odorless, sweet-tasting compound that is found in antifreeze products, coolants, preservatives, and glycerin substitutes.

PHARMACOKINETICS AND PATHOPHYSIOLOGY

- Ethylene glycol undergoes rapid absorption from the gastrointestinal tract, and initial signs of intoxication may occur as early as 30 minutes postingestion.
- As with the other alcohols, it undergoes hepatic metabolism via alcohol dehydrogenase to form various toxic metabolites, glycolaldehyde, glycolic acid, and ultimately oxalate, which is excreted through the kidney.

- The hallmark of ethylene glycol toxicity is a severe anion gap metabolic acidosis due to accumulation of glycolic acid, hypocalcemia, and renal failure, which results from the precipitation of calcium oxalate crystals in the kidney.

CLINICAL PRESENTATION

- The clinical effects of ethylene glycol toxicity can be divided into three stages:
 - Stage I occurs within the first 12 hours of ingestion, with CNS symptoms similar to that experienced with ethanol.
 - As with methanol toxicity, patients can demonstrate an anion gap acidosis with an elevated osmol gap.
 - In one-third of cases, calcium oxalate crystals will be discovered in the urine, a finding considered strongly suggestive of ethylene glycol poisoning.
 - Stage II occurs within 12 to 36 hours after ingestion and is characterized by rapidly progressive tachypnea, cyanosis, pulmonary edema, adult respiratory distress syndrome, and cardiomegaly. Death is most common during this stage.
 - Stage III occurs 2 to 3 days postingestion and is heralded by flank pain, oliguria, proteinuria, anuria, and renal failure.
- Ethylene glycol poisoning is possible in any inebriated patient lacking an odor of ethanol who has severe acidosis, oxalate crystalluria, hematuria, or renal failure. In a child with a metabolic acidosis of an unclear etiology, this diagnosis should be considered

LABORATORY

- Indicated laboratory studies include complete blood cell count, serum electrolytes, blood glucose, calcium, creatine kinase, serum ethanol and ethylene glycol, an arterial blood gas, BUN and serum creatinine, serum osmolality, and urine for crystals, protein, and blood. Both anion and osmolal gaps are calculated.
- Since fluorescein is present in many antifreeze products, fluorescence of the patient's urine, gastric aspirate, or perioral area when exposed to light from a Wood's lamp may be a valuable diagnostic clue.

MANAGEMENT

- Gastric lavage may be useful in patients presenting within 1 hour of ingestion. Activated charcoal can be administered if there are coingestants, although there are no studies documenting its effectiveness in ethylene glycol toxicity.

TABLE 111-1 Toxic Alcohol Antidotes

METHANOL	ETHYLENE GLYCOL
Fomepizole	Fomepizole
Folate	Thiamine
	Pyridoxine
Ethanol	Ethanol

- Patients who develop seizures are treated with standard doses of benzodiazepines and phenobarbital.
- The alcohol dehydrogenase inhibitor fomepizole is used for treatment of ethylene glycol poisoning. Indications include a metabolic acidosis or an ethylene glycol level >20 mg/dL. In cases where a significant ingestion is suspected, fomepizole therapy should not be delayed pending an ethylene glycol level.
- Ethanol competitively binds alcohol dehydrogenase with an affinity 100 times greater than ethylene glycol and slows the accumulation of toxic metabolites; it is an alternative to therapy with fomepizole. If an intravenous preparation of ethanol is unavailable, patients can be loaded orally to achieve an ethanol level of 100 to 150 mg/dL. Since toxic ethanol levels result in profound hypoglycemia in small children, serial glucose measurements are monitored.
- Bicarbonate administration is recommended for patients with pH < 7.20. Serum calcium levels are monitored and hypocalcemia is treated with calcium gluconate. Additionally, thiamine and pyridoxine (vitamin B6) are recommended in ethylene glycol poisonings in order to shunt or reroute the metabolism of ethylene glycol toward less toxic metabolites (Table 111-1).
- Hemodialysis effectively removes ethylene glycol, as well as its toxic metabolite glycolic acid. Hemodialysis is indicated in the setting of metabolic acidosis not responsive to bicarbonate administration, pulmonary edema, and renal failure. Serum ethylene glycol level >50 mg/dL, regardless of clinical signs, is an indication of hemodialysis in patients treated with ethanol.
- Hemodialysis may not be necessary for cases of ethylene glycol poisoning that can be treated with fomepizole as blocking therapy before acidosis or renal dysfunction develops.

ISOPROPANOL

- Isopropanol is a common solvent and disinfectant with CNS-depressant properties similar to ethanol. The majority of pediatric exposures (up to 90%) occur in children less than 6 years of age. Exposure from isopropyl alcohol occurs more frequently in these children than ethanol, methanol, or ethylene glycol ingestions.

- Toxicity results from both accidental and intentional ingestions, as well as inhalation and dermal exposures in young children given “rubbing alcohol” sponge baths for fever.

PHARMACOKINETICS AND PATHOPHYSIOLOGY

- Isopropanol is rapidly absorbed across the gastric mucosa, with acute intoxication occurring within 30 minutes of ingestion. It is metabolized by alcohol dehydrogenase, but, unlike the other alcohols, is not metabolized to an acidic end product.
- Isopropanol is converted to the CNS depressant acetone. Respiratory elimination of the acetone causes a fruity-acetone odor on the patient’s breath similar to diabetic ketoacidosis.
- Isopropanol is a potent inebriant that is about twice as intoxicating as ethanol; a level of 50 mg/dL is comparable to an ethanol level of 100 mg/dL.

CLINICAL PRESENTATION

- Isopropanol-intoxicated patients are lethargic or comatose, hypotensive, and tachycardiac, with the characteristic breath odor of rubbing alcohol or acetone.
- Coma develops at levels >100 mg/dL. Hypotension results from peripheral vasodilation and cardiac depression.
- Gastrointestinal irritation with acute abdominal pain and hematemesis can also occur. With isopropanol, unlike the other toxic alcohols, acidosis, ophthalmologic changes, and renal failure are absent.
- Like ethanol, methanol, and ethylene glycol, isopropanol can produce a significant osmolal gap (Table 111-2).

LABORATORY

- Patients are tested for the presence of acetonemia and acetonuria. Unlike diabetic ketoacidosis, the acetone

TABLE 111-2 Comparisons of Toxic Alcohols

PARAMETER	ETHYLENE		
	METHANOL	GLYCOL	ISOPROPANOL
Anion gap	+	+	–
Acidosis			
Osmolal gap	+	+	+
CNS depression	+	+	+
Eye findings	+	–	–
Renal failure	–	+	–
Ketones	–	–	+
Oxalate crystals	–	+	–

is typically found in the absence of glucosuria, hyperglycemia, or acidemia.

- Other laboratory studies include a complete blood cell count, serum electrolytes, an arterial blood gas, blood glucose, serum ethanol and isopropanol levels, serum osmolarity, and BUN and creatinine.
- Isopropanol levels >400 mg/dL correspond to severe, life-threatening toxicity.

MANAGEMENT

- Patients are managed with particular attention paid to the integrity of the airway. Hypotension is treated with intravenous crystalloid.
- Isopropanol is so rapidly absorbed from the gastrointestinal tract that gastric decontamination with a nasogastric tube is unlikely to be of any benefit. Activated charcoal for isopropanol poisonings not indicated unless mixed with a coingestant.
- No alcohol dehydrogenase inhibition is indicated since the metabolite acetone is relatively nontoxic and excreted through the lungs.
- Hemodialysis is effective in removing isopropanol, but is reserved for prolonged coma, hypotension, and isopropanol levels >400 to 500 mg/dL.
- Typically, patients progress well with supportive care alone.

DISPOSITION

- Isopropanol-intoxicated patients who are lethargic should be admitted, while asymptomatic children may be observed in the emergency department.
- Ingestion of over three swallows (15 mL) of 70% isopropanol by a 10-kg child is an indication for several hours of observation.

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QUESTIONS

1. A 2-year-old child is brought to the ED following ingestion of a green colored, mint flavored mouthwash approximately 2 hours ago. The child has stable vital signs, a normal pulse oximetry but is noted to be “sleepy” in triage. A management priority in this child would be which of the following?
 - A. Rapid administration of activated charcoal.
 - B. A stat urine toxicology screen.
 - C. Bedside glucose measurement.
 - D. Observation and supportive care alone.
 - E. Airway control and endotracheal intubation.
2. A 5-year-old is brought by paramedics for evaluation of altered mental status. On arrival to the ED, you note a fruity odor on the breath. Which of the following would be consistent with an isopropanol ingestion?
 - A. A serum glucose over 500.
 - B. Acute visual changes.
 - C. A metabolic acidosis with elevated anion gap.
 - D. Renal failure and urinary crystals.
 - E. A positive serum acetone measurement.
3. You suspect methanol intoxication in a child brought to the ED following a windshield washer fluid ingestion. The patient has a normal mental status, stable vital signs and demonstrates a mild acidosis on arterial blood gas. You are told by your pharmacy that they no longer stock ethanol preparations for toxic alcohol therapy. Other alternative treatment options include which of the following?
 - A. Flumazenil administration
 - B. Fomepizole therapy

- C. Pyridoxine administration
 D. Emergency hemodialysis
 E. Deferoxime chelation therapy
4. A 17-year-old child presents following an ingestion of antifreeze in an apparent suicide attempt. Upon presentation, he is lethargic and unresponsive to painful stimulation. Following endotracheal intubation, an arterial blood gas demonstrates a pH of 6.95. What is the optimal therapy of choice in this patient?
- A. Intravenous ethanol therapy
 B. Fomepizole administration
 C. Naloxone administration
 D. Emergent hemodialysis
 E. Insulin-dextrose rescue therapy

ANSWERS

1. C. Over the counter mouthwashes can contain significant amounts of alcohol. In a child with suspected ethanol intoxication, the most critical laboratory tests are a serum ethanol and glucose level. Children are often hypoglycemic due to their limited hepatic glycogen stores, as compared to adults. Convulsions and death have been reported in children secondary to alcohol-induced hypoglycemia.
2. E. An elevated serum acetone, normal glucose, associated with a fruity breath odor should increase suspicion for an isopropanol or rubbing alcohol ingestion. While isopropanol may give an elevated osmolar gap, it does not typically produce a metabolic acidosis with elevated anion gap as with the other toxic alcohols. Acute visual changes are more consistent with methanol poisoning, renal failure and urinary crystals are classic for ethylene glycol toxicity.
3. B. The alcohol dehydrogenase inhibitor fomepizole is now the antidote of choice for methanol and ethylene glycol poisoning. This antidote has essentially replaced ethanol therapy. Indications for hemodialysis include visual impairment, renal failure, methanol levels over 50 mg/dL, and refractory metabolic acidosis. Flumazenil is the antidote for benzodiazepine overdose. Pyridoxime is used in ethylene glycol and INH toxicity. Deferoxime chelation therapy is the antidote of choice for severe iron poisoning.
4. D. This patient is in critical condition from ethylene glycol poisoning. The only hope for his survival is emergent hemodialysis. Although ethanol or fomepizole therapy may be indicated earlier in the course, this patient's profound acidosis points to a late stage poisoning with an accumulation of toxic metabolites only cleared by hemodialysis. Naloxone is indicated with opioid overdose. Insulin-dextrose rescue therapy is a new modality indicated for the treatment of unstable calcium channel blocker toxicity.

112 ORGANOPHOSPHATES AND CARBAMATES

Leon Gussow

HIGH-YIELD FACTS

- Several factors make these pediatric cases of organophosphate or carbamate toxicity difficult to diagnose. Often the child is too young to give a history, and the parents might not suspect that oral or topical pesticide exposure has occurred. In addition, the typical respiratory and gastrointestinal symptoms of the cholinergic syndrome may be mistaken for those of common childhood illnesses: bronchitis, pneumonia, upper respiratory infection, or gastroenteritis. Finally, some studies have indicated that well-recognized manifestations of cholinergic toxicity in adults—for example, bradycardia and muscle fasciculations—occur in only a minority of pediatric cases, whereas tonic-clonic seizures occur more frequently in children.

PATHOPHYSIOLOGY

- Organophosphates poison acetylcholinesterase, the enzyme that breaks down and inactivates acetylcholine. Acetylcholine is the chemical messenger at junctions where nerves connect to skeletal and smooth muscle, and to secretory glands. The subsequent buildup of acetylcholine causes hyperstimulation of areas of the nervous system that contain cholinergic receptors.
- There are two major classifications of cholinergic receptors:
 - Muscarinic receptors, found on glands and involuntary smooth muscle.
 - Nicotinic receptors, found on voluntary skeletal muscle and some autonomic ganglia. There are also central cholinergic receptors found within the brain.

CLINICAL PRESENTATION

- The initial signs and symptoms of cholinergic toxicity are often muscarinic. A helpful mnemonic to remember these muscarinic manifestations is DUMBELS:
 - Diarrhea, Diaphoresis
 - Urination
 - Miosis
 - Bronchorrhea, bronchospasm
 - Emesis
 - Lacrimation
 - Salivation
- Although miosis and the overall increase in secretions—saliva, sweat, and tears—can help suggest the diagnosis, the life-threatening effects are really bronchospasm and bronchorrea—the so-called killer Bs.
- Nicotinic effects of excess acetylcholine at the neuromuscular junction include muscle spasm and fasciculations, followed by weakness or paralysis as the muscle fatigues. Bronchorrhea, bronchospasm, and respiratory muscle weakness can combine to cause respiratory failure requiring intubation and assisted ventilation.
- A review of 37 cases of children diagnosed with organophosphate or carbamate toxicity found that the following signs and symptoms occurred most frequently:
 - Miosis (73%)
 - Excessive salivation (70%)
 - Muscle weakness (68%)
 - Lethargy (54%)
 - Tachycardia (49%).

LABORATORY STUDIES

- History, suggestive clinical signs and symptoms, and a high index of suspicion are the keys to making the diagnosis of organophosphate toxicity. The combination of miosis and increased salivation is relatively specific for exposure to a cholinergic agent such as organophosphate. There is no readily available laboratory test that can indicate the diagnosis during initial management.
- Pulse oximetry can help evaluate oxygenation status. The cardiac monitor can detect tachycardia or bradycardia, either of which can be seen following organophosphate exposure. A chest radiograph may show aspiration pneumonitis, which is especially likely if the organophosphate preparation included a hydrocarbon vehicle. Noncardiogenic pulmonary edema may also be seen.

TREATMENT

- Adequate external decontamination of a patient with organophosphate toxicity is critical to prevent continued exposure. All clothing and jewelry should be removed and placed in well-sealed plastic bags. Contaminated skin should be irrigated with copious amounts of water, or *gently* washed with soap and water.
- To prevent secondary contamination, all members of the medical team who are directly treating or decontaminating the patient should wear adequate protective gear. Simple surgical masks and gowns do *not* provide sufficient protection. Butyl rubber gloves and aprons are more effective options. If the patient is vomiting, gastric contents may contain the organophosphate agent and be a source of secondary contamination.
- Neither gastric lavage nor activated charcoal has any demonstrated benefit in the setting of organophosphate ingestion.
- The immediate life-threat in severe organophosphate poisoning is respiratory failure from the combined muscarinic effects of bronchospasm and bronchorrea. Both of these can be alleviated by the antimuscarinic antidote atropine sulfate. The initial dose of atropine is 0.02 mg/kg intravenously (IV). In severe cases, this dose should be doubled every 5 minutes until pulmonary secretions dry up, bronchospasm resolves, and the child can be oxygenated and ventilated. There is no maximum dose of atropine. Frequently, in these cases, surprisingly large doses are required.
- The antidote pralidoxime chloride should be given to treat moderate-to-severe cholinergic toxicity from organophosphates or an unknown agent. The dose is 25 to 50 mg/kg IV over 30 minutes. Pralidoxime regenerates acetylcholinesterase by removing organophosphate from the enzyme's active site. The dose can be repeated at 6 hours intervals, or a continuous infusion of 10 mg/kg/h can be started after the initial load. Pralidoxime is not indicated in known carbamate exposure, but should be administered in cases of significant cholinergic toxicity where the exact agent is not known.
- Diazepam is the treatment of choice for organophosphate-induced seizures.

CARBAMATES

- Carbamate insecticides such as aldicarb and carbaryl (sevin) also inactivate acetylcholinesterase. Unlike organophosphates, however, this inactivation is not permanent. Functional enzyme activity is often

- largely restored within 8 hours, with red blood cell cholinesterase completely restored within 48 hours.
- Carbamate toxicity is primarily restricted to muscarinic effects. Carbamates are much less likely than organophosphates to cause CNS effects, since they do not penetrate the blood–brain barrier well. Nicotinic manifestations are also uncommon. However, children with severe carbamate poisoning can develop mental status depression and occasionally seizures.
 - The initial management of the muscarinic effects of carbamate toxicity is similar to that for organophosphates, with stabilization of the airway and breathing, and adequate decontamination of the patient. Atropine is administered as indicated for muscarinic manifestations. Because carbamates have a relatively short duration of action, pralidoxime is unlikely to be of benefit.

- C. Atropine
 - D. Pralidoxime
 - E. Ipratropium
3. A 5-year-old male is brought in to the ED after being exposed to an unknown pesticide after playing in his garage and spilling an unlabeled container onto his skin. He has been nauseated and vomiting several times. The father knows that it is a pesticide in the container but does not remember the brand name. In treating this patient you understand that the major difference between the toxicity of organophosphate and carbamate insecticides is that carbamates
 - A. Do not cause bronchorrhea and bronchospasm.
 - B. Do not require treatment with atropine.
 - C. Do require unusually large doses of pralidoxime.
 - D. Do not permanently inactivate the enzyme acetylcholinesterase.
 - E. Rapidly penetrate the blood-brain barrier.

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QUESTIONS

1. The most important threats to life after organophosphate pesticide ingestion include which of the following?
 - A. Excess respiratory secretions and bronchospasm.
 - B. Seizures.
 - C. Muscle paralysis.
 - D. Central nervous system depression.
 - E. Bradycardia.
2. A 6-year-old boy was helping his grandfather spray fruit trees when he begins to complain of nausea, vomiting, diarrhea, and crampy abdominal pain. He is driven to a doctor's office where he is found to be tearing and has copious secretions from his nose and mouth. On examination, he is noted to be wheezing. Following decontamination which of the following would be the most appropriate initial management?
 - A. Albuterol
 - B. Epinephrine

ANSWERS

1. A. The most important life-threat immediately following ingestion of organophosphate pesticides are the muscarinic “killer Bs” effects such as bronchorrhea and bronchospasm. The antidote that can dry up the respiratory secretions and relax the airway smooth muscle is atropine.
2. C. The immediate life-threat in severe organophosphate poisoning is respiratory failure from the combined muscarinic effects of bronchospasm and bronchorrhea. Both of these can be alleviated by the antimuscarinic antidote atropine. The antidote pralidoxime should be given to treat moderate-to-severe cholinergic toxicity and usually is given after atropine. Albuterol and epinephrine are not indicated but ipratropium can be used to assist in drying secretions.
3. D. Carbamate toxicity is primarily limited to muscarinic effects, including bronchorrhea and bronchospasm. These can and should be treated with atropine. Since carbamates do not cause permanent inactivation of the enzyme acetylcholinesterase, pralidoxime is usually not required. However, if it appears the patient is toxic from a cholinergic pesticide but it is not clear initially whether this is an organophosphate or a carbamate, it would be reasonable to administer pralidoxime. Carbamates generally cause fewer CNS manifestations than do organophosphates because they do not readily penetrate the blood–brain barrier.

113 CAUSTICS

Jenny J. Lu

Trevonne M. Thompson

PATHOPHYSIOLOGY

- Severity of injury depends on:
 - The agent type, concentration, volume, pH, and titratable acid (or alkaline) reserve (TAR).
 - TAR: amount of a xenobiotic required to neutralize or raise the pH of a caustic to that of physiologic tissues.
 - Duration of contact with tissues.
 - Presence or absence of contents in the stomach.
- Solids tend to produce intense localized oropharyngeal or upper esophageal injury.
- Liquids tend to produce circumferential lesions in the distal esophagus.
- Three major pathophysiologic phases of caustic injuries.
 - Phase 1: acute inflammatory stage.
 - Phase 2: latent granulation phase.
 - Phase 3: chronic cicatrization phase.

ALKALI BURNS

- Alkali burns cause *liquefaction necrosis*, resulting in a deep penetration injury.
- Tend to produce injury very rapidly.
- Most accidental ingestions of household grade bleach appear to do well with supportive care.
- “Lye” refers to a specific alkali, commonly sodium hydroxide, or potassium hydroxide.
- Liquid lye can cause severe esophageal injuries with minimal associated oropharyngeal findings.
- Ingestion of small quantities of a base can cause severe damage without ever reaching the stomach.

ACID BURNS

- Acid burns cause *coagulation necrosis* with severe injury to superficial tissues.
- Acid injury, in contrast to alkali injury, may continue for up to 90 minutes after the ingestion.
- An acid can reach the stomach without causing significant oral or esophageal injury.

PRESENTATION AND DIAGNOSIS

- Accurate ingestion history important.

TABLE 113-1 Internet Resources

Household Products Database (National Institutes of Health)	http://householdproducts.nlm.nih.gov/index.htm
OSHA/EPA Occupational Chemical Database	http://www.osha.gov/web/dep/chemicaldata/
Agency for Toxic Substances and Disease Registry	http://www.atsdr.cdc.gov/toxfaq.html
THOMSON POISONDEX® System	http://www.micromedex.com/products/poisondex/

- Additional resources.
 - Actual container, including labels, of the ingested agent.
 - Manufacturers can sometimes be contacted to identify ingredients.
 - Material safety data sheets (MSDS) can often be found on the internet for various compounds.
 - The regional poison control center can assist with identifying product contents.
 - Many other resources are also available (Table 113-1).
- Clinical presentation is variable, from asymptomatic to shock
- Dysphagia, drooling, vomiting, hematemesis, dyspnea, chest and abdominal pain, and melena are indicative of severe injury
- Life-threatening airway edema may occur
 - Stridor, dyspnea, and dysphonia indicate upper airway compromise.
 - Blind nasotracheal intubation is contraindicated.

MANAGEMENT

- Attention to the airway and recognition of esophageal or gastric perforation are top priorities.
- Induced emesis is absolutely contraindicated.
- Supportive care is the mainstay of therapy.
- Patient should not be fed.
- Gastrointestinal decontamination with activated charcoal is unlikely to be of benefit and will obscure endoscopic visualization.
- Surgical intervention is required if any signs of perforation or peritonitis are present.
- The role of steroid therapy to reduce the incidence of esophageal strictures remains controversial.
 - Beneficial most likely with second-degree injuries.

LABORATORY AND RADIOLOGY STUDIES

- Oxygen saturation, arterial blood gas and chest radiograph when indicated.

- Complete blood count, electrolytes, renal function, coagulation profile, and serum lactate
- Abdominal radiographs when indicated.
- Early endoscopy helps define the extent of the injury and establish prognosis.

SPECIAL CONCERNS

CAUSTIC EYE INJURIES

- Caustic eye injuries can have devastating consequences, including blindness.
- Irrigate the eye for 15–20 minutes with saline, lactated Ringer's solution, or tap water.
- Ophthalmologist consultation is recommended for all ophthalmic caustic injuries.

BUTTON BATTERIES

- Contain metallic salts in a concentrated alkaline medium.
- Vast majority of patients do well.
- Pressure necrosis at the site where the battery becomes lodged can occur.
- Rarely, if the battery does break open, the caustic contents may cause ulceration, perforation, or fistula formation.
- Injury from electrical current can also occur.
- Urgent removal if
 - Battery lodged in the esophagus or airway.
 - Battery lodged in nasal passages.
- Asymptomatic patients with a battery distal to the esophagus can be discharged, followed as outpatient with serial radiographs.

HYDROFLUOROIC ACID

- Severe pain and deep penetration despite minimal skin findings are the hallmarks.
- The mechanism of injury involves liquefaction necrosis and the formation of insoluble calcium and magnesium salts.
- Oral ingestions are frequently fatal.
- May develop systemic acidosis, hypocalcemia, hypomagnesemia, and hyperkalemia.
 - Hypocalcemia may require large amounts of calcium.
 - Intra-arterial calcium gluconate infusion can be considered for refractory cases.
- Calcium gluconate gel can be applied topically to HF burns.

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QUESTIONS

- A 5-year-old male is brought in by his parents after he swallowed a household bleach cleaning product. Which of the following procedures is absolutely contraindicated in a caustic ingestion?
 - Administration of activated charcoal
 - Hemodialysis
 - Induction of emesis
 - Dilution with milk or water
 - All of the above
- A 2-year-old child has ingested a button battery. In which of the following clinical scenarios is urgent removal required?
 - Battery is visualized in the duodenum.
 - Battery is visualized in the stomach.
 - Battery is visualized in the esophagus.
 - Battery is visualized in the rectum.
 - Urgent removal is not required in any of these situations.
- Hydrofluoric acid exposures are notable for
 - Severe pain with oral, ocular, or dermal exposures.
 - The potential to produce significant electrolyte disturbances.
 - Often requiring large amounts of calcium repletion.
 - Reports of death even after seemingly small exposures.
 - All of the above.

ANSWERS

1. C. Induction of emesis is absolutely contraindicated in a caustic ingestion as further injury to the esophagus and oral cavity may occur due to re-exposure to the agent. Activated charcoal is not contraindicated but is of no value and may obscure endoscopic visualization. Hemodialysis is not contraindicated but is rarely a useful procedure in the caustic ingestion unless severe acid–base or electrolyte disturbances are present and uncorrectable. Dilution with milk or water is likely most beneficial within the first seconds after a caustic ingestion but care should be taken not to cause gastric distention and vomiting from giving excessive volumes of fluid.
2. C. Urgent removal of button batteries is required when they are lodged in the esophagus, airway or nasal passage. If they have passed into the stomach, the child can be discharged for outpatient management and followed closely with serial radiographs.
3. E. All of these statements are true of hydrofluoric acid ingestions. Immediate and attentive care should be given to even seemingly minor exposures to hydrofluoric acids.

114 HYDROCARBONS*Trevonne M. Thompson***CLASSIFICATION AND PROPERTIES**

- Viscosity, surface tension, and volatility are the important properties used to assess the toxicity of liquid hydrocarbons.
- Viscosity is the measurement of a liquid's resistance to flow.
- Volatility describes the tendency of a liquid to become a gas.
- Surface tension describes the property of adherence of a liquid compound along a surface.

PATHOPHYSIOLOGY

- Aspiration and subsequent pulmonary toxicity is the primary concern after hydrocarbon ingestions.
- Viscosity, surface tension, and volatility determine the risk of aspiration after a hydrocarbon ingestion.
- Compounds with low viscosity, low surface tension, and high volatility have a higher risk of aspiration and subsequent pulmonary toxicity.

- Inhalation of hydrocarbon vapor can lead to CNS depression, a desirable effect for hydrocarbon abusers.
- Sniffing is the direct inhalation of hydrocarbon vapor.
- Huffing is saturating a cloth with a hydrocarbon and inhaling the vapors.
- Bagging is placing a hydrocarbon in a plastic bag and inhaling from the open end.
- Hydrocarbon exposure, via any route, may induce ventricular dysrhythmias.
- Sudden sniffing death syndrome is a sudden cardiac death associated with volatile hydrocarbon abuse.

CLINICAL PRESENTATION

- Depends on the route and amount of the exposure, from asymptomatic to fulminant respiratory distress or fatal dysrhythmia.
- Coughing, gagging, choking, and vomiting after a hydrocarbon ingestion is presumptive of aspiration.
- Signs of aspiration include tachypnea, crackles, bronchospasm, hemoptysis, hypoxia, acute lung injury, or respiratory failure.
- Radiographic findings are protean, from increased bronchovascular marking to focal consolidations.
- Cardiac findings range from mild tachycardia to ventricular tachydysrhythmias.
- Victims of sudden sniffing death syndrome present in full cardiac arrest.
- The typical CNS finding is mental status depression.

MANAGEMENT

- The mainstay of treatment for a hydrocarbon exposure is supportive care.
- Asymptomatic patients do not need early radiography.
- Perform chest x-ray if any of these are present: choking, coughing, gagging, vomiting, any respiratory sign or symptom.
- There is typically no role for gastrointestinal decontamination.

DISPOSITION

- Symptomatic patients should be admitted and observed until symptom resolution (Fig. 114-1).
- Asymptomatic patients should be observed for 6 hours after ingestion.
- If there is no tachypnea, hypoxia, abnormal pulmonary findings and a normal chest x-ray 6 hours after ingestion, the patient may be safely discharged.

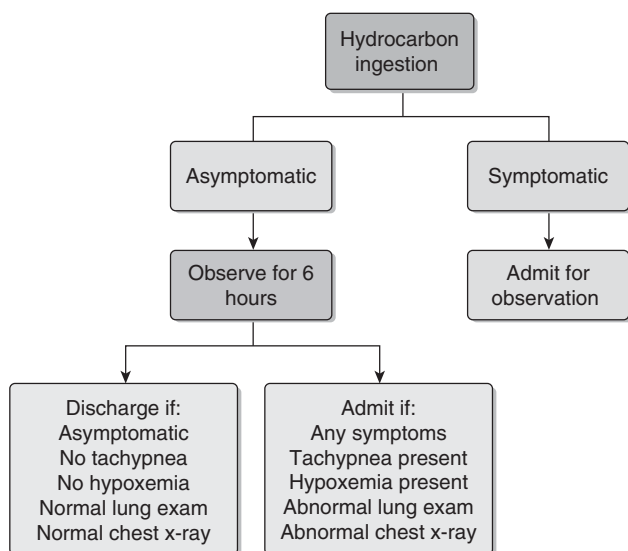


FIG. 114-1. Disposition algorithm.

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QUESTIONS

- Which of the following combination of physical properties of hydrocarbons are important characteristics to understand in assessing a patient who ingested a hydrocarbon?
 - Surface tension, volatility, and viscosity.
 - Surface tension, molecular weight, and color.

- Viscosity, boiling point, and surface tension.
 - Volatility, specific gravity, and miscibility in water.
 - Specific gravity, boiling point, and molecular weight.
- A 3-year-old child presents after accidentally ingesting gasoline. Which is the primary clinical concern when assessing the patient who ingested a hydrocarbon?
 - Cardiac toxicity
 - Pulmonary toxicity
 - GI toxicity
 - Neurologic toxicity
 - Reproductive toxicity
 - A 2-year-old male ingested lighter fluid while playing in the backyard. The mainstay of treatment for this hydrocarbon ingestion is which of the following?
 - Antidotal therapy with *n*-acetylcysteine.
 - Gastrointestinal decontamination with activated charcoal.
 - Supportive care.
 - Whole bowel irrigation.
 - Urinary alkalinization.
 - A 13-year-old male presents to the ED after being brought in by his parents because they caught him sniffing fumes from a bag. Inhalation of hydrocarbon vapors by sniffing, bagging, or huffing place the patient at risk for which disease process?
 - Sudden inhalational psychotic syndrome.
 - Chronic death syndrome.
 - Acute sniffing chest syndrome.
 - Sudden sniffing death syndrome.
 - Acute tachydysrhythmia syndrome.

ANSWERS

- A. Surface tension, volatility, and viscosity are the properties important in assessing a hydrocarbon ingestion. Options B, C, D, and E do not contain the correct combination of properties.
- B. Pulmonary toxicity is the primary clinical concern. There is a concern for cardiac toxicity, specifically sudden cardiac death. This syndrome, when it occurs, tends to occur in the setting of a hydrocarbon abuser who inhales hydrocarbon vapors. This does not typically occur in the ingestion setting. Neurologic toxicity, specifically CNS depression, may be present; however, pulmonary toxicity is the primary concern.
- C. The mainstay of therapy for hydrocarbon ingestions is supportive care. There is no role for gastrointestinal decontamination, *n*-acetylcysteine, or whole bowel irrigation.

4. D. The other conditions do not exist. Sudden sniffing death syndrome present in full cardiac arrest and is associated with volatile hydrocarbon abuse.

115 RODENTICIDES

Arthur Kubic

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INTRODUCTION

- Ingestion of rodenticides by children account for a significant number of exposures reported to poison centers around the country. Trivial, unintentional pediatric ingestions resulted in no clinical effects in the overwhelming majority of cases.
- The EPA currently approves four rodenticides for indoor use: anticoagulants, cholecalciferol, bromethalin, and zinc phosphide (Table 115-1).
- Ingestion of even a small quantity of outdated, illegal, or unapproved products designed for indoor use (eg, strychnine, arsenic, white phosphorus, or sodium monofluoroacetate) can pose a significant danger to children (Table 115-2).

DEVELOPMENTAL CONSIDERATIONS

- Children younger than 6 years of age comprise the largest population at risk of rodenticide poisoning
- Children with developmental delay or those with a history of pica are at greater risk of a large scale or chronic ingestion.
- Adolescents and young adults may consume rodenticides intentionally in a suicide attempt. Mentally ill patients have been known to chronically eat these products as well.

PATHOPHYSIOLOGY

- Prior to 1980, most anticoagulant rodenticide exposures involved a warfarin-containing product. Superwarfarins (long acting anticoagulants) were developed later, and are 100 times more potent than warfarin.
- The long-acting anticoagulants are classified in two groups: the 4-hydroxy coumarins (eg, brodifacoum, bromodiolone, and difethialone) and the indandiones (eg chlorophacinone, diphacinone)

- These anticoagulant rodenticides inhibit K 2–3 epoxide reductase, which leads to reduction of blood clotting factors II, VII, IX, and X.
- The anticoagulant effects will become evident after three to four factor VII half-lives (15–20 hours) after ingestion.
- Extrapolating from data in rats, the minimum toxic dose would be approximately 1.5 mg of brodifacoum for a 10-kg child or 20 to 30 g of a 0.005% bait.

PHARMACOLOGY/TOXICOLOGY

- These agents are well absorbed orally.
- Prolonged duration of action and half-life of the superwarfarins are attributed to their high-lipid solubility and high concentration in the liver.
- Because of these extremely long half-lives, large, symptomatic ingestions require weeks to months of medical care.

CLINICAL FINDINGS

- Bleeding following unintentional ingestion of anticoagulant rodenticides by pediatric patients is unlikely, although mild PT prolongation has been noted.
- Clinical evidence of bleeding may occur following acute intentional ingestions of large amounts or repeated, chronic ingestions as in the case of pica, child abuse, or Munchausen syndrome, by proxy.
- Signs and symptoms may range from minor to life threatening bleeding such as easy bruising, epistaxis, gingival bleeding, petechiae, hematuria, hematemesis, melena, and hemoptysis, extremity pain associated with compartment syndrome, vaginal bleeding, and intracerebral hemorrhage.

LABORATORY AND DIAGNOSTIC TESTING

- No coagulation studies or other laboratory testing is necessary in the asymptomatic pediatric patient following a single unintentional ingestion of a small amount of an anticoagulant rodenticide.
- Asymptomatic patients with unintentional ingestions of 1 mg or more of active ingredient should be evaluated for coagulopathy at 48 to 72 hours post exposure.
- In symptomatic patients or those demonstrating abnormal coagulation profiles, consider obtaining the following: PT/PTT or INR every 6 to 12 hours until the patient is stabilized, serial hemoglobin's and hematocrit, urinalysis to assess for hematuria, stool

TABLE 115-1 Other Non-Anticoagulant Indoor Rodenticides

RODENTICIDE	TOXICOLOGY/CLINICAL FINDINGS	MANAGEMENT/ANTIDOTES
Bromethalin Bait 0.01% Assault*, Clout All Weather Bait*, Fastrac*, Real Kill Rat and Mouse Killer*, Top Gun*, Vengeance*	Uncouples oxidative phosphorylation resulting in decreased ATP production and increased fluid accumulation which interrupts nerve impulse conduction with resultant increased pressure on nerve axons; there is no established human lethal dose. SX: large ingestions may cause headaches, confusion, tremors, myoclonic jerking, seizure, cerebral edema, or coma. Toxicity may be delayed 8–12 h or longer due to conversion to a more active metabolite	Activated charcoal following ingestion of a large amount; no specific antidote; supportive care including measures to correct cerebral edema, i.e., hyperventilation, mannitol or furosemide, dexamethasone; benzodiazepines/ phenobarbital for seizures; monitor cerebral spinal fluid pressure
Cholecalciferol Bait 0.075% Quintox*, Rampage*	Mobilizes calcium from bones producing hypercalcemia, osteomalacia, and metastatic calcification of the cardiovascular system, kidneys, stomach, and lungs; toxicity may occur from a single large ingestion or chronic consumption; death occurs in animals in 2–5 d; however, serious human poisonings or fatalities from these rodent baits have yet to be reported. SX: anorexia, nausea, vomiting, diarrhea or constipation, headache, fatigue, weakness, hypercalcemia, hyperphosphatemia, cardiac dysrhythmias, myocardial infarction, renal tubular injury	Treat for hypercalcemia as clinically indicated
Zinc phosphide Granules 2% Eraze*, Mole Nots*, and Mr. Rat Guard* Note: aluminum phosphide is a fumigant not sold for household use	Converted to phosphine gas when in contact with acid or moisture; direct cellular toxin causing multiorgan injury by inhibiting cytochrome C oxidase, thus blocking the electron transport chain. In one case series of 21 patients, those ingesting under 1 g had a favorable outcome; lethal dose: 4 g in an adult; SX: nausea, profuse vomiting and diarrhea which may be bloody, a decaying fish odor may be noted, headache, cough, tachypnea, dyspnea, dizziness, tremulousness, hypotension, shock, hypocalcemia, tetany, pulmonary edema, convulsions, cardiac dysrhythmias, renal damage, hepatotoxicity, acute pancreatitis, coma; death may occur in 12–24 h	GI decontamination is controversial; lavage may release phosphine gas; activated charcoal may be given but is of questionable value; antacids, H2 blockers, and proton pump inhibitors may be considered; ED staff should work in a well-ventilated area; no specific antidote; provide intensive supportive care including intubation at earliest sign of pulmonary edema, which may be delayed 24–72 h; give benzodiazepines/ phenobarbital for seizures; monitor electrolytes, glucose, calcium, renal and hepatic function tests, magnesium, ABGS or pulse oximetry, chest x-rays

hemocult test, type and screen (type and cross if significant bleeding present), liver function test to rule out hepatotoxicity as a cause of coagulopathy, and head CT if mental status changes are present. Endoscopy may be considered for the assessment of significant GI bleeding.

- Assays for long-lasting anticoagulants may aid in confirmation of the diagnosis, but they are not routinely followed or used to guide therapy.

MANAGEMENT

- Identify the rodenticide product exactly (ie, by the brand name, active ingredient and concentration). Anticoagulant products will not cause rapid onset of symptoms. Any child presenting with early onset gastrointestinal, neurologic, or cardiovascular signs or symptoms should raise the suspicion that a highly toxic product may have been ingested.
- No GI decontamination procedures are necessary following unintentional trivial ingestions of anticoagulant rodenticides. For patients presenting soon

after intentional ingestions of large amounts, give one dose of activated charcoal without a cathartic. Do not perform gastric lavage or induce vomiting in any patient actively bleeding or demonstrating an elevated PT or INR.

- No definitive, clinically proven method of enhanced elimination exists for these agents
- Phytonadione (vitamin K₁) acts as a specific antidote for warfarin and long acting anticoagulants by competing with these agents to initiate formation of depleted clotting factors II, VII, IX, and X.
- Vitamin K₁ therapy is indicated for any patient experiencing active bleeding or with a significantly elevated PT or INR. Prophylactic therapy with vitamin K₁ is not indicated in asymptomatic patients with normal coagulation studies.
- Oral vitamin K₁, usually given three to four times daily, is the safest method of administration and has an onset of effect ranging from 6 to 12 hours.
- Intramuscular and subcutaneous administration are also possible; however, due to the risk of hematoma, subcutaneous administration may be the safer route.

TABLE 115-2 Outdated, Illegal, and Unapproved Rodenticides for Indoor Use

RODENTICIDE ^o	TOXICOLOGY/CLINICAL FINDINGS	MANAGEMENT/ANTIDOTES
α -naphthylthiourea (2) ANTU Bait 1%–3% Bontu [®]	Damages pulmonary epithelium causing pulmonary edema; human lethal dose is estimated to be more than 4 g/kg with no known reported human fatalities; SX: dyspnea, cyanosis, noncardiogenic pulmonary edema and effusions, hypothermia; pulmonary edema may be delayed by 24–72 h	Activated charcoal; no known antidote; symptomatic care with oxygen and ventilatory support; monitor ABGs
Arsenic (1) Arsenic trioxide	Combines with sulfhydryl (-SH) groups in many essential cellular proteins and enzymes; estimated fatal dose 1–4 mg/kg; SX: garlic like breath odor, profuse vomiting and diarrhea, hypotension and shock, cardiac dysrhythmias, renal tubular damage, pulmonary edema, delirium, seizures, coma; delayed SX: peripheral neuropathy, alopecia, Mees lines, blood dyscrasias	Consider orogastric lavage as indicated, activated charcoal; whole bowel irrigation if abdominal radiograph is positive; obtain blood, spot urine, and 24 h urine for arsenic; monitor electrolytes, EKG, renal function tests, CBC; antidotes: IM dimercaprol or po succimer; supportive care with IV fluids, pressors, antidysrhythmics, hemodialysis
Barium carbonate (1)	Soluble barium salts lower serum potassium and raise intracellular potassium; estimated lethal dose 20–30 mg/kg; barium sulfate is nontoxic; SX: nausea, vomiting, diarrhea, paresthesias, weakness, paralysis, hypoglycemia, rhabdomyolysis, dysrhythmias, cardiac/respiratory failure	Consider orogastric lavage as indicated; frequent serum potassium levels with IV potassium replacement as necessary; supportive care with antidysrhythmics; no specific antidote; barium level may confirm diagnosis
Chloralose (2) (α -chloralose)	Has sedative effects similar to chloral hydrate and stimulant effect similar to strychnine; human toxic dose: 1–4 g, infants: 20 mg/kg; SX: increased salivation, sedation, coma, respiratory depression, myoclonus, seizures, hypotension, hypo- or hyperthermia, acidosis	Activated charcoal; no specific antidote; supportive care for respiratory failure, hypotension, rhabdomyolysis; benzodiazepines/phenobarbital for seizures
Calcium cyanide	This solid fumigant rodenticide reacts with moisture in the air to form highly toxic HCN gas. CN inhibits cytochrome oxidase enzymes halting aerobic cellular respiration. Estimated lethal dose of HCN: 50–100 mg. SX: skin flushing, anxiety, tachypnea, tachycardia, headache, progressing to bradycardia, hypotension, agitation, stupor, coma, seizures, and lactic acidosis	Administer 100% oxygen and provide ventilatory support. Consider activated charcoal following ingestion of CN salts. Antidotes to consider include the CN Antidote Kit or the Cyanokit.
Phosphorus (1) White or Yellow Stearns Chemical Paste [®] 2.5%	Protoplasmic poison causing direct cell injury leading to multiorgan failure; lethal dose: 1 mg/kg; mixed with peanut butter as bait; (note: red phosphorous found in matches is nontoxic) SX: bloody emesis, burns to GI tract, vomitus and stools may appear “smoking” or luminescent and have a garlic like odor, delirium, coma, shock, hypocalcemia, hypoglycemia, pulmonary edema, hemorrhage, cardiovascular collapse; delayed SX: myocardial, hepatic, and renal damage; dermally may cause partial and full thickness burns	Lavage and activated charcoal are usually not performed as phosphorous is corrosive; no specific antidote; supportive care with IV fluids and blood products; standard burn care; administer narcotics (e.g., morphine) for pain control; N-acetylcysteine may be hepatoprotective; ED staff must wear personal protective equipment to avoid contact with phosphorous
PNU (1) N-3-pyridylmethyl-N- p-nitro-phenyl urea, Vacor [®] Bait 0.5%, 2%	Destroys pancreatic β cells via interference with niacinamide (nicotinamide) metabolism; lethal dose: 5 mg/kg; product introduced in 1975 and withdrawn in 1979; SX: nausea, vomiting (peanut odor), hyperglycemia, diabetic ketoacidosis, sensory motor and autonomic neuropathies, GI perforation, cardiac dysrhythmias; permanent SX: insulin-dependent diabetes mellitus and postural hypotension	Orogastric lavage and activated charcoal; antidote: early IM or IV niacinamide (nicotinamide) may prevent toxicity, however, parenteral products are not available in the United States; niacin (nicotinic acid) may not be effective; treat hyperglycemic ketoacidosis with insulin; monitor for GI perforation; mineralocorticoids (e.g., fludrocortisone) for persistent postural hypotension
Red Squill (3) Urginea maritima Bait 100%, or as 4.5% extract in bait	Contains scillaren A and B, which are cardiac glycosides; two bulbs have been fatal to an adult; intensely nauseating causing rapid vomiting which limits toxicity; SX: large amounts may cause nausea, vomiting, hyperkalemia, A-V block dysrhythmias, however, cardiac toxicity is rare	Activated charcoal following large ingestions; monitor vital signs, serum potassium, and EKG; antidotes: atropine and digoxin immune Fab
Sodium mono-fluoroacetate (1) Compound 1080 [®] and Sodium fluoro-acetamide Compound 1081 [®]	SMFA is converted to fluorocitric acid which blocks the tricarboxylic cycle of the Krebs cycle; estimated lethal dose 2–10 mg/kg, however, 1 mg may cause serious toxicity; SMFA is a white crystalline powder combined with nigrosin black dye as a colorant; SX: nausea, vomiting, diarrhea, seizures, acidosis, cardiac dysrhythmias, hypotension, hypocalcemia, hypokalemia, respiratory depression, coma	Orogastric lavage and activated charcoal; supportive care for hypotension, acidosis, dysrhythmias, and seizures; IV calcium gluconate for hypocalcemia; no known effective antidote.

(Continued)

TABLE 115-2 Outdated, Illegal, and Unapproved Rodenticides for Indoor Use (Continued)

RODENTICIDE ^a	TOXICOLOGY/CLINICAL FINDINGS	MANAGEMENT/ANTIDOTES
Strychnine (1) Bait 0.3%–0.5%	Acts by antagonizing glycine, an inhibitory neurotransmitter in the postsynaptic motor neurons of the spinal cord; lethal dose 1–2 mg/kg, but may be as low as 5–10 mg in a child; SX: rapid onset of nausea, vomiting, apprehension, painful tonic-tetanic spasms, trismus, opisthotonos, “risus sardonicus” (facial grimacing), seizures, respiratory paralysis, lactic acidosis, rhabdomyolysis, hyperthermia, cardiac arrest; patient is conscious with normal mentation until hypoxia or acidosis leads to CNS depression	Activated charcoal; avoid any stimulus that may trigger seizures; treat seizures with benzodiazepines, phenobarbital; severe cases may need paralyzing agents with ventilator support; supportive care for acidosis, respiratory failure, rhabdomyolysis, and renal failure; no specific antidote, multiple dose activated charcoal possibly beneficial
Rodenticide: TETS (3) Tetramine (tetramethylenedisulfotetramine) Dushuqiang	TETS binds noncompetitively and irreversibly to GABA receptors on neuronal cell membranes and blocks chloride channels. Illegally imported from China. Two samples were analyzed to have 6.4% and 13.8%, respectively. LD50 in mammals: 0.10–0.3 mg/kg; 7–10 mg is lethal in humans. SX: Refractory seizures, coma, and possible ischemic changes on EKG	Information is limited. No specific antidote is available. Supportive care. Seizures were refractory to benzodiazepines and phenobarbital in one pediatric case. Animal studies in China suggest benefit from IV pyridoxine and DMSA. Patients in China have been treated with charcoal hemoperfusion and hemodialysis
Thallium sulfate (1)	Interferes with oxidative phosphorylation by binding with mitochondria sulfhydryl groups; banned in 1965; lethal dose 1 g; SX: nausea, bloody vomiting, and diarrhea followed by ileus, painful sensory neuropathy, respiratory failure, delirium, seizures, renal failure, optic neuritis, muscle weakness, lethargy, coma; delayed SX: alopecia, Mees lines, neuropathies	Orogastric lavage and activated charcoal as clinically indicated; thallium may appear radio-opaque on an abdominal radiograph, obtain blood and 24 h urine collection for thallium; Prussian blue (Radiogardase [®]) interrupts enterohepatic and enteroenteric circulation and enhances fecal elimination; multiple dose charcoal may also enhance elimination; supportive care including IV fluids, blood products, hemodialysis
Tres Pasitos (1) Aldicarb	Acts as a reversible inhibitor of cholinesterase enzymes; product is an approved carbamate insecticide sold illegally as a rodenticide; smuggled into the United States from the Dominican Republic; LD50 in rats: 1 mg/kg; Muscarinic SX: “SLUDGE ^{BAM} ,” Nicotinic SX: tachycardia, mydriasis, weakness, fasciculations, respiratory failure; CNS SX: coma, seizures	Activated charcoal; antidotes: IV atropine sulfate to reverse muscarinic symptoms, benzodiazepines for seizures

- Intravenous phytonadione offers the fastest onset of action (as little as 1–2 hours). However, IV administration is rarely used except for the most severe cases due to potential anaphylactoid reactions including hypotension, flushing, cyanosis, dizziness, dyspnea, cardiac, and/or respiratory arrest.
- Following large ingestions of superwarfarin products, antidotal therapy with large doses of vitamin K₁ (phytonadione), ie, 50 to 100 mg/d, may be required for weeks to months.
- For clinically significant active bleeding, FFP is essential to replenish all clotting factors except platelets. Packed red cells, which do not supply clotting factors, may be given to correct severe anemia secondary to hemorrhage.

DISPOSITION

- The asymptomatic pediatric patient with a reliable history of a “taste” amount of anticoagulant may be discharged from the emergency department with

- instructions provided to caregivers to observe for signs of bleeding or easy bruising for next few days.
- Asymptomatic pediatric patients ingesting >1 mg of active long-acting anticoagulant can be discharged and re-evaluated for coagulopathy at 48 to 72 hours post exposure.
- The adolescent patient who presents soon after the intentional ingestion of a large amount of anticoagulant bait is a candidate for activated charcoal, admission to a noncritical or psychiatric care unit, and monitoring of coagulation profile for 48 to 72 hours.
- For the asymptomatic patient presenting several days after a large acute ingestion, or in a patient with a history of chronic consumption with an abnormal coagulation profile, admission to a noncritical care unit with frequent coagulation studies is reasonable.
- For any symptomatic patient who is actively bleeding, admit to a critical care unit for definitive care with FFP and vitamin K₁.

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QUESTIONS

- A small child was seen with a box of green rodenticide pellets identified as a long acting anticoagulant. The child was with the box for only one to two minutes and it is suspected that only a mouthful was ingested. Which of the following is the best management approach?
 - Referral to the ED for activated charcoal then 48 to 72 hour outpatient coagulation studies.
 - Stat initiation of oral vitamin K₁ with home observation for symptoms.
 - Home observation only for any appearance of symptoms.
 - Referral to ED for activated charcoal and admission for 72 hours for inpatient monitoring.
 - Induce emesis at home with syrup of ipecac followed by outpatient coagulation studies in 48 to 72 hours.
- A toddler is suspected of ingesting 30 g (1 oz) of diphacinone 0.005% rat bait. Which of the following is the best approach for this patient?
 - Home observation for symptoms with outpatient coagulation studies at 48 to 72 hours post ingestion.
 - Stat initiation of oral vitamin K₁ with home observation for symptoms.
 - Referral to ED for activated charcoal and admission for 72 hours for inpatient monitoring.
 - Induce emesis at home with syrup of ipecac followed by outpatient coagulation studies in 48 to 72 hours.
 - Home observation only for any appearance of symptoms.

3. An adolescent female presents to the emergency department three days after ingesting a large box of anticoagulant rodenticide bait. She is currently asymptomatic with no bleeding diathesis. All labs are within normal limits with the exception of an INR of 7 on coagulation studies. What is the best antidotal treatment approach for this patient?
 - A. Oral phytonadione (vitamin K₁).
 - B. Subcutaneous phytonadione (vitamin K₁).
 - C. Intravenous phytonadione (vitamin K₁).
 - D. FFP.
 - E. Intramuscular phytonadione (vitamin K₁).
4. A developmentally delayed 6-year old with a history of PICA presents to the emergency department with epistaxis, dark urine, and patchy ecchymosis and he is suspected to have been chronically ingesting anticoagulant rodenticide. Which of the following should be ordered immediately?
 - A. Syrup of ipecac induced emesis.
 - B. Orogastric lavage.
 - C. Send out a rodenticide level to a reference laboratory.
 - D. Obtain a nephrology consultation for possible hemodialysis.
 - E. Stat CBC, urinalysis, PT/PTT, and INR.
5. An adolescent patient with a previous medical history of depression presents to the emergency department with extensive bleeding; hematochesia, and generalized bruising. Labs reveal very low hemoglobin and hematocrit with an immeasurable INR. The patient eventually admits to eating several boxes of anticoagulant rat bait 5 days prior to presentation. Which is the best management approach to this patient?
 - A. Multiple dose activated charcoal and subcutaneous phytonadione (vitamin K₁).
 - B. Multiple dose activated charcoal, intramuscular phytonadione (vitamin K₁) and FFP.
 - C. Intravenous phytonadione (vitamin K₁) and FFP.
 - D. Oral phytonadione.
 - E. FFP.
6. A previously healthy child presents to the emergency department with sudden onset diabetic ketoacidosis, ileus, and orthostatic hypotension. When asked if the child was exposed to any poisons, the mother states that 3 days prior the child was at the grandmother's home playing in the basement where there may have been some very old rat poison. Which product would most likely cause this combination of symptoms?
 - A. Arsenic
 - B. Zinc phosphide
 - C. White phosphorus
 - D. PNU (Vacor™)
 - E. Red Squill
7. Thirty minutes after entering an old barn, a toddler is found in what appears to be status epilepticus. A container with a handwritten note "rat poison" with no brand name or ingredient list is found. Which of the following substances is most likely the causative agent?
 - A. Sodium monofluoroacetate
 - B. Strychnine
 - C. Arsenic
 - D. White phosphorus
 - E. Bromethalin
8. A worried parent brings her asymptomatic child to the emergency department to be "checked out." Two days prior, the family pet dog was seen eating a cake of rodenticide and is now at the animal hospital being treated for extremely high calcium levels and "irregular heartbeat." The child was seen nearby, however not known to have ingested any product per se. Which rodenticide would most likely cause these symptoms?
 - A. Bromethalin
 - B. Zinc phosphide
 - C. Bromodialone
 - D. Cholecalciferol
 - E. Sodium monofluoroacetate
9. An adolescent presents to the emergency department 4 hours after intentionally consuming a container of "rat poison." His symptoms include corrosive gastritis, shock, and pulmonary edema. A distinctive odor of decaying fish is noted on patient's breath and gastric contents. Which rodenticide is the likely causative agent?
 - A. Zinc phosphide
 - B. Chlorphacinone
 - C. Cholecalciferol
 - D. PNU (Vacor™)
 - E. Calcium cyanide
10. A patient presents several weeks after ingesting an unknown rodenticide that was originally purchased outside the United States. The patient demonstrates alopecia, oliguria, neuropathy, and Mees lines on the fingernails. A high suspicion for which two of the following agents exists?
 - A. Sodium monofluoroacetate or calcium cyanide
 - B. Zinc phosphide or white phosphorus
 - C. Thallium sulfate or arsenic
 - D. Bromethalin or strychnine
 - E. PNU (Vacor™) or barium carbonate.

ANSWERS

1. C. Unintentional trivial pediatric ingestions of anti-coagulant rodenticides require no GI decontamination, referral to an emergency department, antidote, or laboratory studies. Therefore, answers A, B, D, and E are incorrect selections.
2. A. While decontamination, antidote, or admission to a monitored facility is unnecessary in larger unintentional asymptomatic pediatric ingestions, follow up coagulation studies should be done if the child ingests 1mg or more of active ingredient of an anticoagulant rodenticide.
3. A. Oral vitamin K₁ is the safest route of administration. FFP is normally unnecessary unless active bleeding is present.
4. E. Decontamination in a chronic ingestion is unnecessary. Rodenticide levels while available are not used to guide therapy, and hemodialysis is ineffective in an anticoagulant rodenticide exposure. Stat labs to determine the extent of bleeding and coagulation status are appropriate.
5. C. Intravenous phytonadione and FFP are used in combination for severe life threatening anticoagulation and bleeding due to anticoagulant rodenticides. Caution, however, is advised due to the potential for hypersensitivity reactions. Multiple dose charcoal has no proven benefit and other routes of administration of phytonadione would have slower onset of action.
6. D. Toxicity of PNU (Vacor™) is caused by pancreatic beta cell destruction and autonomic neuropathy. The other four agents demonstrate different toxidromes (Table 115-2)
7. B. Strychnine demonstrates a very rapid onset of CNS excitation. The other substances are characterized by delayed symptoms or other toxidromes (Tables 115-1 and 115-2)
8. D. Hypercalcemia is the primary action of cholecalciferol. The other rodenticides demonstrate other toxicities (Tables 115-1 and 115-2)
9. A. Zinc phosphide causes hemorrhagic gastritis and is broken down to phosphine gas, which accounts for multi-organ toxicity including pulmonary edema and often has a characteristic “decaying fish” odor. The remainder of the rodenticides listed present with different symptomatology (Tables 115-1 and 115-2).
10. C. Arsenic and Thallium are heavy metals with similar toxicity profiles. Neuropathy, nephrotoxicity, and alopecia, are characteristic of this toxicity. The other choices are not associated with the presenting symptoms (Tables 115-1, 115-2).

Prescription Drugs

116 CARDIOTOXINS

Allan R. Mottram

Jerrold Leikin

β-ADRENERGIC BLOCKING AGENTS

- The mortality rate following β-blocker overdose is much lower than that for calcium channel blockers or digoxin, but in terms of absolute numbers they are the second leading cause of death from cardiovascular medications.

PHARMACOLOGY

- The pharmacologic effects of β-blocking drugs are mediated through modulation of intercellular signals and calcium secondary to inhibited adrenergic activation. Although many β-blockers are β₁-selective at therapeutic doses, these drugs have both β₁- and β₂-effects in overdose.
- Intrinsic sympathomimetic properties of some β-blockers cause agonist-antagonist activity, which may blunt the bradycardic response in some patients. The membrane-stabilizing activity characteristic of some β-blockers is a quinidine-like effect, resulting in inhibition of fast sodium channels, decreased contractility, and ventricular arrhythmias.
- β-Blockers with increased intrinsic sympathomimetic activity and decreased membrane-stabilizing properties demonstrate less toxicity than those with increased membrane-stabilizing properties.
- Sotalol is a β-blocker, which has class III antiarrhythmic properties. In overdose, it may prolong the QT interval, resulting in ventricular arrhythmias, including torsades de pointes.

PHARMACOKINETICS

- The absorption, distribution, and elimination of β-blockers vary with the preparation. Extended-release formulations of β-blockers can have a marked delay in the onset of toxic effects. Conversely, standard

release β-blockers are rapidly absorbed. The elimination half-life varies from 2 to 24 hours, but can be significantly increased in overdose.

PATHOPHYSIOLOGY

- Toxicity from acute β-blocker overdose largely results from suppression of the cardiovascular system. Negative inotropic and chronotropic effects result in bradycardia and hypotension. Respiratory compromise in β-blocker overdose can result from cardiogenic shock, decreased respiratory drive, or β₂-antagonist effects. β₂-Blockade causes bronchospasm, and usually affects patients with previously diagnosed asthma. Hypoglycemia can occur secondary to β₂-mediated decrease in glycogenolysis and gluconeogenesis; however, it is not common unless there are associated comorbidities or coingestants. CNS depression can be caused by direct toxicity, hypoxia, hypoglycemia, or shock.

CLINICAL PRESENTATION

- The onset of symptoms most commonly occurs within 1 to 2 hours. Cardiovascular manifestations include hypotension, bradycardia, heart block, and congestive heart failure. Electrocardiographic manifestations of toxicity include sinus bradycardia, prolongation of the PR interval, second- and third-degree AV blockade, and interventricular conduction delays. The QRS may be prolonged with ingestions of β-blockers with membrane-stabilizing effects. Propranolol and sotalol have been associated with ventricular arrhythmias. Deaths from β-blockers toxicity are associated with bradydysrhythmias and asystole; ventricular arrhythmias are less common. Respiratory toxicity includes noncardiogenic pulmonary edema, pulmonary edema, exacerbation of asthma, and decreased respiratory drive. Patients may also present with CNS depression or seizures.

LABORATORY EVALUATION

- All patients with a history of β-blocker ingestion are placed on a cardiac monitor and receive an electrocardiogram (ECG). Laboratory tests for blood levels of β-blockers are not helpful in the acute setting. Serum electrolytes are obtained. Serum glucose is assessed.

MANAGEMENT

- A well-looking patient should not be reassuring, since they can decompensate quickly. Patients with

normal mental status should be decontaminated with activated charcoal.

- Glucagon is the agent of choice in β -blocker ingestions resulting in hypotension and/or bradycardia. Glucagon binds to its own receptor site, triggering cAMP signaling pathways, bypassing the cellular lesion at the β -receptor. An initial bolus of glucagon is administered intravenously at a dose of 0.05 to 0.15 mg/kg IV over 1 minute. If symptoms recur, a repeat bolus is given. An infusion can be started following the bolus dose, with the effective bolus dose infused per hour. The initial effect is seen within several minutes, and should persist for 10 to 15 minutes. Nausea and vomiting are common side effects of glucagon.
- Adrenergic agents are often effective in increasing heart rate, contractility, and peripheral vascular resistance. In cases of severe cardiovascular drug toxicity, large doses may be required. If the response to glucagon is inadequate, epinephrine and dopamine may improve both heart rate and blood pressure. Norepinephrine is effective in situations with low systemic vascular resistance; however, with the myocardial depression seen with severe β -blockade, alternative agents may be more efficacious. Atropine, 0.02 mg/kg IV (minimum single dose 0.1 mg; maximum cumulative dose 1 mg) may be useful for bradycardia.
- Bradycardia and hypotension refractory to pharmacologic intervention may benefit from temporary pacing, although this will not reverse the myocardial depression in severe overdose. Interventions such as intra-aortic balloon pump, extracorporeal membrane oxygenation (ECMO) or cardiac bypass are considerations for patients with toxicity refractory to all other therapy.
- Hemodialysis, hemofiltration, and hemoperfusion are rarely useful in the setting of β -blocker overdose. Hemodialysis can be considered in the setting of renal failure and hemodynamic instability in a drug with low volume of distribution and low protein binding.

DISPOSITION

- A patient with a history of immediate-release β -blocker ingestion is observed on a cardiac monitor for 8 hours after ingestion. Patients who have signs of cardiovascular, respiratory, or CNS toxicity are admitted to an intensive care setting. Patients with a history of ingestion of extended-release preparations or sotalol are admitted and monitored for 24 hours. A patient who ingested an immediate release β -blocker can be medically cleared after the 8-hour observation period if there are no signs of toxicity found by clinical examination, ECG, or cardiac monitoring.

CALCIUM CHANNEL BLOCKERS: HIGH-YIELD FACTS

- The American Association of Poison Control Centers annual report indicated 10,031 calcium channel blocker exposures in 2006. Deaths due to calcium channel blocker overdose have been declining in recent years and rarely occur in the pediatric setting.

PHARMACOLOGY

- Calcium channel blockers are classified as dihydropyridines, phenylalkylamines, or benzothiazepines.
- The clinical effects of the three classes of calcium channel blockers differ for several reasons. They bind at different locations on calcium channel receptor subunits have preference for different resting cell membrane potentials, and bind as a function of channel state. Receptor selectivity translates into the dihydropyridines primarily resulting in vasodilation; the nondihydropyridines have more pronounced effects on cardiac conduction. Verapamil affects myocardial contractility, AV node conduction, peripheral vascular resistance, and is one of the more toxic calcium channel blockers in overdose. In overdose all classes of calcium channel blockers can cause significant peripheral vasodilatation, decreased AV conduction, and decreased myocardial contractility.

PHARMACOKINETICS

- Most calcium channel blockers undergo hepatic metabolism with extensive first pass effect, have a large volume of distribution, and are highly protein bound. The onset of action for immediate release preparations is 30 minutes, with a half-life from 3 to 7 hours; this can be greatly increased in the setting of overdose and with sustained-release preparations.

PATHOPHYSIOLOGY

- The clinical effects of calcium channel blocker overdose can be life threatening. Slowing of the sinus node causes bradycardia. Slowing of conduction can cause heart blocks or asystole. Decreased contractility can cause heart failure and shock. Lowered peripheral vascular resistance leads to hypotension, which may exacerbate the hypotension associated with bradycardia, bradyarrhythmias, and heart failure. Hyperglycemia occurs frequently with significant overdoses, and may correlate with the severity of poisoning. Patients with cardiac disease and those on

other medications that suppress heart rate and contractility may develop severe toxic effects after mild overdose, or even at therapeutic doses.

- In all cases cardiovascular effects predominate. Verapamil and diltiazem typically cause bradycardia and hypotension. Hypotension may be caused by sinoatrial node depression, atrioventricular node depression leading to AV blocks, or decreased peripheral vascular resistance. Nifedipine primarily affects the arterioles, causing decreased peripheral vascular resistance, which leads to hypotension and reflex tachycardia.
- Neurologic and respiratory findings are usually secondary to cardiovascular toxicity and shock. Respiratory effects include decreased respiratory drive, pulmonary edema, and ARDS. Neurologic sequelae include depressed sensorium, cerebral infarction, and seizures. Nausea, vomiting, and constipation can occur.

LABORATORY EVALUATION

- Drug levels for calcium channel blockers are not helpful in an acute overdose. An ECG is obtained. Electrolytes are evaluated, specifically Na^+ , Ca^{2+} , Mg^{2+} , and K^+ . Glucose is evaluated since decreased insulin release can lead to hyperglycemia. An abdominal radiograph may be useful in patients with a history of ingesting sustained-release tablets, since some calcium channel blockers are radiopaque, and concretions can occur.

MANAGEMENT

- Patients with normal mental status should be decontaminated with activated charcoal. Whole bowel irrigation can be considered for asymptomatic patients who present early after overdosing on a sustained-release formulation, but it should be used with great caution as it may complicate the management of patients who subsequently become hypotensive.
- Following initial resuscitation, therapy focuses on enhancing calcium channel function. However, treatment may have little effect when the calcium channel is severely poisoned. Calcium salts increase extracellular calcium concentration, and may reverse hypotension because of vasodilation, especially in less severe overdoses. However, improvement is usually transient in the serious overdose setting, and there is little or no effect on heart rate or conduction. Atropine may be helpful for patients with symptomatic bradycardia or heart block. Isoproterenol or

pacemaker devices may be useful. A trial of glucagon is reasonable when coingestion with a β -blocker is suspected. It is not as effective for calcium channel blocker poisoning as it is for β -blocker poisoning.

- Vasopressors should be utilized early for patients who do not respond to intravenous fluid, calcium, and atropine. An agent with combined α - and β -effects, such as high-dose dopamine or norepinephrine, is appropriate. Phenylephrine and dobutamine may also be effective. More than one agent may be required.
- Hyperinsulinemia–euglycemia therapy (HIE) should be considered early in the critically ill patient. Efficacy of HIE is likely attributable to the metabolic effects of insulin which result in improvement in blood pressure, systolic and diastolic myocardial performance, and survival time.
- The protocol for HIE is 1 unit/kg regular insulin intravenous bolus followed by 0.5 units/kg/hr intravenous infusion. An intravenous dextrose bolus of 0.25 gm/kg, followed by an infusion of 0.5 g/kg/h may be initiated; however, patients with significant poisoning are not expected to develop hypoglycemia. Serial blood sugar determinations are followed, and the dextrose infusion adjusted accordingly. Potassium is monitored and replaced to maintain serum potassium levels at 2.8 to 3.2 mEq/L.
- Interventions such as intra-aortic balloon pump, ECMO or cardiac bypass are considerations for patients with toxicity refractory to all other therapy. Most calcium channel antagonists have a large volume of distribution, are highly protein bound, and subject to hepatic metabolism making them poor candidates for extracorporeal removal.
- Patients with a history of sustained-release ingestion are observed for at least 24 hours. Those patients with no signs of toxicity, no history of sustained-release ingestion, and no ECG abnormalities can be observed for 8 hours after the time of ingestion.

DIGOXIN

- Digoxin is used today for the treatment of congestive heart failure and supraventricular dysrhythmias. Historically, mortality because of digoxin overdose has been related to the type of cardiac arrhythmia induced by toxicity and the degree of associated hyperkalemia. Mortality rates of 68% for patients exhibiting digoxin-induced sustained ventricular tachycardia and 100% for ventricular fibrillation were noted prior to the development of digoxin immune Fab fragments.

PHARMACOLOGY/PATHOPHYSIOLOGY

- Digoxin is a positive inotrope that increases the force and velocity of myocardial contractions. In the failing heart, it can increase the cardiac output and decrease elevated end-diastolic pressures.
- At toxic concentrations, it is felt that intracellular calcium concentrations are markedly increased, and that the membrane potential is unstable, which leads to dysrhythmias.
- There are numerous factors that predispose the patient to digoxin toxicity, the most common of which is electrolyte imbalance. Both hypokalemia and hyperkalemia can increase the possibility of developing digoxin toxicity. Hyperkalemia in particular can result in significant conduction delays. Hypokalemia is common in patients on diuretic therapy and can predispose patients to the effects of chronic digoxin toxicity. Hypomagnesemia, hypercalcemia, renal

insufficiency, and underlying heart disease all predispose to digoxin toxicity.

CLINICAL PRESENTATION

- The presentation of digoxin toxicity is highly varied, and depends largely on whether it results from an acute overdose or is a manifestation of chronic toxicity (Fig. 116-1).
- In the acute setting, patients tend to have more dramatic, clinical, and laboratory parameters than in chronic toxicity. Symptoms can be abrupt, with severe nausea, vomiting, and diarrhea. Associated complaints include weakness, headache, paresthesias, and altered color perception. Cardiovascular symptoms include palpitations and dizziness that may be secondary to hypotension. Movement disorders may also be present.

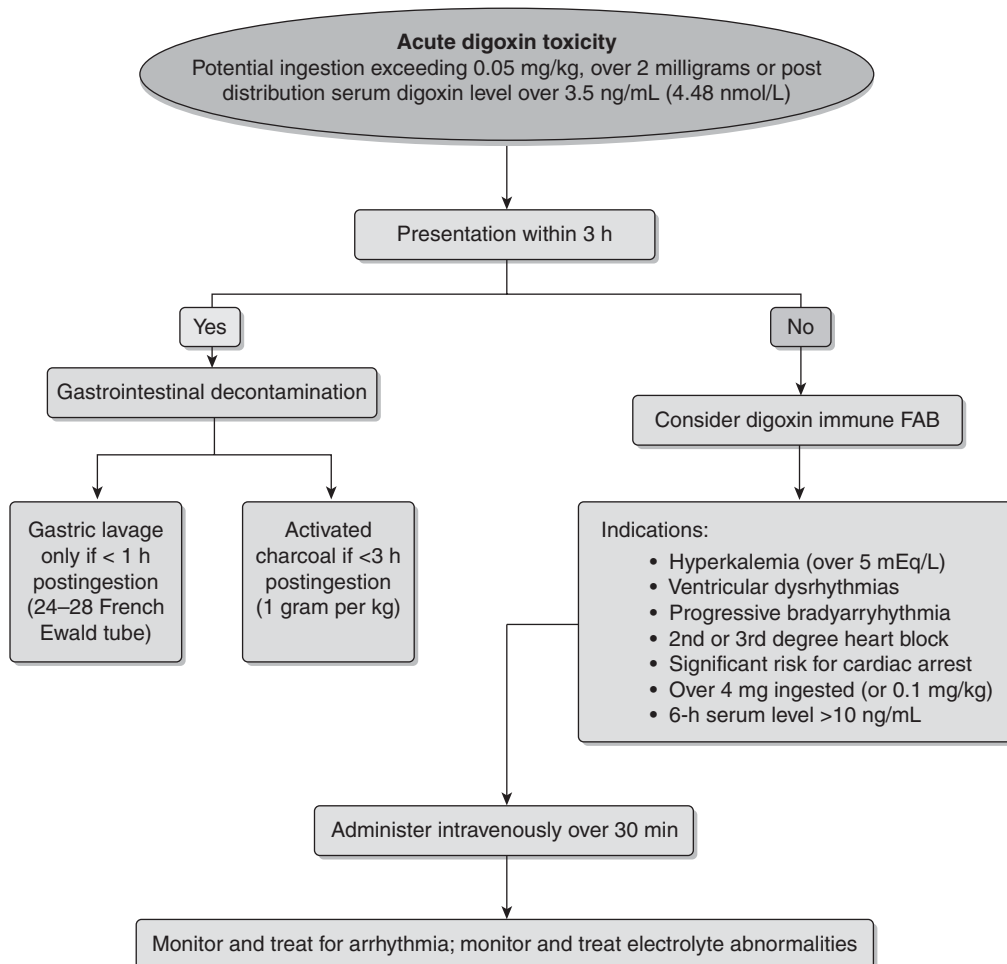


FIG. 116-1. Flow chart for initial evaluation and treatment of acute digoxin toxicity.

- Patients with chronic toxicity tend to have more vague complaints, although many of the symptoms of acute overdose also occur. Malaise, anorexia, and low-grade nausea and vomiting are common. Patients with chronic toxicity tend to be more symptomatic at lower levels than those with acute overdoses.
- Cardiovascular toxicity is the most important factor in determining morbidity and mortality. There are multiple dysrhythmias associated with digoxin toxicity, the most common being frequent premature ventricular beats. Other dysrhythmias can be supraventricular, nodal, or ventricular. Common disturbances are junctional escape beats and accelerated junctional rhythm, paroxysmal atrial tachycardia with AV block, and AV block of varying degrees. There is no single pathognomonic rhythm. Lethal cardiac disturbances rarely occur in children with normal hearts, but serious AV conduction disturbances can occur.

DIAGNOSIS

- A history of the exact amount of digoxin ingested is extremely helpful. A dose greater than 0.1 mg/kg is an indication that serious consequences can occur.
- A serum digoxin level is indicated whenever there is clinical suspicion of toxicity. In an overdose situation, the level is most accurate if obtained ≥ 6 hours after the ingestion. The therapeutic digoxin range is between 0.8 and 1.8 ng/mL. Unfortunately, there is poor correlation between the digoxin level and clinical manifestations. In an acute overdose, a level as high as 2.6 ng/mL does not correlate well with toxicity. In a chronic overdose, toxicity can occur at lower levels. The fatality rate approaches 50% when the serum digoxin level exceeds 6 ng/mL.
- Other necessary laboratory studies include a complete blood count, serum electrolytes, calcium, magnesium, blood urea nitrogen, and creatinine. Cardiac monitoring is essential, as is a 12-lead electrocardiogram.

MANAGEMENT

- Activated charcoal is indicated as a single dose. Multiple doses of charcoal have been reported to be of value for digitoxin preparations in which there is avid enterohepatic circulation, and may be of value for digoxin. However, the advent of Digibind has supplanted consideration for enhanced elimination with MDAC. Whole-bowel irrigation should be avoided in patients with potential for hemodynamic instability.

ANTIDOTAL THERAPY

- Digoxin immune Fab fragments are specific anti-digoxin antibodies derived from sheep. In order to decrease the risk of immunogenicity, only the Fab fragment is used. Specific indications include an ingestion of greater than 0.1 mg/kg, a digoxin level of greater than 10.0 ng/mL, potassium greater than 5 mEq/L, or the presence of a life-threatening dysrhythmia. In chronic digoxin poisoning significant toxicity may occur at much lower serum levels. Standard modalities to treat hyperkalemia may also be used, with the exception of calcium salts. In the face of digoxin toxicity, the administration of calcium may exacerbate the development of dysrhythmias.
- The dose of Fab fragments is based either on the amount ingested or on the serum level. Each vial of Fab fragments contains 38 mg of protein that will bind 0.6 mg of digoxin. Specific guidelines for dosing Fab fragments are available on the package insert.
- Allergic reactions to Fab fragments are rare. Skin testing can be performed, but is usually not necessary. In cases where Fab fragments have been effective, results have been achieved 30 minutes to 4 hours after administration. After administration of Fab fragments, subsequent digoxin levels will be falsely elevated for several days, because the bound digoxin is measured along with the free drug. Certain laboratories can assay free digoxin levels, which avoids this problem.
- In addition to the administration of Fab fragments, standard treatment of dysrhythmias or AV blocks is indicated.
- Atropine or temporary pacing may be necessary to temporize while Fab fragments are taking effect. Cardioversion and lidocaine are appropriate in the event of ventricular tachycardia or fibrillation. Treatment with intravenous phenytoin or magnesium sulfate has been shown to be useful in digoxin-induced tachydysrhythmias. Drugs to avoid in the treatment of digoxin-induced cardiac toxicity include calcium, bretylium tosylate, sotalol, isoproterenol, and quinidine. Direct-current cardioversion should only be used as a last resort for unstable, life-threatening arrhythmias. If utilized, it should be dosed at the lowest energy possible.
- Diuresis, hemodialysis, and hemoperfusion do not aid in the removal of digoxin or digitoxin. Plasma exchange is also not useful.

DISPOSITION

Children with trivial ingestions (less than 0.05 mg/kg) who are asymptomatic and have no detectable levels of digoxin 4 hours after the ingestion can be discharged

from the emergency department after 6 hours of observation. Any child with signs or symptoms of toxicity is admitted to a pediatric intensive care unit.

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QUESTIONS

- An 18-month-old child presents with a history of taking some of her grandmother's blood pressure medication. The examination of the bottle indicates that the medication is an immediate-release beta-blocker. Her blood pressure is 60/palpation and pulse rate is 50. What is the agent of choice in for the initial management of this patient?
 - Atropine
 - Calcium gluconate
 - Dopamine
 - Naloxone
 - Glucagon
- Which cardiac rhythm is pathognomonic for digoxin toxicity in the setting of an acute ingestion?
 - Supraventricular tachycardia
 - Junctional escape beats
 - Accelerated junctional rhythm
 - Paroxysmal atrial tachycardia with block
 - None
- A 2-year-old child presents after an accidental ingestion of his father's antihypertensive medication identified as a calcium channel blocker. He presents hypotensive and bradycardic. Which of the following is correct in the treatment of this patient with a calcium channel blocker overdose?
 - Atropine is contraindicated.
 - High dose insulin can be helpful.
 - Treatment with calcium salts is invariably effective in reversing hypotension.
 - Vasopressors are never indicated.
 - Hemodialysis is always effective when other therapies fail.

4. A 3-year-old female with congenital heart disease is on daily doses of digoxin. Her parents bring her to the ED after the patient vomited three times, had several bouts of diarrhea and complained of a headache. Her serum digoxin level is 6 ng/mL. She is noted to have a paroxysmal atrial tachycardia with AV block. Which of the following is true regarding digitalis toxicity in this patient?
 - A. It is not affected by the level of serum potassium.
 - B. It is associated with vague complaints when toxicity is of a chronic nature.
 - C. Rarely associated with premature ventricular beats.
 - D. It is associated with higher serum digoxin levels in chronic versus acute toxicity.
 - E. Morbidity and mortality are related to CNS toxicity.
5. In the above patient which of the following is true regarding her treatment options?
 - A. Needs emergent dialysis.
 - B. Requires the administration of calcium salts.
 - C. Requires digoxin immune Fab fragments in a dose-dependent fashion.
 - D. Direct-current cardioversion is recommended for her tachydysrhythmias.
 - E. Does not depend on the amount of digoxin ingested.

ANSWERS

1. E. Glucagon is the agent of choice in beta-blocker ingestions resulting in hypotension or bradycardia. Atropine may be used for persistent bradycardia but is not the first line choice. Likewise, dopamine may be used to counter hypotension and bradycardia if glucagon is ineffective. Calcium is used for calcium channel blocker ingestions. Naloxone has no use in beta-blocker ingestions.
2. E. While all of these rhythms may occur with digoxin toxicity, none are pathognomonic.
3. B. HIE therapy should be considered early in the critically ill patient. The efficacy of HIE is likely attributable to the metabolic effects of insulin which result in improvement in blood pressure, systolic and diastolic myocardial performance, and survival time. Atropine may be helpful for patients with symptomatic bradycardia or heart block. Calcium salts increase extracellular calcium concentration, and may reverse hypotension because of vasodilation, especially in less severe overdoses. However, improvement is usually transient in the serious overdose setting, and there is little or no effect on heart rate or conduction. Vasopressors should be utilized

early for patients who do not respond to intravenous fluid, calcium, and atropine. Most calcium channel antagonists have a large volume of distribution, are highly protein bound, and subject to hepatic metabolism making them poor candidates for hemodialysis.

4. B. In the acute setting, patients tend to have more dramatic clinical, and laboratory parameters than in chronic toxicity. Patients with chronic toxicity tend to have more vague complaints. There are numerous factors that predispose the patient to digoxin toxicity, the most common of which is electrolyte imbalance. Both hypokalemia and hyperkalemia can increase the possibility of developing digoxin toxicity. Cardiovascular toxicity is the most important factor in determining morbidity and mortality. There are multiple dysrhythmias associated with digoxin toxicity, the most common being frequent premature ventricular beats. Patients with chronic toxicity tend to be more symptomatic at lower levels than those with acute overdoses.
5. C. Digoxin immune Fab fragments are specific antidigoxin antibodies and specific indications for use include an ingestion of greater than 0.1 mg/kg, a digoxin level of greater than 10.0 ng/mL, potassium greater than 5 mEq/L, or the presence of a life-threatening dysrhythmia. In chronic digoxin poisoning significant toxicity may occur at much lower serum levels. Standard modalities to treat hyperkalemia may also be used, with the exception of calcium salts. In the face of digoxin toxicity the administration of calcium may exacerbate the development of dysrhythmias. The dose of Fab fragments is based either on the amount ingested or on the serum level. Diuresis, hemodialysis, and hemoperfusion do not aid in the removal of digoxin or digitoxin. Direct-current cardioversion should only be used as a last resort for unstable, life-threatening arrhythmias. If utilized, it should be dosed at the lowest energy possible.

117 PRESCRIPTION DRUGS: ANTIDEPRESSANTS

Michael R. Christian

INTRODUCTION

- Antidepressants represent approximately 20% of all prescription medications in the United States.
- There is an increasing number of antidepressants available for use (see Table 117-1).
- While much less toxic than tricyclic antidepressants (TCAs), selective serotonin reuptake inhibitors

TABLE 117-1 Antidepressant Class

TCA
Amitriptyline
Imipramine
Nortriptyline
SSRI
Citalopram
Escitalopram
Fluoxetine
Fluvoxamine
Paroxetine
Sertraline
Atypicals
SNRI (Serotonin Norepinephrine Reuptake Inhibitor)
Duloxetine
Venlafaxine
NDRI (Norepinephrine Dopamine Reuptake Inhibitor)
Bupropion
Miscellaneous
Trazodone (serotonin agonist, α -antagonist)
Mirtazepine (SSRI and α_2 -blocker)

(SSRs) and atypical antidepressants have caused deaths in the pediatric population.

TRICYCLIC ANTIDEPRESSANTS

PHARMACOLOGY AND PATHOPHYSIOLOGY

- TCAs inhibit presynaptic reuptake of norepinephrine and serotonin. In overdose, TCAs have a wide variety of pharmacologic effects which result in clinical toxicity.
- Quinidinelike effect: QT and QRS prolongation often followed by dysrhythmias.
- Norepinephrine reuptake inhibition: tachycardia, hypertension, and seizures. Hypotension may follow secondary to depletion of monoamine stores.
- Alpha-blockade: hypotension due to a decrease of vasomotor tone.
- Anticholinergic effects: hyperthermia, tachycardia, mydriasis, dry mucous membranes, decreased gastrointestinal motility, urinary retention, and altered mental status ranging from agitation to seizures to coma.

CLINICAL MANIFESTATIONS & DIAGNOSIS

- Patients will manifest a variety of symptoms due to the pharmacologic effects of TCAs.
- It is critical to recognize that patients may initially appear stable followed by a rapid deterioration, which usually occurs within 2 hours of ingestion.

- Qualitative drug screens may help in diagnosis. However, quantitative serum levels are of low clinical utility.

MANAGEMENT

- Intubation is necessary for patients with depressed mental status or an absent gag reflex.
- Intubated patients should be hyperventilated to a pH of 7.45–7.5.
- Hypotension should be treated initially with boluses of crystalloid followed by a continuous infusion of norepinephrine if necessary.
- An EKG should be performed immediately and cardiac monitoring is essential.
- QRS duration between 100–160 ms is associated with a higher risk of seizures and dysrhythmias.
- If the QRS duration exceeds 120 ms, 1–2 mEq/kg of sodium bicarbonate should be given as a bolus repeatedly until the QRS duration is less than 100 ms, the arterial pH exceeds 7.5, or the patient becomes hypernatremic.
- In patients with ventricular tachycardia, lidocaine can be considered.
- Seizures are generally self-limited but can be treated with a benzodiazepine. However, seizures are an ominous sign of impending cardiovascular toxicity.

DISPOSITION

- Patients without tachycardia or other anticholinergic effect, QRS widening, or drowsiness during a 6-hour observation periods may be safely discharged.

SELECTIVE SEROTONIN REUPTAKE INHIBITORS

PHARMACOLOGY & PATHOPHYSIOLOGY

- SSRIs act primarily to inhibit the reuptake of serotonin. Because of this selectivity, SSRIs are much less toxic than TCAs in overdose.

CLINICAL MANIFESTATIONS & DIAGNOSIS

- SSRIs can cause a variety of symptoms secondary to serotonergic excess including central nervous system (dizziness, lethargy), cardiovascular (tachycardia), and gastrointestinal (vomiting) effects.
- In extreme cases, which are usually associated with coingestants, serotonin syndrome may occur.

- Serotonin syndrome is manifested by altered mental status, hyperreflexia, rigidity, tremor, and hyperthermia, which can lead to rhabdomyolysis, renal failure, hepatic failure, and hyperthermia-induced cellular dysfunction.

MANAGEMENT

- Treatment generally consists of supportive care.
- Benzodiazepines should be liberally used to treat agitation, tachycardia, and tremor.
- Hyperthermic patients should be actively cooled.
- An EKG should be obtained. Citalopram and escitalopram have been associated with QRS and QT prolongation, which can be delayed.
- The use of cyproheptadine, a serotonin antagonist, can be considered but its benefit beyond supportive care is unproven.

DISPOSITION

- Patients with severe toxicity should be admitted to the ICU.
- Asymptomatic patients with a normal EKG may be safely discharged after a 6–12 hour observation period.

ATYPICAL ANTIDEPRESSANTS

PHARMACOLOGY & PATHOPHYSIOLOGY

- Serotonin–norepinephrine reuptake inhibitors (SNRIs) like venlafaxine and duloxetine inhibit serotonin and norepinephrine reuptake.
- Dopamine–dopamine reuptake inhibitors (NDRIs) like bupropion inhibit norepinephrine and dopamine uptake.
- Trazadone is a serotonin agonist and an alpha-antagonist.
- Mirtazapine acts as a SSRI and alpha 2-blocker.

CLINICAL MANIFESTATIONS, DIAGNOSIS, & TREATMENT

- Treatment generally consists of supportive care.
- In addition to symptoms that are seen with SSRIs, SNRIs, and NDRIs can also cause seizures and quinidinelike effects, similar to TCAs, which can be treated with repeated boluses of sodium bicarbonate.
- Trazadone and mirtazapine in overdose usually cause a depressed mental status (due to excess serotonin)

and hypotension (due to alpha-blockade). Priapism has been associated with use of trazadone.

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QUESTIONS

1. The cardiovascular toxicity of TCAs is due to which of the following pharmacologic effects?
 - A. Quinidinelike effect
 - B. Anticholinergic side effects
 - C. Blockade of norepinephrine uptake
 - D. Alpha-adrenergic blockade
 - E. All of the above
2. A 15-year-old female with a history of depression is brought to the ED 2 hours after reportedly ingesting an unknown quantity of amitriptyline. On presentation, she is alert with stable vital signs and has no complaints. She now denies ingestion. Which of the following statements regarding her expected ED course and management is correct?
 - A. Her mental status, symptomatology, and cardiac rhythm should be observed for 6 hours, and if no abnormality is detected, she can safely be discharged pending psychiatric evaluation.
 - B. Since there is no change in mental status or cardiac rhythm on presentation, she is unlikely to develop a problem, and she can safely be referred to psychiatry without further medical attention.
 - C. She must be admitted for continued observation since toxic effects are often delayed for up to 24 hours.

- D. Activated charcoal is unlikely to be effective since the patient did not present within 1 hour of reported ingestion.
- E. In spite of the delayed presentation, ipecac for induction of emesis is indicated to reduce the load of potentially highly toxic drug.
3. A 16-year-old male is brought to the ED by his parents after taking 20 to 30 tablets of his mother's amitriptyline. On presentation, his airway is clear but he is somewhat somnolent. Blood pressure is 90/50, heart rate is 130, and respiratory rate is 20. His temperature is 101°F. An EKG is performed and reveals tachycardia with wide QRS complexes. What is the most appropriate course of action?
- A. Initiate a dopamine drip at 2 to 5 $\mu\text{g}/\text{kg}/\text{min}$.
- B. Administer activated charcoal, 50 g PO and continue to observe.
- C. Intubate, hyperventilate, and administer sodium bicarbonate, 1–2 mEq/kg as a bolus.
- D. Physostigmine 0.02 mg/kg slow IV push.
- E. Phenytoin 18 mg/kg IV over 1 hour as a prophylaxis for seizures.
4. A 16-year-old female with a history of depression ingested an unknown amount of her fluoxetine and may have had access to her father's phenelzine. Upon arrival, the patient's vital signs are: temperature = 104.5°F, blood pressure = 162/95, heart rate = 127, respiratory rate = 20, and oxygen saturation = 98% on room air. On exam, the patient is very agitated, unable to answer questions appropriately, tachycardic, exhibits 4+ patellar reflexes bilaterally, and is vomiting. Which of the following is the most critical action in the patient's immediate treatment?
- A. Administer cyproheptadine 12 mg PO.
- B. Administer haloperidol 5 mg IV for agitation.
- C. Administer activated charcoal 50 g PO.
- D. Initiate aggressive supportive care including active cooling and liberal use of benzodiazepines for agitation and tachycardia.
- E. Initiate aggressive supportive care including intubation, hyperventilation to a pH of 7.5, and boluses of sodium bicarbonate 1–2 mEq/kg.
5. In the previous patient, a urine dipstick was performed which was remarkable for 3+ blood but the urine microscopy was negative for RBCs. Which study should be performed next?
- A. Urine culture.
- B. Renal ultrasound.
- C. Noncontrast CT scan of abdomen and pelvis.
- D. Creatine phosphokinase level.
- E. Coagulation studies.
6. An 8-year-old male accidentally ingested some of his mother's antidepressant medication that she kept in a Tic-Tac container. The mother does not remember the name of her medication. The clinical pharmacist is contacting the patient's pharmacy to determine which medication the patient had been prescribed. The patient is somnolent but arousable with normal vital signs. An EKG is normal except for a QTc of 512 ms. Which of the following medications is the most likely to have been ingested?
- A. Fluoxetine
- B. Escitalopram
- C. Paroxetine
- D. Sertraline
- E. Fluvoxamine
7. An overdose of which of the following medications is most likely to require treatment with boluses of sodium bicarbonate 1–2 mEq/kg IV?
- A. Venlafaxine
- B. Fluoxetine
- C. Mirtazapine
- D. Trazadone
- E. Sertraline
8. A 16-year-old male with a history of depression presents to the ED complaining of a painful erection for 5 hours. The patient does not remember the name of his medication for depression but denies taking any additional medications. Which of the following antidepressants is the patient most likely taking?
- A. Amitriptyline
- B. Mirtazapine
- C. Trazadone
- D. Sertraline
- E. Phenelzine

ANSWERS

1. E. The quinidinelike effect is responsible for cardiac dysrhythmias but the anticholinergic effects, blockade of norepinephrine reuptake, and α -blockade all contribute to the tachycardia and hypertension.
2. A. The majority of patients who develop life-threatening problems do so within 2 hours of arrival in the ED. Toxic patients are likely to arrive appearing clinically stable and then may suddenly deteriorate. If no toxic effects are observed during a 6-hour period, medical monitoring can be discontinued. Psychiatric evaluation is indicated to assess for suicidal risk. Ipecac is contraindicated due to the risk for sudden deterioration, but charcoal is indicated even for delayed presentations, since the anticholinergic effects of the drug results in decreased gut motility.

3. C. If the QRS complex is greater than 100 ms, most investigators agree that alkalinization is indicated. This can be accomplished by a combination of intubation and hyperventilation in addition to IV boluses of sodium bicarbonate. The goal of alkalinization is to achieve a pH between 7.45 and 7.5. The initial approach to hypotension is fluid administration, followed by vasopressors if fluids are ineffective. There is a theoretical advantage to norepinephrine over dopamine due to its ability to reverse the alpha-blockade caused by the drug. Seizures are usually self-limited and may not require treatment. Phenytoin is not effective prophylactically. Physostigmine should be reserved for patients who do not respond to first-line therapy for seizures, dysrhythmias, or severe hypertension.
4. D. In cases of serotonin syndrome, aggressive supportive care including active cooling and benzodiazepines is critical. The use of cyproheptadine, which is only available orally, is contraindicated in patients who are vomiting. In addition, its benefit beyond aggressive supportive care is questionable. Haloperidol is of little use in this scenario and may be harmful. Activated charcoal should not be administered in a patient who is actively vomiting. Intubation, hyperventilation to a pH of 7.5, and boluses of sodium bicarbonate 1–2 mEq/kg is the preferred treatment in patients who have toxicity from ingestion of TCAs.
5. D. The patient is most likely in rhabdomyolysis. The heme group in myoglobin will cause a urine dipstick to be positive for blood, but the urine microscopy will be negative for RBCs. Aggressive resuscitation with crystalloid is critical and urinary alkalinization may be necessary. A urine culture, coagulation studies, or renal imaging will not help in the diagnosis of rhabdomyolysis.
6. B. Of the SSRIs, citalopram and escitalopram are the most likely to cause a prolonged QT interval.
7. A. Of the antidepressants listed, venlafaxine is the most likely to cause a quinidinelike effect requiring treatment with sodium bicarbonate.
8. C. Of the antidepressants listed, trazadone is the only medication associated with priapism.

118 NEUROLEPTICS

Timothy B. Erickson

GENERAL

- Neuroleptics or phenothiazines are the group of major tranquilizers or antipsychotic drugs that are therapeutically designed to treat schizophrenia and other psychiatric disorders (Table 118-1).

PATHOPHYSIOLOGY

- Neuroleptics act by blocking dopaminergic, alphaadrenergic, muscarinic, histaminic, and serotonergic neuroreceptors. Blockade of the dopamine receptors not only results in the desired behavior modification, but also produces extrapyramidal side effects, such as dystonic reactions.
- Alpha-adrenergic blockade produces peripheral vasodilation and orthostatic hypotension. Muscarinic blockade results in anticholinergic properties such as sedation, tachycardia, flushed or dry skin, urinary retention, and delayed GI motility.
- Neuroleptics also cause a membrane depressant action or quinidine-like effect that alters myocardial contractility, and can result in conduction defects. Although the mechanism of toxicity in neuroleptics resembles that of tricyclic antidepressants, serious cardiac dysrhythmias, refractory hypotension, respiratory depression, and seizures are uncommon.

TABLE 118-1 Common Typical Neuroleptics

CHEMICAL CLASS	GENERIC NAME	PROPRIETARY NAME
Phenothiazines		
Aliphatics	Chlorpromazine	Thorazine
	Promethazine	Phenergan
	Acepromazine (veterinary)	Aceprotabs
Piperazines	Prochlorperazine	Compazine
	Trifluoperazine	Stelazine
	Fluphenazine	Prolixin
	Perphenazine	Trilafon
Piperidines	Mesoridazine	Serentil
	Thioridazine	Mellaril
Butyrophenones	Droperidol	Inapsine
	Haloperidol	Haldol
Thioxanthenes	Thiothixene	Navane
Indoles	Molindone	Mobane
Dibenzoxazepines	Loxapine	Loxitane
Diphenylbutylpiperidines	Pimozide	Orap

DYSTONIC REACTIONS

CLINICAL PRESENTATION

- Acute dystonia is an unpredictable side effect of neuroleptics, and it occurs in approximately 10% of overdoses. It can also occur as an idiosyncratic reaction following a single therapeutic dose of a neuroleptic. These reactions are characterized by slurred speech, dysarthria, confusion, dysphagia, hypertonicity, tremors, and muscle restlessness. Of the neuroleptics, prochlorperazine most often causes acute dystonia.
- Other reactions or dyskinesias include oculogyric crisis (upward gaze), torticollis (neck twisting), facial grimacing, opisthotonos (scoliosis), and tortipelvic gait disturbances. Symptoms usually begin within the first 5 to 30 hours after ingestion.
- In recent years several new neuroleptic agents have become very popular, including clozapine, olanzapine, risperidone, quetiapine, ziprasidone, aripiprazole, and paliperidone. (Table 118-2) This group of agents produces a lower incidence of extrapyramidal side effects than previous agents.

MANAGEMENT

- If a child exhibits signs of acute muscular dystonia, intravenous diphenhydramine (2 mg/kg up to 50 mg over several minutes) is rapidly administered. Improvement usually occurs within 15 minutes.
- Alternatively, the patient can be given benztropine intramuscularly in a dose of 0.05 to 0.1 mg/kg (up to 2 mg, not to exceed 8 mg over 24 hours).

ACUTE OVERDOSE

CLINICAL PRESENTATION

- Following an acute overdose of neuroleptics, mild CNS depression is common, usually occurring within 1 to 2 hours of the ingestion. Children are more susceptible to these sedative effects than adults. In the overdose setting, respiratory depression can occur, but rarely requires aggressive airway management.
- Phenothiazines tend to lower a patient's seizure threshold, though the actual incidence of seizures in acute overdose is low.
- Like the tricyclic antidepressants, poisoning from neuroleptics can result in orthostatic hypotension and cardiac dysrhythmias, particularly with henothiazines. Sinus tachycardia is the most common dysrhythmia,

TABLE 118-2 Atypical Neuroleptic Agents

CHEMICAL CLASS	GENERIC NAME	PROPRIETARY NAME
Unclassified	Aripiprazole	Abilify
Dibenzodiazepines	Clozapine	Clozaril
	Olanzapine	Zyprexa
Dibenzothiazepines	Quetiapine	Seroquel
Benzisoxazoles	Risperidone	Risperdal
Benzisothiazoyl	Ziprasidone	Geodone
Unclassified	Paliperidone	Invega
Atypical Neuroleptics		
Aripiprazole	During clinical trials, one case of an 18-mo-old child ingesting 15 mg of aripiprazole and 2 mg of Ativan was reported to be "uneventful."	
Clozapine	Case reports of as little as 50 mg in children, ranging in age from 21 mo to 4 y, have resulted in CNS depression, ataxia, and tachycardia. Death was reported in a 15-y-old following an intentional overdose of an unknown amount of clozapine.	
Olanzapine	Case reports of 10–100 mg ingestions in children have resulted in agitation, CNS depression, EPS, and hypersalivation. A 15-y-old required intubation following ingestion of 115 mg of olanzapine along with carbamazepine. In one report, onset of symptoms did not occur until 10 h postingestion.	
Quetiapine	A single case report of a 1300 mg ingestion in an 11-y-old resulted only in mental status changes.	
Risperidone	Several published series and case reports of pediatric risperidone ingestions, in doses ranging from 1 to 110 mg, revealed somnolence, agitation, hypotension, tachycardia, and EPS to be the most common symptoms of overdose.	
Ziprasidone	During premarketing trials, there were 10 accidental or intentional poisonings, with the highest dose reported as 3240 mg. That patient only experienced mild sedation, slurred speech, and transient hypertension.	

but QT interval prolongation can sometimes be noted on electrocardiogram.

- Mydriasis may occur; however, miosis is common due to alpha-adrenergic blockade.
- Due to the anticholinergic properties of the neuroleptics, the patient may also exhibit decreased GI motility, urinary retention, hyperthermia, and dry or flushed skin.
- Therapeutic phenothiazine use has been associated with sleep apnea and sudden death in infants.

LABORATORY TESTS

- Although serum phenothiazine levels can be obtained to confirm ingestion, levels correlate poorly with clinical effects, making their utility negligible.
- Baseline laboratory tests include complete blood count, electrolytes, renal function, and glucose. Urine should be collected for myoglobin, particularly if the patient is hyperthermic.
- Due to potential neuroleptic-induced cardiotoxicity, an electrocardiogram is indicated.

MANAGEMENT

- Initial management of an acute neuroleptic overdose includes stabilizing the airway and circulation.
- If the patient remains hypotensive despite adequate amounts of IV fluid, a vasopressor with alpha-agonist activity, such as norepinephrine, may be considered.
- Because of the potential cardiotoxicity of phenothiazines, patients require close cardiac monitoring. Alpha-1 receptor blockade, along with direct myocardial depression, may cause significant hypotension following overdose.
- Sinus tachycardia is the most common dysrhythmia associated with neuroleptic toxicity; however, supraventricular and ventricular tachydysrhythmias may occur. Piperidine phenothiazines and the butyrophenones, possess quinidine-like effects on the myocardium.
- Since phenothiazine toxicity classically demonstrates central nervous system depression and pupillary miosis, adequate doses of naloxone can be administered to treat potential coexistent opioid toxicity.
- Gastric lavage may be considered if the patient presents within 1 hour of ingestion. Activated charcoal may be administered following a recent ingestion.
- No specific antidote exists for acute neuroleptic poisoning and hemodialysis is not efficacious. Most children presenting after acute neuroleptic toxicity do well with supportive care alone.

NEUROLEPTIC MALIGNANT SYNDROME

CLINICAL PRESENTATION

- Less than 1% of patients exhibit the life-threatening extrapyramidal dysfunction known as the neuroleptic malignant syndrome (NMS), characterized by skeletal muscle rigidity, coma, and severe hyperthermia following the use of phenothiazines or haloperidol.
- This syndrome can occur following acute overdose, chronic therapy, or idiosyncratically following a single dose of a neuroleptic.

- Patients with NMS will present with a constellation of clinical manifestations including features from each of the following symptom groups:
 - Temperature dysregulation: fever, which may be life-threatening.
 - Altered mental status: agitation, confusion and delirium, which may be insidious in onset, and ultimately to lethargy, obtundation, and coma.
 - Autonomic instability: fluctuating vital signs, including tachycardia, and hypotension or hypertension.
 - Musculoskeletal: generalized “lead pipe” rigidity, myoclonus, and tremors
 - Other: diaphoresis, dehydration, tachypnea, respiratory failure, rhabdomyolysis, and renal failure.

MANAGEMENT

- The neuroleptic syndrome results in a high mortality rate and is treated aggressively with rapid cooling and administration of dantrolene, which acts peripherally by treating skeletal muscle rigidity.
- Oral bromocriptine (a direct dopamine agonist) has been successfully used alone and in conjunction with dantrolene to successfully treat adult patients with NMS.

DISPOSITION

- Any symptomatic child presenting with acute neuroleptic poisoning is admitted and observed for CNS and respiratory depression as well as for cardiotoxicity or thermoregulatory problems.
- Patients with minor, asymptomatic ingestions can be observed for up to 6 hours. If the patient is discharged, caretakers should be advised to watch for the signs of delayed dystonic reactions.
- If the child has been treated successfully for acute dystonia with either diphenhydramine or benztropine, a 2- to 3-day course of oral diphenhydramine is indicated, since many of the neuroleptics have a long duration of action.

ATYPICAL ANTIPSYCHOTICS

- The existing literature indicates these medications are relatively safe when taken in overdose, particularly when coingestants are not involved.
- They generally have a safer therapeutic and overdose profile than first-generation antipsychotic medications, but many adverse and toxic effects still need to be considered in therapeutic monitoring and overdose management (Table 118-2).

- Toxicological exposures and fatalities associated with atypical antipsychotics continue to increase in the United States and the toxicological potential of these agents in children may be underestimated.

CLOZAPINE

- Clozapine can cause salivation, CNS depression, agitation, seizures, and rarely cardiac disturbances. It is known for its potential to precipitate clinically significant agranulocytosis during chronic therapy and following acute overdose.

OLANAZAPINE

- In an overdose setting, olanzapine can cause CNS depression, which may be prolonged require airway protection, miosis, QT interval prolongation and ketosis.

QUETIAPINE

- Quetiapine toxicity results in agitation, depressed sensorium, hypotension, tachycardia, and QT interval prolongation as been reported in large overdoses.

RESPERIDONE

- CNS depression described as transient lethargy, hypotension, and tachycardia have been reported in adolescent overdoses.

ZIPRASIDONE

- Overdose of ziprasidone may cause sedation, and QT interval prolongation. Serious neurological or cardiovascular complications are uncommon, but morbidity and mortality has been reported with mixed ingestions.

ARIPIRAZOLE

- Overdose may cause sedation in children for several hours, drooling, flaccid facial muscles, mild hypotension, ataxia, tremor, and vomiting. Toxicity is generally not associated with QT interval prolongation, dysrhythmias, or seizures.

PALIPERIDONE

- Increases prolactin levels and is available in extended-release forms. Side effects include restlessness, movement disorders, tachycardia, and insomnia.

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QUESTIONS

1. The NMS is characterized by which of the following findings?
 - A. Muscular flaccidity.
 - B. Central nervous system hyperexcitability.
 - C. Dysarthria.
 - D. Severe hyperthermia.
 - E. History of neuroleptic use in the presence of high environmental/ambient temperatures.
2. Following the administration of prochlorperazine to a 5-year-old child for excessive vomiting, the patient develops torticollis, tongue protrusion, restlessness, and dysarthria. What is the best option for treatment for this child?
 - A. Dextrose
 - B. Diphenhydramine

- C. Flumazenil
D. Naloxone
E. Halperidol
3. A 15-year-old has recently been prescribed a newer atypical antipsychotic agent which generally has a safer therapeutic and overdose profile than first-generation antipsychotic medications. Adverse and toxic effects still need to be considered in therapeutic monitoring and overdose management. Which of the following toxic effects is characteristic of these newer antipsychotic agents?
A. Seizure activity
B. Profound hyperthermia
C. QT interval prolongation on ECG
D. Muscular rigidity
E. Hypotension
4. In the setting of an acute overdose, children are more susceptible than adults to which type of neuroleptic effects?
A. Orthostatic changes
B. Cardiac dysrhythmias
C. Pupillary changes
D. GI effects
E. Sedative effects

ANSWERS

1. D. NMS is characterized by skeletal muscle rigidity, coma, and severe hyperthermia. It is unrelated to ambient temperature. Dysarthria is characteristic of acute dystonic reaction that may develop in response to use of a neuroleptic but is not related to the development of NMS.
2. B. If a child exhibits signs of acute muscular dystonia, intravenous diphenhydramine should be rapidly administered. Improvement usually occurs within 15 minutes. Alternatively, the patient can be given benztropine intramuscularly. Halperidol will exacerbate acute dystonic reactions. Flumazenil the antidote for benzodiazepine overdose. Naloxone is used for rapid opioid toxicity reversal.
3. C. Overdoses of the atypical or newer antipsychotics such as olanzapine, quetiapine, and ziprasidone have been shown to cause QT interval prolongation on ECG. Rarely do they cause seizure activity unless taken with other more toxic coingestants. Hyperthermia, hypotension, and muscular rigidity are more characteristic of the classic, older neuroleptic agents such as phenothiazines.
4. E. Children are more susceptible to the sedative effects than adults. All the listed effects can occur.

119 ISONIAZID TOXICITY

Jenny J. Lu

Theodore Toerne

HIGH-YIELD FACTS

- INH has been first-line treatment against active tuberculosis (TB) and prophylactic therapy for positive tuberculin skin test reactions.
- Estimated 9.2 million new cases of TB in 2006, an increase from 9.1 million cases in 2005 (World Health Organization).
- 354 INH exposures in US in 2005, with 76 in children younger than 6 years and 127 cases in children between the ages of 6 and 19 years (2005 Annual Report of the Poison Control Centers Toxic Exposure Surveillance System).

PHARMACOLOGY

- Isoniazid or isonicotinic acid hydrazide.
 - Structurally similar to nicotinic acid (niacin), nicotinamideadenosine dinucleotide (NAD), and pyridoxine (vitamin B6).
 - Disrupts mycolic acid synthesis, which is essential to the mycobacterial cell wall.
- Highly water-soluble, less than 10% protein binding, with an apparent volume of distribution of 0.6 L/kg.
- Serum concentrations peak in about 2 hours.
- Metabolic degradation of INH is complex and occurs primarily by hepatic acetylation.
 - Slow acetylators
 - Autosomal recessive for the acetylation gene.
 - Potential to have higher peak plasma concentrations, and thus higher risk for toxic side effects.
 - 50–60% of Caucasians and Blacks are slow acetylators, while up to 90% of Asians and Inuits are rapid inactivators.
- Several interactions with drugs, including theophylline, phenytoin, warfarin, valproate, and carbamazepine.

PATHOPHYSIOLOGY

- Two main mechanisms of INH toxicity deplete gamma-aminobutyric acid (GABA), the primary inhibitory neurotransmitter in the CNS.
 - Inhibition of pyridoxine phosphokinase, which converts pyridoxine to its active form, pyridoxal-5'-phosphate, required for conversion of glutamic acid to GABA.

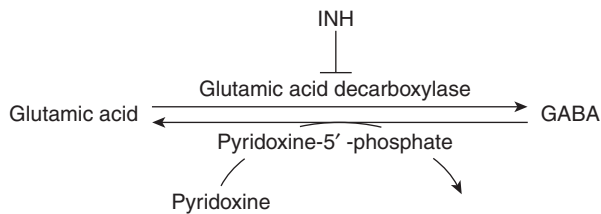


FIG 119-1 Mechanisms of INH toxicity.

- Inhibition of glutamic acid decarboxylase also required for the synthesis of GABA.
- Depletion of GABA leading to increased CNS excitability is believed to be the etiology of INH-induced seizures (Fig. 119–1).
- Metabolic acidosis associated with INH toxicity appears to be caused by increased serum lactate levels, secondary to intense seizure-induced muscle activity and less lactate clearance.
- Toxic dose of INH is highly variable.
 - Normal pediatric therapeutic dose is 10 to 20 mg/kg/d, up to 300 mg/d.
 - Acute ingestion of as little as 1.5 g of INH can lead to neurotoxicity, 6 to 10 g may be fatal, and more than 10 g is usually fatal without medical intervention.
 - Doses larger than 30 mg/kg often produce seizures.
 - Ingestion of more than 80 to 150 mg/kg can lead rapidly to death.

DIAGNOSIS

CLINICAL PRESENTATION OF ACUTE TOXICITY

- Because of rapid gastrointestinal absorption, symptoms of INH toxicity typically occur within 30 minutes to 1 hour postingestion.
 - Initial symptoms and signs: nausea, vomiting, fever, dizziness, ataxia, altered sensorium, and slurred speech.
- Triad of toxicity: refractory seizures, metabolic acidosis, and coma.
 - Hyperthermia, hypotension, hyperglycemia, ketonemia, and ketonuria have also been observed in overdose.
- Diagnosis of a toxic INH ingestion in the convulsing pediatric patient is frequently missed and the antidotal treatment delayed.
 - A high index of suspicion for INH toxicity is necessary.
- INH levels may help confirm diagnosis, but usually unavailable in the acute management.

TREATMENT

STABILIZATION

- Airway control, stabilization of breathing and circulation, and seizure management are top priorities.

DECONTAMINATION

- Orogastric lavage.
 - Contraindicated because of the potential risk for seizures and consequent complications with an unprotected airway.
- Activated charcoal.
 - Could be considered if the patient presents *within minutes* after an ingestion, although the risk of seizures and aspiration remains.
 - Endotracheal intubation should never be performed for the sole purpose of administering activated charcoal.
- No role for whole bowel irrigation

ANTIDOTAL THERAPY

- Correction of GABA deficiency through the administration of pyridoxine (B_6) is the cornerstone of treatment in INH toxicity.
- Commercially available in 1g/10 mL vials, pyridoxine is mixed in a 5% or 10% solution with $D_5 W$.
- Pyridoxine dose.
 - If the INH dose ingested is known: gram-for-gram basis, administered over 5 to 10 minutes.
 - If the dose ingested is unknown: 70 mg/kg up to a total of 5 g, and repeated at 5 to 10 minute intervals until the seizures are controlled.
 - Crushed pyridoxine tablets may be given orally or as a slurry through a nasogastric tube.
- In general, for toxicant-induced seizures, benzodiazepines such as diazepam and lorazepam are the first line of treatment.
 - In INH-induced seizures, because of depletion of GABA, pyridoxine may be the only effective therapy.
 - Short-acting barbiturates could be used if the seizures are unresponsive to benzodiazepines or pyridoxine.

ENHANCED ELIMINATION

- INH is dialyzable, but hemodialysis is usually unnecessary if adequate doses of pyridoxine and benzodiazepines have been given.

DISPOSITION

- All patients suspected of ingesting INH require close observation in a setting with capabilities for rapid airway and seizure management.
- Asymptomatic patients who remain completely symptom-free after 8 hours following an alleged isolated ingestion with INH may be medically cleared in the ED.
- Symptomatic patients require admission to an intensive care unit.

CHRONIC TOXICITY

- Uncommon in the normal pediatric population and is usually restricted to children receiving active or prophylactic treatment.
- Nausea, vomiting, fever, abdominal pain, or pruritus may herald hepatic insult that, if not treated, can progress to fulminant hepatic failure.
- Chronic INH use associated with optic neuritis, hepatitis, peripheral neuropathy, a pellagralike syndrome of dermatitis, diarrhea, dementia, and a variety of psychological reactions

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QUESTIONS

1. A convulsing 16-year-old male is brought to the emergency department (ED) by ambulance. A history of recent TB treatment is obtained from the family but his medications and doses are unknown. The initial recommended dose of pyridoxine is which of the following?
 - A. 1 g
 - B. 3 g
 - C. 5 g
 - D. 7 g
 - E. 9 g
2. The primary mechanism of toxicity of INH involves which of the following?
 - A. Histamine release.
 - B. Depletion of GABA.
 - C. Inhibition of cytochrome oxidase.
 - D. Inhibition of aldehyde dehydrogenase.
3. A 3-year-old female presents in status epilepticus. She was found with her mother's empty isoniazide pill bottle, which was recently filled. Which of the following medications is the treatment of choice in this patient?
 - A. Lorazepam
 - B. Pentobarbital
 - C. Pyridoxine
 - D. Phenytoin
 - E. Propofol

ANSWERS

1. C. The recommended dose of pyridoxine to be given in an overdose when the ingested dose of INH is known is on a “gram administered per gram ingested” basis. When unknown, 5 g of pyridoxine should be given and repeated in 5 to 10 minutes intervals until termination of seizures. INH may be given intravenously or as a slurry through a nasogastric tube. The other answer choices are technically not incorrect. As hospital supplies of pyridoxine are often limited, the smaller doses should still be given without hesitation if that is all that is available.
2. B. The overall effect of INH is inhibition of the enzymes pyridoxine phosphokinase and glutamic

acid decarboxylase, which have the overall effect of decreasing GABA, the primary inhibitory neurotransmitter. INH toxicity does not occur through the other mechanisms listed.

3. C. Pyridoxine (B6) is the corner stone of treatment for isoniazid toxicity. Seizures in this setting are typically refractory to anticonvulsants.

120 CARBON MONOXIDE POISONING

Sean M. Bryant

- Carbon monoxide (CO) is a colorless, odorless, and tasteless, nonirritating gas. Poisoning may occur secondary to fires, a faulty furnace, motor (automobile, boat, chainsaw, motorcycle, etc) exhaust and methylene chloride metabolism in the liver. Mortality has historically been high, but delayed cognitive effects are the most detrimental results in survivors.

PATHOPHYSIOLOGY

- CO creates a functional anemia.
 - CO binds to hemoglobin 240 times the affinity of oxygen.
 - CO binds to myoglobin and cytochrome C oxidase.
- Ischemic-reperfusion injury to the brain occurs.
 - Nitric oxide interaction occurs resulting in a cascade of events.
 - Endothelial injury and platelet activation occurs.
 - Oxygen free radicals are produced.
 - Lipid peroxidation and apoptosis results.
- Children may be more susceptible to CO poisoning than adults.
 - Increased metabolic rate.
 - Higher minute ventilation.
 - Fetal hemoglobin binds CO much more avidly than adult hemoglobin.

CLINICAL PRESENTATION

- Nonspecific signs and symptoms
 - Headache
 - Dizziness
 - Nausea/Vomiting
 - Fatigue
 - Irritability
 - Tachypnea
 - Tachycardia

- Confusion
- Seizures
- Syncope
- Coma
- Death

LABORATORY STUDIES

- Carboxyhemoglobin (COHb) concentration (cooximeter).
 - Either venous or arterial.
 - Quantifies proportion of CO bound to hemoglobin.
 - Does not measure tissue levels.
 - Normal levels may be less than 5%.
 - Low levels do not rule out poisoning.
 - Consider “time” from exposure and use of oxygen prior to measure.
 - Delayed measurement may be low even though patient is very ill.
- Other diagnostic tests.
 - Blood gas analysis and electrolytes.
 - Acid-base status may yield an anion gap metabolic acidosis.
 - pO₂ will remain normal (as will oxygen saturation).
 - Myoglobin (urine) and creatinine phosphokinase (serum).
 - Rhabdomyolysis can occur after prolonged down time
 - Computed tomography or magnetic resonance imaging of brain
 - Low density changes in globus pallidus
 - Chest x-ray and EKG nonspecific but may detect pulmonary or cardiac effects (especially in fire victims).

TREATMENT

- Supportive care
 - Airway, breathing, and circulation.
 - Address seizures with benzodiazepines.
 - Treat fire victims and patients with prolonged down time as appropriate.
- Oxygen
 - High flow, nonrebreather mask (if not on ventilator).
 - Half-life of CO.
 - 4–6 hours on room air.
 - 90 minutes with 100% oxygen.
 - Consider continuing oxygen administration until COHb is less than 5% and patient is clinically well and stable.
 - Pregnant patients require longer oxygen therapy.
 - 5 × normal secondary to prolonged CO elimination.
- Hyperbaric oxygen (HBO).
 - Half-life of CO is 23 minutes on average.

- Not instituted solely to decrease COHb concentration.
- Theoretical goal is to prevent delayed neurologic sequelae.
- Consider for:
 - any report of central nervous system abnormalities (syncope, altered consciousness, seizures, abnormal neurologic examination);
 - pregnancy and any symptoms;
 - Some centers use elevated levels (>25) as criteria for HBO.
- HBO protocol
 - Varies among centers.
 - May be one session and discharge.
 - Child may be admitted to have 3 sessions within 24 hours.
- Risks
 - Increased pressure in sinuses and middle ear.
 - Seizures and pneumothorax (rare).

DISPOSITION

- Discharge
 - Asymptomatic
 - COHb < 5%
 - After insuring proper education about CO poisoning.
 - With instructions to have gas company or fire department interrogate living space.
- Admit
 - If abnormal neurologic examination (or history of).
 - Any patient with end organ injury or acidosis.
 - All who remain symptomatic after HBO.
 - Pregnant patients requiring fetal monitoring.
 - Any patient returning to an “unsafe” living environment.
- Follow-up
 - Neurologic reassessment.
 - Delayed neurologic sequelae may occur up to 2 months from exposure.
 - Persistent headaches
 - Memory lapses
 - Irritability
 - Personality changes
 - Parkinsonism
- Consultation
 - Regional poison center or medical toxicologist.
 - Disposition inquiries, indications for HBO, and closest HBO centers.

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Weaver LK, Hopkins RO, Chan KJ, et al: Hyperbaric oxygen for acute carbon monoxide poisoning. *N Engl J Med* 347:1057–1067, 2002.

QUESTIONS

1. A 5-year-old male presents to the ED complaining of headache, nausea, vomiting, fatigue, and dizziness. The nurse informs you that multiple family members that live in the same house are also being seen in the ED complaining of having “flu-like” symptoms. Which of the following would be the best course of action?
 - A. Discharge them home with instructions for lots of fluids and rest.
 - B. Admit to the hospital.
 - C. Ask the caretakers if they own a CO detector.
 - D. Set up for emergent hyperbaric oxygen therapy.
 - E. Check for the CT scan of the head of each family member.
2. A mother and 2-year-old child present to the ED with headaches and vomiting after turning on a furnace in their home as the weather became colder. The child might be more affected with this exposure than the mother because of which of the following?
 - A. The child sleeps more than an adult.
 - B. The child lives closer to the ground than the adult.
 - C. The child’s liver is not as effective in detoxifying CO as the adult.
 - D. The child’s minute ventilation is greater than that of the mother.
 - E. The child’s brain is not fully developed.
3. A child is brought to the ED with dizziness, headache, nausea, and vomiting. You obtain a history of excessive playing in the basement where the father has been cutting wood with a chainsaw. A COHb concentration is 8% for the child. Which of the following is true?
 - A. CO poisoning is ruled out because the parents are smokers.
 - B. The child will be fine because the level is less than 25%.
 - C. A COHb concentration less than 10% is always normal.

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- D. The child requires emergent HBO.
E. Further history should be obtained.
4. A 2-year-old patient presents after having been brought in from a house fire. Her pulse oximeter reads 99%. The pO_2 is also reportedly normal. Which of the following is true?
A. The patient could be CO poisoned.
B. CO poisoning is ruled out because the pulse oximeter is normal.
C. CO poisoning is ruled out because the pO_2 is normal.
D. CO poisoning does not occur from house fires.
E. A CT scan of the head should be ordered to look for a CVA.
5. A 16-year-old female arrives after being found in the garage with the car running. She has tried to commit suicide several times in the past. Her COHb concentration is 14%. She complains of severe headache, dizziness, and nausea but had no loss of consciousness. What is the next appropriate course of action?
A. Send her for HBO.
B. Check a CT scan of the head.
C. Check a pregnancy test.
D. Do a lumbar puncture.
E. Administer antiemetics.
6. A 4-year-old boy is brought to the ED for “near drowning.” He was swimming behind a houseboat in the local lake. He is now awake and alert but earlier was unconscious. Which of the following is true?
A. Since this is summertime, CO poisoning is ruled out.
B. Checking a COHb level would help “confirm” exposure.
C. HBO would not be warranted because he is currently awake.
D. Discharging the patient without further workup would be appropriate.
E. Houseboats are not a source of CO poisoning.
7. A 6-year-old female arrives to the ED with a family concerned about CO poisoning. Their CO alarm has been going off sporadically. Her COHb concentration is 12%. There is no history of neurologic changes but she has had a headache for the last 2 days. Which of the following is the best course of action?
A. Discharge the patient home.
B. Instruct the family to call the fire department and/or gas company.
C. Administer oxygen via nasal cannula.
D. Ignore the rest of the family members.
E. Send the patient for HBO.
8. A 15-year-old male is brought to the ED with obtundation requiring intubation. A COHb concentration is 35%. He is markedly hypotensive and an EKG reflects ischemic changes. What is the next best course of action?
A. Minimize oxygen delivery to prevent diminishing the respiratory drive.
B. Stabilize the patient’s cardiovascular status.
C. Rapidly transfer for HBO.
D. Check serial COHb levels to insure they are decreasing.
E. Disregard the EKG findings they are rate related changes.
9. A 14-year-old male was working on his motorcycle in a closed barn. He was found down and brought to the ED. The motorcycle, according to the paramedics, had no gas in it. Which of the following is true regarding CO poisoning?
A. The patient only warrants HBO if the COHb level is greater than 25%.
B. A COHb level of 5% rules out CO poisoning.
C. Regardless of the level, CO poisoning is high on the differential list.
D. The lack of gasoline in the motorcycle rules out CO poisoning.
E. If the pulse oximeter is normal, then CO poisoning is ruled out.
10. A family of 5 presents after a relative dropped by and noted that nobody would answer the doorbell. All patients were hard to arouse and one had vomited on the floor. The patients are now awake and alert with complaints of headache in the ED. What is the next best appropriate action?
A. Discharge the patients back home.
B. Call the regional poison center for location of the nearest HBO facility (preferably a multi-place chamber to fit the whole family in).
C. Administer 100% oxygen until all COHb levels are zero.
D. Give acetaminophen to treat the headaches.
E. Send the patient with vomiting for HBO.

ANSWERS

1. C. CO detectors have proven worthy to prevent poisoning. If the patient does not have one, further inquiry into possible CO poisoning may be warranted.
2. D. Minute ventilation is greater in children and may result in greater exposure than an adult. The liver has no detoxifying function for CO. CO has a similar specific gravity as oxygen, therefore, with disperse throughout a room rather than be more dense near the floor.

3. E. This is a confirmed history of CO exposure. Further history of any CNS abnormalities (syncope, seizure, and coma) would help determine if the patient would warrant hyperbaric oxygen therapy. Also, further history of how chronic the exposure has been, other family members affected, and potential utility of normobaric oxygen therapy may be warranted to alleviate symptomatology. Follow up would also be appropriate.
4. A. House fires are a common source of CO poisoning. CO poisoning regularly presents with normal oxygen saturation and pO_2 . A CT scan may show basal ganglia densities in the globus pallidus but is not indicated routinely for CO poisoning.
5. C. Even though the patient has typical signs and symptoms with a confirmed COHb concentration, she does not warrant HBO, but her fetus would. A lumbar puncture would only be helpful for working up meningitis in this case, which is unlikely. High flow oxygen would be better than antiemetics, especially before the pregnancy test comes back.
6. B. This patient would warrant HBO for CO poisoning if the CO level confirms exposure (as long as he arrives before the CO is eliminated) because he had loss of consciousness. Further workup to confirm an etiology of the near drowning is appropriate. Houseboats are reported to be sources of exposure, especially when swimming behind or underneath them.
7. B. This patient is in danger of re-exposure when going home. They should all be checked for confirmatory levels and administered high flow oxygen via a non-rebreather mask until the level is less than 5% and they are all asymptomatic. HBO is only indicated with neurologic changes, which are absent in this case.
8. B. Patients who are “unstable” should not be transferred solely for HBO unless they are going to a higher level of care (ie, a cardiologist is only available at the transferring facility). 100% Oxygen will eliminate CO with a half life of approximately 90 minutes. CO binds to myoglobin as well as the electron transport chain, which may result in myocardial infarctions.
9. C. The fact that the motorcycle is out of gas may reflect a massive exposure with unknown down time. Additionally, the COHb concentration might be regarded as low if the patient is far out from exposure. Since he was found unconscious, the patient warrants HBO if that is the culprit.
10. B. A multiplace chamber can treat multiple patients simultaneously and would be ideal. The patients should not be sent back to the home until it has been assessed for the CO source. Oxygen will alleviate symptoms of poisoning but may not get rid of all CO (ie, a worthy goal would be to drive the COHb to less than 5%).

Drugs of Abuse

121 OPIOIDS

Timothy B. Erickson

INTRODUCTION

- Opioids are naturally occurring or synthetic drugs that have activity similar to that of opium or morphine.
- Opioids are used clinically for analgesia, anesthesia, as cough suppressants, and to alleviate diarrhea.

AGE AND DEVELOPMENTAL CONSIDERATIONS

- Neonates can experience lethargy at birth if there was recent maternal opioid use, or if large doses of an opioid agent were iatrogenically administered to the mother during labor. The neonate is prone withdrawal symptoms during the newborn period if the mother exhibited chronic dependency during her later prenatal stages.
- Toddlers are susceptible to opioid poisoning if their environment permits exposure, most often from unintentional ingestions.
- Powdered heroin, methadone and codeine tablets, long acting morphine derivatives, and fentanyl patches may be readily available to the younger child.
- Methadone is one of the more potentially toxic opioids to toddlers and the incidence of accidental ingestion is on the rise resulting serious poisonings from small doses.
- Adolescents tend to experiment with different routes of exposure, such as inhalational (smoking), intranasal (snorting), ingestion, or intravenous administration.
- The opioids may have synergistic effects with other drug combinations such as ethanol, benzodiazepines, and GHB (gamma hydroxybutyrate), or opposing effects when mixed with sympathomimetic agents like cocaine and amphetamines.
- Chronically addicted teenagers are also prone to acute withdrawal states, particularly when treated with the antidote naloxone.

PHARMACOLOGY/TOXICOKINETICS

- Opioid activity resembles the body's three endogenous opioid peptides: enkephalins, endorphins, and dynorphins.
- The three main receptors are mu (μ), kappa (κ), and delta (δ).
 - Most analgesia results from supraspinal μ_1 receptors. It is the only opioid receptor in the brain and is primarily responsible for the opioid-induced euphoria and sedation.
 - μ_2 is responsible for spinal analgesia, respiratory depression, miosis, physical dependence, and decreased gut motility.
 - κ_1 produces spinal analgesia, and miosis (although less pronounced than μ_2).
 - κ_2 results in dysphoria and disorientation.
 - The delta receptor produces spinal analgesia.
- Durations of effects range from fentanyl, with a duration of approximating 1 hour, to methadone, which lasts up to 24–48 hours.
- Many of the other oral agents, such as codeine, sustained release morphine, (eg, MS cotin, oxycontin), oxycodone, and Lomotil (an antidiarrheal agent containing diphenoxylate and atropine), will also demonstrate a delayed effect of up to 4 to 12 hours.
- The development of tolerance is characterized by a shortened duration and a decreased intensity of analgesia, euphoria, and sedation, which creates the need to consume progressively larger doses to attain the desired effect.
- Physical dependence refers to physiologic and psychological changes that necessitate the continued presence of a drug in order to prevent a withdrawal syndrome. Although unpleasant, withdrawal from narcotics is rarely life threatening (Table 121-1).

PATHOPHYSIOLOGY

- Opioid-induced respiratory depression is primarily mediated through the μ_2 receptors. When opioid agonists bind to these receptors, the ventilatory drive is reduced by diminishing the sensitivity of the medullary chemoreceptors to hypercapnea.
- Acute lung injury, classically described with severe opioid overdose, results from hypoxia secondary to ventilatory compromise, which causes precapillary pulmonary hypertension. Increased pulmonary capillary permeability causes an extensive fluid leak. This variation of acute lung injury (described in other sources as “noncardiogenic pulmonary edema”) may also be due to direct hypersensitivity or alveolar membrane toxicity.

TABLE 121-1 Common Opioids with Generic and Trade Names

GENERIC	TRADE NAMES
Morphine	MS-Contin, Oramorph SR, MSIR, Roxanol, Kadian, RMS
Hydromorphone	Dilaudid
Codeine	Tylenol #3, #4, Tylenol with codeine
Oxycodone	OxyContin, Oxyl, Percodan (ASA) Percocet (APAP)
Hydrocodone	Anexsia, Hycodan, Hycomine, Lorcet, Lortab, Tussinex, Tylox, Vicodin, Vicoprofen
Meperidine	Demeral, Mepergan, MPPP, MPTP
Methadone	Dolophinel, ORLMM
Buprenorphine	Buprenex
Propoxyphene	Darvon
Pentazocine	Talwin, Talwin Nx (with naloxone)
Butorphanol	Stadol, Torbugesic, Torbutol
Fentanyl	Sublimaze (80 × more potent than morphine)
Fentanyl patch	Durgesic patch
Fentanyl citrate	Actiq (solid on a stick for oral use)
Sufentanil Sufenta	(1000 × more potent than morphine)
Carfentanil Wildnil	(10 000 × more potent than MS)

- Morphine specifically causes stimulation of parasympathetic pupilloconstrictor neurons in the oculomotor nerve. Other opioids mediate inhibitory neurotransmission, causing hyperpolarization of inhibitory neurons to the parasympathetic neurons, causing the classic “pinpoint pupil” associated with opioid use.
- Constipation is a common side effect of both therapeutic, chronic, and recreational opioid use.
- Certain opioids cause cardiotoxicity via conduction system dysfunction. Propoxyphene blocks myocardial sodium channels, with a quinidine-like effect similar to cyclic antidepressants. Methadone toxicity may prolong the QT interval inducing torsades de pointes.

CLINICAL PRESENTATION

- Opioid poisoning classically presents with an altered level of consciousness. The triad of acute toxicity consists of CNS depression, respiratory depression, and pupillary constriction or “pinpoint pupils.” CNS depression ranges from mild sedation to stupor and coma.
- Patients are typically hypotensive, hypothermic, bradycardic, and hyporeflexic.
- In large overdoses, the respiratory toxicity can also cause severe hypoxia, hypercarbia, and acute lung injury.
- While miotic or “pinpoint” pupils are a classic opioid-induced clinical finding, mydriasis, or pupillary dilation has been described with meperidine, propoxyphene and diphenoxylate-atropine (lomotil) overdoses.

- Lomotil overdose presents with a two-phase toxicity. Phase 1 manifests with anticholinergic symptoms such as dry mouth, flushing, and mydriasis; phase 2 consists of opioid effects of respiratory and central nervous system depression with associated miosis.
- Less common effects of opioid toxicity include generalized seizure activity following overdose of propoxyphene, meperidine, tramadol, fentanyl, or pentazocine. Neonates receiving continuous intravenous morphine can also suffer seizures from mere toxicity or during acute opioid withdrawal.
- Dermatologic effects include flushing and pruritus secondary to histamine release. Gastrointestinal effects include decreased gut motility and constipation.

LABORATORY AND DIAGNOSTIC TESTING

- Patients presenting with suspected opioid toxicity should have their oxygenation status continually monitored. If severe respiratory compromise and hypoxia continue despite antidote therapy and oxygen administration, an arterial blood gas analysis is obtained to rule out hypercarbia and acidosis.
- If a child presents with a depressed level of consciousness, a rapid bedside serum glucose is measured to rule out hypoglycemia. If head trauma is suspected, a CAT scan of the brain is indicated.
- With severe respiratory distress, acute lung injury or aspiration pneumonitis should be suspected, and confirmed with a chest radiograph.
- With a propoxyphene and methadone overdose, an electrocardiogram to assess the QRS complex is indicated.
- Serum toxicology screens are not helpful in the initial management of the opioid overdosed child. These assays may only detect opioid compounds for up to 6 hours.
- Urinary qualitative screens may be useful in ruling-out an opioid exposure in a child presenting with altered mental status. These are typically positive for up to 48–72 hours postingestion.
- Most urinary screens lack sensitivity, and may not detect many of the synthetic opioids including methadone, hydrocodone, propoxyphene, and fentanyl. Toxicology screens may also result in “false positive” screens following dietary ingestion of poppy seeds.
- Along with other routine baseline laboratory tests, serum acetaminophen and salicylate levels should be measured, since many opioid compounds such as hydrocodone and codeine also contain these common analgesics.

MANAGEMENT

- The primary management of opioid poisoning includes stabilization of the airway and administration of the pure opioid antagonist naloxone. If adequate doses of this antidote are given in a timely fashion, intubation can be avoided, since the onset of action for naloxone is usually within 1 minute of administration.
- If there is no response to naloxone, and oxygenation cannot be maintained with bag-valve-mask ventilations, the patient's airway is secured by endotracheal intubation.
- In addition to intravenous administration, naloxone can be given via the endotracheal tube, subcutaneously, intramuscularly, intranasally or by nebulizer, with a comparably rapid onset of action. Clinical trials have demonstrated that slower absorption via the subcutaneous route was offset by the delay in establishing intravenous access, particularly in young children.
- In the overdose setting, the dose of naloxone is 0.1 mg/kg in children from birth to 5 years of age or in children weighing less than 20 kg. In older children, or in the setting of life-threatening toxicity, a rapid 2-mg dose is recommended.
- If the patient experiences acute withdrawal symptoms, general supportive care measures are indicated, since opioid withdrawal is not a life-threatening situation.
- In the newborn setting, withdrawal seizures have been well documented in neonates born to opioid dependent mothers.
- Naloxone's duration of action is 20–30 minutes, which is shorter than most opioid agents, except fentanyl. Repeated doses may be indicated, particularly with opioids having longer durations of action, such as oxycodone, methadone, and lomotil (diphenoxylate).
- The new longer acting antagonist nalmefene may be useful in younger, nonopioid dependent children who are exposed to longer acting agents. Doses of 0.5 to 2.0 mg of nalmefene have been reported to be safe and effective, with a duration of effect up to 8 hours.
- In the setting of chronically addicted adolescents, a shorter acting antagonist like naloxone would be a more humane approach, since withdrawal symptoms would be shorter in duration.
- In patients who have ingested oral narcotics, gastric lavage may be performed if the patient presents within 1 hour after the ingestion. However, attention must be paid to the patient's mental status due to the risk of respiratory depression.
- An initial dose of activated charcoal is advised following recent oral ingestions, particularly in opioids that have delayed absorption, such as diphenoxylate-atropine (lomoltil) and sustained-release morphine products.
- Children who present as body packers or stuffers of heroin or opioid-containing drugs may require gastrointestinal decontamination with whole bowel irrigation using PEG (polyethylene glycol) solution to enhance elimination of drug packets. Contraindications to whole bowel irrigation include unstable vital signs, respiratory compromise, and lack of bowel sounds or gut motility.
- Activated charcoal should be given prior to whole bowel irrigation to adsorb any leaking drug from the packets. Charcoal is noted in the rectal effluent indicates successful whole bowel irrigation.

DISPOSITION

- Any young pediatric patient presenting with CNS and respiratory depression from opioid poisoning that is responsive to naloxone should be admitted for observation.
- Most of the opioids demonstrate longer duration of action than naloxone and repetitive dosing or continuous naloxone infusion may be required.
- Long-acting agents such as methadone and the anti-diarrheal agent lomotil are much more likely to demonstrate recurrence of central nervous system and respiratory depression. Because of the potential delays in toxicity, young children should be observed in a monitored setting for at least 24 hours.
- In patients who continue to demonstrate respiratory compromise or require repeated doses of naloxone, it would be prudent to observe these patients for a longer period of time.
- Adolescent patients presenting with CNS depression from heroin overdose who quickly respond to naloxone administration can be discharged home with reliable caretakers after 4–6 hours of observation. In these patients demonstrating chronic addiction patterns, detoxification programs and appropriate counseling referrals should be arranged.

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QUESTIONS

- Opioid poisoning is classically associated with which of the following clinical scenarios?
 - CNS excitation, tachycardia, hyperthermia, and dilated pupils.
 - Cardiotoxicity, seizures, and hypotension.
 - Hyperthermia and muscular rigidity.
 - CNS depression, depressed respiratory drive, bradycardia, and pinpoint pupils.
 - Vomiting, diarrhea, diaphoresis, bradycardia, and bronchosecretions.
- A 3-year-old child presents with respiratory depression and unresponsiveness following ingestion of her uncle's methadone at home. Which of the following treatment options is the most optimal for this patient?
 - Rapid endotracheal intubation.
 - Intravenous dextrose.
 - Intravenous physostigmine.
 - Intravenous naloxone administration and 4 hours observation in the ED.
 - Intravenous naloxone administration and admission for 24 hours of observation.
- An 18-month-old child presents comatose after being found "chewing on her grandmother's medication patch." The grandmother has a history of metastatic breast cancer. Which of the following is the most likely agent responsible for this child's clinical presentation?
 - Clonidine
 - Nitroglycerin
 - Methylsalicylate
 - Nicotine
 - Fentanyl

ANSWERS

- D. Opioids classically present with CNS depression, depressed respiratory drive bradycardia and pinpoint or miotic pupils. Sympathomimetic agents such as

cocaine and amphetamines present with CNS excitation, tachycardia, hyperthermia, and dilated pupils (mydriasis). Cardiotoxicity, seizures, and hypotension are more consistent with cyclic antidepressant overdose. Hyperthermia and muscular rigidity is a classic finding in neuroleptic malignant syndrome (NMS). Organophosphate insecticide exposure causes vomiting, diarrhea, diaphoresis, bradycardia, and bronchosecretions.

- E. This 3-year-old child needs rapid antidote administration of the opioid antagonist naloxone (nalmeferine) would be an acceptable alternative reversal agent). Because of the long half-life of methadone, repeat doses of naloxone may be required and this child should be admitted for 24 hours. Timely administration of naloxone often obviates the need for endotracheal intubation in these types of patients. Physostigmine is the antidote or reversal agent for pure anticholinergic overdose such as diphenhydramine and jimson weed.
- E. The most likely agent responsible for this child's depressed mental status is fentanyl. This opioid drug is commonly used in patch form for the treatment of chronic pain and for cancer patients. Clonidine patches could also lead to a depressed sensorium and respiratory drive, but is classically used for the management of hypertension and opioid withdrawal. Nicotine toxicity would result in CNS excitation, tachycardia, and potential seizure activity. Nitroglycerin patches may lead to hypotension. Significant salicylate toxicity has been reported in children and adults from dermal absorption after using these patches excessively for musculoskeletal pain.

122 COCAINE TOXICITY

Michael R. Christian

Steven E. Aks

INTRODUCTION

- Cocaine abuse continues to be a pervasive problem. Pediatric patients usually suffer toxicity when exposed to cocaine used by others.

PHARMACOLOGY

- Cocaine is a sympathomimetic that blocks fast sodium channels and inhibits reuptake of norepinephrine, epinephrine, and dopamine.

- Cocaine is rapidly absorbed from mucous membranes, lung tissue, and gastrointestinal tract.

PATHOPHYSIOLOGY & CLINICAL MANIFESTATIONS

- Central nervous system effects: agitation, hallucinations, seizures, and ischemic or hemorrhagic strokes. Pediatric patients may present paradoxically lethargic.
- Cardiovascular effects: hypertension, hyperthermia resulting in rhabdomyolysis, sinus tachycardia, supraventricular tachycardias, ventricular tachycardia, and myocardial ischemia (myocardial infarction has been reported in patients as young as 17 years of age). Quinidinelike effects secondary to the blockade of fast sodium channels may also occur.
- Pulmonary effects: exacerbation of asthma, pneumothorax, pneumomediastinum, pulmonary infarction, and respiratory failure.
- Gastrointestinal effects: abdominal pain, hemorrhagic diarrhea, and shock possible if cocaine ingested.

DIAGNOSIS

- Cocaine toxicity is primarily a clinical diagnosis based on history and physical exam.
- Urine toxicology screen for benzoylecgonine, a metabolite of cocaine, may be positive for up to 72 hours after exposure.
- Serum levels of cocaine correlate poorly with severity of symptoms.
- Cardiac monitoring is essential. An EKG should be obtained to evaluate for dysrhythmias.
- Useful labs include a complete blood count, electrolytes, glucose, blood urea nitrogen, creatinine, and creatine phosphokinase in patients with suspected rhabdomyolysis.
- In patients with chest pain or hypoxia, a chest radiograph should be performed to evaluate for pneumothorax, pneumomediastinum, or pulmonary infarction.
- A noncontrast CT head should be obtained in patients with a headache or focal neurologic deficits.

MANAGEMENT

- Care is largely supportive.
- Hyperthermic patients should be actively cooled.
- Benzodiazepines should be administered liberally for moderate to severe agitation, hypertension, and seizures.

- Nitroprusside or phentolamine can be considered in patients with severe hypertension.
- Quinidinelike effects may manifest as QRS prolongation and a terminal R-wave in aVR, which can herald severe dysrhythmias. Similar to the quinidinelike effects seen with TCAs, QRS prolongation should be treated with boluses of sodium bicarbonate 1 to 2 mEq/kg until QRS < 100 ms, pH is 7.5, or patient becomes hypernatremic.
- After adequate fluid resuscitation, consider urinary alkalization in patients with rhabdomyolysis.
- Activated charcoal or whole bowel irrigation can be considered in body stuffers (who haphazardly ingest packets of cocaine when confronted by law enforcement) or body packers (who ingest large amounts of well-packaged drug with high purity for transport). Symptomatic body packers may require emergent surgical intervention.

DISPOSITION

- Asymptomatic or mildly symptomatic patients may be discharged after a 4–6 hour observation period.
- Patients with severe symptomatology require monitoring in a critical care setting.
- Body packers should be monitored until all packets have been passed.

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QUESTIONS

1. The clinical effects of cocaine most closely resemble which of the following toxidromes?
 - A. Anticholinergic toxicity
 - B. Parasympathetic toxicity

- C. Sympathomimetic stimulation
 D. Cholinergic excess
 E. Narcotic syndrome
2. An 18-year-old male presents to the ED with a complaint of chest pain. He admits to cocaine use four hours prior to arrival. Which of the following statements is correct regarding his work-up?
 A. Urine toxicology screen is not helpful.
 B. Blood levels for cocaine correlate closely to hypermetabolic signs and symptoms.
 C. EKG has been shown to be unnecessary due to the low prevalence of coronary artery disease in this age group.
 D. Chest radiograph is useful to exclude pneumothorax, pneumomediastinum, or infiltrate.
 E. Noncontrast CT scan of the brain is indicated to rule out the possibility of CVA.
3. A 16-year-old male is brought to the ED by paramedics after being observed to be acting in a threatening and aggressive manner in a public place. He is uncooperative and trashing about on the ambulance cart. His blood pressure is 145/95 with a pulse rate of 130 and a respiratory rate of 30. The most appropriate initial pharmacologic intervention in the patient is which of the following?
 A. Labetalol
 B. Metoprolol
 C. Haloperidol
 D. Midazolam
 E. Chlorpromazine
4. After the patient above is medicated, the temperature is noted to be 106°F. His toxicology screen is positive for cocaine only. The microscopic urinalysis is completely normal but the urine dip reveals moderate blood. What further diagnostic studies are indicated in this patient?
 A. Noncontrast CT scan of the abdomen.
 B. Serum creatine phosphokinase (CPK) and urine myoglobin.
 C. Noncontrast CT scan of the brain.
 D. CK-MB and Troponin-I.
 E. Arterial blood gases and blood lactate.
5. A 12-year-old female is brought by EMS from the international airport. The patient is obtunded with a temperature of 104.5°F, blood pressure of 145/95, heart rate of 138, respiratory rate of 30, and an oxygen saturation of 98% on a nonrebreather mask. A chest radiograph is normal but an abdominal radiograph reveals many radiopaque foreign bodies distributed throughout the small and large bowel. In addition to aggressive supportive care including airway management, fluid resuscitation, and active cooling, which of the following actions is the most critical in the immediate management of this patient?
 A. Labetalol drip titrated to control the patient's blood pressure and heart rate.
 B. Administration of activated charcoal.
 C. Initiation of whole bowel irrigation.
 D. Urine toxicology screen.
 E. Emergent surgical consultation.
6. In the previous patient, a stat EKG is performed which reveals a heart rate of 136 bpm, a PR interval of 123 ms, a QRS of 148 ms, and a QTc of 495 ms. Which therapy is indicated?
 A. Sodium bicarbonate bolus
 B. Labetalol drip titrated to control the patient's blood pressure and heart rate
 C. Lorazepam
 D. Phentolamine
 E. Procainamide
7. The patient from the previous question is accompanied by her mother and her 14-year-old brother who are also evaluated. The 14-year-old male is asymptomatic with normal physical exam and EKG but an abdominal radiograph reveals many radiopaque foreign bodies distributed throughout the small and large bowel. Urine toxicology is negative for cocaine. How should this patient be managed?
 A. Patient may be safely discharged with a reliable caretaker.
 B. Patient should be observed for 4–6 hours in the ED and discharged with a reliable caretaker if asymptomatic.
 C. Patient should undergo emergent endoscopy for removal of packets.
 D. Whole bowel irrigation should be administered in a monitored setting until all packets have been passed.
 E. Patient should undergo surgical laparotomy.
8. A 9-year-old male is brought to the ED by his parents for his third asthma exacerbation in a week. According to the patient's pharmacy, the parents have been getting his prescriptions for albuterol, salmeterol, and fluticasone refilled as prescribed over the last six months. The patient's wheezing is improved after administration of bronchodilators and oral steroids. A detailed social history reveals that the patient's father smokes crack-cocaine daily. The patient's urine toxicology screen is positive for cocaine. Which of the following is most likely to decrease the frequency of asthma exacerbations in this patient?
 A. Addition of inhaled montelukast.
 B. Prolonged oral steroid taper.
 C. Removing exposure to father's cocaine.

- D. Improve patient adherence to medication regimen.
- E. Substituting albuterol for levalbuterol.

ANSWERS

1. C. Cocaine toxicity presents with signs and symptoms of sympathomimetic stimulation. This toxidrome may be difficult to distinguish from that caused by anticholinergic toxicity. However, unlike sympathomimetic toxicity, anticholinergics cause urinary retention and decreased bowel sounds.
2. D. Chest radiographs are indicated to rule out pulmonary pathology. Urine toxicology screen is useful to confirm the ingestion and rule out coingestants. Blood levels of cocaine and cocaine metabolites correlate poorly with signs and symptoms. An EKG is indicated, regardless of age, since myocardial ischemia and infarction have been described in otherwise healthy individuals as young as 17 years of age with normal coronary arteries. A CT scan of the brain is not indicated on a routine basis but is indicated when there is severe headache or neurologic deficit.
3. D. Moderate to severe agitation responds to benzodiazepines, which are also the drug of choice for seizures. Benzodiazepines are also effective treatment for most patients with mild to moderate hypertension. In more severe cases, labetalol, which has both alpha- and beta-blocking characteristics, has been effective, as has sodium nitropruside. Beta-stimulation can exacerbate hypertension. Haloperidol and chlorpromazine lower the seizure threshold and are relatively contraindicated.
4. B. The hyperactivity of cocaine toxicity can result in rhabdomyolysis with or without associated hyperthermia. It is suspected when a urine dipstick is positive for blood (actually myoglobin) but microscopy is negative for red blood cells. The syndrome is confirmed by the presence of CPK in the blood and myoglobin in the urine, both of which are produced by muscle breakdown. This patient would be treated with aggressive cooling and alkalinization of the urine. Myoglobin is prevented from precipitating out in the renal tubules by maintaining an alkaline urine, thus preventing the development of renal failure.
5. E. Emergent surgical consultation is critical. Given the patient's history, toxidrome, and abdominal radiograph findings, the patient is most likely an involuntary body packer. Body packers often ingest large amounts of pure drug for illegal transport. If a single package ruptures, the effects can be devastating. This patient should undergo emergent exploratory laparotomy for removal of ingested drugs. A labetalol drip will be ineffective. Cocaine is a powerful vasoconstrictor and often causes bowel necrosis and perforation in this setting; activated charcoal and whole bowel irrigation are strictly contraindicated. A urine toxicology screen will delay definitive management and may even be negative if the metabolite of cocaine, benzoylecgonine, has not yet been excreted in the urine.
6. A. The patient is exhibiting the quinidinelike effects of cocaine secondary to the blockade of fast sodium channels, which is exhibited on the EKG by QRS prolongation. Sodium bicarbonate boluses should be used to overcome this blockade of sodium channels. Lorazepam, labetalol, and phentolamine may improve the patient's hypertension and tachycardia, but have little to no effect on the quinidinelike effect and subsequent dysrhythmias. Procainamide also blocks sodium channels and would exacerbate the effect of cocaine.
7. D. This patient should undergo whole bowel irrigation in a monitored setting until all packets have been passed. The patient should not be discharged because a delayed rupture of packets may occur. Endoscopy can result in the rupture of packets and should be avoided. Asymptomatic patients do not require an invasive laparotomy.
8. C. Cocaine and its adulterants are potent bronchoconstrictors. In patients with frequent exacerbations, it is important to perform a thorough social history. Avoidance will likely improve patient's symptoms drastically. With the information obtained from the pharmacy, the patient is probably compliant with the medication regimen. A prolonged oral prednisone taper, addition of montelukast, or substitution of albuterol for levalbuterol is unlikely to improve patient's frequency of symptoms as much as the avoidance of exposure to crack-cocaine.

123 PHENCYCLIDINE & KETAMINE

Matthew Valento

INTRODUCTION

- Phencyclidine (PCP) is a dissociative anesthetic commonly abused as a recreational drug. It is available on the street in many forms and is often used concurrently with other drugs such as marijuana and cocaine. Ketamine is structurally related to PCP and

is widely employed as a clinical anesthetic, though it is also frequently diverted from legal practices for illicit street use.

PHARMACOLOGY

- PCP and ketamine are piperidine derivatives that are rapidly absorbed and distributed into the central nervous system (CNS).
- Both drugs dissociate the somatosensory cortex from higher CNS centers, leading to diminished responses to external stimuli.

CLINICAL PRESENTATION

- Neurologic effects include significant psychomotor abnormalities, including agitation, ataxia, confusion, nystagmus (horizontal, vertical, or rotatory), and psychosis.
- Agitation can progress to violence, loss of pain perception, and delusions of superhuman strength.
- Mild sympathomimetic effects may be present.
- A fluctuating level of consciousness is often observed.
- Ketamine may be associated with “emergence reactions” as the patient recovers from its anesthetic effects, characterized by confusion, bizarre behavior, and hallucinations.

DIAGNOSIS

- Diagnosis of PCP or ketamine intoxication is based primarily on clinical presentation.
- Dextromethorphan may cause a false positive result for PCP on many urine drug assays.

MANAGEMENT

- Supportive care, with a focus on controlling agitation, is the mainstay of treatment.
- Liberal use of benzodiazepines is recommended, in combination with haloperidol and physical restraints, if necessary.
- Phenothiazines (eg, chlorpromazine) should be avoided as they may lower the seizure threshold, worsen hyperthermia, and cause hypotension.

DISPOSITION

- Patients with prolonged coma, hyperthermia or significant sympathomimetic toxicity, rhabdomyolysis,

or unstable vital signs should be admitted to an intensive care unit.

- Patient with mild clinical toxicity not requiring large amounts of sedation can be observed in the emergency department (ED) for 6–8 hours until return of baseline mental status.

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QUESTIONS

1. A 15-year-old male presents extremely agitated and combative to the ED after his friends state he used PCP at a party. Which of the following treatment modalities represents appropriate management for a patient with PCP intoxication?
 - A. Gastric lavage to remove drug from the stomach.
 - B. Adjustment of the urinary pH to a more acidic state, allowing greater renal elimination of the drug.
 - C. Repeated doses of fluphenazine until agitation is controlled.
 - D. Repeated doses of lorazepam and physical restraints if needed.
 - E. Place the patient in a dark room and calmly attempt to “talk the patient down.”
2. A 16-year-old girl is brought to the ED by her parents because she is confused and acting abnormally after taking an unknown substance. In trying to determine the agent used, which of the following is a classic finding of PCP or Ketamine intoxication?
 - A. Psychomotor retardation
 - B. Hypothermia
 - C. Bidirectional nystagmus
 - D. Respiratory depression
 - E. Unreactive pupils

ANSWERS

1. D. Benzodiazepines, such as lorazepam or diazepam, are an effective pharmacologic intervention for controlling agitation in patients with PCP intoxication. Physical restraints are often necessary. Phenothiazines are contraindicated as they may precipitate seizures and cause hypotension. PCP is rapidly absorbed from the gut and attempts at decontamination (lavage, whole bowel irrigation) will likely be difficult in an agitated, combative patient. Attempts to reason with these patients or “talk them down” will be unsuccessful and may be dangerous to the caregiver. Urinary acidification, while historically suggested as a treatment modality, is not recommended as it may enhance myoglobin precipitation in renal tubules and worsen nephrotoxicity.
2. C. Horizontal, vertical, and rotary nystagmus can be present and is a classic finding. Motor activity is usually extreme and great strength may be exhibited. Hyperthermia may result. There is no respiratory depression. Pupils may be miotic or mydriatic but remain reactive to light.

124 AMPHETAMINES*James Rhee***BACKGROUND**

- Amphetamines are the group of stimulants that have a similar chemical structure.
- A number of different modifications to the underlying chemical structure have led to a variety of compounds with various stimulant and hallucinogenic effects.
- Methamphetamine and 3,4-methylenedioxymethamphetamine (MDMA) are amphetamines that have become leading drugs of abuse in the United States.

PHARMACOLOGY

- In general, amphetamines easily cross the blood–brain barrier.
- Frequent use may lead to an accumulation of drug in the body, prolonging the half-life and duration of effect.
- Amphetamines cause generalized monoamine release which results in increased activation of monoamine receptors—notably, dopamine and adrenergic receptors.

- MDMA can lead to significant activation of serotonin receptors as well.

CLINICAL MANIFESTATIONS

- Amphetamine toxicity leads to generalized sympathetic stimulation.
- Symptoms of amphetamine toxicity include tachycardia, diaphoresis, hypertension, tremor, tachypnea, mydriasis, agitation, hallucinations, euphoria, and paranoia.
- MDMA can induce bruxism—grinding of the teeth.
- Serotonin syndrome (a syndrome of neuromuscular excitability with associated autonomic instability, altered mental status, and hyperthermia due to increased serotonin activity) has been described with MDMA use.
- A patient presenting with evidence of clonus or hyperreflexia in the setting of MDMA use should alert the clinician to the possibility of serotonin syndrome.
- Hyponatremia is a recognized complication of MDMA use—due to increased release of vasopressin that results in the increased retention of free water by the body.
- When extreme, the ensuing hyponatremia can cause cerebral edema, seizures, and death.
- Severe complications associated with amphetamine toxicity include rhabdomyolysis, hyperthermia, seizures, stroke, intracerebral hemorrhage, acute coronary syndrome, and malignant cardiac dysrhythmias.

DIAGNOSTIC TESTING

- Routine laboratory and other diagnostic studies are generally not helpful in diagnosing amphetamine toxicity.
- A metabolic profile, complete blood count, creatine phosphokinase, and an electrocardiogram may be performed to assess for complications of amphetamine toxicity.
- In the setting of altered mental status, a computed tomography of the head and follow-up lumbar puncture with cerebrospinal fluid analysis should be performed to evaluate for intracerebral hemorrhage and to exclude other etiologies of the patient’s condition.
- A urine drug screen may help confirm amphetamine exposure.
- A number of agents can cause a positive amphetamine urine drug screen including pseudoephedrine, phenylpropanolamine, selegiline, and ephedra.
- Some of the modified amphetamines can result in a negative urine drug screen for amphetamines.

- It is best to consult with the drug screen manufacturer to understand the limitations of the specific amphetamine immunoassay used at a particular institution.

MANAGEMENT

- Initial management should focus on measures to support airway patency, adequate respirations, and effective circulation.
- The administration of activated charcoal should be considered in children who present within an hour of an oral exposure to amphetamines.
- If there is a possibility of airway compromise, activated charcoal should be withheld unless adequate protection of the patient's airway is ensured.
- Typical symptoms of amphetamine toxicity (eg, tachycardia, hypertension, and agitation) can be managed with benzodiazepines as needed for effect.
- Large doses of benzodiazepines may be needed for severe toxicity.
- Benzodiazepines are also the first-line agent for serotonin syndrome and amphetamine-induced seizures.
- For protracted seizure activity, another sedative-hypnotic agent such as phenobarbital or propofol may be needed.
- Phenytoin is generally not effective for toxin-induced seizures.

HYPONATREMIA

- Evaluation for hyponatremia is warranted in seizing patients with a history of MDMA exposure
- The treatment in this setting, after controlling seizure activity, is the correction of the underlying hyponatremia.

HYPERTHERMIA

- Hyperthermia in the setting of amphetamine toxicity is a very poor prognostic sign and should be attended to immediately with aggressive active cooling measures.
- Sedation with benzodiazepines or other sedative-hypnotic agent is necessary to limit the body's heat production.
- In extreme conditions, neuromuscular blockade with a nondepolarizing paralytic agent may be necessary.
- Continuous bedside electroencephalography should be performed if neuromuscular blockade is necessary.

OTHER COMPLICATIONS

- Rhabdomyolysis, stroke, intracerebral hemorrhage, acute coronary syndrome, and malignant cardiac dysrhythmias are all other complications that may occur with amphetamine toxicity.
- Avoid β -blockers.
- Manage these complications in standard fashion.

DISPOSITION

- Children who are asymptomatic or with mild toxicity should be observed in a controlled medical setting for 4 to 6 hours.
- Circumstances surrounding the exposure of these patients should be considered when planning disposition.
- Patient with moderate-to-severe symptoms and complications should be admitted for further observation and management.

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QUESTIONS

1. A 16-year-old male patient presents to the emergency department from a rave with generalized seizure activity. His seizures persist in the emergency

- department. His vital signs include a blood pressure of 180/100 and a heart rate of 140. His pupils are dilated, and his skin is diaphoretic. Which of the following actions would be the next best intervention for the patient?
- Administer charcoal.
 - Infuse hypertonic saline solution.
 - Establish a definitive airway.
 - Order a urine drug screen.
 - Perform a lumbar puncture.
2. A 2-year-old female patient is brought in by the police after she was found in a house where others were manufacturing methamphetamine. She starts to have generalized seizures. Which of the following drugs should be used first to help manage the seizure?
- Lorazepam
 - Phenytoin
 - Fosphenytoin
 - Phenobarbital
 - Isoniazid
3. A 16-year-old female patient presents from a party with dilated pupils, diaphoretic skin, tachycardia, agitation, and hypertension. Which of the following vital signs is most predictive of a poor prognosis?
- Heart rate greater than 120.
 - Respiratory rate greater than 30.
 - Core body temperature greater than 105.
 - Systolic blood pressure greater than 180.
 - Diastolic blood pressure greater than 110.
4. A 14-year-old male patient comes to the emergency department after having seizures. He has no previous history of seizures. His friends report that he was out late last night at a party and took some ecstasy for the first time. Which of the following electrolyte abnormalities would most likely explain this patient's presentation?
- Hypermagnesemia
 - Hypocalcemia
 - Hypokalemia
 - Hypernatremia
 - Hyponatremia
5. A 2-year-old male patient presents with significant tachycardia following amphetamine exposure. The pediatric cardiologist wants to give him a medication to control his rate. Which of the following medications is contraindicated in this patient?
- Amlodipine
 - Diltiazem
 - Verapamil
 - Esmolol
 - Digoxin

ANSWERS

- C. Establishing a definitive airway in this patient is the first priority. Just like when approaching any emergent patient, careful attention to airway, breathing, and circulation take precedence over any other management options.
- A. Lorazepam is the treatment of choice in this patient. Benzodiazepines should be considered a first-line agent for any toxin-induced seizures. This includes amphetamine-induced seizures. Benzodiazepines are also used to treat the typical symptoms of amphetamine toxicity such as tachycardia, hypertension, and agitation.
- C. Core body temperature greater than 105. The presence of hyperthermia is associated with increased mortality and is a very poor prognostic sign. Addressing the hyperthermia promptly with active cooling measures in a patient with severe amphetamine toxicity is extremely important. Sedation with benzodiazepines or other sedative-hypnotic agents may be necessary to limit the body's heat production. In extreme situations a neuromuscular blockade with a nondepolarizing paralytic agent may be necessary.
- E. Hyponatremia. Ecstasy (MDMA) can induce hyponatremia. This condition is often exacerbated by the consumption of a large amount of water.
- D. Esmolol is contraindicated in this patient. Beta-blockers are contraindicated in amphetamine toxicity as it may lead to unopposed alpha-adrenergic activation.

125 GAMMA-HYDROXYBUTYRATE

Jenny J. Lu

Timothy B. Erickson

HIGH-YIELD FACTS

- Originally investigated in the 1960s as a general anesthetic, but never widely accepted in the United States.
- Used in the 1980s by bodybuilders for purported anabolic steroid effects but banned by FDA after several reports of toxicity.
- Despite ban by FDA, recreational use of GHB by adolescents and young adults in the 1990s at rave parties increased, particularly in Europe.
 - Sold illicitly under various street names (Table 125-1).

TABLE 125-1 “Street” and Chemical Names for Gamma-Hydroxybutyrate

COMMON STREET NAMES	COMMON CHEMICAL NAMES
Liquid X	4-hydroxy butyrate
Grievous Bodily Harm	Gamma hydrate
Cherry Meth	Gamma-hydroxybutyrate sodium
Organic Quaalude	Gamma-hydroxybutyric acid
Easy Lay	Gamma-OH
Georgia Home Boy	Sodium oxybate
Insom-X	Sodium oxybutyrate
Liquid E	
Gamma G	

- GHB gained notoriety in recent years as a drug used to facilitate sexual assault.
- Main clinical use today is for the treatment of cataplexy in patients with narcolepsy.

PHARMACOLOGY

- Available as a clear liquid, white powder, tablet, or capsule.
 - For oral ingestions, peak concentrations occur within 25 to 45 minutes.
- Duration of effect is approximately 1 to 2.5 hours after anesthetic doses of 50 to 60 mg/kg and 2.5 hours after accidental overdoses.
- Plasma levels are usually undetectable within 4 to 6 hours after therapeutic doses.
- Volume of distribution is variable; not protein bound.

PATHOPHYSIOLOGY

- GHB is also a naturally occurring neuromodulator with a number of effects:
 - Inhibition of dopamine release.
 - Increase in endogenous opioids throughout the brain.
- Structurally an analog of the inhibitory neurotransmitter gamma-aminobutyric acid (GABA).
 - Ability to traverse the blood–brain barrier.
 - Agonist activity at the GHB and GABA (B) receptors.
- Abrupt loss of consciousness and apnea caused by exogenous GHB.
- Several GHB analogues and precursors exist.
 - Common precursors include gamma-butyrolactone (GBL) and 1,4-butanediol (found in industrial solvents such as paint thinners, floor strippers, consumer nail care products).
 - Can be metabolized to GHB and cause similar, but delayed, clinical effects.

- GHB precursor recently found in toys after causing clinical toxicity.

DIAGNOSIS

CLINICAL PRESENTATION OF ACUTE TOXICITY

- Main effects: Mental status depression.
 - Other signs and symptoms of GHB overdose: vomiting, sedation, and respiratory depression; unpredictable sudden awakening from a coma just prior to endotracheal intubation has been anecdotally described.
 - Although classically considered to be a CNS depressant, agitation, excitement, and combativeness have been reported.
- Patients may present to the emergency department with a history of recreational drug use, an accidental exposure to a GHB precursor-containing product, or as the victim of an alleged poisoning.
 - A high index of suspicion is required to make the diagnosis
 - Ecstasy and ethanol are common coingestants.
- Blood and urine specimens must be sent to one of a few national reference laboratories.
 - Results not timely enough to be clinically useful.
 - Due to rapid metabolism and elimination, the window for detection of GHB is very narrow.
 - GHB concentrations may have returned to endogenous levels by the time a specimen is obtained (normal or undetectable by 8 and 12 hours following an ingestion).
 - Can make proof of GHB poisoning problematic in cases of alleged sexual assault.
 - Appropriate cutoff levels and careful interpretation of results required to distinguish between endogenous and exogenous GHB concentrations.

TREATMENT

STABILIZATION

- Attentive supportive care, airway control, and stabilization of breathing and circulation should be the priorities.

DECONTAMINATION

- No role for gastric lavage or activated charcoal administration for isolated ingestions of GHB.
 - Significant amounts of the drug are unlikely to be present in the stomach by the time of presentation.

- Risk of pulmonary aspiration from the abrupt loss of consciousness observed with GHB intoxication.
- Activated charcoal for other coingestions can be considered if the patient's airway is protected and the patient is not combative.

ANTIDOTAL THERAPY

- No specific antidote is available for GHB poisoning.
- Treatment is centered around supportive care.
- Naloxone, flumazenil, and physostigmine have not been demonstrated to be effective in reversing GHB-induced coma.
 - Use of physostigmine has been associated with adverse cardiovascular effects.

ENHANCED ELIMINATION

- No role exists for hemodialysis or hemoperfusion for the isolated GHB overdose.

DISPOSITION

- Patients requiring ventilatory support or other significant interventions should be admitted.
- Patients whose symptoms have completely resolved and who are accompanied by reliable family members can potentially be considered for discharge after a period of observation.
- Consultation with a poison center is prudent for management and disposition issues.

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Zvosec DL, Smith SW, Litonjua R, et al: Physostigmine for gamma-hydroxybutyrate coma: inefficacy, adverse events, and review. *Clin Toxicol* 45:261–265, 2007.

QUESTIONS

1. A 16-year-old female is brought in to the ED after ingesting an unknown substance slipped in her drink and suspected by friends as a “date rape” drug. Which of the following effects may occur following GHB intoxication in this patient?
 - A. Apnea
 - B. Sedation
 - C. Coma followed by abrupt awakening
 - D. Agitation
 - E. All of the above
2. A 15-year-old male is brought in to the ED heavily sedated after ingesting GHB at a school event. Which of the following is false regarding GHB testing?
 - A. Window of detection is narrow.
 - B. Detected on most hospital urine drug screens.
 - C. Results of serum levels are unavailable in the acute clinical setting of intoxication.
 - D. Serum cut-off levels and interpretation of results are challenging.
3. A 16-year-old female is brought to the ED by a friend with a history of ingesting GHB at a party. Her vital signs are stable but she is comatose. Which of the following is likely to be true in this patient?
 - A. Naloxone is an effective antidote.
 - B. Physostigmine is an effective antidote.
 - C. Management is supportive.
 - D. Gastric lavage is urgently indicated.

ANSWERS

1. E. Although classically thought to be a CNS depressant, agitation, excitement, and combativeness have all been reported with GHB intoxication. Abrupt and unpredictable awakening from a comatose state just prior to endotracheal intubation has been anecdotally reported.
2. B. Most hospitals do not test for GHB. Blood or urine samples must be sent to one of a few national reference laboratories. Measuring GHB levels can be problematic due to the narrow window of detection and to difficulties interpreting low or undetectable levels because of individual variations in endogenous GHB levels. It is not clinically useful in the acute intoxication setting to send a GHB level.

3. C. Management in this case is purely supportive. Neither naloxone nor physostigmine are effective antidotes for GHB and there is no role for gastric lavage in this case.

126 LEAD POISONING

Mark B. Mycyk

INTRODUCTION

- The average blood lead level of American children has decreased by more than 80% since the 1970s because of early screening initiatives and environmental hazard reduction. Lead poisoning still affects people of all ages and classes, but the prevalence remains highest in inner-city underprivileged children. The current CDC blood action level is 10 µg/dL.

LEAD SOURCES

- Old household paint (and associated dust) is the most common source of lead poisoning.
- Other sources include contaminated drinking water, lead-painted toys from overseas, lead-glazed pottery, folk remedies, and lead brought home from the parents' workplace.
- Immigrant and refugee children are at high risk for occult lead poisoning.

PHARMACOKINETICS/ PATHOPHYSIOLOGY

- Up to 50% of ingested lead is absorbed by the gastrointestinal tract.
- Up to 30% of lead dust is absorbed through the respiratory tract.
- Absorbed lead resides in RBCs, soft tissue, and bone.
- Target organs of lead are the nervous system, bone marrow, and kidneys.
- The brain is most susceptible to lead damage in children < 2 years old.

CLINICAL MANIFESTATIONS

- Symptoms and signs of lead toxicity are often subtle and nonspecific, such as abdominal cramping, poor appetite, diminished energy, poor concentration, and various aches.
- The most likely cause for ED referral is a high screening blood lead level (BLL).
- Periodic screening is important for at-risk children, especially <2 years of age.
- Lead toxicity is grossly correlated with BLL (Table 126-1).
- Lead encephalopathy presents with ataxia, vomiting, lethargy, stupor, seizures, or coma.
- Microcytic anemia frequently coexists with lead poisoning.
- Low EP or ZPP suggests recent acute exposure, elevated EP, or ZPP suggests chronic exposure.
- Radiographic lead lines in long bones occur with BLLs >45 µg/dL.

TABLE 126-1 Class of Child, Toxic Effect, and Recommended Action According to Blood Lead Measurement

CLASS	BLOOD LEAD LEVEL (µg/dL) ^a	TOXIC EFFECT	RECOMMENDED ACTION
I	0–5	No noticeable effect	
	5–9	Inhibition of ALAD	
IIA	10–14		Rescreen every 3 mo; educate parents
IIB	15–19	Inhibition of ferrochelatase	Retest in 3 mo, nutritional and educational intervention, environmental investigation
III	20–44	Reduced growth, hearing, nerve conduction, neuropsychological deficits, reduced heme synthetase, increased EP, urine d-ALA	All of the above plus retest 1 wk–1 mo
IV	45–69	Anemia, abdominal colic, reduced IQ, lead lines in x-ray	All of the above plus chelation: oral DMSA or IV CaNa ₂ EDTA
V	>70	Encephalopathy risk, nephropathy (>100 µ/dL)	Medical emergency: chelate with BAL plus EDTA, increased ICP precautions

^aConversion factor: 1.0 µg/dL = 0.04826 mmol/L.

ALAD, aminolevulinic acid dehydratase; EP, erythrocyte protoporphyrin; ALA, aminolevulinic acid.

With permission from Centers for Disease Control and Prevention. Screening young children for lead poisoning: Guidance for state and local public health officials. US Dept. of Health and Human Services, Public Health Service, *Federal Register*, February 21, 1997.

- Ingested paint chips can be seen on flat abdominal x-rays.
- Basophilic stippling, a “classic” finding on a CBC, occurs uncommonly.

MANAGEMENT

- The most important step is identifying the lead source and removing the patient from the source.
- Paint chips in the abdomen warrant decontamination with whole bowel irrigation.
- BLLs between 10 and 45 $\mu\text{g}/\text{dL}$ warrant follow-up testing and environmental evaluation.
- The current CDC blood action level is 10 $\mu\text{g}/\text{dL}$.
- Children with BLLs 45 to 69 $\mu\text{g}/\text{dL}$ may be treated with oral dimercaptosuccinic acid (DMSA) as an outpatient.
- Children with BLLs >70 $\mu\text{g}/\text{dL}$ warrant hospitalization and treatment with DMSA, CaNa_2EDTA , or BAL in conjunction with CaNa_2EDTA .
- Rebound BLL elevation is common after chelation therapy and follow-up testing is essential.
- Consultation with a toxicologist, poison center, or lead specialist is essential.

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QUESTIONS

1. A 2-year old is being evaluated for a microcytic anemia and blood tests are done in the work up to identify the cause. She is subsequently referred to the ED with a confirmed toxic serum lead level

and initiation of treatment. Which of the following should be considered a potential source of lead poisoning in this patient and asked about in the history?

- A. Household paint dust
 - B. Folk remedy
 - C. Contaminated drinking water
 - D. Lead dust from the parent’s workplace
 - E. All of the above
2. During your evaluation of a 3-year old for a fall, you notice growth arrest lines (“lead lines”) on the forearm x-ray. A BLL will not be available until tomorrow. That child’s BLL is at least how high based on this radiographic finding?
 - A. 10 mcg/dL
 - B. 20 mcg/dL
 - C. 40 mcg/dL
 - D. 45 mcg/dL
 - E. 50 mcg/dL
 3. A 12-month-old male has a routine screening BLL drawn and is found to have a markedly elevated level of 40 mcg/dL. Which of the following is an oral chelator used for lead poisoning?
 - A. BAL
 - B. $\text{Ca-Na}_2\text{EDTA}$
 - C. Desferrioxamine
 - D. DMSA
 - E. Hydroxocobalamin
 4. The most likely reason for ED evaluation for lead poisoning in the 21st century is which of the following reasons?
 - A. Referral for high outpatient screening BLL.
 - B. Infant with seizures.
 - C. Kindergarten child with fever.
 - D. Teenager with new ataxia.
 - E. Symptomatic anemia.

ANSWERS

1. E. The most common source of lead poisoning in children is old household paint and the associated dust. Exposure can also come from contaminated drinking water, lead-painted toys, lead-glazed pottery, folk remedies, and lead brought home from the parents’ workplace.
2. D. Radiographic lead lines in long bones occur with BLLs of generally greater than 45 $\mu\text{g}/\text{dL}$.
3. D. DMSA is an oral chelator used to treat elevated BLLs. The current CDC blood action level is 10 $\mu\text{g}/\text{dL}$. CaNa_2EDTA and BAL are both chelators but are given parenterally. CaNa_2EDTA is given IV and BAL is given IM.

4. A. The most likely cause for ED referral in this day and age is for a high screening blood lead levels necessitating treatment. Symptoms and signs of lead toxicity are often subtle and nonspecific, such as abdominal cramping, poor appetite, diminished energy, poor concentration, and various aches.

127 IRON

Michael R. Christian

Steven E. Aks

INTRODUCTION

- Previously, iron toxicity caused more pediatric deaths than any other poison. However, the FDA has since made an advisory recommendation requiring products containing more than 30 mg of elemental iron be packaged in blister packs, which has decreased the incidence of iron toxicity. However, prenatal vitamins containing significant amounts of elemental iron are still dispensed in bottles. Today, pediatric death from iron toxicity is rare, but these cases may still be seen.

PHARMACOLOGY & PATHOPHYSIOLOGY

- Iron is absorbed through the gastrointestinal tract in the ferrous (Fe^{2+}) state, oxidized to the ferric (Fe^{3+}) state, and bound to transferrin.
- Toxicity occurs when the transferring binding capacity is exceeded, which is determined by the amount of elemental iron ingested. Ingestions exceeding 40 mg/kg are associated with significant toxicity; ingestions exceeding 60 mg/kg are associated with death.
- The proportion of elemental iron varies significantly depending upon the preparation (see Table 127-1).
- Iron is a potent catalyst of free radical generation, which causes oxidative damage to blood vessel walls, gastrointestinal tract, liver, kidneys, lungs, and heart.

CLINICAL MANIFESTATIONS

- Stage 1 (0–6 hours): varies from mild nausea and vomiting to hematemesis, hemorrhagic diarrhea, altered mental status, and hypotension, depending upon the amount of elemental iron ingested.

TABLE 127-1 Iron Preparations

IRON PREPARATION	ELEMENTAL IRON (%)
Ferrous sulfate	20
Ferrous fumarate	33
Ferrous gluconate	12

- Stage 2 (6–12 hours): gastrointestinal symptoms may improve during this quiescent or “danger” phase.
- Stage 3 (12–24 hours): severe toxicity manifested by gastrointestinal hemorrhage, renal failure, hepatic failure, altered mental status, severe metabolic acidosis, and cardiovascular collapse.
- Stage 4 (24–48 hours): primarily hepatotoxicity with transaminitis followed by fulminant hepatic failure.
- Stage 5 (4–6 weeks): gastrointestinal strictures are a possible late complication with significant morbidity.

DIAGNOSIS

- Complete blood count, metabolic panel, hepatic panel, serum iron level, and an abdominal radiograph should be obtained.
- Metabolic acidosis, serum glucose >150 mg/dL, and white blood cell count >15,000 are associated with toxicity.
- Serum iron levels should be obtained between two and six hours after ingestion. Levels >500 $\mu\text{g}/\text{dL}$ are associated with toxicity and levels >1,000 $\mu\text{g}/\text{dL}$ are potentially fatal.

MANAGEMENT

- In patients with pills visible on abdominal radiograph, consider whole bowel irrigation with polyethylene glycol solution via nasogastric tube at 25 mL/kg/hr in children or 1–2 L/hr in adolescents until clear rectal effluent.
- Active gastrointestinal bleeding, ileus, and bowel obstruction are contraindications to whole bowel irrigation.
- Chelation with deferoxamine at 15 mg/kg/hr IV is indicated in patients with a serum iron of >500 $\mu\text{g}/\text{dL}$ and a metabolic acidosis.
- Resolution of metabolic acidosis is a more reliable endpoint than reduction of serum iron level or the disappearance of the classic “vin rose” urine that is occasionally seen with therapy. Do not administer deferoxamine beyond 24 hours because of the increased risk of acute lung injury. Deferoxamine can also cause significant hypotension.

TABLE 127-2 Poison Center-Based Triage Guidelines

INGESTED DOSE (mg/kg)	TREATMENT RECOMMENDATION
<30	Observe at home
>30–40	If symptomatic, refer to a health care facility
>40 mg/kg	Refer to health care facility

Klein-Schwartz W, Odega GM, Gorman RL, et al. Assessment of management guidelines in acute iron ingestion. *Clin Pediatr.* 1990;29:316–321. Copyright © 1990 by SAGE Publications. Reprinted by Permission of SAGE Publications.

DISPOSITION

- Most pediatric patients with exposure to iron do not need to be evaluated at a healthcare facility (see Table 127-2).
- Asymptomatic patients, with peak serum iron levels less than 500 µg/dL approximately four hours postingestion, can be discharged with supervision by reliable caretakers.
- Patients with metabolic acidosis, hypotension, shock, or requiring treatment with deferoxamine should be monitored in the critical care setting.
- Patients with moderate to severe toxicity require follow-up to evaluate for delayed gastrointestinal strictures.

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QUESTIONS

1. A 2-year-old female presents to the ED approximately six hours after ingesting 30 tablets of 325 mg prenatal ferrous sulfate. The parents state that several hours after ingestion, the child vomited once but now appears to be improved. Your advice to the parents should be which of the following?
 - A. The danger period is over. The child will do well.
 - B. The child is in the danger phase.
 - C. If no pills are identified by the abdominal radiograph, the child is in no danger.
 - D. The child should undergo gastric lavage.
 - E. Activated charcoal should be given to the child.
2. Which of the following iron levels would suggest life-threatening toxicity?
 - A. Greater than 100 to 200 µg/dL
 - B. Greater than 200 to 300 µg/dL
 - C. Greater than 300 to 400 µg/dL
 - D. Greater than 400 to 500 µg/dL
 - E. Greater than 500 µg/dL
3. A 3-year-old male presents after an acute ingestion of iron tablets and is found to have an iron level greater than 600 µg/dL. He has been vomiting and hypotensive. The child will be treated with deferoxamine. All of the following are true regarding deferoxamine *except*:
 - A. Indicated if a peak iron level is 300 to 350 µg/dL.
 - B. Administered at six-hour intervals.
 - C. Intravenous administration is used for severe toxicity.
 - D. The end point of chelation is when the color of the patient's urine turns from yellow to red.
 - E. Hypotension is a common side effect of intravenous therapy.
4. Which formulation of iron has the highest percentage of elemental iron?
 - A. Ferrous sulfate
 - B. Ferrous sulfate suspension
 - C. Ferrous fumarate
 - D. Ferrous gluconate
 - E. Multivitamin
5. The mother of a 6-year-old male calls the ED and states that her child was found playing with her iron pills in blister packaging approximately 15 minutes ago. Ten tablets of ferrous sulfate 325 mg are missing. The child has had no episodes of emesis or diarrhea. The patient is alert and interactive. Her child weighed 55 pounds (25 kg) at his well-child visit last week. You should advise the mother to do which of the following?
 - A. Bring her child to the ED immediately.
 - B. Induce vomiting with ipecac.
 - C. Call 911.
 - D. Bring child to ED if he has emesis or diarrhea.
 - E. Administer 15 mL of calcium carbonate.
6. A 16-year-old female intentionally ingested approximately 40 tablets of 325 mg ferrous sulfate approximately 5 hours prior to arrival. Previously, the patient had multiple episodes of nausea, vomiting, and diarrhea, but these symptoms have resolved. The patient has a temperature of 37.3°F, blood pressure

of 108/67, heart rate of 110, respiratory rate of 20, and an oxygen saturation of 98% on room air. Exam is remarkable for mild, diffuse abdominal tenderness to palpation without rebound or guarding. Labs are remarkable for a WBC of 18,000, bicarbonate of 18 mEq/L, glucose of 160 mg/dL, and a serum iron level of 672 $\mu\text{g/dL}$. A KUB reveals multiple radiopaque foreign bodies in the small bowel. In addition to aggressive supportive care, which of the following should be performed in this patient?

- A. Orogastric lavage.
 - B. Administer syrup of ipecac.
 - C. Placement of NG and administration of polyethylene glycol at 1–2 L/hour until clear rectal effluent.
 - D. Order the patient to drink polyethylene glycol at 1–2 L/hour until clear rectal effluent.
 - E. Administration of activated charcoal 50 g orally followed by additional doses of 25 g every 6 hours.
7. In the previous patient, gastrointestinal decontamination is performed and deferoxamine is started at 15 mg/kg/hr. Over the next 20–48 hours, what end organ is at the highest risk of toxicity?
- A. Kidney
 - B. Liver
 - C. Thyroid
 - D. Lungs
 - E. Central nervous system
8. In the previous patient, she has a follow-up appointment as an outpatient approximately four weeks after her ingestion. The patient is sent from the clinic to the ED for admission because she has been unable to tolerate solid food for 2–3 days. What is the most likely etiology of the patient's symptoms?
- A. GI stricture formation.
 - B. Delayed hepatotoxicity
 - C. Pancreatic insufficiency.
 - D. CNS toxicity causing dysphagia.
 - E. Malignancy.

ANSWERS

1. B. Stage 1 of iron toxicity begins at time of ingestion and lasts for 6 hours. Severe ingestions may present with nausea, vomiting, diarrhea, hematemesis, altered mental status, and possibly hypotension. Stage 2 occurs at 6 to 12 hours postingestion and is referred to as the quiescent or “danger” phase because the child can appear to be improving or asymptomatic. Stage 3, from 12 to 24 hours postingestion, is marked by major symptoms of toxicity including

bleeding, cardiovascular collapse, altered mental status, renal and hepatic failure, and metabolic acidosis. Stage 4 is a latent or recovery phase. Although iron pills may be identified by radiograph, the physician should not rely on the absence of pill identification to continue treatment. Time of presentation of this child is too late for gastric lavage. Lavage must be done 1 to 2 hours postingestion in order to be effective. Activated charcoal is not effective in absorbing iron in the gastrointestinal tract.

2. E. A level greater than 350 $\mu\text{g/dL}$ is considered toxic if the patient is symptomatic, but levels greater than 500 $\mu\text{g/dL}$ suggest potentially life-threatening toxicity regardless of symptoms.
3. D. Classically, the endpoint of chelation was reached when the color of the patient's urine turns from wine red, a.k.a. “vin rose” to yellow (normal). However, the resolution of metabolic acidosis is now thought to be a more reliable endpoint.
4. C. Ferrous fumarate contains 33% elemental iron, ferrous sulfate contains 20% elemental iron, ferrous gluconate contains 12% elemental iron, and multivitamins typically contain <5% of elemental iron.
5. D. Due to an FDA-mandated change in the packaging of iron supplements, iron rarely causes deaths in the pediatric population. When dealing with iron, it is important to estimate the amount of elemental iron ingested in the worst-case scenario. The patient could have ingested ten tablets of 325 mg ferrous sulfate, which contains 20% elemental iron. Therefore, the patient could have ingested 650 mg or 26 mg/kg of elemental iron. In this case, it would be reasonable and safe to observe the patient at home. There is no role for ipecac in pediatric poisonings. Calcium carbonate (mylanta) has no role in the treatment of iron ingestion.
6. C. This patient clearly has a significant ingestion of iron and whole bowel irrigation should be performed. It is very unlikely that this patient, or any patient for that matter, will be able to drink 1–2 L of polyethylene glycol every hour. It is necessary to administer the polyethylene glycol via an NG. Orogastric lavage and syrup of ipecac have little to no role in the poisoned patient. Activated charcoal does not bind metals like iron.
7. B. In stages 3 and 4 of iron toxicity, patients are mostly likely to die from the complications of hepatic failure.
8. A. In stage 5 of iron toxicity, patients may form gastrointestinal strictures (usually at the gastric pylorus), which have significant morbidity.

Environmental Poisons

128 CYANIDE POISONING

Mark B. Mycyk

INTRODUCTION

- Cyanide poisoning is unusual in the United States and very rare among children. Smoke inhalation from building fires is the most common source for hydrogen cyanide from combustible synthetic materials. Ingestion of cyanide is less common and may occur from acetonitrile in nail removal agents, potassium cyanide in metal cleaning solutions imported from Asia, and cyanogenic glycosides found in the seeds and pits of certain plants such as apples, apricots, and peaches.

PHARMACOPATHOLOGY

- Inhaled hydrogen cyanide gas results in toxicity within seconds to minutes.
- Ingested cyanide salts may result in toxicity within minutes to a few hours.
- Cyanide primarily causes tissue hypoxia at cytochrome $a-a_3$ in the mitochondria.
- Cyanide disrupts ATP production and oxygen utilization.
- Cyanide shifts the oxygen-hemoglobin dissociation curve to the left.
- Lactic acidosis results from anaerobic metabolism.
- Critical early targets of cyanide are the brain and heart since both are dependent on oxygen.
- Endogenous rhodanase (sulfurtransferase) converts cyanide to less toxic thiocyanate.
- Rhodanase activity is enhanced in the presence of thiosulfate.
- Hydroxocobalamin (vitamin B_{12a}), converts cyanide to nontoxic cyanocobalamin (vitamin B_{12}).

CLINICAL PRESENTATION

- Inhalation of cyanide gas causes loss of consciousness within seconds.

- Oral exposure causes symptoms from minutes to several hours after ingestion. Early symptoms include headache, anxiety, confusion, blurred vision, palpitations, nausea, and vomiting.
- Early clinical signs of poisoning are mixed: CNS stimulation or depression, tachycardia or bradycardia, hypertension or hypotension. Ocular exam reveals dilated pupils and bright red retinal veins on funduscopy.
- Late signs of poisoning are seizures, coma, apnea, cardiac arrhythmias, and complete cardiovascular collapse.
- The smell of bitter almonds may be detected in some cases.
- The presence of cyanosis is a relatively late finding, since cyanide typically causes a leftward shift of the oxygen dissociation curve.

LABORATORY EVALUATION

- Whole blood cyanide levels are not available emergently.
- Arterial blood gases show a significant metabolic acidosis. A simultaneous venous blood gas analysis for comparison may demonstrate a diminished arterial-venous O_2 difference (AO_2-VO_2 approaching zero) from impaired oxygen delivery.
- An elevated lactate or anion gap acidosis are common.
- Numerous electrocardiographic changes may occur in cyanide toxicity.
- Elevated carboxyhemoglobin levels correlate with cyanide levels in smoke inhalation victims.

TREATMENT

- Airway management with 100% oxygen.
- Intravenous access. IV fluids for hypotension.
- The Taylor Antidote Kit contains amyl nitrite, sodium nitrite, and sodium thiosulfate.
- Hypotension is common with nitrite therapy and methemoglobin levels should be monitored.
- Sodium thiosulfate provides a sulfur donor to enhance rhodanase-mediated conversion of cyanomethemoglobin to less toxic thiocyanate.
- Hydroxocobalamin (a precursor to vitamin B_{12}) detoxifies cyanide to form nontoxic cyanocobalamin. Hydroxocobalamin is not associated with hypotension or excessive methemoglobinemia. Red discoloration of the skin is common with hydroxocobalamin.

DISPOSITION

- Patients requiring antidotal treatment require intensive care unit admission.
- Ingested cyanogenic glycosides require at least 6 hours observation.
- Acetonitrile ingestions require 12 to 24 hours observation because of delayed metabolism to cyanide.

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QUESTIONS

1. A 10-year-old male is brought in after being trapped in a clothing store that caught fire. He is unresponsive at the scene. What test result often correlates with significant cyanide poisoning from a house/building fire?
 - A. Anemia
 - B. Elevated bicarbonate
 - C. Elevated carboxyhemoglobin
 - D. Hypokalemia
 - E. Hyponatremia
2. A 5-year-old presents in the ED after being in a house fire. He had a seizure prior to arrival and is comatose. He smells of bitter almonds. Since cyanide levels cannot be obtained emergently, what laboratory test is highly suggestive of cyanide poisoning?
 - A. Elevated CPK
 - B. Elevated lactate
 - C. Elevated pH
 - D. Hematuria
 - E. Hypocalcemia

3. In the previous patient you decide to treat on the clinical presentation and suggestive lab findings for cyanide toxicity with the Taylor Antidote Kit. What is the most common adverse effect of hydroxocobalamin?
 - A. Headache
 - B. Hypotension
 - C. Nystagmus
 - D. Pruritis
 - E. Red skin discoloration
4. Which of the following tests should be closely monitored in patients who received the Taylor Antidote Kit?
 - A. Base excess
 - B. ECG
 - C. Methemoglobin
 - D. Sulfhemoglobin
 - E. Urine output

ANSWERS

1. C. Elevated carboxyhemoglobin levels correlate with cyanide levels in smoke inhalation victims. In children, smoke inhalation from building fires is the most common source for hydrogen cyanide from combustible synthetic materials.
2. B. Whole blood cyanide levels are not available emergently. Arterial blood gases show a significant metabolic acidosis and an elevated lactate or anion gap acidosis are common and helpful in making the diagnosis.
3. E. Hydroxocobalamin (a precursor to vitamin B₁₂) detoxifies cyanide to form nontoxic cyanocobalamin. Hydroxocobalamin is not associated with hypotension or excessive methemoglobinemia. Red discoloration of the skin is common with hydroxocobalamin.
4. C. Hypotension is common with nitrite therapy and methemoglobin levels should be monitored during administration.

129 MUSHROOM POISONING

Matthew Valento

INTRODUCTION

- Mushroom poisoning is a rare but potentially life-threatening clinical scenario. Classification of the ingested mushroom can be difficult, as mushroom species vary widely and toxin concentrations can differ depending on locale, season, and the part of

the mushroom ingested. As such, identification of North American mushroom poisoning relies primarily on clinical features. If samples or pictures of the suspected toxic mushroom are available, the regional poison center may be able to assist in identification.

CLASSIFICATION

- Poisonous mushrooms can be classified into ten groups based on the primary toxin:
 - Cyclopeptides
 - Gyromitrin
 - Muscarine
 - Coprine
 - Ibotenic acid and muscimol
 - Psilocybin
 - Gastrointestinal irritant
 - Orellanine
 - Allenic norleucine
 - Myotoxin

CYCLOPEPTIDE GROUP

- Mushrooms containing cyclopeptide amatoxins are responsible for the most mushroom fatalities in North American and worldwide.
- Include species from genus *Amanita*, *Galerina*, and *Lepiota*.
- Commonly encountered species include *Amanita phalloides* (the “death cap”), *Amanita verna* (“death angel”), and *Amanita virosa* (“destroying angel”).
- Produce severe delayed (>6 hours postingestion) gastroenteritis followed by hepatic and renal toxicity.
- Clinical presentation characterized by three stages:
 - Severe gastroenteritis 6–24 hours postingestion.
 - An apparent quiescent phase with resolution of GI symptoms, though hepatocellular damage continues.
 - The final phase, 36–72 hours postingestion, is characterized by hepatotoxicity, jaundice, hypoglycemia, nephrotoxicity, coagulopathy, and death.
- Supportive care, including aggressive fluid/electrolyte replacement, is a mainstay of treatment.
- Multiple pharmacologic interventions have been attempted, with limited success. The following agents should be considered, if available:
 - Activated charcoal, 1 g/kg every 2–4 hours.
 - *N*-acetylcysteine.
 - Silymarin/silybinin (limited availability in North America).
- Severe toxicity may require liver transplantation.

GYROMITRIN GROUP

- This group contains *Gyromitra* species, including *Gyromitra esculenta* (the “false morel”).
- Ingestion of mushrooms containing gyromitrin produces monomethylhydrazine, which inhibits pyridoxal phosphate, leading to decreased gamma aminobutyric acid (GABA) function.
- Produces gastroenteritis, abdominal pain, diffuse muscle cramping 5–10 hours postingestion.
- The clinical hallmark of severe toxicity is intractable seizures, similar to isoniazid poisoning.
- Treatment includes activated charcoal and fluid/electrolyte replacement.
- Seizures should be treated with pyridoxine, 70 mg/kg IV, until resolution.

MUSCARINE GROUP

- Several *Clitocybe* and *Inocybe* mushrooms, as well as some *Amanita* species, are included in this group.
- Poisoning produces typical signs/symptoms of muscarine-induced cholinergic toxicity, including salivation, lacrimation, urination, diarrhea, and bronchorrhea/bronchospasm.
- Muscarine does not cross the blood–brain barrier; as such, central signs are not seen.
- Symptoms are generally mild and self-limiting, and rarely require specific therapies.
- Atropine can be used for severe cholinergic toxicity.

COPRINE GROUP

- *Coprinus* mushrooms, also known as “inky caps,” are nontoxic when ingested in the absence of alcohol.
- The primary metabolite of coprine, 1-aminocyclopropanol, produces a disulfiram-like reaction in the presence of ethanol.
- Patient may present with flushing, paresthesias, diaphoresis, nausea, and vomiting.
- Treatment is supportive and toxicity is usually mild.

IBOTENIC ACID AND MUSCIMOL GROUP

- The group includes *Amanita muscaria*, *Amanita pantherina*, and *Amanita gemmata*, with brilliant red or tan caps resembling mushrooms commonly depicted in children’s books.
- The toxins ibotenic acid and muscimol are psychoactive isoxazoles, producing somnolence, hallucinations, dysphoria, and confusion 0.5–2 hours postingestions.

- Seizures may be observed in pediatric ingestions.
- Supportive care is the primary treatment, with benzodiazepines for significant agitation or seizures.

PSILOCYBIN GROUP

- These mushrooms, including some *Psilocybe*, *Conocybe*, and *Panaeolus* species, contain the hallucinogenic, proserotonergic indole psilocybin.
- Ingestion produces rapid onset of ataxia, visual hallucinations, illusions, and mood lability.
- Patients rarely may present with tachycardia, tremor, and significant agitation.
- Treatment is supportive, with benzodiazepines for agitation.

GASTROINTESTINAL IRRITANT GROUP

- This large, commonly encountered group of “little brown mushrooms” contains a variety of poorly defined toxins.
- Gastrointestinal toxicity begins shortly (0.5–3 hours) postingestion.
- Symptoms are generally mild and require only supportive care.
- A rare occurrence after ingestion of *Paxillus involutus* is a mild GI syndrome followed by immune-mediated hemolytic anemia, hemoglobinuria, and renal failure.

ORELLANINE GROUP

- Includes *Cortinarius* mushrooms containing orellanine, a nephrotoxin similar to paraquat.
- Initial symptoms occur 24–26 hours postingestion and include headache, chills, nausea, vomiting, and flank pain.
- Days to weeks later, patients may develop interstitial nephritis and oliguric renal failure, which may require dialysis or renal transplant.
- Treatment includes fluid management with follow-up monitoring for nephrotoxicity.

ALLENIC NORLEUCINE GROUP

- *Amanita smithiana*, found in the Pacific Northwest and resembling the edible pine mushroom matsutake, is the primary member of this group.
- Patient present with gastrointestinal symptoms, malaise, headache, and dizziness 0.5–12 hours postingestion.

- Acute renal failure develops 4–6 days later, often with concurrent elevations in ALT and lactate dehydrogenase (LDH).
- Activated charcoal should be considered during the early stage of toxicity.
- Patient may require hemodialysis.

MYOTOXIN GROUP

- A small number of cases of severe rhabdomyolysis following ingestion of *Trichloma equestre* has been described.
- Symptoms include fatigue, weakness, nausea, and myalgias 24–72 hours postingestion, with concurrent large elevations in serum creatine phosphokinase (CPK).
- May progress to pulmonary congestion, myocarditis, dysrhythmias, and death.
- The toxin has not been identified, and treatment is primarily supportive.

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QUESTIONS

1. A 3-year-old female is brought to the emergency department (ED) by her family after eating several mushrooms believed to be *Amanita phalloides* (“death cap”) while out in the woods on a camping trip. Which clinical feature is most consistent with poisoning from a mushroom of this cyclopeptide group and might be expected in this patient?
 - A. Severe nausea, vomiting, and diarrhea 2 hours postingestion.
 - B. Intractable seizures.
 - C. Interstitial nephritis.
 - D. Visual hallucinations and agitation.
 - E. Gastroenteritis 10 hour postingestion, followed by elevated liver enzymes.

2. A 17-year-old male presents to the ED after several witnessed generalized tonic-clonic seizures. A family member states that earlier in the day he went mushroom hunting for morels with his uncle. What antidotal therapy should be considered if seizures continue?
- N*-acetylcysteine
 - Activated charcoal
 - Pyridoxine
 - Silybinin
 - Phenytoin
3. Which of the following does not correctly pair the mushroom toxin with its potential clinical feature?
- Orellanine: oliguric renal failure
 - Ibotenic acid: hallucinations
 - Cyclopeptides: hepatotoxicity
 - Coprine: rhabdomyolysis
 - Gyromitrin: seizures

ANSWERS

1. E. Cyclopeptide-containing mushrooms classically produce a delayed (>6 hours postingestion) gastroenteritis. Hepatotoxicity and renal failure may follow 2–3 days later. Intractable seizures are a sign of toxicity from mushrooms containing gyromitrin. Ingestion of mushrooms containing orellanine can produce interstitial nephritis and renal failure. Visual hallucinations often develop soon after ingesting mushrooms containing the psychoactive indole psilocybin.
2. C. Ingestion of *G esculenta*, the “false morel,” may produce intractable seizures, primarily through inhibition of pyridoxal phosphate enzyme systems. Pyridoxine, 70 mg/kg IV, should be administered. *N*-acetylcysteine and silybinin have been used with limited success to treat toxicity from cyclopeptide-containing mushrooms. Activated charcoal, while a standard treatment modality for some mushroom poisonings, is not safe to administer to a patient with active seizure activity. Phenytoin and other antiepileptics are not recommended for toxin-induced seizures.
3. D. All of the answers are correct except D. Coprine-containing mushrooms can produce a disulfiram-like reaction when ingested with alcohol. Severe rhabdomyolysis has been described after ingestion of *T equestre*; the toxin is unknown.

130 POISONOUS PLANTS

Ejaaz A. Kalimullah
Andrea G. Carlson

INTRODUCTION

- Evaluation of plant exposure presents may be challenging for health care provider.
- Information regarding the species of plant as well as the amount ingested is often lacking.
- The degree of toxicity expected may vary depending on the particular part of the plant ingested, the growth stage of the plant, and prior processing of the plant material.
- Considerable overlap exists in the clinical manifestations of toxicity of many plants.
- While serious toxicity is absent in most exposures, a number of plants are mildly toxic and a few are harmful even in small amounts.

EPIDEMIOLOGY

- The American Association of Poison Control Centers (AAPCC) database catalogs 64,236 human exposures to plants in 2006, accounting for 2.7% of all reported human cases.
- 44,710 plant exposures were reported in children ≤5 years of age.
- Only a single fatality was attributed to a plant or herbal/botanical product in the 2006 AAPCC National Poison Data System annual report.
- Nearly 70% of plant ingestions are by children younger than 6 years.
- Plant ingestions by toddlers and other young children often involve trivial amounts found while exploring their environment.
- Older children and adolescents may intentionally consume specific plant species for their purported psychoactive effects, and may be at higher risk for morbidity.

IDENTIFICATION OF PLANTS

- Plant name and/or accurate species information is often difficult to ascertain.
- Useful historical information includes whether the plant is an indoor or outdoor variety, and a description of the plant's flower, stem, leaves, height, and location.

- Consultation with a botanist, medical toxicologist, or poison control center is recommended whenever assistance is needed.
- Transmission of digital color images can be used to assist consultants.
- Rough recognition of taxonomic families of poisonous plants and/or comparison of the species in question to field guide pictures may help exclude potentially life-threatening plant exposures.

SPECIFIC TOXIC PLANTS

MUCOSAL IRRITANTS

- A wide variety of plant species cause mucous membrane irritation due to microscopic needle-shaped bundles of calcium oxalate (raphides).
- Raphides release the calcium oxalate crystals on mastication, resulting in damage to mucous membranes, immediate burning, and inflammation.
- Plants containing calcium oxalate include: philodendron (*Philodendron* sp.), dumbcane (*Dieffenbachia*), peace lily (*Spathiphyllum* sp.), pothos (*Epipremnum aureum*), caladium (*Caladium bicolor*), calla lily (*Zantedeschia aethiopica*), Jack-in-the-pulpit (*Arisaema triphyllum*), elephant's ear (*Colocasia esculenta*), and skunk cabbage (*Symplocarpus foetidus*).
- Treatment is symptomatic with demulcents, analgesics as needed, and antihistamines for associated pruritis or local dermal irritation.
- Exposures to various *Capsicum* species such as the chili pepper (*Capsicum annuum*) also constitute an important source of mucosal irritant exposures.
- Clinical effects consist of intense burning pain, hyperalgesia, irritation, and erythema. Profuse lacrimation and conjunctival inflammation may occur with ocular exposure.
- In some cases, contact dermatitis can develop and symptoms can persist for hours to days.
- Treatment is focused on decontamination and pain control. While cold-water irrigation can provide relief, capsaicin is poorly soluble. Alternatives suggested in the medical literature include: vinegar, topical antacids, vegetable oil, and 2% lidocaine gel.

GASTROINTESTINAL IRRITANTS

AMERICAN MISTLETOE (*PHORADENDRON SEROTINUM*, *PHORADENDRON LEUCARPUM*)

- American Mistletoe pediatric accidental ingestions are largely asymptomatic, with essentially no major morbidity.

- Reported symptoms are mostly gastrointestinal (GI) upset, with symptoms developing within six hours of exposure.
- Treatment is supportive and decontamination for small (≤ 5 berries or ≤ 3 leaves) ingestions is unnecessary.

POINSETTIA (*EUPHORBIA PULCHERRIMA*)

- Poinsettia (*E pulcherrima*) exposures also tend to lack significant morbidity.
- Most accidental pediatric poinsettia exposures can be managed without ED referral given the rarity of significant symptoms.
- Management requires no more than supportive care and GI decontamination has not been shown to be of benefit.

HOLLY (*ILEX* SP.)

- *Ilex opaca* (American holly) and *Ilex aquifolium* (English holly) can also cause GI irritation. The plants produce green berries, which mature into red berries that are attractive to small children.
- The holly berries are likely responsible for toxicity and large ingestions can cause nausea, vomiting, abdominal cramping, and diarrhea.
- Small (≤ 2 berries) accidental ingestions are unlikely to produce significant symptoms. Basic supportive care with attention to hydration status is sufficient.

POKEWEED (*PHYTOLACCA AMERICANA*)

- Pokeweed contains toxic saponins (phytolaccatoxin, phytolaccagenin), as well as a lymphotropic mitogen.
- All parts of the plant, particularly the root, are toxic. Poisoning primarily causes nausea, vomiting, abdominal cramps, and diarrhea.
- Severe cases with heme positive stools, tachycardia, and hypotension have also been reported.
- Rare cases with electrocardiographic (EKG) changes have been reported in adults, usually in the setting of significant GI symptoms.
- Supportive care is the mainstay of treatment and decontamination with activated charcoal may be considered in nonvomiting patients. Admission may be required for observation and continued care in the setting of large or markedly symptomatic pokeweed ingestions.

SYSTEMIC TOXINS

PLANTS CONTAINING ANTICHOLINERGIC SUBSTANCES

- Jimsonweed (*Datura stramonium*), deadly nightshade (*Atropa belladonna*), and black henbane (*H niger*) are some of the plants that contain muscarinic antagonist

- alkaloids such as atropine, hyoscyamine, and scopolamine.
- Jimsonweed grows wild throughout the United States, and its seeds have been abused by adolescents and teenagers due to their psychotropic effects.
 - An anticholinergic toxidrome with tachycardia, mydriasis, dry skin and mucous membranes, hypoactive bowel sounds, urinary retention, and hyperthermia may be noted.
 - Central anticholinergic effects result in altered mental status, ranging from somnolence to severe agitation and delirium with hallucinations.
 - Agitation can be initially treated with benzodiazepines. Hyperthermia should be treated supportively with cooling measures.
 - Physostigmine has been safely and effectively utilized as an antidote in anticholinergic plant poisoning, usually in cases with refractory delirium, hyperthermia, seizures, or significant arrhythmias.
 - Patients with marked delirium, seizures, cardiopulmonary instability, or those treated with physostigmine should be admitted to an intensive care unit.

PLANTS CONTAINING SOLANINE

- Plants containing solanaceous alkaloids grow throughout the United States, and include the black nightshade (*Solanum nigrum*), Jerusalem cherry (*Solanum pseudocapsicum*), bittersweet (*Solanum dulcamara*), and potato (*Solanum tuberosum*).
- Ingestion of sun-greened potatoes or uncooked potato sprouts may also cause illness as they contain α -solanine and the related glycoalkaloid, α -chaconine.
- Small ingestions generally result in no more than self-limited GI effects. Severe intoxications can manifest with CNS and respiratory depression, hyperthermia, bradycardia, hypotension, and tachycardia.
- Treatment is supportive in nature.

PLANTS CONTAINING CARDIAC GLYCOSIDES

- Foxglove (*Digitalis purpurea*), lily-of-the-valley (*Convallaria majalis*), common oleander (*Nerium oleander*), and yellow oleander (*Thevetia peruviana*) all contain digitalis-like glycosides.
- All parts of these plants, particularly the seeds in the case of yellow oleander, are potentially toxic.
- Children with small exploratory ingestions of whole plant material from lily-of-the-valley or common oleander are unlikely to develop toxicity, but ingestion of only a few yellow oleander seeds can produce life-threatening poisoning.
- Patients may present with nausea, vomiting, dizziness, diarrhea, and abdominal pain.

- EKG findings following yellow oleander poisoning often include sinus bradycardia and conduction defects affecting the sinus or AV node.
- Hyperkalemia may also be noted. While a detectable serum digoxin level can help prove exposure, the absolute level cannot be used to guide antidotal therapy.
- Treatment with antidigoxin Fab fragments is safe and can restore sinus rhythm, correct bradycardia, and ameliorate hyperkalemia.
- Asymptomatic patients following nontrivial ingestions should have serial EKGs, repeat electrolyte determinations, and be observed for 12 hours.

PLANTS CONTAINING SODIUM CHANNEL ACTIVATORS

- Monkshood (*Aconitum* sp.), false hellebore (*Veratrum* sp.), azalea (*Rhododendron* sp.), rhododendron (*Rhododendron* sp.), and mountain laurel (*Kalmia latifolia*) all contain various toxins capable of stabilizing the open form of voltage-dependent sodium channels within the excitable membranes of neuronal and cardiac tissues.
- Significant exposures can cause vomiting, paresthesias, muscle weakness, hypotension, and seizures.
- Bradydysrhythmias, conduction blocks, and tachydysrhythmias have also been described.
- No antidote exists and therapy is supportive care driven. Intubation and circulatory support with IV fluids and vasopressors may be needed.
- Ventricular dysrhythmias necessitating antidysrhythmic therapy or electrical cardioversion may occur.
- Bradycardia or AV block may respond to atropine, although temporary pacing has been required.
- While death is rare even in adults, and large ingestions are less likely in children, pediatric cases with significant toxicity have been reported.

YEW (*TAXUS* SP.)

- The seeds and leaves of the yew plant, but not the fleshy red aril, contain the cardiotoxic alkaloids taxine A and B.
- Deaths have been rarely reported in the medical literature, typically in the setting of suicidal ingestions.
- A poison center review of >11,000, largely pediatric, yew exposures revealed that the majority of cases (92.5%) were asymptomatic and no deaths were reported.
- The most frequently encountered symptoms were GI upset (65.5%), dermal irritation (8.3%), hypotension or arrhythmias (6%), and seizures (6%).
- There is no antidote and treatment is supportive care-driven.

TOBACCO (*Nicotiana* sp.)

- Cases of significant morbidity from tobacco exposure reported in the literature result from exposure to green tobacco leaves or highly concentrated nicotine preparations.
- Most pediatric exposures involve the exploratory ingestion of cigarettes, cigarette butts, or nicotine gum. Ingestion of ≥ 2 whole cigarettes or ≥ 6 cigarette butts is more likely to result in apparent symptomatology.
- Vomiting is commonly seen and patients without spontaneous vomiting in the first hour are unlikely to have ingested a toxic amount.
- A biphasic course can be seen with initial tachycardia, mydriasis, hypertension, tremor, and seizures. This stimulatory phase may be followed by autonomic and neuromuscular blockade from persistent stimulation, resulting in fasciculations and skeletal muscle paralysis.
- Death is uncommon and usually results from respiratory arrest or cardiovascular collapse.
- Treatment is supportive, and most patients may be safely discharged after brief observation.

POISON HEMLOCK (*Conium maculatum*)

- Poison hemlock can be found throughout the United States.
- The plant contains coniine as well as other nicotinic alkaloids.
- Manifestations of toxicity are similar to those of nicotine, with an initial stimulatory phase that may include tachycardia, diaphoresis, tremor, and seizures.
- The subsequent depressant phase may involve bradycardia, hypotension, muscular paralysis, and coma.
- Initial GI symptoms are often prominent.
- Supportive care is the mainstay of therapy. While death occurs rarely, it usually results from respiratory compromise.
- Asymptomatic patients who present with a possible ingestion should be observed for 4–6 hours.

WATER HEMLOCK (*Cicuta* sp.)

- Water hemlock is thought to be the most toxic plant in North America.
- It contains cicutoxin and other toxic C_{17} -polyacetylenes throughout, with the highest concentrations in the root.
- Cicutoxin is a highly potent convulsant due to its antagonism of $GABA_A$ receptors.
- Patients may rapidly develop recurrent seizures, respiratory distress, and rhabdomyolysis following

initial GI complaints. Death may result from cardiopulmonary arrest complicating status epilepticus.

- Treatment consists of aggressive supportive care with early attention to definitive airway management and rapid escalation of anticonvulsant therapy.
- Benzodiazepines and phenobarbital should be employed, while phenytoin is unlikely to be beneficial.
- Symptomatic patients should be admitted to an intensive care unit and at least 4–6 hours of observation is indicated for all patients with suspected ingestions.

PLANTS CONTAINING TOXALBUMINS

- The castor bean (*Ricinus communis*) and rosary pea or jequirty bean (*Abrus precatorius*) contain the toxalbumins ricin and abrin, respectively.
- Less potent toxalbumins are also found in the physic nut (*Jatropha curcas*), black locust tree (*Robinia pseudoacacia*), and European mistletoe (*Viscum album*).
- Due to reported fatalities after the ingestion of small numbers (range ~2–20) of castor beans, ricin is regarded as highly toxic.
- Liberation of ricin and abrin from the seeds typically requires mastication or other mechanical preparation.
- Ricin and abrin both inhibit protein synthesis and exert pronounced effects on the GI tract.
- Vomiting and diarrhea leading to dehydration and delayed shock may occur. Castor bean ingestion has also been associated with GI bleeding and hemolysis.
- Onset of symptoms after ingestion is usually within 4–6 hours, but may take up to 10 hours.
- There is no antidote available. Symptomatic patients will require aggressive IV fluid resuscitation and may need vasopressor support.

PLANTS CONTAINING COLCHICINE

- Both the autumn crocus (*Colchicum autumnale*) and the glory lily (*Gloriosa superba*) are members of the Lily family that contain the antimetabolic agent, colchicine.
- Fatalities have been reported in modern medical literature after the mistaken ingestion of colchicine-containing plants.
- Acutely, nausea, vomiting, abdominal pain, and diarrhea may result.
- In more severe intoxications, delayed effects may be seen, including GI hemorrhage, bone marrow suppression, multiorgan failure, and cardiovascular collapse.
- There is no commercially available antidote, and prolonged supportive care may be required.

- Initial care entails circulatory support and early decontamination whenever possible, given the potential for significant morbidity and lack of an available antidote.
- Given the variable onset of symptoms (2–12 hours), all asymptomatic patients merit observation for an extended period.

ACKEE (*BLIGHIA SAPIDA*)

- Ackee fruit is a staple of the Jamaican diet and is grown in the West Indies, Florida, and Hawaii.
- The unripe fruit and seeds contain the toxins, hypoglycin A and B, which inhibit metabolic pathways and can cause profound hypoglycemia.
- The illness, which also manifests with severe GI distress and CNS derangements, is known as Jamaican vomiting sickness.
- Lethargy, metabolic acidosis without ketonemia, seizures, coma, and hepatic steatosis may be noted.
- Fatalities are more common in children, possibly due to lower liver glycogen stores and a greater tendency to hypoglycemia.
- Treatment requires hospital admission, careful attention to blood glucose levels, antiemetics, IV hydration, and symptom-driven supportive care.

PSYCHOACTIVE PLANTS

- A variety of plants and plant products have been abused by adolescents and teenagers for their hallucinogenic effects.
- The seeds of morning glory (*Ipomea violacea*), Hawaiian baby woodrose (*Argyrea nervosa*), and Hawaiian woodrose (*Merremia tuberosa*) contain lysergic acid amides and can cause effects similar to lysergic acid diethylamide (LSD).
- Peyote (*Lophophora williamsii*) is a small, spineless cactus found in the southwestern United States and northern Mexico. The tops of the cactus are sliced off and dried, forming brown “buttons” that have a high content of mescaline.
- *Salvia divinorum* is an herb native to southern Mexico that contains the potent hallucinogen, salvinorin A.
- Most cases of hallucinogen intoxication are self-limited and symptoms generally subside in 4–6 hours.
- Intoxication may cause agitation and anxiety, requiring sedation with benzodiazepines. Patients should receive supportive treatment in a quiet, nonthreatening environment.

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QUESTIONS

- Which of the following is true regarding pediatric plant exposures?
 - Pediatric plant ingestions in the United States are a common cause of serious poisoning.
 - Most pediatric plant ingestions involve children younger than 6 years of age.
 - Accurate history regarding the specific plant species involved and amount ingested is usually readily provided to the clinician.
 - Exploratory plant ingestions are uncommon in toddlers.
 - Fatalities due to plant exposures are frequently reported to the AAPCC National Poison Data System.
- Which of the following is most accurate regarding mucosal irritant plants?
 - Calcium oxalate crystals released from raphides on mastication result in damage to mucous membranes, immediate burning, and inflammation.
 - The philodendron and dumbcane do not contain raphides.
 - The clinical effects noted in philodendron and dumbcane ingestions are due to their capsaicin content.
 - The clinical effects noted in *Capsicum* species ingestions are primarily due to calcium oxalate containing raphides.
 - Capsaicin has excellent water solubility, making decontamination easy.
- A 3-year-old male presents to the emergency department after consuming two berries from an *Ilex opaca* (American holly) plant in an exploratory ingestion. Which statement most accurately describes his expected clinical course?
 - He will likely need hospitalization for further care.
 - Cardiovascular toxicity should be expected.
 - GI bleeding is likely.
 - He may experience mild GI upset.
 - Prolonged observation in the emergency department is indicated.
- A 16-year-old male is brought to the emergency department by ambulance due to altered mental status. His vital signs are: temperature 38°C, pulse 128, respiratory rate 20, and blood pressure 140/94. He is agitated, with dilated pupils and minimal axillary sweat. Which of the following is a possible toxicologic explanation?
 - Sympathomimetic toxidrome resulting from black nightshade (*S. nigrum*) ingestion.
 - Sympathomimetic toxidrome resulting from deadly nightshade (*A. belladonna*) ingestion.
 - Anticholinergic toxidrome resulting from Jimsonweed (*D. stramonium*) ingestion.
 - Cholinergic toxidrome resulting from Jimsonweed (*D. stramonium*) ingestion.
 - Cholinergic toxidrome resulting from deadly nightshade (*A. belladonna*) ingestion.
- A 5-year-old female presents to the emergency department with bradycardia after ingesting several yellow oleander (*T. peruviana*) seeds. Her EKG reveals the presence of AV conduction abnormalities. Which of the following is true?
 - Atropine is more effective than antidigoxin Fab fragments for treating significant cardiac arrhythmias due to yellow oleander poisoning.
 - Her serum digoxin level can be used to help guide antidigoxin Fab fragment dosing.
 - Antidigoxin Fab fragments would not be of value in treating any concurrent hyperkalemia.
 - Severe poisoning does not typically occur from yellow oleander ingestions, given the low levels of digitalis-like glycosides present.
 - Treatment with antidigoxin Fab fragments is safe and effective for significant cardiac arrhythmias due to yellow oleander poisoning.
- A 3-year old male is brought to the emergency department by his parents after being seen eating two cigarette butts. Which of the following is correct regarding his anticipated clinical course?
 - He will likely experience seizures.
 - He should be admitted to the hospital for further care.
 - The onset of vomiting is often delayed in serious nicotine ingestions.
 - Patients who ingest ≥ 2 whole cigarettes or ≥ 6 cigarette butts are more likely to develop symptoms.
 - Active GI decontamination is needed.
- After foraging for edible roots on a camping trip, a 15-year-old male falls ill with GI upset followed by tachycardia, diaphoresis, tremor, and seizures. By the time he is brought to the emergency departments, he is comatose, bradycardic, and hypotensive. Which of the following is most accurate?
 - His symptoms are consistent with foxglove (*D. purpurea*) poisoning.
 - His symptoms are consistent with poison hemlock (*C. maculatum*) ingestion.
 - His symptoms are consistent with pokeweed (*P. americana*) ingestion.
 - These symptoms represent an anticholinergic toxidrome caused by Jimsonweed (*D. stramonium*).
 - These symptoms are typically seen in early castor bean (*R. communis*) poisoning.

8. While on a rafting trip, a 16-year-old female begins having seizures shortly after consuming a “wild carrot.” She is evacuated to a regional medical center and is noted to have refractory seizure activity. Which of the following is correct?
- She most likely ingested water hemlock (*Cicuta* sp.).
 - She most likely ingested yellow oleander (*T peruviana*).
 - Her symptoms are consistent with pokeweed (*P americana*) ingestion.
 - Phenytoin is the first-line anticonvulsant for managing her seizures.
 - The plant she ingested contains cicutoxin, a highly potent convulsant due to its agonism of GABA_A receptors.
9. In a suicide attempt, a 17-year-old female chews and swallows more than twenty castor beans (*R communis*). Which of the following is most accurate?
- She is unlikely to develop any symptoms.
 - Ricin exerts pronounced effects on the GI tract by inhibiting microtubule polymerization and acting as a mitotic poison.
 - GI bleeding has not been reported after ricin ingestion.
 - Seeds swallowed whole are unlikely to produce toxicity.
 - Hypotension in the setting of ricin ingestion is usually due to vasodilation.
10. A 4-year old male develops vomiting, lethargy, hypoglycemia, and metabolic acidosis after being fed unripe Ackee fruit. Which of the following is true regarding his illness?
- Hypoglycemia more commonly occurs in adults.
 - Fatalities due to Jamaican vomiting sickness are more common in adults.
 - Jamaican vomiting sickness is caused by the toxins, hypoglycin A and B.
 - Hepatic steatosis has not been reported in association with Jamaican vomiting sickness.
 - Ackee fruit is not grown in the United States.
- botanical product in the 2006 AAPCC National Poison Data System annual report. Specific plant species information and accurate quantification of the exposure is frequently difficult to ascertain in plant exposure cases.
- A. Calcium oxalate containing raphides are found in multiple plants, including the philodendron, dumbcane, peace lily, pothos, caladium, calla lily, Jack-in-the-pulpit, elephant’s ear, and skunk cabbage. The clinical effects noted in philodendron and dumbcane ingestions are due to raphides. Clinical effects noted in *Capsicum* species ingestions are significantly mediated by their capsaicin content. Capsaicin is poorly water soluble and effective decontamination can be challenging.
 - D. *Ilex opaca* (American holly), along with American mistletoe (*P serotinum*, *P leucarpum*) and the poinsettia (*E pulcherrima*), are commonly encountered GI irritant plants. They generally do not cause serious poisoning and most trivial exposures do not require observation or hospital admission. Cardiovascular toxicity or GI bleeding would not be expected.
 - C. The elevated temperature, tachycardia, mydriasis, agitation, and decreased axillary sweat are consistent with an anticholinergic toxidrome. Jimsonweed (*D stramonium*), deadly nightshade (*A belladonna*), and black henbane (*H niger*) all contain anticholinergic alkaloids. The black nightshade (*S nigrum*) plant contains solanaceous alkaloids and does not cause a classic sympathomimetic toxidrome. Decreased axillary sweat, hypoactive bowel sounds, and dry mucous membranes are features that may help discriminate between the anticholinergic and sympathomimetic toxidromes.
 - E. Of plants containing digitalis-like glycosides, yellow oleander is likely responsible for the greatest number of fatalities worldwide. Ingestion of only a few yellow oleander seeds can produce life-threatening poisoning due to the relatively high content of cardiac glycosides. While the serum digoxin level can help prove exposure, the absolute level cannot be used to guide antidotal therapy. Treatment with antidigoxin Fab fragments is a safe and effective antidote for significant cardiac arrhythmias. It has been shown to restore sinus rhythm, correct bradycardia, and ameliorate hyperkalemia.
 - D. Patients who ingest ≥ 2 whole cigarettes or ≥ 6 cigarette butts are more likely to develop symptoms. Symptoms usually develop within 30–90 minutes in children after ingesting tobacco products. Vomiting is commonly seen and patients

ANSWERS

- B. Most pediatric plant ingestions involve children younger than 6 years of age. Exploratory plant ingestions are not uncommon in toddlers. However, plant exposures are neither a common cause of serious poisoning or fatalities in the United States. Only a single fatality was attributed to a plant or herbal/

without spontaneous vomiting in the first hour are unlikely to have ingested a toxic amount. Active decontamination is usually not indicated given the frequent presence of vomiting in tobacco product ingestions. While tachycardia, mydriasis, hypertension, tremor, and seizures may be seen in the stimulatory phase of nicotine intoxication, most patients can be safely discharged after brief observation.

7. B. Manifestations of poison hemlock toxicity can include an initial stimulatory phase with tachycardia, diaphoresis, tremor, and seizures. The subsequent depressant phase may involve bradycardia, hypotension, muscular paralysis, and coma. Initial GI symptoms are often prominent. Foxglove contains digitalis-like cardiac glycosides and predominantly causes cardiac arrhythmias. Pokeweed poisoning primarily causes nausea, vomiting, abdominal cramps, and diarrhea. While Jimsonweed does cause an anticholinergic toxidrome, the described clinical presentation is not consistent with an anticholinergic picture. Vomiting and diarrhea leading to dehydration and delayed shock is typically described in severe ricin poisoning.
8. A. Water hemlock is easily confused with the wild carrot (*Daucus carota*) or water parsnip (*Pastinaca sativa*). The plant contains cicutoxin and other toxic C₁₇-polyacetylenes. Cicutoxin is a highly potent convulsant due to its antagonism of GABA_A receptors. Death may result from cardiopulmonary arrest complicating status epilepticus. Benzodiazepines and phenobarbital should be employed initially, while phenytoin is unlikely to be beneficial. Yellow oleander contains digitalis-like cardiac glycosides and predominantly causes cardiac arrhythmias. Pokeweed poisoning primarily causes nausea, vomiting, abdominal cramps, and diarrhea.
9. D. Due to reported fatalities after the ingestion of small numbers (range ~2–20) of castor beans, ricin is regarded as highly toxic. However, the seeds must be chewed to liberate the ricin. Seeds swallowed whole are unlikely to produce toxicity. Ricin inhibits ribosomal protein synthesis and exerts pronounced effects on the GI tract. After oral exposure, vomiting, and diarrhea leading to dehydration, GI bleeding, and hypovolemic shock may occur.
10. C. Ackee fruit is grown in the West Indies, Florida, and Hawaii. The unripe fruit and seeds contain the toxins, hypoglycin A and B, which inhibit metabolic pathways and can cause profound hypoglycemia. Patients can develop hepatic steatosis similar to that of Reye's syndrome. Fatalities are more common in children, possibly due to lower liver glycogen stores and a greater tendency to hypoglycemia.

131 LETHAL TOXINS IN SMALL DOSES

Leon Gussow

INTRODUCTION

- There are a number of prescriptions and over-the-counter preparations that can cause extreme toxicity, even fatality, in a toddler after ingestion of a single dose. The emergency physician must be familiar with these highly toxic agents.
- This chapter, while not exhaustive, will discuss some of these dangerous overdoses.

CAMPHOR

- Camphor is present in many over-the-counter liniments and cold preparations, such as Campho-Phenique, Ben-Gay, Vicks Vaporub, Absorbine, and Tiger Balm. Camphor has long been used as an anti-pruritic, rubefacient, and antiseptic.
- Camphor has a strong, unmistakable odor and a pungent taste that some children find appealing. It is a rapidly acting neurotoxin, producing both CNS excitation and depression. As little as 1 g has been reported to cause death in a 19-month-old child.
- Clinical symptoms begin rapidly with onset 5 to 120 minutes after ingestion. Initially, a feeling of generalized warmth progresses to pharyngeal and epigastric burning. Mental status changes can follow: confusion, restlessness, delirium, and hallucinations. CNS depression with coma and hypoventilation can occur. Muscle twitching and fasciculations may herald the onset of seizures, which have also been reported to occur suddenly, without preceding symptoms.
- Management of camphor ingestion consists of supportive care with an emphasis on airway protection and seizure control. Because camphor is so rapidly absorbed gastric aspiration is unlikely to improve outcome. The use of ipecac is contraindicated. Seizures not responsive to benzodiazepines can be treated with phenobarbital.
- Asymptomatic patients should be observed for 6 hours after ingestion prior to discharge from the emergency department.

BENZOCAINE

- Benzocaine is present in many local anesthetics, including first aid ointments and infant teething formulas such as Baby Orajel and Americaine Topical

Anesthetic First Aid Ointment. Exposure can be from oral ingestion or dermal absorption.

- Benzocaine is metabolized to aniline and nitrosobenzene, both of which can cause methemoglobinemia, especially in infants younger than 4 months, who are deficient in methemoglobin reductase.
- Methemoglobinemia has occurred in an infant after an ingestion of 100 mg of benzocaine, the amount in one-quarter teaspoon of Baby Orajel.
- Clinical signs and symptoms typically begin within 30 minutes to 6 hours after ingestion, with tachycardia, tachypnea, and a characteristic cyanosis that does not respond to oxygen. In more severe exposures, agitation, hypoxia, metabolic acidosis, lethargy, stupor, and coma may supervene. Seizures can also occur.
- Treatment of benzocaine exposure consists principally of general support and, in selected cases, the administration of antidote. Neither gastric lavage nor administration of activated charcoal is mandatory in this setting, since they have not been demonstrated to improve clinical outcome.
- The antidote for patients with methemoglobinemia is methylene blue. Indications for use include methemoglobin levels >30% or signs of respiratory distress or altered mental status. The dose—1 to 2 mg/kg IV over 5 minutes—is repeated in 1 to 2 hours if symptoms persist. Isolated cyanosis is not an indication for methylene blue, because it often occurs at low methemoglobin levels, is usually well tolerated, and resolves spontaneously.

LOMOTIL

- Lomotil is an antidiarrheal preparation that combines an opiate (diphenoxylate) with an anticholinergic (atropine).
- Respiratory depression can recur as late as 24 hours after Lomotil ingestion. There appears to be no correlation between the dose ingested and the severity of symptoms.
- Any child with known or suspected ingestion of any amount of Lomotil is admitted and monitored for at least 24 hours, no matter what the initial clinical condition.
- Although patients often present with a confusing mix of opioid and anticholinergic signs and symptoms, opioid effects are always seen in overdose and often predominate. Atropine-induced anticholinergic symptoms can occur before, during, or after opioid effects, or may not occur at all.
- Initial manifestations of Lomotil overdose in children include drowsiness, lethargy or excitement, dyspnea, irritability, miosis, hypotonia or rigidity, and urinary

retention. In severe cases the patient may present with coma, respiratory depression, hypoxia, and seizures.

- Management of Lomotil poisoning includes admission of all patients and careful observation for 24 hours. Neither induced emesis nor gastric lavage is indicated.
- Respiratory depression and coma are treated with intravenous naloxone (0.1 mg/kg), which may have to be repeated frequently. A maintenance dose of naloxone can be given, starting with two-thirds of the bolus dose that initially produced the desired response administered each hour, titrated to clinical condition. When naloxone is given, anticholinergic symptoms may emerge.

CHLOROQUINE AND HYDROXYCHLOROQUINE

- Chloroquine has been used since the 1940s for the treatment and prevention of malaria. Additional uses currently include the treatment of extraintestinal amebiasis and some connective tissue diseases such as SLE and rheumatoid arthritis.
- Chloroquine is a powerful rapidly acting cardiotoxin capable of causing sudden cardiopulmonary collapse. The interval between ingestion and cardiac arrest is often less than 2 hours. Even a small amount can be life threatening in a toddler.
- Chloroquine causes myocardial depression and vasodilation, producing sudden profound hypotension. Automaticity and conductivity of myocardium are also impaired, resulting in bradycardia and ventricular escape rhythms. The electrocardiogram can show sinus bradycardia, widened QRS, prolonged intraventricular conduction time, T-wave changes, ST depression, prolonged QT, complete heart block, ventricular tachycardia, or ventricular fibrillation. Neurotoxicity induced by chloroquine often presents as drowsiness and lethargy, followed by excitability. Seizures and coma can occur.
- The physician treating chloroquine toxicity should be prepared to manage sudden cardiac or respiratory arrest. Intubation and supported ventilation may be required. Blood pressure is maintained with intravenous fluids and pressors. Class IA antiarrhythmics (quinidine, procainamide, disopyramide) are contraindicated. Because chloroquine is rapidly absorbed from the gastrointestinal tract, gastric lavage is unlikely to improve clinical outcome. Chloroquine is well absorbed by activated charcoal, which may be considered if it can be administered within 30 to 60 minutes of ingestion. As with theophylline toxicity, hypokalemia can occur because of ion transport into

cells. Aggressive repletion of potassium in this setting has in some cases led to severe hyperkalemia. Recent evidence suggests that early mechanical ventilation with administration of high-dose diazepam and epinephrine may be lifesaving in severe cases of chloroquine toxicity in adults. However, this therapy has not been well described in the pediatric population.

- Any child who has ingested chloroquine should be referred immediately to a medical facility for observation and cardiac monitoring. If no coingestants are involved, a patient who is asymptomatic for 6 hours can be safely discharged after consideration of the social situation and potential for repeat exposure. Any symptoms or EKG changes require admission.
- There is scant literature concerning hydroxychloroquine overdose in children. Although considered less toxic than chloroquine, hydroxychloroquine has similar cardiac and neurologic effects. The most prudent course would be to handle hydroxychloroquine ingestion with a similar approach to that outlined above for chloroquine.

METHYL SALICYLATE

- Methyl salicylate is a concentrated liquid that is absorbed quickly and can produce early-onset severe salicylate toxicity. It is found in many topical liniments (Ben Gay, Icy Hot Balm) and in oil of wintergreen food flavoring.
- One teaspoon of oil of wintergreen contains 7 g of salicylate (equivalent to 21 aspirin tablets).
- Since ingestion of less than one teaspoon has killed a child, any ingestion of these preparations is potentially serious. Clinical presentation and treatment of this overdose is similar to that of other types of salicylate poisoning.

IMIDAZOLINE DECONGESTANTS

- Imidazoline decongestants are found in a wide variety of over-the-counter nasal sprays and eye drops.
- Imidazolines are α_2 adrenergic agonists. When ingested, they cause opioid-like effects: hypotension, bradycardia, miosis, CNS depression, and respiratory depression.
- Because of the interplay between the central and peripheral actions of the imidazolines, overdose can present with a variable and changing clinical picture. Bradycardia can alternate with tachycardia, hypotension with hypertension, and lethargy with agitation. Hypoglycemia and hypothermia have both been reported.

- Imidazoline-induced CNS depression may respond to naloxone. Clinically significant bradycardia can be treated with atropine. Asymptomatic children should be observed for 6 hours.

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QUESTIONS

1. A 16-month-old male is brought to the ED after ingesting Bengay and presents confused and restless. Which of the following are true concerning camphor toxicity?
 - A. Seizures are always preceded by muscle twitching and fasciculations.
 - B. The initial treatment for camphor-induced seizures is a benzodiazepine.
 - C. Resistant seizures induced by camphor can be treated with phenytoin.
 - D. Children who ingest camphor must be observed for a minimum of 24 hours.
 - E. Induced emesis with ipecac is recommended if a child presents within 1 hour of ingesting camphor.
2. A 12-month-old female is brought to the ED by a baby sitter after ingesting Baby Orajel. She is tachycardic, tachypneic, and cyanotic. Which of the following is true concerning benzocaine toxicity?
 - A. Children under 4 months of age are relatively resistant to the development of methemoglobinemia.
 - B. Cyanosis that responds to supplemental oxygen is characteristic.

- C. A child who presents within 1 hour of benzocaine ingestion should be given a dose of activated charcoal.
 - D. Treatment with the antidote methylene blue is not required if the methemoglobin level is less than 30%.
 - E. Cyanosis without other symptoms is not an indication for methylene blue.
3. Indications for hospital admission for possible Lomotil toxicity include:
- A. Known ingestion of more than 1 cc of Lomotil.
 - B. Suspected ingestion of more than 1 cc of Lomotil.
 - C. Lethargy.
 - D. Drowsiness.
 - E. All of the above.

ANSWERS

- 1. B. After ingestion of camphor, seizures can occur suddenly without being heralded with twitching or fasciculations. Camphor is rapidly absorbed and manifestations of toxicity usually occur within 2 hours; a child who is asymptomatic after 6 hours of observation in the emergency department may be discharged. Ipecac is no longer recommended for any ingestion; it is an especially bad idea after camphor exposure, since seizures may occur suddenly. The initial drug of choice for camphor-induced seizures is a benzodiazepine; as a general rule, phenytoin is not effective against toxin-induced seizures.
- 2. E. Children under 4 months of age are unusually susceptible to developing benzocaine-induced methemoglobinemia, since they are deficient in methemoglobin reductase. Cyanosis that does not respond to supplemental oxygen is characteristic of methemoglobinemia. Treatment with activated charcoal is never mandatory, and has never been proven to improve clinical outcomes in these patients. Children with significant symptoms from methemoglobinemia such as respiratory distress or altered mental status should be treated with the antidote methylene blue even if the methemoglobin level is less than 30%. Cyanosis alone is not an indication for the antidote.
- 3. E. Any child with a known or suspected ingestion of Lomotil should be observed for 24 hours, even if asymptomatic.

132 CHEMICAL WEAPONS: NERVE AGENTS AND VESICANTS

Leon Gussow

- Children are even more susceptible than adults to the effects of chemical weapons, especially those that are inhaled or absorbed through the skin. A child's surface area relative to his or her body mass is greater than that of an adult, increasing dermal absorption of chemical agent. In addition, the child's higher metabolic and respiratory rates increase the uptake of inhaled vapors. Finally, metabolic pathways that detoxify or eliminate poisons may be underdeveloped.

NERVE AGENTS

- Nerve agents, highly toxic organophosphate compounds, are essentially much more potent versions of the organophosphate insecticides. Nerve agents that have been manufactured and stockpiled in the past include tabun, sarin, and soman. VX is the most potent of these agents, with a potentially fatal liquid exposure involving as little as one drop applied to the skin of an adult; this lethal amount would be proportionally much less in children.
- Nerve agents are powerful inhibitors of the enzyme acetylcholinesterase (AChE). This enzyme normally serves to modulate the actions of acetylcholine, a neurotransmitter that is found throughout the peripheral and central nervous systems. With the enzyme blocked, acetylcholine accumulates and the cholinergic receptors become overstimulated in an uncontrolled manner. There are three major classifications of cholinergic receptors and actions: 1 *Muscarinic*: cause increased secretion from glands and contraction of involuntary smooth muscle. Major clinical muscarinic effects are bronchorrhea and bronchospasm. 2 *Nicotinic*: found at the neuromuscular junction of voluntary skeletal muscle. Major clinical nicotinic effects include fasciculation, muscle weakness, and paralysis. 3 *Central*: Hyperstimulation of cholinergic receptors in the brain causes seizures, coma, and central apnea.
- Bronchorrhea and bronchospasm, respiratory muscle weakness, and central apnea can all lead to death from respiratory failure.
- There are three antidotes for significant nerve agent exposure, one dealing with each of three types of cholinergic receptors: 1 *Muscarinic*: the antimuscarinic agent *atropine* will dry up respiratory secretions and

relieve bronchospasm, allowing the patient to be ventilated and oxygenated. The initial dose is 0.05 to 0.10 mg/kg IV or IM (maximum 5 mg). This can be repeated every 5 minutes until ventilation and oxygenation are possible. 2 *Nicotinic: Pralidoxime chloride (2-PAM)* removes nerve agent from the active site on AChE) reactivating the enzyme. This helps reverse the *nicotinic* effects of nerve agents such as muscle weakness. 3 *Central: Diazepam* is indicated not only to treat nerve agent-induced seizures, but also for any severe exposure with *central* manifestations.

SULFUR MUSTARD

- The blistering agent *sulfur mustard* is an oily liquid with an odor similar to that of garlic, horseradish, or mustard.
- Mustard quickly damages cellular components such as nucleic acids and proteins, causing cell death. Rapidly dividing cells, such as the hematopoietic components of bone marrow and the linings of the gastrointestinal and respiratory tracts, are most sensitive to mustard's effects.
- Clinical effects are dose dependent. At low doses, skin irritation and blistering occurs; at higher doses, systemic toxicity can also be seen. While tissue damage occurs within minutes of exposure, initial signs and symptoms are typically delayed for several hours.
- Skin findings include erythema that progresses to blister formation over 24 hours. Warm, moist areas such as the axillae and groin are particularly susceptible.
- Ocular manifestations are similarly delayed and include lid edema, conjunctival injections, and, with severe exposure, corneal ulceration.
- Respiratory involvement begins with sore throat, cough, and hoarseness, and can in rare instances of overwhelming exposure progress to pulmonary edema. Early tachypnea or dyspnea suggests a poor prognosis.
- Treatment for exposure to mustard begins with skin and eye decontamination. Although tissue damage occurs almost immediately upon contact with mustard liquid or vapor, late decontamination is still indicated to minimize systemic absorption and to prevent secondary contamination of rescue or medical personnel. Copious irrigation with water and a mild detergent constitutes appropriate dermal decontamination.
- There are no specific antidotes for sulfur mustard exposure. In contrast to thermal burns, mustard injury does not generally cause massive fluid loss. Death most frequently occurs 5 to 10 days after exposure, usually from pulmonary insufficiency or infection.

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QUESTIONS

1. Multiple children are brought in to your ED after a suspected nerve agent exposure while on the subway. Which of the following would you *not* anticipate from the muscarinic effects of nerve agents?
 - A. Salivation
 - B. Tearing
 - C. Diaphoresis
 - D. Increased upper airway resistance
 - E. Skeletal muscle paralysis
2. You are working in a refugee camp when several children are brought in after exposure to sulfur mustard. Which of the following statements is correct concerning mustard vesicant agents in these exposed patients?
 - A. Cellular damage is delayed for several hours after sulfur mustard contacts the skin.
 - B. Moist areas of the axilla and groin are relatively resistant to mustard-induced injury.
 - C. Although mustard agents cause extreme eye discomfort, they never cause permanent injury.
 - D. There is no specific antidote for sulfur mustard exposure.
 - E. Since damage from sulfur mustard occurs immediately upon contact, external decontamination of exposed victims is not necessary.

ANSWERS

1. E. Muscarinic receptors are found where autonomic nerves connect to secretory glands and involuntary smooth muscle. Therefore, the cholinergic nerve agents will cause increased secretions and uncontrolled contraction of muscles in the gut and respiratory tract. The muscarinic effects of nerve agents can

be remembered using the mnemonic *SLUGBAM*: Salivation, Lacrimation, Urination, Gastrointestinal emptying (vomiting and diarrhea), Bronchospasm and bronchorrhea, Abdominal pain, and Meiosis. Skeletal muscle paralysis is a nicotinic effect of nerve agents.

2. D. Damage from exposure to sulfur mustard occurs immediately upon contact; however, dermal manifestations of this damage may be delayed for several

hours. Moist areas such as the axilla and groin are particularly susceptible to injury. Although rare, ocular contact with sulfur mustard can result in permanent injury and visual impairment. External decontamination of victims of sulfur mustard exposure is critical to prevent spread of the agent to other parts of the body, as well as secondary contamination of health care workers. There is no specific antidote for sulfur mustard exposure.

133 **HUMAN AND ANIMAL BITES**

David A. Townes

EPIDEMIOLOGY

- Estimated two million bite wounds are reported each year in the United States.
- Bite wounds account for approximately 1% of all emergency department visits and result in numerous hospitalizations.
- Bite injuries are clustered in the summer months.
- Dogs account for the majority of bites, followed by cats, with the remainder divided among a variety of animal species.
- More than half of these injuries occur in children.
- Boys tend to be bitten more often than girls.

HISTORY AND PHYSICAL EXAMINATION

- Proper management of bite wounds begins with a thorough history including what type of animal caused the wound and the age of the wound.
- One must also elicit host factors that may affect wound healing, especially a history of diabetes, peripheral vascular disease, chronic use of glucocorticoids or other immunocompromised states.
- Physical examination should include a full examination and exploration of the wound noting the type of wound (laceration, crush, or puncture) and the extent of involvement of deep structures.
- If the wound occurs over a joint, the joint should be examined through the full range of motion.
- When appropriate, radiographs should be obtained to look for fractures, foreign bodies and air in the joint or soft tissues.

- Computed tomography of the head should be considered in dog bites to the scalp.
- Careful attention should be paid to signs of infection, including erythema, swelling, discharge, lymphadenopathy, or pain on passive range of motion.

WOUND CARE

- The most common complication associated with bite wounds is wound infection.
- While there is some variability among studies, infection rates as high as 30% for dog bites, 80% for cat bites and 60% for human bites have been reported.
- One of the best methods of reducing the risk of infection is adequate irrigation of the wound. An acceptable method is to irrigate the wound with 1 to 2 L of normal saline through a 19- or 20-gauge vascular catheter or similar irrigation device.
- The wound should be debrided as needed.
- The decision to close the wound depends on its age, type, and location. Under no circumstances should a bite wound that appears infected be closed.
- In most cases, bite wounds on the hand should be left open because of the high potential for morbidity if these wounds become infected.
- Dog bites may usually be safely closed if they are not located on the hand.
- Cat bites, which are usually puncture wounds, should not be closed because they cannot be adequately cleaned. Cat bites that are lacerations rather than puncture wounds may usually be closed if they are not on the hand. Extending puncture wounds to better irrigate them remains controversial and should be decided on an individual basis.
- In general, bite wounds that are more than 8 to 12-hours-old should be left open. Potentially disfiguring bite wounds on the face may be closed even when more than 12 hours old; however, these

patients must be followed very carefully for evidence of infection.

- All bite wounds treated on an outpatient basis should be reevaluated within 48 hours.

ANTIBIOTICS

- Bite wounds that have evidence of infection should be treated with antibiotics.
- The use of prophylactic antibiotics remains controversial and the type of animal, location of the wound and host factors must be considered.
 - Bite wounds on the hands and feet should generally be treated with prophylactic antibiotics, while those on the face and scalp do not generally require treatment with prophylactic antibiotics, as they are less likely to become infected.
 - Bite wounds caused by cats and humans should generally be treated with prophylactic antibiotics, while those caused by dogs and rodents may not need prophylactic treatment.
 - If the decision to treat with prophylactic antibiotics is made, the initial treatment should be for 3 to 7 days, at the end of which, if there is no evidence of infection, the wound is very unlikely to become infected and the antibiotics may be discontinued.
- The organisms responsible for bite wound infections are often from the animal's oral flora but may also include the host's skin flora. Approximately one-third of bite wound infections demonstrate multiple organisms.
 - Dog bites tend to become infected with *Staphylococcus aureus*, *Streptococcus* species and *Pasteurella canis*, but *Pseudomonas* species, *Enterobacter cloacae* and many others have been identified.
 - Cat bites are likely to become infected with *Pasteurella multocida*.
 - Human bite wounds tend to become infected with *S aureus*, *Streptococcus* species, and *Eikenella corrodens*.
- *Pasteurella* species cause a rapidly developing infection with signs and symptoms apparent in less than 24 hours. *Pasteurella* species are covered by penicillin, ampicillin, amoxicillin, amoxicillin/clavulanic acid, second- and third-generation cephalosporins, doxycycline, trimethoprim-sulfamethoxazole, clarithromycin, and azithromycin. First-generation cephalosporins, dicloxacillin and erythromycin have poor activity against *Pasteurella* species.
- Delay in signs and symptoms of infection for more than 24 hours should lead the physician to consider other etiologic agents such as *Staphylococcus* or *Streptococcus*

species. In the case of dog and cat bites where *Pasteurella* species is not suspected, *Staphylococcus* and *Streptococcus* species may be covered by dicloxacillin or a first-generation cephalosporin. Due to the increasing incidence of methicillin-resistant *S aureus* (MRSA), consideration may also be given to using trimethoprim-sulfamethoxazole or clindamycin.

- Empiric therapy for dog and cat bites should include a β -lactam antibiotic and a β -lactamase inhibitor, a second-generation cephalosporin with anaerobic activity or a combination of penicillin and a first-generation cephalosporin.
- In the case of human bite wounds, penicillin or amoxicillin/clavulanic acid can be used to cover *Eikenella* and dicloxacillin can be used to cover *Staphylococcus* and *Streptococcus*.
- Amoxicillin/clavulanic acid is generally considered both first line prophylaxis and treatment for bite wounds. Initial treatment for all infected bite wounds should be 10 to 14 days. It may be necessary to use a two-antibiotic regimen for bite wounds in patients who cannot tolerate certain antibiotics.

RABIES PROPHYLAXIS

- Rabies is a viral infection transmitted in the saliva of infected animals and may lead to encephalomyelitis. It is almost universally fatal and accounts for an estimated 55,000 deaths worldwide annually.
- Rabies deaths in the United States are rare due in part to the relatively low prevalence and widespread availability of postexposure prophylaxis.
- In determining the need for rabies postexposure prophylaxis, the physician must consider the type of animal causing the injury and the prevalence of rabies in the region.
- In the case of bats, postexposure prophylaxis is indicated for any bite, scratch, or mucous membrane exposure if the animal cannot be collected and tested. Postexposure prophylaxis may be indicated in cases in which contact is likely to have occurred but is not documented including a child sleeping in a room where a bat is found.
- In the case of dogs, cats and ferrets, the animal should be captured and quarantined for 10 days.
 - If the quarantined animal remains healthy, no treatment is necessary.
 - If the quarantined animal becomes ill or if rabies is suspected, the animal should be sacrificed and the brain examined for evidence of rabies. If the quarantined animal is infected, the child should be immediately vaccinated.

- If the animal cannot be located, decisions regarding prophylaxis must be based on the prevalence of rabies in the area and the species of biting animal.
- Most other carnivores including skunks, raccoons and foxes should be considered infected unless proven negative by laboratory testing. Livestock, large rodents, lagomorphs and other mammals should be considered on an individual case basis.
- Postexposure prophylaxis includes cleaning of the wound, administration of one of the two available licensed vaccines, and administration of human rabies immune globulin (HRIG).
 - As of 2010, the recommendation for the schedule of postexposure vaccine administration in the United States has changed from the previously recommended five 1-mL intramuscular injections on day 0, 3, 7, 14, and 28 to four 1-mL intramuscular injections on day 0, 3, 7, and 14. This new recommendation is in conflict with the current package insert for both licensed vaccines in the United States and with some international recommendations.
 - The previously recommended five 1-mL intramuscular injections on days 0, 3, 7, 14, and 28 are still recommended for immunocompromised patients.
 - Vaccine should be administered in the deltoid or anterolateral thigh and should not be administered in the gluteal area.
 - If feasible, the entire volume of HRIG should be infiltrated into the wound and the surrounding area with any remaining volume administered intramuscularly at a site remote from the vaccine administration.
 - The same syringe should not be used to administer the vaccine and the HRIG.
- The World Health Organization (WHO) has promoted the use of pre-exposure prophylaxis in toddlers and children living in highly endemic areas.
 - Pre-exposure prophylaxis includes three doses of HDCV administered on days 0, 7, 21, or 28.
 - Postexposure prophylaxis in individuals who received pre-exposure prophylaxis includes the administration of two doses of HDCV on days 0 and 3, but no administration of HRIG, as it is not indicated in individuals who have received pre-exposure prophylaxis.

TETANUS PROPHYLAXIS

- Tetanus immunoprophylaxis should also be administered in patients with bite wounds if not current.

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QUESTIONS

1. Comparing dogs and cats, which of the following is true?
 - A. Cats account for more bite wounds but fewer bite wound infections.
 - B. Cats account for more bite wounds and more bite wound infections.
 - C. Dogs account for more bite wounds but fewer bite wound infections.
 - D. Dogs account for more bite wounds and more bite wound infections.
 - E. Bites wounds and bite wound infections are equal among dogs and cats.
2. A 5-year-old male presents after being bitten by a stray dog. He has a 2 cm × 5 mm linear laceration to his posterior calf with no active bleeding. The best method to reduce the likelihood of a wound infection is
 - A. Administer prophylactic antibiotics
 - B. Irrigation of the wound with 1 to 2 liters of normal saline through a 19- or 20-gauge vascular catheter or similar irrigation device.
 - C. Administer rabies vaccine.
 - D. Administer HRIG.
 - E. Administer tetanus immunoprophylaxis.
3. A 6-year-old female presents to your ED with several superficial lacerations to her hand and a small puncture wound after being bitten by a cat.

Which of the following are true regarding wounds from *Pasteurella* species?

- A. Generally cause a rapidly developing infection with signs and symptoms apparent in less than 24 hours.
 - B. Are common in cat bite wound infections but rare in dog bite wound infections.
 - C. Are sensitive to first-generation cephalosporins.
 - D. Are common in human bite wound infections.
 - E. Generally cause a slowly developing infection with signs and symptoms apparent in greater than 24 hours.
4. A healthy 10-year-old male presents after being bitten by a bat trapped in the attic of his home. According to the most recent guidelines for immunocompetent patients in the United States, rabies postexposure prophylaxis would include which of the following for this patient?
- A. Administration of the vaccine in five 1-mL intramuscular injections on day 0, 3, 7, 14, and 28 and administration HRIG.
 - B. Administration of the vaccine in five 1-mL intramuscular injections on day 0, 3, 7, 14, and 28 without administration HRIG.
 - C. Administration of the vaccine in four 1-mL intramuscular injections on day 0, 3, 7, and 14 without administration HRIG.
 - D. Administration of the vaccine in four 1-mL intramuscular injections on day 0, 3, 7, and 14 and administration HRIG.
 - E. Administration of a single 1-mL intramuscular injection of vaccine along with HRIG.
5. A 16-year-old female is traveling to a high-risk area for rabies in Africa with her parents and receives pre-exposure prophylaxis as recommended by the CDC. During her travels in Africa, she is bitten by a stray dog. Rabies postexposure prophylaxis in this patient who had pre-exposure prophylaxis includes which of the following?
- A. Administration of four doses of the vaccine on day 0, 3, 7, and 14, without administration HRIG.
 - B. Administration of two doses of vaccine on days 0 and 3, and administration of HRIG.
 - C. Administration of HRIG only.
 - D. Administration of four doses of the vaccine on day 0, 3, 7, and 14, and administration HRIG.
 - E. Administration of two doses of vaccine on days 0 and 3, without administration of HRIG.

ANSWERS

1. C. Dogs account for the majority of bite wounds seen in the emergency department followed by cats with the

remainder divided among other animal species. Bites are clustered in the summer months with boys bitten more often than girls. Cat bite wounds tend to become infected at a higher rate than dog bite wounds.

2. B. While certain bite wounds will require administration of prophylactic antibiotics, rabies vaccine and HRIG, the best way to reduce the incidence of bite wound infection is cleaning of the wound by copious irrigation.
3. A. *Pasteurella* species are found in both cat and dog bite wound infections. It is not a common pathogen in human bite wound infections. It causes a rapidly developing infection with signs and symptoms apparent in less than 24 hours. Delay in signs and symptoms of infection for more than 24 hours should lead the physician to consider other etiologic agents such as *Staphylococcus* or *Streptococcus* species. First-generation cephalosporins, dicloxacillin and erythromycin have poor activity against *Pasteurella* species.
4. D. In 2010, the advisory committee on immunization practices (ACIP) released new recommendations for rabies postexposure prophylaxis that decreased the number of 1-mL intramuscular injections from five, on day 0, 3, 7, 14, and 28 to four, on day 0, 3, 7, and 14. Recommendations for the administration of HRIG were not changed from the previous recommendations.
5. E. Rabies pre-exposure prophylaxis includes three doses of vaccine administered on days 0, 7, 21, or 28. In individuals who have received appropriate pre-exposure prophylaxis, postexposure prophylaxis consists of administration of two doses of vaccine on days 0 and 3, without administration of HRIG.

134 SNAKE ENVENOMATIONS

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EPIDEMIOLOGY

- Venomous snakes are classified into two families: Viperidae and Elapidae.
- Crotalinae is a subfamily of Viperidae, better known as pit vipers, due to the heat sensing organs on either side of the head.
 - The Crotalinae subfamily includes three genera: *Crotalus* (rattlesnakes), *Agkistrodon* (copperheads and cottonmouths), and *Sistrurus* (massasaugas).

- Copperhead (*Agkistrodon contortrix*) and cottonmouth (*Agkistrodon piscivorus*) snakes are primarily found in the southern and eastern US.
- It is estimated that 8,000 crotaline envenomations occur in the United States annually. Approximately 20% of these involve patients under 20 years of age with 5–6 annual fatalities.

PIT VIPERS

ANATOMY

- Pit vipers possess a triangular or arrow-shaped head, whereas nonpoisonous North American snakes have a smooth, tapered body and narrow head.
- Crotalids have facial pits between the nostril and eye that serve as heat and vibration sensors, enabling the snake to locate prey.
- While nonpoisonous snakes typically possess round pupils, pit vipers have vertical or elliptical pupils.
- Members of the genus *Crotalus* are further characterized by tail rattles and a single row of ventral anal scales.

PATHOPHYSIOLOGY

- Since snakes are defensive animals and rarely attack, they will remain immobile or even attempt to retreat if given the opportunity.
- Bites most commonly occur in small children who are paralyzed with fear or in individuals who handle and harass the snake. Because of their small body weight, infants and young children are relatively more vulnerable to severe envenomation.
- The severity of envenomation also depends on the location of the bite.
 - Bites on the head, neck, or trunk are more severe than extremity bites.
 - Direct envenomation into an artery or vein is associated with a much higher mortality rate.
- When envenomation occurs, the smaller pediatric patient is generally exposed to a larger milligram per kilogram venom load.
- The basic principles of treatment do not change depending on the child's age or developmental stage.
- Intravenous antivenom is always the first-line therapy and dosing should be targeted toward the potential venom load, as opposed to the patient's kilogram weight.
- Venom is a complex mixture of enzymes that primarily function to immobilize, kill, and digest the snake's prey.
 - Proteolytic enzymes cause muscle and subcutaneous necrosis due to a trypsin-like action.
 - Hyaluronidase decreases the viscosity of connective tissue.

- Phospholipase provokes histamine release from mast cells.
- Thrombin-like amino acid esterases act as defibrinating anticoagulants.
- The major toxic effects occur within the surrounding tissue, blood vessels, and blood components.

CLINICAL PRESENTATION

- Local cutaneous changes classically include 1 or 2 puncture marks with pain and swelling at the site, while nonvenomous snakes usually leave a horseshoe-shaped imprint of multiple teeth marks.
- Approximately 25% of all pit viper bites are considered “dry bites” resulting in no toxicity.
- With severe envenomation, swelling, and edema may involve the entire extremity within one hour. Ecchymosis, hemorrhagic vesicles, and petechiae may appear within several hours.
- Systemic signs and symptoms include paresthesias of the scalp, periorbital fasciculations, weakness, diaphoresis, nausea, dizziness, and a “minty” or metallic taste in the mouth.
- Severe bites can result in coagulopathies, thrombocytopenia, and disseminated intravascular coagulation (DIC).
- Rapid hypotension and shock, with pulmonary edema and renal and cardiac dysfunction, can also result, particularly if the victim suffers a direct intravenous envenomation.

PREHOSPITAL MANAGEMENT

- The victim's extremity should be immobilized and physical activity minimized.
- To maintain renal flow and intravascular volume, oral fluids are vigorously administered.
- Overaggressive first aid measures can be dangerous and may exacerbate limb morbidity.
 - Incision and suction of the bite wound with the human mouth may result in tissue damage or infection.
 - Mechanical suction devices exist and have been anecdotally reported to remove up to one-third of the venom if used immediately after envenomation, but no well-designed human clinical trials exist which support their use.
 - Cryotherapy can lead to further wound necrosis and is not currently recommended.
 - Electric shock therapy as a first aid treatment of snakebites is not recommended.
- Optimal therapy consists of placing the patient at rest with the affected extremity raised to the cardiac level.

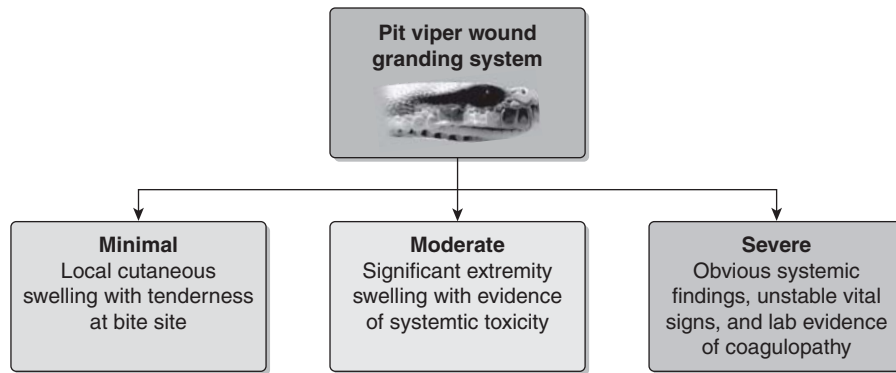


FIG. 134-1. Pit viper wound grading system.

- Emergency evacuation should be arranged as quickly as possible for transport to the closest facility with access to antivenom therapy.
- The wound site should be measured and leading edges marked, so that symptom progression can be judged upon hospital arrival.
- Intravenous access is obtained if possible and narcotic analgesics administered.
- Crotaline snakebite wounds are generally graded as minimal, moderate, or severe based on the degree of envenomation, which can ultimately guide therapy (Fig. 134-1).

DIAGNOSTIC STUDIES

- Laboratory studies recommended with rattlesnake bites include a complete blood count including platelets, prothrombin time, or international normalized ratio (INR), partial thromboplastin time, fibrinogen level, and fibrin degradation products.
- Abnormal hematologic parameters are considered evidence of systemic toxicity and should be incorporated along with clinical examination into the decision to administer antivenom.
- If initial laboratory testing is normal and minor local tissue swelling and pain are present, it is acceptable to reevaluate these hematologic laboratory values in 2 to 6 hours.
- Other laboratory parameters, such as chemistry panels, creatinine phosphokinase (CPK), and urinalysis, should be monitored if there is evidence of rhabdomyolysis, myoglobinuria, or renal insufficiency.

HOSPITAL MANAGEMENT

- Patients presenting after rattlesnake envenomation should be given tetanus prophylaxis if indicated.

- Affected extremities should be elevated to the level of the heart and any previously placed constriction bands or wraps removed.
- Intravenous access in an unaffected extremity should be established for the delivery of antivenom as well as analgesic medications.
- The liberal use of narcotic agents is often necessary to control pain.
- Prophylactic antibiotics are generally not recommended since rattlesnake venom possesses its own bacteriostatic properties. With evidence of infection or a history of human mouth suction to the wound, initiation of a first-generation cephalosporin or amoxicillin-clavulanate is indicated.
- Prophylactic fasciotomy and digital dermatomy for crotaline snakebite treatment are discouraged and rarely indicated. A true compartment syndrome is unlikely following rattlesnake envenomation, since rattlesnake strikes generally place venom subcutaneously, not subfascially. The preferred treatment for significant limb swelling is intravenous antivenom. Surgical therapy should only be considered in cases where elevated compartment pressures have been well documented despite aggressive intravenous antivenom therapy.
- Surgical debridement of devitalized tissues or amputation of necrotic digits may become necessary but should be delayed until complete wound stabilization.

ANTIVENOM THERAPY

CROTALIDAE POLYVALENT ANTIVENIN

- Equine-derived crotaline polyvalent antivenin is an older whole IgG preparation for treatment of rattlesnake envenomation in North and South America and is still available in many hospitals.

- It is effective against envenomations from rattlesnakes, cottonmouths, copperheads, fer-de-lance, cantiles, and South American bushmasters.
- The amount of antivenin administered depends on the severity of the envenomation. In general, if the envenomation is rated as minimal, 5 vials are routinely administered; in moderate cases, 10 vials are used; and in severe cases 15 to 20 vials are administered.
- The amount of antivenin administered can also vary depending on the species and geographic distribution of the snake.
- In comparison with adults, pediatric patients are given proportionately more antivenin, since children receive a greater amount of venom per kilogram of body weight.
- Polyvalent antivenin is most efficacious if given within 4 to 6 hours of the bite. It is of less value if delayed for 8 hours, and is of questionable value after 24 hours.
- Prior to any antivenin administration, skin testing may be performed with dilute horse serum (usually included in the antivenin kit), given subcutaneously.
- In the setting of a severe envenomation, patients with positive skin reactions can still receive the antivenin, although close monitoring for anaphylaxis and pretreatment with diphenhydramine and glucocorticoids, as well as epinephrine, may be required.
- Serum sickness, a flu-like syndrome with fever, malaise, arthralgias, lymphadenopathy, rash, pruritus, and urticaria, usually develops 10 to 20 days after antivenin administration, with symptoms proportionate to the number of vials given. It is generally self-limited and effectively treated with antihistamines and a short course of methylprednisolone.

CROTALINE FAB ANTIVENOM

- Ovine (sheep serum) preparation that is highly purified and consists of only the smaller Fab antibody fragments.
 - Crotaline Fab is equally effective and safer than the older polyvalent antivenin product resulting in a significant reduction in the rates of allergic reaction.
 - However, in the event that Fab antivenom is not available, therapy with the traditional equine formulation may still be instituted.
 - As with the older antivenin, the dosing is based on venom load as opposed to the kilogram weight of the patient.
 - Patients with envenomation symptoms should initially receive 4–6 vials of Crotaline Fab regardless of the child's size. Symptoms should be reassessed hourly and antivenom redosed at 2–4 vials until symptoms have stabilized or improved.
- While the severity of side effects associated with the new crotaline Fab antivenom is much lower than that of equine-based antivenom, patients should still be observed closely for anaphylactoid reactions. The incidence of serum sickness is low.
 - Most reactions can be easily treated by slowing the infusion rate and administering intravenous diphenhydramine.

DISPOSITION

- Asymptomatic patients presenting after a crotaline strike should be observed for a minimum of eight hours following the injury. If no symptoms or signs of envenomation develop, the patient may be safely discharged with the diagnosis of a “dry” (nonenvenomated) bite.
- One exception to this rule would include patients with envenomation by a Mojave rattlesnake (*Crotalus scutulatus*). These snakes have been associated with delayed onset of significant neurotoxic symptoms, so patients should be admitted and observed for 24 hours.
- All patients with symptoms of envenomation should be admitted for further antivenom therapy, wound care, and monitoring. These patients require admission to an intensive care setting that allows for frequent wound checks, as well as frequent antivenom and analgesic dosing.
- Wound checks including extremity measurements should be performed hourly during the initial phase of treatment until symptoms have stabilized.

CORAL SNAKES

- Two members of the coral snake family (Elapidae) are indigenous to the United States. The western coral snake (*Micrurus euryxanthus*) found in Arizona and New Mexico and the more venomous eastern coral snake (*Micrurus fulvius*) found in the Carolinas and the Gulf states.
- Coral snakes account for only 1% to 2% of annual snakebites in the United States.
- Coral snakes have rounded heads and circular pupils similar to many nonpoisonous species.
- The coral snake is often mistaken for certain varieties of the nonpoisonous king snake because both have red, yellow, and black rings. The old adage “red on yellow, kill a fellow; red on black, venom lack” helps distinguish the venomous coral snake from the nonpoisonous scarlet king snake.
- It is important to note that these distinguishing patterns are only accurate in North America. Highly venomous South American coral snakes and other snakes worldwide may have red and black adjacent bands.

- The much smaller fangs of a coral snake may leave little evidence of envenomation; therefore, any suspicion of an elapid bite warrants medical evaluation.

CLINICAL PRESENTATION

- As with other Elapidae serpents, the venom of the coral snake is primarily neurotoxic.
- The bite site will initially exhibit local cutaneous edema, swelling, and tenderness. Within several hours, the patient may experience paresthesias, vomiting, weakness, diplopia, fasciculations, confusion, and occasionally respiratory depression.
- Convulsions have been observed in smaller children. The fatality rate from eastern coral snake bites is as high as 10%.

MANAGEMENT

- Coral snake bites are treated aggressively, since a significant bite can lead to neurological and respiratory depression within 24 hours.
- Antivenin is administered early in the treatment course. The coral snake antivenin is effective against bites of the eastern coral snake, but not against western coral snake bites. However, the venom of the western coral snake is less toxic than that of its eastern counterpart.
- Three to five vials of the antivenin are generally recommended following skin testing. As with the *Crotalid* polyvalent antivenin, adverse side effects include anaphylaxis and serum sickness.

DISPOSITION

- Any child who has sustained a documented bite from a coral snake is admitted to the intensive care unit for airway management and appropriate antivenin administration for a 24- to 48-hour period.

EXOTIC SNAKES

- Several bites occur each year from nonindigenous snakes. Many of these snakes are illegally imported into the United States as exotic pets or purchased over the internet from international distributors.
- Physicians encountering victims of exotic snake envenomation may receive assistance in treatment by calling the local zoo's herpetologist or regional poison control center. The general approach is local

wound care, supportive treatment, and specific antivenin therapy, if available.

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QUESTIONS

1. A 5-year-old female is brought in the ED after sustaining a snake bite while hiking in the woods with her family. Which of the following statements is correct regarding snake envenomations?
 - A. There is only one family of venomous snakes indigenous to the United States.
 - B. The venom of coral snakes is primarily neurotoxic.
 - C. Incision of the bite and suctioning out the venom is indicated in pit viper bites.
 - D. Tourniquets have been shown to be effective when applied immediately after a venomous snake bite.
 - E. Antivenin should be given to all venomous snake bites.

2. A 10-year-old male presents to the ED with a history of being bitten on the arm by a rattlesnake while on a camping trip with his family. He appears ill with significant progressive swelling to his left arm, nausea, and dizziness. He is hypotensive and tachycardic. Which of the following would be appropriate in the management of this patient?
 - A. Fluid restriction.
 - B. Observation in the ED for 6 to 12 hours.
 - C. Administration of Crotalidae antivenin.
 - D. Admission to the general pediatric ward.
 - E. Avoidance of venipuncture for routine blood work.
3. Which of the following is correct regarding snake envenomations?
 - A. Pediatric patients are given proportionately less antivenin than adults.
 - B. Antivenin is highly efficacious if given after 24 hours of the bite.
 - C. A coral snake in the United States with red bands next to black is most likely to be venomous.
 - D. The venom of western coral snakes is more toxic than the eastern and has an effective antivenin.
 - E. If the patient suffers a snake bite from a nonindigenous snake the local zoo herpetologist is a good resource to be consulted.
4. A 6-year-old presents after being bitten by a cottonmouth snake. These bites in children have which of the following characteristics?
 - A. Children are less vulnerable to severe envenomations than adults.
 - B. Bites on the lower extremity are more severe than on the head or trunk.
 - C. Severe bites can result in coagulopathies and DIC.
 - D. The fatality rate is as high as 20% even with optimal treatment.
 - E. The hallmark of pit viper bite is multiple teeth marks with no pain or swelling.
5. A 7-year-old male presents after being bitten by a coral snake. He develops pain and swelling at the site followed by paresthesias. Which of the following is correct regarding coral snake envenomations?
 - A. Coral snakes are distinguished by a triangular head, facial pits, and vertical pupils.
 - B. They are dull in color typically green or brown.
 - C. Symptoms may include cardiac dysrhythmias.
 - D. Antivenin should be administered if an envenomation is suspected due to potential respiratory compromise.
 - E. There are no serious side effects to the administration of antivenin.

ANSWERS

1. B. The venom of coral snakes is primarily neurotoxic. The two families of venomous snakes indigenous to the United States are Crotalidae (pit vipers) and Elapidae (coral snakes). Incision and suctioning of the snake bite and tourniquet placement is contraindicated. Antivenin therapy is guided by the type of snake and the severity of symptoms.
2. C. This bite would be considered moderate to severe. Crotalidae antivenin is the fundamental treatment for pit viper envenomation. There is now a Crotalidae polyvalent Fab antivenom available. To maintain renal flow and intravascular volume, fluids should be given. If the child exhibits moderate to severe envenomation, has evidence of coagulopathy, or requires antivenin administration, admission to an intensive care unit is indicated. Laboratory studies such as a CBC, INR coagulation profile, and U/A should be obtained for evaluation of systemic effects.
3. E. Information obtained by calling the local zoo may be extremely helpful when dealing with exotic snake bites. In comparison with adults, pediatric patients are given proportionately *more* antivenin since children receive a greater amount of venom per kg of body weight. Antivenin is most efficacious if given within 4 to 6 hours of the bite and of less value with delays in treatment. The mnemonic for coral snakes to help distinguish poisonous snakes is “red on yellow, kill a fellow; red on black, venom lack.” The venom of the western coral snake is *less* toxic than the eastern and the antivenin is not effective for western coral snake bites.
4. C. Cottonmouth snakes are pit vipers. A severe bite can result in coagulopathies and DIC. Because of their small body weight, infants and young children are relatively more vulnerable to severe envenomation. Bites on the head or trunk are more severe than bites to the extremities. The overall mortality rate is generally good at less than 1% if the antivenin is given in adequate amounts without delay. The hallmark of pit viper bites is fang marks with local pain and swelling.
5. D. Antivenin should be administered if an envenomation is suspected due to potential respiratory compromise. Pit vipers classically possess a triangular head, facial pits, and vertical pupils. Coral snakes are brightly colored with black, red, and yellow rings. The venom is primarily neurotoxic and not cardiotoxic. Adverse side effects of the antivenin include anaphylaxis and serum sickness.

135 SPIDER AND ARTHROPOD BITES

Timothy B. Erickson
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BLACK WIDOW SPIDERS

ANATOMY

- The *Lcatrodectus* genus is a common spider found in temperate and tropical Zones of the world.
- Female black widow spiders are 20 times larger in size, darker in color, and more toxic than their male counterparts.
- Black widows are described as charcoal or black with eight eyes, eight legs, fangs and poison glands, and a characteristic red hourglass mark on the ventral aspect of the abdomen.

PATHOPHYSIOLOGY

- The female black widow is generally considered poisonous to humans; whereas the males are not due to their small jaws and minimal number of poison glands. Black widows spiders control the amount of venom they inject; an estimated 15% of bites to humans are nonenvenomating.
- The toxin facilitates exocytosis of synaptic vesicles and the release of the neurotransmitters norepinephrine, gamma-aminobutyric acid, and acetylcholine. It also causes degeneration of motor endplates, resulting in denervation.
- The venom destabilizes nerve cell membranes by opening ion channels, causing a massive influx of calcium into the cell, which may lead to hypocalcemia.

CLINICAL PRESENTATION

- Black widow bites usually produce a pinprick sensation but often go unnoticed. Within the first two hours after the bite, the site may develop redness, cyanosis, urticaria, or a characteristic halo-shaped target lesion.
- Local symptoms may be followed by generalized symptoms of pain in regional lymph nodes, chest, abdomen, and lower back. Pain classically descends down the lower extremities with burning sensation on the soles of the feet.
- Abdominal rigidity and vomiting can be quite severe and may even be mistaken for a surgical emergency.
- Infants may present with erythema of wound areas, irritability, constant crying, sialorrhea, agitation,

and seizures. School-aged children and adolescents describe pain at the wound site, abdominal and thoracic pain, muscle spasms, and fine tremors.

- Flexor spasm of the limbs can cause the patient to assume a fetal position while writhing in pain. Severe symptoms include hypertension, sweating, salivation, dyspnea with increased bronchosecretions, and convulsions.
- Death is uncommon, but may occur from respiratory or cardiac failure, with an overall mortality of <5%.

MANAGEMENT

- Local wound care is appropriate and pain at the bite site may be relieved with early application of ice. Tetanus prophylaxis should be updated if needed, but antibiotics are unnecessary unless there is evidence of a wound infection.
- Oral analgesics may be of benefit, but parenteral analgesics, such as morphine, may be required if the pain is generalized. Muscle spasms may require oral or parenteral benzodiazepines.
- Calcium gluconate has little clinical benefit following black widow envenomation.
- In extreme cases with severe symptoms, lactrodectus antivenin is recommended. The antivenin has been associated with a relatively low rate of allergic reactions.
- The use of lactrodectus-specific antivenin is restricted to patients with severe envenomation and no allergic contraindications, and in whom opioids and benzodiazepines are ineffective. Young children with severe toxicity should receive antivenin early in the clinical course.
- Patients receiving antivenin may experience flu-like symptoms or serum sickness 1–3 weeks following treatment. This entity is generally self-limited and responsive to antihistamines and prednisolone.

DISPOSITION

- Any symptomatic pediatric patient who has suffered a bite from a black widow spider is admitted for observation and pain control.
- With cardiopulmonary compromise or convulsions, the child is admitted to the intensive care unit for stabilization and antivenin administration.

BROWN RECLUSE SPIDERS

INTRODUCTION

- *Loxosceles* spiders are known to be reclusive nocturnal hunters. Victims typically are bitten on the extremities while rummaging in confined spaces like

a closet or and attic or while putting on a shoe or using a blanket that a spider is trapped in.

- The Hobo spider (*Tegenaria agrestis*), originally from Europe and central Asia, now resides in the Pacific Northwest of the United States and adjacent Canada. Similar symptomatology leads many to incorrectly attribute Hobo spider bites to that of the brown recluse, which is not indigenous to the north-west US.

ANATOMY

- The brown recluse gets its name because of its brown or fawn colored body. It has a characteristic violin or fiddle-shaped marking on the dorsal cephalothorax.

PATHOPHYSIOLOGY

- The venom of the *Loxosceles* is more potent than that of the rattlesnake and can cause extensive skin necrosis.
- The venom acts directly on the cell wall, causing immediate injury and cell death. It contains the calcium dependent enzyme sphingomyelinase D which, along with C-reactive protein, has a direct lytic effect on red blood cells.
- Following cell wall damage an intravascular coagulation process causes a cascade of clotting abnormalities and local polymorphonuclear leukocyte infiltration, culminating in a necrotic ulcer.

CLINICAL PRESENTATION

- Most brown recluse bites occur in predawn hours and are often painless.
- The clinical response to loxoscelism ranges from cutaneous irritation (necrotic arachnidism) to a life-threatening systemic reaction. Most signs and symptoms of envenomation are localized to the bite area.
 - The majority result in nothing more than a local reaction and resolve spontaneously. Classically, erythema surrounds a dull blue-grey macule circumscribed by a ring or halo of pallor. The color difference is important in identifying necrotic arachnidism. Gradually, over 3 to 4 days, the wound forms a necrotic base with a central black eschar. In 7 to 14 days, the wound develops a full necrotic ulceration.
 - The systemic reaction, is much less common than the cutaneous reaction, and is associated with a higher morbidity. The reaction rarely correlates with the severity of the cutaneous lesion. Within

24 to 72 hours following the envenomation, the patient experiences fever, chills, myalgias, and arthralgias. In severe systemic reactions, the patient may suffer coagulopathies, hypotension, jaundice, disseminated intravascular coagulation (DIC), convulsions, renal failure, and hemolytic anemia. In rare cases, a patient may succumb to a fatal systemic reaction.

MANAGEMENT

- The proper management of envenomation by the brown recluse spider depends on whether the reaction is local or systemic.
- Proper care includes wound cleansing, immobilization and elevation of the affected extremity to reduce pain and swelling. Early application of ice to the bite area will lessen the local wound reaction, whereas heat will exacerbate the symptoms.
- Tetanus should be immediately updated, but antibiotics are only indicated if there is a secondary wound infection. Antihistamines and analgesics prove to be beneficial, especially in children.
- Early excision of ulcers and steroid injections are not recommended. Complications of early surgical intervention include recurrent wound breakdown and long term distal extremity dysfunction.
- Delayed excision of ulcers after the necrotic process has subsided, followed by secondary closure is the preferred management.
- The polymorphonuclear leukocyte inhibitor dapsone was advocated to diminish scarring and subsequent surgical complications. Its use, however, has not proven effective in any large study with human or animal models and it may induce methemoglobinemia and hemolytic anemia in children with G6PD deficiency.
- Hyperbaric oxygen treatment for brown recluse envenomations has little clinical evidence supporting its use.
- Systemic effects of brown recluse spider bites are rare but can be life threatening.
- The patient must be monitored closely for the development of DIC. Transfusion of RBCs and platelets may be necessary. Glucocorticoids may provide a protective effect on the RBC membrane, slowing hemolysis. Urine alkalization with bicarbonate may lessen renal damage if the patient is experiencing acute hemolysis.
- Although it is not commercially available in the United States, there is ongoing research with brown recluse antivenom. There is little evidence to support its efficacy, particularly against local cutaneous effects.

DISPOSITION

- Patients with a rapidly expanding lesion or necrotic area with evidence of hemolysis are hospitalized.
- Patients who are asymptotic following a period of observation in the emergency department and have normal baseline laboratory values may be discharged home with close outpatient follow-up and wound care within 24 to 48 hours.

TARANTULAS

- Tarantulas are widely feared because they are the largest of all spiders.
- Found predominantly in the deserts of the western United States, these large, hairy spiders are relatively harmless. They are extremely shy and bite only when vigorously provoked or roughly handled. Their bites usually cause minimal pain and surrounding edema with little or no necrosis and no serious systemic effects.
- Treatment of bites consists of local wound care and tetanus prophylaxis. The patient can be treated with antihistamines and topical glucocorticoids.
- Hairs on tarantulas can be flicked off in large numbers as a defense mechanism. These hairs are capable of producing urticaria and pruritis that may persist for several weeks. They may also get into the eyes and cause keratoconjunctivitis or ophthalmia nodosa.

SCORPIONS

- Epidemiologically, scorpions are the most significant of all arachnid envenomations, resulting in adult morbidity and pediatric mortality. In the United States, there have been no reported deaths from scorpion stings in more than 25 years, but they remain a public health concern throughout the South and Southwest.

ANATOMY

- The scorpion has a pair of anterior legs with pinchers, a segmented body, and a long, mobile tail equipped with a stinger.
- Only the southwestern desert scorpion (*Centruroides exilicauda*, formerly *Centruroides sculpturatus*) poses a serious health threat in the United States.
- Also called bark scorpions because they cling to the bottom of fallen brush and trees, they are brownish in color and are most active at night. The chitin shell of this scorpion will fluoresce under an ultraviolet or Wood's lamp, helping in identification.

PATHOPHYSIOLOGY

- *Centruroides* venoms cause spontaneous depolarization of nerves of both the sympathetic and parasympathetic nervous systems.

CLINICAL PRESENTATION

- Unless the scorpion is identified, the diagnosis is based on clinical symptoms.
- Most victims will have only local pain, tenderness, and tingling. Young children and those who suffer more serious envenomations may manifest the venom effects as overstimulation of the sympathetic, parasympathetic, and central nervous systems.
- Elevation of all the vital signs usually occurs within an hour of envenomation, and tachydysrhythmias may develop.
- Dysconjugate, "roving" eye movements are very common in children, along with other neurological findings, including muscle fasciculations, weakness, agitation, and opisthotonos. Less common findings are ataxia, respiratory distress, and seizures.

MANAGEMENT

- The treatment of *Centruroides* envenomations is supportive. Cool compresses and analgesics are used for the local symptoms and pain. Wound care and tetanus prophylaxis are indicated.
- Benzodiazepines may be helpful for agitation and muscle spasms.
- Advanced life support and airway control are essential for more severe envenomations.
- In the United States, a *Centruroides*-specific, goat-derived antivenom has been available in very limited supply. Immediate and delayed allergic reactions including serum sickness are possible.
- *Centruroides*-specific antivenom should be reserved for cases of severe systemic toxicity. Consultation with a toxicologist experienced in scorpion envenomation is recommended before using antivenom. One to two vials of antivenom in cases of severe toxicity can lead to rapid resolution of symptoms.

HYMENOPTERA

- The order Hymenoptera includes bees, vespids (hornets and wasps), and fire ants.
- These insects cause one-third of all reported envenomations in the United States and an estimated 50 to 150 annual deaths.

- Hymenoptera venoms possess intrinsic toxicity, but it is their ability to sensitize the victim and cause subsequent anaphylactic reactions that makes them lethal.

BEEES AND VESPIDIS

- Honeybees (*Apis mellifera*) are not intrinsically aggressive; they usually sting defensively when stepped on.
- The honeybee's stinger is a modified ovipositor (only females sting) that is connected to a venom sac. Honeybees lose their barbed stinger after stinging and die.
- Africanized honeybees, or "killer bees," (*Apis mellifera scutellata*) are now found in Texas, Arizona, California, and most of the temperate southeastern and southwestern states.
 - Although the toxicity of their venom is equal to that of their native counterpart, they are far more aggressive.
 - Massive numbers of stings from an attack of Africanized bees can result in multisystem damage and death from severe venom toxicity. Most patients of massive envenomation suffer acute tubular necrosis or renal involvement with myoglobinuria. In swarms, these bees can overwhelm and kill even healthy nonallergic victims.
- The most common hornets in the United States are the yellow jackets (*Vespa pennsylvanica*). Yellow jackets are usually seen around garbage cans, beverage containers, and various foods. They are extremely aggressive and sting with little provocation.
- Wasps (*Polistes annularis*) have a formidable sting and build their nests in the eaves of buildings. These vespids are carnivorous and able to use their smooth stingers multiple times.

PATHOPHYSIOLOGY

- Four possible reactions are seen after hymenoptera stings:
 - *Local reactions:* These are the most common reactions resulting from the vasoactive effects of the venom and are generally mild. The most common response includes pain, mild erythema, edema, and pruritus at the sting site. There are no systemic signs or symptoms, but a severe local reaction may involve one or more contiguous joints. Local reactions occurring in the mouth or throat can produce swelling that may lead to upper airway obstruction, especially in younger children.

- *Toxic reaction:* A systemic toxic reaction from venom may occur when a patient suffers from multiple stings. Africanized bees are notorious for such attacks, but an aggressive native hive may elicit a similar response. Symptoms of a toxic reaction may resemble anaphylaxis, but gastrointestinal manifestations, light-headedness, and syncope more commonly occur. Headache, fever, drowsiness, involuntary muscle spasms, edema without urticaria, and convulsions may also ensue. Severe envenomations may lead to respiratory insufficiency and arrest. Hepatic failure, rhabdomyolysis and DIC have been reported in both adult and pediatric victims.
- *Anaphylactic reaction:* A generalized systemic allergic may occur after envenomation. Generalized systemic reactions to Hymenoptera venom are thought to occur from an immunoglobulin E (IgE)-mediated mechanism, leading to the release of pharmacologically active mediators within mast cells and basophils. Symptoms are often mild, but severe reactions can lead to death within minutes. Unlike the toxic reaction, there is no correlation between systemic allergic reactions and the number of stings. The majority of allergic reactions occur within the first 10–15 minutes, and nearly all occur within 6 hours. Fatalities that occur within the first hour of the sting usually result from airway obstruction or hypotension. Initial symptoms typically consist of ocular pruritus, facial flushing, and generalized urticaria. Symptoms may intensify rapidly with chest or throat constriction, wheezing, dyspnea, abdominal cramping, diarrhea, vomiting, vertigo, fever, laryngeal stridor, syncope, and shock.
- *Delayed reaction:* A delayed reaction, appearing 1–2 weeks after a sting consists of serum sickness-like signs and symptoms of fever, malaise, headache, urticaria, lymphadenopathy, and polyarthritis. This reaction is believed to be immune complex-mediated.

MANAGEMENT

- If present, the embedded stinger should be removed manually.
- Treatment is symptomatic, with ice or cold compresses and an antihistamine.
- In more severe local reactions, there is a more sustained inflammatory response; the swelling may spread to the entire extremity and persist for several days. A short course of prednisone may decrease the duration of symptoms.
- Toxic reactions reflect the effects of multiple stings (usually over 25 to 50 stings).

- Systemic reactions occur in approximately 1% of Hymenoptera stings. They range from mild, nonlife-threatening cutaneous reactions to classic anaphylactic shock.
- The mainstay of treatment is epinephrine. Epinephrine counteracts the bronchospastic and vasodilatory effects of histamine. Epinephrine can be given as a subcutaneous injection (0.01 mL/kg of 1:1000 solution; not to exceed 0.3 mL). In more severe reactions, the intravenous or endotracheal route is preferred (0.1 mL/kg of 1:10,000 solution).
- Early intubation is indicated if there is evidence of severe laryngeal edema or stridor, because airway obstruction is the leading cause of death in anaphylaxis.
- Antihistamines should be given early, but not as a substitute for epinephrine. An H₂-receptor blocker (e.g., cimetidine or ranitidine), in addition to an H₁-receptor blocker (diphenhydramine), may aid in inhibiting the vasodilatory effects of histamine.
- Adjunctive therapy for bronchospasm might include inhaled beta-2 agonists (e.g., albuterol) and intravenous aminophylline.
- When hypotension is present, vigorous isotonic fluid resuscitation should be instituted.
- Glucocorticoids should be given for their anti-inflammatory effects as well as their effect of preventing the late-phase response. A delayed serum sickness-like reaction may appear 10 to 14 days following the initial sting. This immune complex disorder may be treated with a short course of prednisone.
- Venom immunotherapy desensitization is very effective in preventing further systemic reactions, with 95% to 100% protection after 3 months of treatment. Referral to an allergist is indicated for any child who has experienced life-threatening respiratory symptoms or hypotension.

DISPOSITION

- patients who have had a systemic reaction should be instructed to wear protective clothing and avoid Hymenoptera-infested habitats.
- Portable epinephrine kits (Epi-Pen and Epi-Pen Jr) should be prescribed prior to the patient leaving the emergency department.
- The patient should be urged to carry the kit at all times and to use epinephrine for any systemic symptoms.
- Even if symptoms are mild, the patient should still seek emergency care.
- The patient should also be instructed to wear a medical alert tag.

IMPORTED FIRE ANTS

- Five known species of fire ants belonging to the genus *Solenopsis* are found in the United States.
- Colony mounds are found most commonly in yards, playgrounds, and open fields.
- Fire ants are aggressive insects with no natural enemies. They are social insects and tend to attack in swarms, with multiple stings the norm.
- Stings are more common among children and occur most frequently on their ankles and feet during the summer months.

PATHOPHYSIOLOGY

- Fire ants first bite the victim with powerful mandibles, then, if undisturbed, will arch the body and swivel around the attached mandibles to sting the victim repeatedly with the stinger.
- This produces a characteristic circular pattern of papules/stings around two central punctures.
- Fierce fire ant attacks ensue in response to an alarming pheromone released by an individual or group of ants.
- Fire ant venoms produce a sharp, burning sensation, hence the name.
- The venoms have cytotoxic, bactericidal, insecticidal, and hemolytic properties. They also activate the complement pathway and promote histamine release. Fire ant venoms are immunogenic and result in sensitization of the sting victim and the risk of future anaphylaxis.

CLINICAL PRESENTATION

- Clinical manifestations are predominantly local dermatologic reactions.
- The initial bites and stings cause burning pain associated with circular wheals or papules around the central hemorrhagic punctures. The wheal-and-flare reactions resolve within 1 hour, but then develop into sterile pustules within 24 hours. The pustules slough off over 48 to 72 hours, leaving shallow ulcerated lesions.
- The pustules are intensely pruritic and often become contaminated after the victim scratches the lesions. These secondary infections are usually minor but may cause considerable morbidity.
- Between 15% and 50% of victims develop more severe local reactions, characterized by an exaggerated wheal-and-flare response followed by the development of erythema, edema, and induration.
- These lesions are intensely pruritic, may resemble cellulitis, and persist for 24 to 72 hours before subsiding.

MANAGEMENT

- Topical glucocorticoid ointments, local anesthetic creams, and oral antihistamines may be useful for the itching associated with these reactions.
- No intervention has been shown to prevent or resolve the pustules, and treatment consists of local conservative measures: application of ice or cool compresses for symptomatic relief and gentle, frequent cleansing of the affected areas to prevent secondary infections.
- Anaphylaxis may occur several hours after a sting and is known to occur more frequently in children than in adults. Immunotherapy may be appropriate for persons with severe hypersensitivity to fire ant venom or those who have had a previous anaphylactic reaction to a fire ant sting.

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QUESTIONS

1. An 18-year-old normally healthy patient presents with sudden onset of severe diffuse abdominal pain with nausea and vomiting. He is febrile and his

vital signs are stable. His abdominal examination reveals marked rigidity. He denies urinary symptoms, change in bowel habits, trauma, or previous episodes of abdominal pain. On examination, the right leg is noted to have a halo-shaped target lesion. Which of the following would be the most appropriate treatment for this patient?

- A. CT scan of the abdomen and pelvis.
 - B. Immediate surgical consult for exploratory laparotomy.
 - C. Administration of antivenin.
 - D. Parenteral opiates, muscle relaxants, and treatment of vital sign abnormalities.
 - E. Analgesics, fluid hydration and an intravenous pyelogram.
2. A 20-year-old woman presents with the complaint of being bitten by a spider while in the attic moving some old blankets. She now has swelling, erythema, and has a central blue-grey macule with a surrounding ring of pallor. She denies any other symptoms. The most appropriate treatment of this wound would be:
 - A. Apply heat, and give topical antibiotics.
 - B. Apply local heat, analgesics, and consider antivenom.
 - C. Apply ice, dapsone, and early excisional treatment.
 - D. Apply ice, local wound care and delayed excision.
 - E. Apply ice, dapsone, and steroids.
 3. A 16-year-old presents to the ED very anxious because he was just bitten by a pet tarantula that he recently found on a camping trip in Arizona. He had mild tenderness at the bite site but denies symptoms currently. Appropriate treatment would be?
 - A. Make cruciate incisions and suction to withdraw the venom.
 - B. Apply ice, analgesics, and administer dapsone.
 - C. Apply ice, analgesics, and muscle relaxants.
 - D. Reassure the patient that this is a harmless bite.
 - E. Apply ice, analgesics, consider antivenom.
 4. A 6-year-old patient recently treated for a brown recluse spider bite develops shortness of breath. She is noted to be cyanotic and hypoxic refractory to oxygen therapy. The nurse notes that her blood is brown colored when drawing her labs. What complication do you suspect in this patient?
 - A. Hemolytic anemia.
 - B. Methemoglobinemia from treatment.
 - C. DIC.
 - D. Severe systemic reaction to the spider bite.
 - E. Acute adult respiratory distress syndrome.
 5. Which of the following is true regarding scorpion stings?
 - A. There have been no reported deaths in the US from scorpion stings over the past 25 years.

- B. There is no known antivenom.
- C. Hyperbaric oxygen has been shown to be effective in severe cases.
- D. Dapsone may be considered for severe local reactions.
- E. Scorpion stings are harmless and typically asymptomatic.

ANSWERS

1. D. This patient has a characteristic black widow spider bite, which can present with abdominal rigidity and vomiting mimicking a surgical emergency. The bite may produce a pinprick or burning sensation but may go unnoticed as in this patient. Within the first few hours, the site may develop redness, cyanosis, urticaria, or a characteristic halo-shaped target lesion. Treatment includes early application of ice, parenteral opioids such as morphine, muscle relaxants such as diazepam, and possibly calcium gluconate, while stabilizing vital signs. The use of the *Lactrodectus*-specific antivenom is restricted to severe envenomations refractory to other treatments.
2. D. This patient has a classic scenario for a brown recluse spider bite. She has a local cutaneous reaction. Classically, erythema surrounds a dull blue-grey macule circumscribed by a ring or halo of pallor. This may progress to form a necrotic base with a central black eschar. The wound should be cleansed, tetanus status addressed, and the extremity immobilized to reduce pain. Early ice application lessens the local reaction whereas heat will exacerbate symptoms. Early excision can cause complications such as recurrent wound breakdown and is not recommended; rather, delayed excision after several weeks is recommended with secondary closure and skin grafting as needed.
3. D. Tarantulas, the largest of all spiders are relatively harmless. They are found in the western United States. Local wound care, tetanus prophylaxis as needed and reassurance would be appropriate management.
4. B. This patient has methemoglobinemia, a known complication of dapsone. Dapsone, a polymorphonuclear leukocyte inhibitor, has been recommended for brown recluse spider bites in the past to decrease the amount of scarring and subsequent surgical complications. Its use is no longer recommended since it has not been proven effective in any large study with human or animal controls.
5. A. Worldwide, scorpions are responsible for thousands of deaths annually but in the United States there have been no reported deaths in more than 25 years. A hyperimmune goat antivenom is available

for severe envenomations with potentially life-threatening symptoms. Most victims have local pain, tenderness, and tingling. More severe cases may have dysconjugate “roving” eye movements and muscle fasciculations, weakness, agitation, and opisthotonus. Treatment is supportive with cool compresses, analgesics, tetanus prophylaxis, and wound care. Hyperbaric oxygen therapy and dapsone are not indicated.

136 MARINE ENVENOMATIONS

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INTRODUCTION

- Hazardous marine life can be classified into four major groups:
 - Venomous bites and stings, such as those inflicted by scorpion-fish and the Portuguese man-o'-war.
 - Shock injuries, as from electric eels.
 - Traumatogenic bites (such as from sharks and barracudas).
 - Toxic ingestions or fish poisoning.
- Toddlers are most likely to be envenomed in shallow waters and are typically unable to give a detailed or reliable history.
- Young children may step on poisonous marine animals or handle them, resulting in extremity stings.
- Adolescents are more adventurous and frequent deeper waters as surfers, ocean swimmers, snorkelers, and scuba divers. This age group is also more susceptible to intoxication with ethanol or recreational drugs.

COELENTERATES

- Coelenterates (*Phylum cnidaria*) include jellyfish, sea anemones, and corals.
- Jellyfish stings are the most common marine envenomations.
- A commonly encountered jellyfish is the sea nettle (*Chrysaora quinquecirrha*), which is widely distributed in temperate and tropical waters.
- One of the more feared jellyfish is the Portuguese man-o'-war (*Physalia physalis*). Its tentacles can reach up to 30 m in length.
- The deadliest and most venomous of coelenterates is the box jellyfish, or sea wasp, of Australia.

PATHOPHYSIOLOGY

- Coelenterates envenomate with organelles called nematocysts, which contain venom-bearing threads that reside within specialized epithelial cells on the tentacles.
- Each nematocyst is a capsule with a folded eversible tubule, carrying a variety of toxins with neurologic, cytolytic, and enzymatic effects. Upon contact or when encountering a change in osmolality, these threads are everted from the nematocysts in order to be thrust into the prey.
- When a human is stung, the penetration reaches into the innervated and vascular dermis. Both living and dead coelenterates can envenomate, as can fragmented tentacles and “unfired” nematocysts on the skin.
- The venom in nematocytes is potentially dermatonecrotic, myotoxic, cardiotoxic, neurotoxic, and hemolytic.

CLINICAL PRESENTATION

- Mild coelenterate envenomation from true jellyfish or sea nettles generally causes local pruritus and characteristic linear, spiral, and painful urticarial lesions.
- The lesions often blister and there is localized surrounding edema. The pain and stinging sensation occurs instantly, peaks within 60 minutes and may persist for hours.
- Systemic symptoms from Portuguese man-o’-war stings may include nausea, vomiting, dysphagia, muscle cramps, myalgias, arthralgias, diaphoresis, and weakness.
- Hemolysis and renal failure have been described following man-o’-war stings in pediatric patients. Other severe systemic symptoms include hemolysis, dysrhythmias, cardiovascular collapse, respiratory distress, paralysis, seizures, coma, and death.
- The vast majority of *Chironex fleckeri* stings are not life-threatening, but present with painful skin welts as the major finding. However, fatalities that do occur usually do so within 5–20 minutes of the envenomation.

MANAGEMENT

- Treatment includes reassurance of the victim and immobilization of the injured part. Ice may provide some analgesia.
- The area is rinsed with sterile saline or seawater to maintain a condition isosmolar to seawater and to wash off unfired nematocysts. Fresh water is not recommended because it is hypoosmolar and often activates unfired nematocysts.

- To inactivate nematocysts remaining on the skin, alter the pH by soaking the wounds with a weak acid like household vinegar (5% acetic acid solution).
- The inactivated nematocysts are then removed by gentle shaving or scraping.
- If the victim shows signs and symptoms of anaphylaxis, treat appropriately. In most cases, analgesics and antihistamines are helpful.
- Tetanus immunization is indicated, but prophylactic antibiotics are not.
- Sea anemones and corals are sessile creatures that cause local urticarial reactions upon contact.
- Contact with hard (true) corals may cause lacerations that are treated with vigorous local wound care, topical antiseptics, and tetanus prophylaxis.
- If a child is envenomated by an Australian box jellyfish, antivenin against Chironex is indicated if available. The rapid onset of cardiotoxicity with severe envenomations suggests that the antivenin should be given without delay and in proper doses to be life-saving.

DISPOSITION

- Mild stings responsive to vinegar can be managed at home after a 3 to 4 hour observation. Children with systemic toxicity or inadequate pain control despite local wound treatment should be kept for observation.
- Any child envenomed by a box jellyfish should be kept for observation for 24 hours. Symptomatic patients may require antivenin.

VENOMOUS FISH

- *Stingrays* are the most commonly encountered venomous fish, with more than 2000 stings reported annually. Eleven species of stingrays are found in the United States coastal waters.
- They are flat, round-bodied fishes that burrow underneath the sand. When startled or stepped on, the stingray thrusts its spiny tail upward and forward, driving its barb (a venom-laden stinging apparatus) into the body (foot, lower extremity, or chest if you hover too close over them) of the victim.
- When entering or wading in shallow water, perform the “Stingray Shuffle”: drag and shuffle your feet rather than step as you would on land; this reduces the likelihood that you will accidentally step on a buried stingray; by startling it and giving it a chance to move out of your path. When diving over sandy bottoms in deeper water, scope out the bottom and watch for the oval or rhomboidal outline that could indicate a buried stingray before touching the bottom. If a stingray consistently turns to face you or raises its

stinger-bearing tail above its back like a scorpion, it would be prudent to back off.

- *Varieties of scorpion-fish* include zebra fish and lionfish (*Pterois*), scorpion-fish (*Scorpaena*), and stonefish (*Synanceja*), in increasing order of venom toxicity. Lionfish are increasingly popular as aquarium pets. Ocean entry and handling them without protective equipment are the common mechanisms of injury.
- Catfish are found in both fresh and salt water. Stings occur from spines contained within an integumentary sheath on their dorsal or pectoral fins. The hands and forearms of fishermen and seafood handlers are the most common sting sites.

PATHOPHYSIOLOGY

- Stingrays have one to four venomous spines or barbs on the dorsum of a whip-like tail. The spines are retro serrated, so they anchor into the flesh securely and may become difficult to remove.
- As the barb is withdrawn, the sheath surrounding it milks the venom glands, releasing the venom. Parts of the sheath may be torn away and remain in the wound.
- The venom is intensely active, partially heat-labile and causes varying degrees of local tissue necrosis and cardiovascular disturbances.
- Scorpionfish have venomous spines on the dorsal, anal and pelvic fins. This venom is also partially heat-labile. Stonefish have dorsal spines harboring one of the most toxic fish venoms. Both the scorpion and stonefish are not easily disturbed by shuffling your feet, so stay away, and exercise caution.

CLINICAL PRESENTATION

- The best way to treat a stingray spine injury is not to allow it to happen in the first place. With stingrays, intense pain out of proportion to the apparent injury is the initial finding, peaking within 1 hour and lasting up to 48 hours. Signs and symptoms are usually limited to the injured area. The barb may be present or absent. Bleeding is usually not life threatening, but weakness, nausea, anxiety, and syncope have been reported. Death has occurred from a fatal chest wound from hovering too close.
- Envenomations from lionfish, scorpionfish, and stonefish cause immediate intense pain that peaks within 60 to 90 minutes and persists for up to 12 hours.
- Local erythema or blanching, edema and paresthesias may persist for weeks. Systemic findings include

nausea and vomiting, weakness, dizziness, and respiratory distress.

- Stonefish venom, a potent neurotoxin, can cause dyspnea, hypotension, and cardiovascular collapse within 1 hour and death within 6 hours. Local necrosis and severe pain may persist for days.
- With catfish stings, burning and throbbing sensation occurs immediately, but usually resolves within 60 to 90 minutes. The discomfort may last up to 48 hours. Systemic symptoms are rarely reported.

MANAGEMENT

- Stingrays
 - Treatment includes irrigation with sterile saline to dilute the venom and remove sheath fragments.
 - The spine of the stingray including the venom glands is typically difficult to remove from the victim and radiographs may be necessary to locate the spine or retained fragments.
 - The injured part should be immersed in hot water, no warmer than 113°F, for 30 to 90 minutes to inactivate any heat-labile venom components.
 - Analgesics are usually required. Tetanus immunization is updated. Because of the penetrating nature of the envenomation, wounds are debrided and left open.
 - Treatment with a broad-spectrum prophylactic antibiotic, such as trimethoprim-sulfamethoxazole (TMP-SMX), ciprofloxacin, a fluoroquinolone or a third-generation cephalosporin, is recommended because of concern for infection by *Vibrio* species as well as *Staphylococcus* and *Streptococcus* spp.
 - ACLS and surgical consult for any chest or abdominal puncture wounds that present with life threatening symptoms or shock.
- Scorpionfish and Lionfish
 - Envenomation is treated with immersion of the affected limb in hot water (113°F) for 30 to 90 minutes, or until pain is relieved.
 - Wounds are irrigated with sterile saline, explored, and cleaned of debris. The wound is left open and treatment with prophylactic antibiotics is initiated.
 - Local treatment for a stonefish sting is the same as that for envenomations by other scorpion fish, with special attention given to maintaining cardiovascular support.
- *Stonefish*: There is a specific stonefish antivenin available in Australia. The antivenin is an equine-derived product and carries the risk for inducing anaphylaxis.

- *Catfish* sting treatment is immediate immersion in hot water (no warmer than 113°F) for pain relief.
 - Catfish spines may penetrate the skin and break off. Sometimes, the spines can be located by routine radiographs. Retained catfish spines should be removed.
 - The wound should be explored and debrided. The puncture wound is left open.
 - Treatment with prophylactic broad-spectrum antibiotics and tetanus prophylaxis are indicated.

DISPOSITION

- Children with mild stings responsive to hot water soaks may be discharged after observation.
- Children not responsive to pain management may have a retained foreign body.
- Children envenomed by stonefish should be monitored in an intensive care setting. If available, antivenin administration is indicated in symptomatic patients.

ECHINODERMS

- Echinoderms are spiny invertebrates that include sea urchins, sea stars, starfish, sand dollars, and sea cucumbers. Sea urchins most often cause medically significant envenomations.

PATHOPHYSIOLOGY

- The spines or pedicellariae of sea urchins produce painful puncture wounds, swelling, purple tattooing of skin to mark point of contact and localized erythema.

CLINICAL PRESENTATION

- The spines can be up to one foot long in the needle-spined urchin. They can easily puncture the skin, break off and be retained.
- Their venom can cause local pain that may persist for days.

MANAGEMENT

- Treatment is immediate immersion in hot water (no warmer than 113°F), careful removal of pedicellariae and spines and local wound care.
- Tetanus immunization and antibiotic prophylaxis are often indicated.

DISPOSITION

- Most sea urchin puncture victims can be discharged home with continued hot water soaks and antibiotic prophylaxis.
- If there is a retained foreign body, follow up evaluation is prudent.

SEA SNAKES

- Sea snakes of the family *Hydrophiidae* are encountered throughout the Indo-Pacific region.
- The yellow-bellied sea snake (*Pelamis platurus*) has the widest distribution ranging from the Indo-Pacific to Africa to Central America. They are among the deadliest snakes in the world, and may bite without provocation. Most bites are associated with net fishing and inadvertent handling.

CLINICAL PRESENTATION

- The venom of the sea snake has neurotoxic, myotoxic, and nephrotoxic effects.
- Most sea snake bites are dry bites with little venom injected.
- With true envenomations, symptoms usually manifest within 30 minutes to 3 hours. Initial symptoms may include muscle spasms and trismus. Severe envenomations may result in acute neurotoxicity with rapid muscular and respiratory paralysis.

MANAGEMENT

- Apply a mild pressure wrap to the involved extremity with a compression bandage and immobilize, in a fashion similar to applying a posterior mold, until the victim receives definitive care. Respiratory support may be required.
- A polyvalent antivenin from Australia is commercially available. If antivenin is administered, the patient should be closely monitored for signs of anaphylaxis and given appropriate doses of diphenhydramine, glucocorticoids, and epinephrine as needed.

DISPOSITION

All documented and suspected sea snake envenomation victims should be monitored in an intensive care setting for possible airway management and antivenin administration (Fig. 136-1).

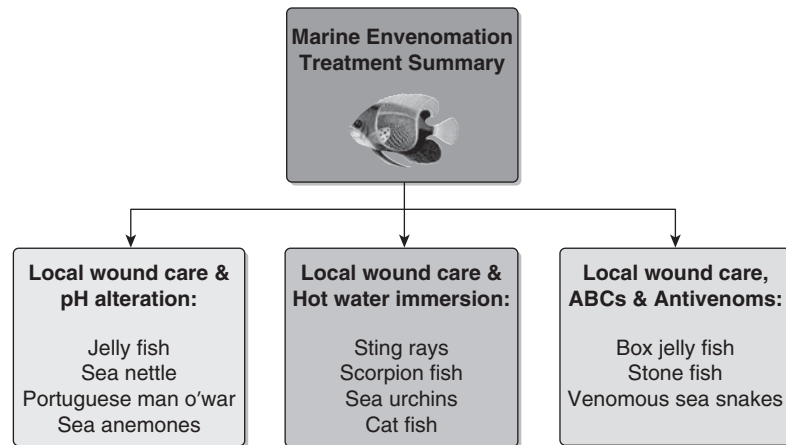


FIG. 136-1. Marine envenomation treatment summary.

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QUESTIONS

- An 18-year-old scuba diver making a beach entry accidentally stepped on a stingray while walking in the shallow waters of an ocean floor. Which of the following would be the most appropriate treatment with this injury?
 - Irrigation with sterile saline, immersion in hot water, analgesics, tetanus antibiotics.
 - Rinsing with vinegar, local wound care, analgesics, tetanus.
 - Surgical debridement to remove the sheath fragments, antibiotics, analgesics, tetanus.
 - Irrigation with sterile saline, analgesics, tetanus, and antibiotics.
 - Irrigation with sterile saline, antivenin, and tetanus.
- A 14-year-old girl is playing on the beach when she steps on a sea urchin. Which of the following is true regarding the treatment of this patient?
 - A specific antivenin exists in Australia.
 - Treatment is by immersion in hot water, removal of spines, administration of tetanus toxoid and antibiotics.
 - Treatment is by irrigation with sterile saline, rinsing with vinegar, removal of spines, administration of tetanus toxoid and antibiotics.
 - Treatment is by removal of spines, administration of tetanus toxoid and antibiotics.
 - No treatment is indicated except for local wound care, tetanus toxoid and antibiotics.
- Children shuffling their feet when entering the oceans can prevent stepping on which of the following marine life?
 - Scorpion fish
 - Stone fish
 - Lion fish
 - Sting Ray
 - All of the above

4. A 12-year-old boy is swimming in the ocean in Florida when he complains of a sudden severe stinging pain to the right leg. He is noted to have multiple linear and spiral painful urticarial lesions to the anterior thigh. He has no systemic symptoms and is otherwise without complaints. Which is the most appropriate treatment for this patient?
- Immerse the area immediately in hot water soaks.
 - Rinse area with sterile saline and apply vinegar to alter the pH.
 - Administer prophylactic antibiotics and topical steroids to the area.
 - Use fresh water to rinse off any unfired nematocysts.
 - Administer antivenom after applying ice packs to the area.
5. A 5-year-old boy sustains a marine envenomation while swimming in the ocean and very quickly develops muscular and respiratory paralysis. He is treated with respirator support, immobilization and mild compression wrapping of the affected limb, antivenin and ICU observation. Which of the following marine fauna sting or bite is consistent with this patient's clinical course and treatment?
- Stingray
 - Zebra fish
 - Jelly fish
 - Sea snake
 - Sea urchin

ANSWERS

- A. Treatment of a stingray wound includes irrigation with sterile saline or seawater to dilute the venom and remove sheath fragments, immersion in hot water for 30 to 90 min to inactivate the heat labile venom, analgesics, tetanus and prophylactic antibiotics such as trimethoprim-sulfamethoxazole, ciprofloxacin, or a third generation cephalosporin.
 - B. Treatment of these injuries includes immersion in hot water, careful removal of spines and vigorous local wound care. Tetanus immunization and broad-spectrum antibiotics prophylaxis is indicated.
 - D. All the marine life forms listed are ocean bottom dwellers, two are stoic predators, but only the stingray has the humility to move out of your way when you shuffle your feet.
 - B. This child has sustained a mild jellyfish sting. The area should be rinsed with sterile saline or seawater to maintain an isosmolar condition in order wash off unfired nematocysts and then soaked in vinegar or 5% acetic acid solution to inactivate them. Fresh water is a hypo-osmolar solution when compared to salt water and may cause spontaneous firing of unfired nematocyst, adding insult to the existing injuries. Prophylactic antibiotics are not indicated. This is not the area for Australian box jellyfish and the sting is mild so antivenom is not indicated.
5. D. The venom of the sea snake has neurotoxic, myotoxic, and nephrotoxic effects. Symptoms usually manifest within 30 minutes to 3 hours. Initial symptoms may include muscle spasms and trismus. Severe envenomations may result in acute neurotoxicity with rapid muscular and respiratory paralysis. Immediate first aid consists of respiratory support, limb wrapping, immobilization and transportation to the nearest medical facility for ICU observation and antivenin therapy.

137 DROWNING

Julie Martino
Mark Mackey

EPIDEMIOLOGY

- Drowning is a process resulting from a primary respiratory impairment due to submersion/immersion in a liquid medium.
- Highest rates are seen in the 0–4 year age range.

PATHOPHYSIOLOGY

- Pulmonary
 - Fresh water and salt water cause hypoxia by different mechanisms (Fig. 137-1).
- Neurologic
 - Hypoxia causes cerebral ischemia and edema.
- Cardiovascular
 - Hypoxia causes sinus bradycardia and peripheral vasoconstriction. The clinical picture appears similar to cardiogenic shock.
- Electrolytes
 - Mixed respiratory and metabolic acidosis is common.
 - Significant electrolyte abnormalities are rare.
 - Watch for renal failure due to acute tubular necrosis (ATN) and rhabdomyolysis.
- Hypothermia
 - Commonly seen in victims.
 - May be neuroprotective.

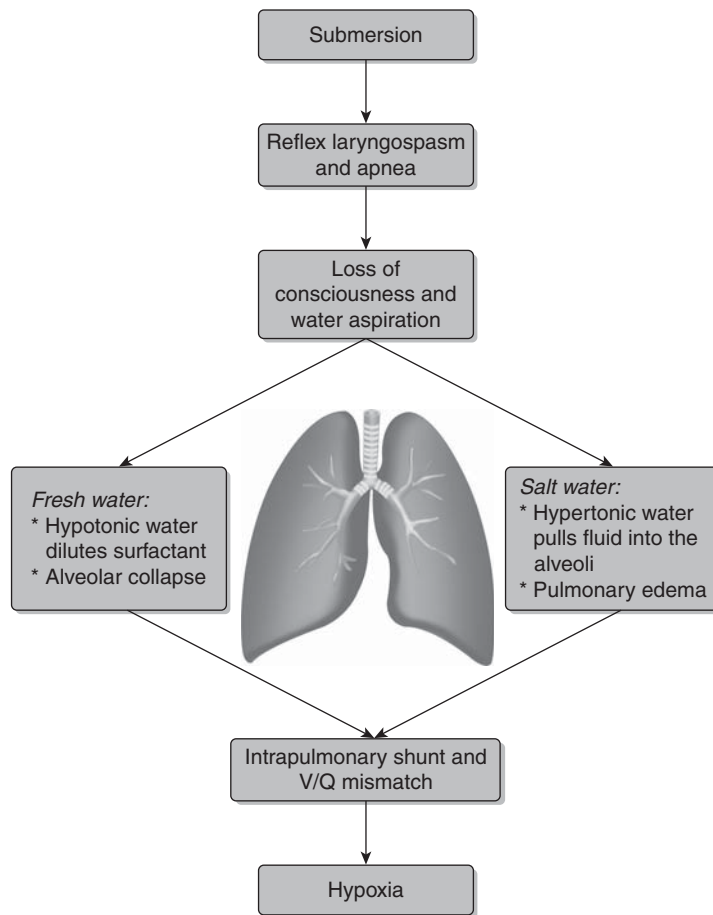


FIG. 137-1. Pathophysiology of drowning.

MANAGEMENT

- Prehospital
 - Rapid rescue from water and CPR.
 - Do not attempt the Heimlich maneuver.
- Emergency Department (ED)
 - Intubation or CPAP/BiPAP.
 - Albuterol as needed for bronchospasm.
 - Obtain chest radiograph and oxygen saturation on all symptomatic patients.
 - Observe even asymptomatic patients at least 6 hours.

PROGNOSIS

- Good prognostic indicators are short submersion time and spontaneous pulse and respirations.
- Approximately 10% of survivors have neurologic damage.

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QUESTIONS

- A 3-year-old boy is found unconscious and pulseless in a public pool. After removing the child from the water, what is the most appropriate next action?
 - Heimlich maneuver
 - Active rewarming
 - CPR
 - Assessment for injury
 - C-spine immobilization
- A 12-year-old girl was submerged underwater for three minutes. She arrives to the ER conscious and complaining of cough. What must be included in her initial workup?
 - Head CT
 - Complete blood count
 - Peak flow
 - Chest x-ray
 - Urinalysis
- The most common ECG abnormality associated with drowning is
 - Sinus tachycardia
 - SVT
 - Sinus bradycardia
 - Torsades
 - AV block
- Paramedics bring a 14-year-old boy from the local beach where he had been rescued from a rip tide current. His Glasgow Coma Scale is 15 and his pulse oximetry reads 85% on room air. Which of the following is indicated in this hypoxic patient?
 - Noninvasive positive pressure ventilation (CPAP or BiPAP)
 - Steroids
 - Antibiotics
 - Diuretics
 - Prone positioning
- A babysitter leaves an 18-month-old girl in the bathtub to answer the phone. When she returns a few minutes later, the child is not breathing. The patient is intubated by EMS and brought to the ED. Which of the following laboratory abnormalities would you expect in this drowning victim?
 - Severe hyponatremia
 - Anemia
 - Elevated carbon monoxide levels
 - Elevated lipase
 - Acidosis

ANSWERS

- C. Rapid initiation of effective CPR improves outcomes. The Heimlich maneuver is no longer recommended as it often results in aspiration. Active rewarming is not indicated in the prehospital setting and passive hypothermia may actually be protective. Assessment for injury and cervical spine immobilization are both important but should occur after airway, breathing and circulation have been addressed.
- D. All symptomatic patients should receive a chest x-ray and pulse oximetry assessment. Head CT is not universally indicated for drowning patients. CBC and urinalysis are only performed as clinically indicated and are often normal. Peak flow has no role in assessment of the drowning patient.
- C. Sinus bradycardia and atrial fibrillation are the most common arrhythmias associated with drowning.
- A. In the awake and cooperative patient, PPV has been shown to improve hypoxia. Neither steroids, antibiotics, diuretics nor prone positioning are routinely recommended.
- E. Victims will often have a mixed respiratory and metabolic acidosis due to hypoventilation and ischemia. Hyponatremia and other electrolyte abnormalities are rarely seen as most drowning victims aspirate less than 10 mL/kg of water. Anemia may occur with significant blood loss from concomitant injury, but it is not a sequela of drowning. Carbon monoxide poisoning is seen with smoke inhalation and pancreatitis may be seen with toxic ingestion but neither occurs with submersion injury.

138 BURNS

Kavitha P. Reddy

EPIDEMIOLOGY

- Burns are the fifth-leading cause of unintentional injury-related deaths in children.
- Children younger than 4 years of age are more likely to have scalding-related injuries.
- Children older than 4 years of age are more likely to have flame-related injuries.

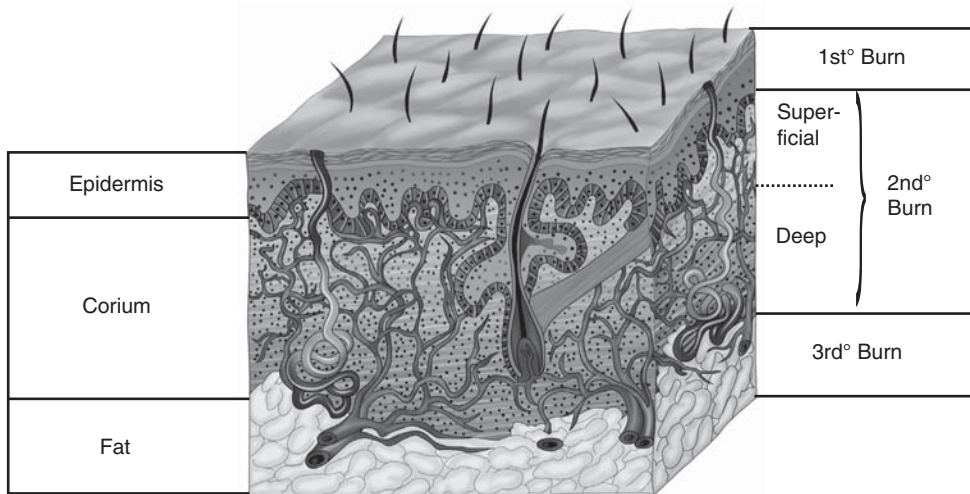


FIG. 138-1. Depth of burn wounds.

- Over the last 20 years, there has been a significant decrease in morbidity and mortality, secondary to prevention guidelines and earlier intervention.

ETIOLOGY

- Thermal: accidental spilling of hot liquids ($>120^{\circ}\text{F}$) near child.
- Flash: accidental ignition of volatile substances near child.
- Flame: house fires.
- Child abuse: injury does not match the mechanism described.

PATHOPHYSIOLOGY

- Destruction of tissue by coagulation necrosis.
- Third-spacing of fluid can result in profound intravascular hypovolemia and shock.
- Injured tissue can serve as a nidus for infection.
- First degree burns involve epidermis only, such as a sunburn (Figs. 138-1 and 138-2)
- Second degree burns or partial-thickness burns involve epidermis and part of dermis (Figs. 138-1 and 138-3).
- Third degree burns or full-thickness burns involve the entire epidermis and dermis with loss of sensation (Figs. 138-1, 138-4, and 138-5).
- Fourth degree burns extend beneath dermis to include muscle, tendon, or bone.

CLINICAL EVALUATION/ MANAGEMENT

- Primary survey focusing on airway and severity of burn.
- If $> 20\%$ body surface area (BSA) involved, establish two large bore peripheral lines and place patient on cardiac and pulse oximetry monitoring.

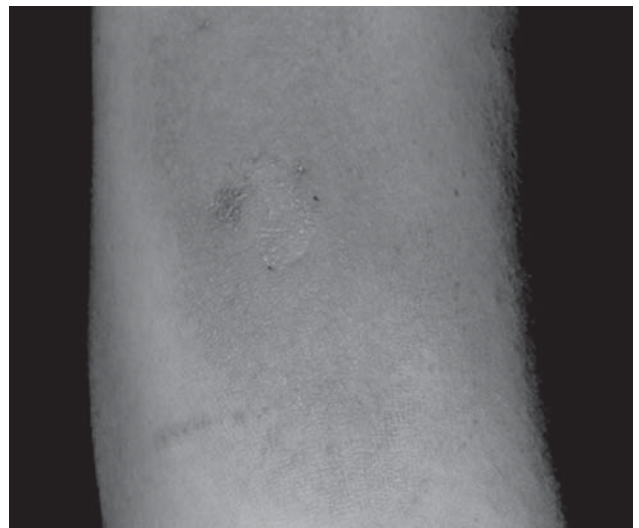


FIG. 138-2. First-degree burn.



FIG. 138-3. Second-degree burn.

- Carbonaceous sputum or singed nasal hairs should alert physician to impending airway edema and need for early intubation.
- Bronchospasm without airway edema can be treated with humidified oxygen, continuous positive airway pressure (CPAP), β -adrenergic agonists, and steroids.
- Secondary survey should include thorough examination and determination of percentage of BSA burned.
 - BSA burned can be calculated using the “rule of nines,” where the head and each upper extremity is approximately 9% BSA, anterior chest, posterior chest, and each lower extremity are 18%



FIG. 138-4. Third-degree burn.



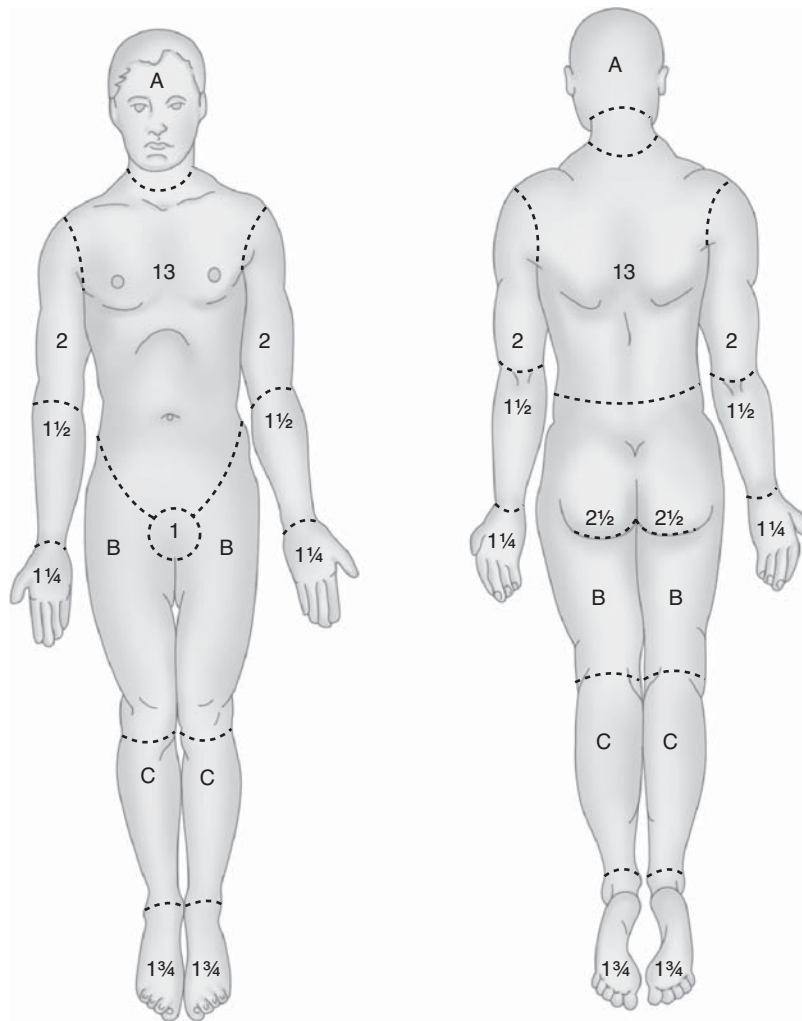
FIG. 138-5. Third-degree burn.

BSA and the perineum is 1% BSA. The Lund and Browder chart gives age-adjusted percentages (Fig. 138-6).

- Look closely for signs of impending compartment syndrome requiring escharotomy.
- Fluid resuscitation using the Parkland formula: 4 ml/kg/% BSA burned, with one-half of this volume, as lactated ringers or normal saline, given over the first 8 hours and second-half given over remaining 16 hours.
- Update tetanus status as needed.
- Pain management with intravenous opioids.
- Morphine sulfate commonly used at starting dose of 0.1 mg/kg IV.
- Initial wound care should consist of covering burns with a dry, sterile sheet. To avoid hypothermia, saline-soaked gauze should be applied only to small burns.
- Management of blisters controversial: large or hemorrhagic blisters can be debrided, but smaller blisters should be left intact
- Application of 1% silver sulfadiazine to wounds is common, but should be avoided on the face due to risk of staining of the skin

DIAGNOSTIC STUDIES

- Complete blood count.
- Chemistries.
- Coagulation studies.
- Arterial blood gas with carbon monoxide level
- Type and crossmatch.
- Creatine kinase, myoglobin and urinalysis to evaluate for rhabdomyolysis in severe burns.
- Chest radiograph.



Relative percentages of areas affected by growth (age in years)

	0	1	5	10	15	Adult
A = half of head	9½	8½	6½	5½	4½	3½
B = half of one thigh	2¾	3¾	4	4¼	4½	4¼
C = half of one leg	2½	2½	2¾	3	3¼	3½

Second degree _____ and

Third degree _____ ”

Total percent burned _____

FIG. 138-6. Classic Lund and Browder chart.

DISPOSITION

- Indications for outpatient management, admission, and transfer to burn center (Table 138-1).
- All outpatient cases should be re-evaluated in 24–48 hours to ensure proper wound healing.

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TABLE 138-1 Guidelines for Burn Triage and Disposition

Outpatient Management
Partial-thickness burn—less than 10% body surface
Full-thickness burn—less than 2% body surface
Inpatient Management
Hospital (other than burn center)
Partial-thickness burn—less than 25% body surface
Full-thickness burn—less than 15% body surface
Partial-thickness burn—face, hands, feet, perineum
Questionable burn wound depth or extent
Chemical burn, minor
Significant coexisting illness or trauma
Inadequate family support
Suspected abuse
Fire in an enclosed space
Burn center
Partial-thickness burn—more than 25% body surface
Full-thickness burn—more than 15% body surface
Full-thickness burn—face, hands, feet, perineum
Respiratory tract injury
Associated major trauma
Major chemical and electrical burns

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QUESTIONS

1. A mother brings her 2-year-old child in for examination after the child sustained a burn. Mom states she was cooking and a pot of boiling water accidentally

fell off the stove onto the child. What clinical findings would raise your suspicion for possible child abuse?

- A. A small blister on the child's right ankle.
 - B. Circumferential, symmetrical burns on the child's legs, in a stocking distribution.
 - C. Erythema and tenderness over the right anterior leg.
 - D. A small irregular area of erythema and blisters over the anterior trunk.
 - E. A region of erythema and blisters on the right cheek.
2. Parents bring their 5-year-old child to the emergency department (ED) after hot soup accidentally fell onto her right arm. They immediately come in after the accident, and you notice the child has erythematous skin of the right forearm with new blister formation. The skin is very tender to palpation. What type of burn does this child have?
 - A. First-degree burn
 - B. Second-degree burn
 - C. Third-degree burn
 - D. Fourth-degree burn
 - E. These findings are not consistent with a burn.
 3. An 11-year-old patient arrives to the ED by ambulance after being pulled from a house fire. The patient is conscious, but reports slight shortness of breath. You note pulse oximetry of 98% on room air, pulse of 95, blood pressure of 119/75, and temperature of 97.8°F. Your medical student notes that the patient is coughing up sputum which looks thick and black. What is the next appropriate step in the management of this patient's airway?
 - A. Place the patient on 2 liters of oxygen via nasal cannula and observe his respiratory status.
 - B. Discoloration of the sputum is most commonly due to smoking and without associated respiratory distress, you should not be concerned.
 - C. Carbonaceous or black sputum suggests impending airway edema and early intubation is necessary.
 - D. Continue to monitor with pulse oximetry, but no supplemental oxygen is necessary at this time.
 - E. Culture the sputum and begin empiric antibiotics
 4. You are caring for a 16-year-old patient who was admitted 24 hours earlier with burns from a house fire. During rounds, your student informs you that the patient's serum creatinine is higher than on admission and that the urine has turned dark. What diagnosis should you be concerned about?
 - A. Myoglobinuria. The patient's urine will show large blood without evidence of red blood cells

- on microscopic analysis. There will also be elevation of the creatinine phosphokinase (CPK).
- B. Dehydration. The BUN/creatinine will be elevated in a ratio of 15–20:1.
 - C. Carbon monoxide. The carboxyhemoglobin level will be high.
 - D. Hyperkalemia. The serum potassium level will be high.
 - E. Urinary tract infection. The urine will show high bacterial content along with increased white blood cells in the urine.
5. You are on call at a trauma center and the paramedics bring you a 10-year-old (45 kg) male involved in a house fire. He is found to have approximately 25% of his BSA burned. According to the Parkland formula, how much fluid should you give this patient over the first 8 hours?
 - A. 1.8 L
 - B. 1.0 L
 - C. 2.25 L
 - D. 4.5 L
 - E. 500 mL
 6. You are working in a community hospital when a 9-year-old female is brought in due to a small second-degree burn on her right anterior trunk. She does not meet criteria for inpatient management and you decide to discharge her home. Before she leaves, how should you dress her wound?
 - A. Clean the wound with hydrogen peroxide and then cover with gauze.
 - B. Place saline-soaked gauze on the wound.
 - C. Apply a topical antibiotic, such as 1% silver sulfadiazine ointment, to the wound and cover with dry, sterile gauze.
 - D. Leave the wound open to the air to ensure faster healing.
 - E. Place povidone iodine on the wound and leave open to air.
 7. You are at work at a small rural hospital and are caring for a 3-year-old male with second-degree burns to 9% of his body which include the scattered areas to anterior chest and left anterior thigh. The patient's pain is well controlled and you are trying to decide on the appropriate disposition. Where would you send this patient?
 - A. Transfer to a burn center.
 - B. This patient needs inpatient management, but no transfer.
 - C. This patient can be discharged home and does not need any follow up due to the small size of the burns.
 - D. Have a burn specialist come into your hospital to see the child.
 - E. Discharge the patient home and have a follow up arranged within two days.
 8. A 6-year-old male presents to your ED, brought in by his father, with complaints of burns to both arms. He appears to have diffuse first-degree burns to both of his forearms. It is circumferential from his elbows to his hands. His neurovascular status is intact. Using the "rule of nines." What percentage of BSA is involved?
 - A. 18%
 - B. 5%
 - C. 2%
 - D. 9%
 - E. 12%
 9. A 5-year-old male is brought into your trauma center after sustaining a full-thickness burn to his perineum. Your resident tells you that only 1% BSA is burned and therefore, he would like to send the patient home with pain medicines, topical antibiotics, and follow up. What is the appropriate disposition for this patient?
 - A. Your resident is correct and you should discharge the patient home.
 - B. Transfer the patient to the nearest burn center.
 - C. Admit the patient to your hospital and give intravenous pain medicines and fluids.
 - D. Admit the patient to psychiatry since this type of wound is commonly self-inflicted.
 - E. Observe the patient in the ED for 6 hours and then discharge him home if there is no evidence of urinary obstruction.

ANSWERS

1. B. Symmetrical stocking distribution burns do not match this mechanism of injury, especially since they are bilateral and circumferential. The most likely explanation for this type of burn would be a submersion injury, which should always raise suspicion for child abuse. Blisters or erythema on the anterior leg, face, or trunk are consistent with hot liquids accidentally falling on the child.
2. B. Second-degree burns are also known as partial-thickness burns. They often involve the epidermis and part of the dermis; therefore, you will see erythema and blister formation. Because the nerve endings are preserved, sensation will be intact, unlike third degree or full-thickness burns which damage nerve endings, rendering the burn painless.
3. C. The danger for victims of house fires does not just involve surface area of burns. Inhalation of

smoke can cause significant damage to the airway. Any sign of singed nasal hair or carbonaceous sputum should alert the physician to impending airway edema as this suggests significant inhalation injury. This finding along with a patient's subjective complaints of shortness of breath should be cause for early intubation. If a physician waits until the patient is in respiratory distress, the airway edema may have progressed too far, making intubation impossible.

4. A. While all of these problems can happen in burn patients, this specific scenario describes the diagnosis of acute tubular necrosis, which occurs because of muscle breakdown with increased myoglobin in the tubules of the kidney. This obstructive pathology leads to necrosis of renal tubule cells and the rise in serum creatinine signals renal failure. Classically, one will find large blood without RBCs on urinalysis, pointing towards myoglobinuria from rhabdomyolysis or muscle cell breakdown (rising CPK).
5. C. The Parkland formula (4 mL/kg/ %BSA burned) is often used to estimate fluid requirements over 24 hours in patients with large surface area burns. In this patient who weighs 45 kg with 25% BSA burned, his fluid requirements over 24 hours would be 4.5 L (4500 mL). The physician should give half of this requirement over the first 8 hours and the remainder over the next 16 hours. The question asks for the amount of fluid to be given over the first 8 hours, therefore the answer in 2.25 L (2250 mL).
6. C. The outpatient management of burn wounds is a topic of debate, but current recommendations suggest applying a topical antibiotic and covering with dry, sterile dressings. Saline-soaked gauze may be used initially for wound care in the ED, but moisture may promote infection if left in place at discharge. Silver sulfadiazine is routinely used on burn wounds, with the exception of the face, as there is some concern about discoloration or scarring. If the patient is being transferred to a burn center where physicians will need to look at the wounds again, wounds are covered with dry, sterile sheets without using dressings at all. Do not use hydrogen peroxide or iodine, as these can cause more tissue destruction.
7. E. For second-degree burns or partial-thickness burns, a patient with less than 10% BSA burned can safely be discharged home. All burn patients should have close follow up to assure healing of the wound without infection. If the patient has between 10 and 25% BSA burned, inpatient management is prob-

ably necessary, but the patient does not necessarily need transfer to a burn center unless there are major injuries or respiratory compromise or the wounds involve the face, hands, feet, or perineum. If the patient has partial-thickness burns involving greater than 25% BSA burned, they should be transferred to a burn center.

8. D. Using the "rule of nines," this patient has 9% BSA burned. Each arm is 9%, therefore, half of the arm is 4.5%. Since both arms are involved, you would multiply this estimate by two, giving you 9%. First degree burns, such as sunburns, do not require hospitalization or transfer. Pain control may be this patient's biggest issue.
9. B. Although this patient has only 1% BSA of burns, he has injury to his perineum. Any partial or full-thickness burn to the perineum requires transfer to a burn center for wound care. It would be inappropriate to keep the patient in your ED or hospital as this patient could suffer long-term consequences from this type of burn. Although it is not clear how he sustained the burn to just this area, the primary concern at this time is management of his wound. Psychiatric and child abuse issues may become a concern after his initial treatment at a burn center.

139 ELECTRICAL AND LIGHTNING INJURIES

Mary Ann Cooper

ELECTRICAL INJURIES

- Electrical injuries may result in massive tissue destruction, changes in growth patterns, and neurologic injury, including chronic pain syndromes and permanent cognitive deficits, affecting the child's ability to learn and become a productive adult.
- Children at most risk are exploring toddlers (12–30 months) and adolescents.
- The majority of victims are male.

ELECTROPHYSIOLOGY

- Voltage and amperage are not predictive of injury or disability.
- Electric field strength is a more useful and accurate concept in explaining and predicting injuries from technical or man-made electricity than the classical

Kouwenhoven factors that have traditionally been cited in the medical literature.

- Electricity is a “volume injury” and does not go along nerves and blood vessels, as taught in the older literature.
- Expect more damage at joints and smaller cross sectional tissues.
- Damage to internal organs may occur and is frequently missed early on.

MECHANISMS OF INJURY

- There are several common mechanisms of injury (Table 139-1).
- Concussive/blunt injury may occur from being thrown or falling.

ANATOMIC SITES OF INJURY

- *Surface burns* may be found in many nonwater-related electrical injuries.
- Deep electrical injuries tend to resemble crush injuries in their level of tissue destruction.
- The most common areas of injury are the hand, skull, and foot.
- Compartment syndromes may occur as edema and vascular thrombosis affects either inflow or outflow of blood to tissue.
- Delayed bleeding and orofacial growth abnormalities may result from oral injuries.
- Current passing directly through *the heart* can induce ventricular fibrillation and a number of arrhythmias.
- *Vascular injuries* include thrombosis, vasculitis with necrosis of large vessels, vasospasm, and late aneurysm formation. Strong peripheral pulses do not guarantee vascular integrity.
- *Acute renal failure* may occur with inadequate initial fluid resuscitation.
- Immediate *CNS* effects include loss of consciousness, agitation, amnesia, deafness, seizures, visual disturbance, and sensory complaints.
- A variety of autonomic disturbances also occur.
- *Cataracts* can be seen in any electrical injury involving the head or neck.

TABLE 139-1 Mechanisms of Electrical Injury

Contact injury
Flash burn
Arc burn
Secondary ignition
Concussive force
Blunt trauma

- Abdominal and other internal organ damage is frequently missed during the early care.
- Blunt trauma, falls, or tetanic muscle contractions can cause *fractures or dislocations*.
- Neurocognitive deficits may occur.

MANAGEMENT

PREHOSPITAL CARE

- Extrication may be extremely dangerous until the power source is safely disconnected.
- Victims should be treated both as burn victims as well as blunt trauma patients.
- Aggressive fluid therapy is essential to sustain circulation and begin diluting myoglobin.

EMERGENCY DEPARTMENT CARE

- The greatest threats to life include cardiac arrhythmias, renal failure from myoglobin and hemoglobin precipitants, and hyperkalemia from massive muscle breakdown.
- Tests to be considered include arterial blood gases, complete blood count, serum electrolytes, blood urea nitrogen, serum creatinine, glucose, type and cross-match and urine for myoglobin. Creatine kinase (CK) is not predictive of the degree of injury.
- Radiographs should be ordered as clinically indicated.
- Adequate fluids should be given to maintain a urine output of 1 to 2 mL/kg/h when pigmentation is present and less after it has cleared.
- A decreasing level of consciousness, unexplained coma, lateralizing signs, or change in mental status necessitates cranial computed tomography (CT) scan to rule out intracranial damage.
- Fasciotomy and escharotomy may be necessary in some cases, particularly if the chest wall is involved. Amputations are common with major injury.
- Compartment syndromes can occur if venous output is blocked by thrombosis and as tissue edema occurs.
- Debridement is best left to a burn surgeon and should be conservative for lip burns.
- Tetanus prophylaxis should be given as needed
- Consultations may be required particularly for severe cases.
- All children with oral injuries require plastic surgery and dental or orthodontic consults.
- Transfer to a burn center may be indicated.

- Lesser injuries and small burns can be treated conservatively with few or no laboratory tests or x-rays.
- Baseline ECG and cardiac monitoring is not indicated for children exposed to household current (120–240 V) unless there was loss of consciousness, tetany, wet skin, transthoracic current flow, or the event was unwitnessed.

PREVENTION

- Extension cords should be in good repair and not used to replace or avoid conduit wiring.
- Unused outlets should be covered with dummy plugs.
- Electrical appliances must be kept away from sinks and bathtubs.
- Electrically operated toys should be age-appropriate and supervised by adults.
- Older children and adolescents can benefit from school safety programs that address the dangers of power lines and transformer substations.

LIGHTNING INJURIES

- Lightning kills more people annually in the United States than any other storm-related phenomenon except floods (Fig. 139-1).
- More males are injured than females at every age and in every country.
- 90% of those injured by lightning survive.
- A significant number of survivors will have disabilities from chronic pain or neurocognitive injury.

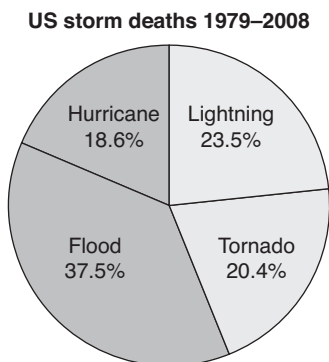


FIG. 139–1. Weather causes of U.S. storm deaths (1979–2008).

MECHANISMS OF INJURY

- Lightning is dangerous for three reasons: electrical effects, heat production, and concussive force.
- Mechanisms of Injury are illustrated in Figure 139-2. See Table 139-2 for the estimated frequency of each mechanism.

TYPES OF INJURIES

- *Cardiac arrest* at the time of the injury is the only proximate cause of death from lightning. Survivors frequently have *anoxic injury*.
- Congestive heart failure, cardiac contusions, and delayed rupture of the heart have also been reported
- ECG changes include nonspecific ST–T wave changes, T-wave changes, axis shift, QT prolongation, and ST-segment elevation and usually resolve gradually.
- *Lung injuries* reported include pulmonary contusion, hemorrhage, pneumothorax, pulmonary edema, and aspiration secondary to altered mental status
- *Arterial spasm and vasomotor instability* result in cool, mottled, pulseless extremities.
- *Keraunoparalysis*, especially of the lower extremities, almost always resolves after a few hours although permanent paresis has been reported.
- *Transient loss of consciousness, retrograde amnesia, and paresthesias* are common.
- Less than one-third of lightning survivors have any *external signs of burns*.
- Other neurologic findings may include seizures, skull fracture, intracerebral hemorrhages, elevated intracranial pressure, cerebellar ataxia, Horner’s syndrome, SIADH, and peripheral nerve damage.
- *Cognitive injury* similar to other blunt head injury commonly occurs but may not be recognized acutely.
- *Deep tissue damage and myoglobinuria* is very rare.
- “Entry” or “exit” areas rarely, if ever, occur with lightning.
- *Postconcussive headaches* lasting weeks to months are common.
- *Cataracts* may occur, developing either immediately or over a prolonged recovery period. *Other eye injuries* are common.
- *Tympanic membrane rupture* is common.
- *Other aural complications* include burns to the ear, ossicular disruption, tinnitus, vertigo, and nystagmus.
- *Gastric upset* similar to postconcussive syndrome is not uncommon for weeks to months after the injury.

- *Psychological sequelae*, including anxiety, sleep disturbances, nocturnal enuresis, depression, PTSD, and cognitive disability, have all been reported.

MANAGEMENT

PREHOSPITAL CARE

- Lightning injury victims should be approached as blunt multiple trauma patients with attention to advanced life-support protocols and cervical spine protection

- “Reverse triage” (resuscitation of the apparently dead) is the rule and best use of manpower.
- Emergency personnel are at risk when working with victims near active thunderstorms.

EMERGENCY DEPARTMENT CARE

- Treatment follows the same guidelines as for all severely injured patients including the ABCs and cervical spine control.
- Search for blunt injuries

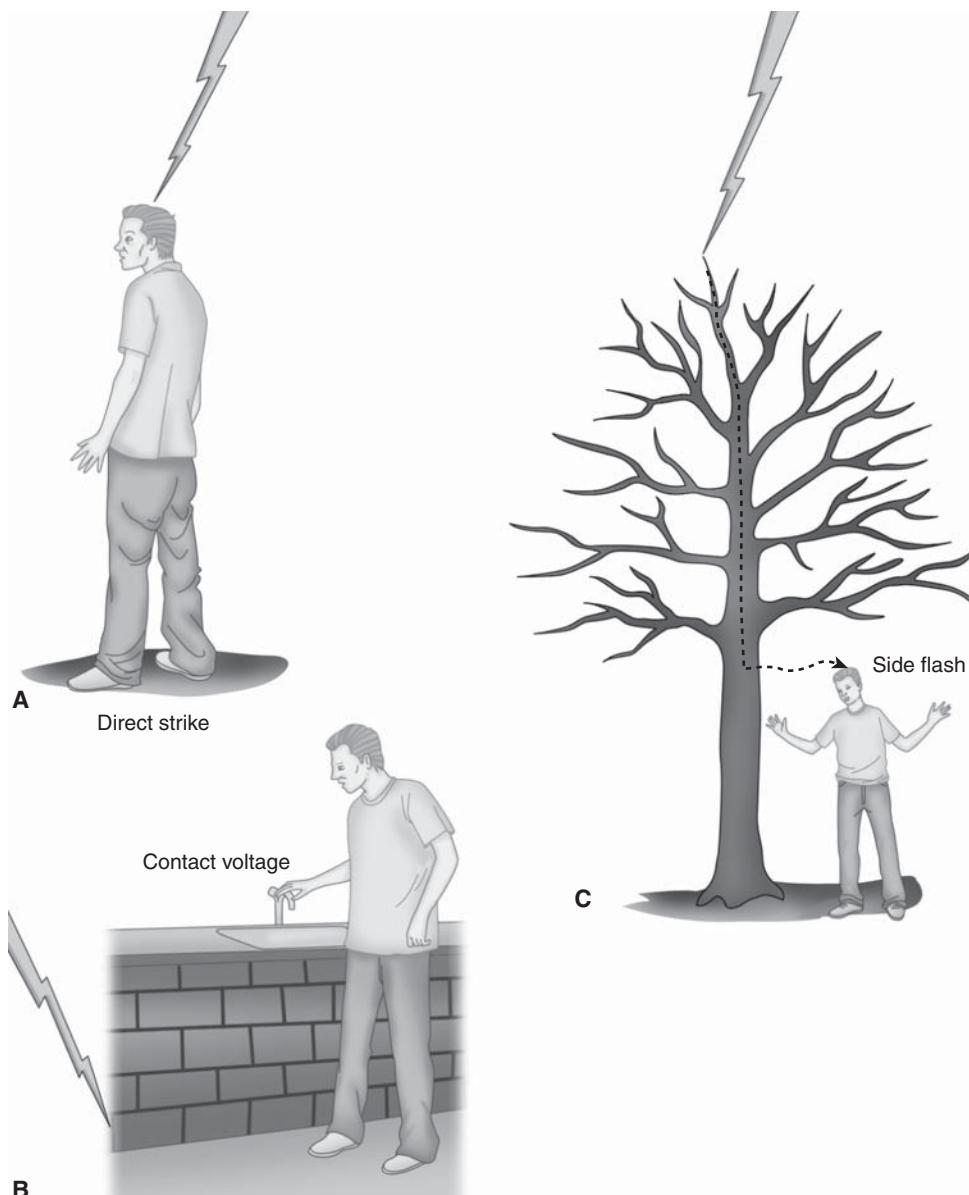


FIG. 139-2. Mechanisms of lightning injury.

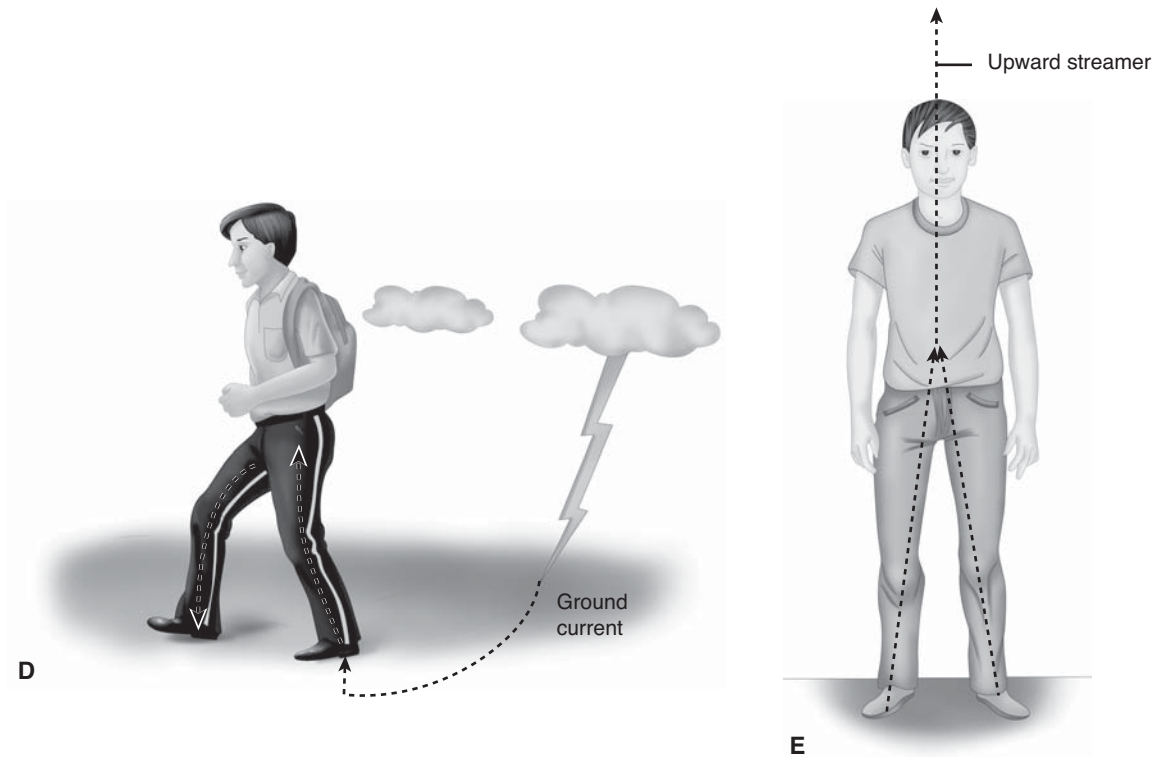


FIG. 139-2. (Continued)

- A baseline ECG is also required. Cardiac monitoring is only indicated if there was a cardiac arrest or abnormal initial ECG.
- Any arrhythmia should be treated by standard protocols.
- Fluid resuscitation must be approached cautiously; central monitoring lines may be helpful.
- Laboratory tests are likely to be completely normal except in the most severely injured.
- Radiographs are done as indicated. Cranial CT scan is indicated for all unconscious patients.
- Burns should be treated by protocol.
- Eye and ear examinations should be done with special attention paid to looking for TM rupture.
- Tetanus prophylaxis may be indicated.

DISPOSITION

- The vast majority of lightning survivors can be discharged from the emergency department.
- Exceptions include postcardiac arrest, unstable or confused patients, or those with inadequate home supervision, and close follow-up care.

SEQUELAE

- Long-term sequelae may include postconcussive syndrome, cognitive disability leading to school performance issues, chronic pain syndromes, delayed and often atypical seizures and disturbances in balance, coordination, mood, sleep, affect, and memory.
- Some of these may be especially difficult to appreciate in adolescents.

TABLE 139-2 Estimated Frequency of Mechanisms of Lightning Injury

MECHANISM	FREQUENCY (%)
Direct strike	3–5
Contact potential	15–25
Side splash/flash	20–30
Ground current	40–50
Upward streamer/leader	10–15

PREVENTION

- “When Thunder Roars, Go Indoors”—No place outside is safe when thunderstorms are in the area.
- The vast majority of lightning injuries are preventable if one knows and follows lightning injury prevention guidelines listed in Table 139-3.

TABLE 139-3 Lightning Injury Prevention Guidelines

Know the weather forecast beforehand if outdoor activities are planned.

Change plans if thunderstorms or severe weather are in the forecast for the area and time of the activity.

If one must be outdoors, have a safety plan thought out that includes a safer place (substantial buildings or fully enclosed metal vehicles) for evacuation. Be sure there is time to reach it.

Have a “weather eye” to the sky to watch for threatening weather.

When Thunder Roars, Go Indoors!
(If thunder is heard, immediately seek a safer area.)

Do not resume outdoor activity until thirty minutes after the last lightning is seen or thunder heard.

When indoors, do not touch conducting materials such as hard-wired phones, game controllers, computers, and plumbing when thunderstorms are in the area.

Adults are always responsible for the safety of children in their care.

A wonderful teaching tool is the *Leon the Lightning Lion Safety Game*, written for preschoolers and nonreaders but also useful for older children and adults. This and other teaching materials, posters, public service announcements by prominent sports figures, and games are available at the *National Lightning Safety Week Web site*.

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QUESTIONS

- EMS brings a 2-year-old to the emergency department (ED). The child has a mouth wound, apparently from sucking on an electrical extension cord. The child is alert, playful and in no distress. Which of the following is the most appropriate treatment for this patient?
 - Aggressive debridement of the burn.
 - Tetanus prophylaxis.
 - Plastic surgery consult.
 - Aggressive fluid resuscitation.
 - Urinalysis for myoglobinuria.
- The most appropriate disposition for the above patient would be which of the following?
 - A 6-hour observation in the ED for cardiac monitoring.
 - Hospital admission to a monitored bed.
 - Transfer to a burn unit.
 - Discharge home to follow up with a pediatric neurologist in 3–6 months.
 - Discharge home with instructions to the parents about delayed bleeding and how to control it.
- Parents bring a 9-year-old child to the ED. She was playing tag in their backyard with friends and received a shock from the electrified fencing used for their three cows. Her vital signs are stable, she has no signs of burns and she appears to be in no distress. Emergent care for this patient should consist of which of the following?
 - Reassurance of the parents and discharge to home.
 - CK level.
 - ECG and cardiac monitoring.
 - Urinalysis for myoglobinuria.
 - Tetanus prophylaxis.
- A 15-year-old child is brought to the ED by his parents. The child tells you that he was shooting insulators on a pole at his parents farm for target practice. When one of the wires fell to the ground more than a dozen yards away, he experienced a tingling shock and was knocked to the ground without loss of consciousness. He was able to get up and walk back to the farmhouse. He is alert and anxious, but has no sign of burns, his vital signs are stable and his pulse is regular. His ED management should include:
 - Discharge with referral to his primary care physician for follow-up.
 - CK level.
 - ECG and cardiac monitoring.
 - Urinalysis for myoglobinuria.
 - Tetanus prophylaxis.

5. EMS brings a 10-year-old boy to the ED. He unintentionally came in contact with a high voltage line when he was trying to retrieve a kite. He is confused and has extensive burns to his right hand, across his trunk and on the sole of his left foot. His care in the ED should include which of the following?
- ATLS protocol and transfer to a burn unit that cares for electrical injuries.
 - Debridement of the burns.
 - ECG and cardiac monitoring.
 - Urinalysis for myoglobinuria.
 - Tetanus prophylaxis.
6. A man brings his 18-year-old wife who is 32 weeks pregnant to the ED after she experienced a tingling feeling on her screened-in porch during a thunderstorm. She had no loss of consciousness or fall. She is alert, oriented and has no burns. Care of the woman should include
- Immediate induction of labor.
 - Referral to L&D for fetal monitoring and evaluation.
 - Fluid resuscitation.
 - ECG and cardiac monitoring.
 - Hospital admission to a monitored bed.
7. An 11-year-old boy is brought to the ED by his mother two days after experiencing a shock in the shower shed at a distant summer camp during a thunderstorm where a nearby power line was downed. He is anxious and complains of muscle pain but has no external signs of injury. It is unclear if he had loss of consciousness. His treatment should include
- Discharge with referral to his primary care physician for follow-up.
 - CK level.
 - ECG and cardiac monitoring.
 - Urinalysis for myoglobinuria.
 - Tetanus prophylaxis.
8. A 17-year-old male is brought to the ED after receiving a shock while camping two days before on an outward bound experience. He was sleeping in a two-man tent and both he and his tent mate (who is not with him) were confused after the shock, nauseated and unable to eat for two days. He began experiencing visual problems, dizziness, and trouble with fine motor movement. His visual acuity in the ED is OD 20/100, OS 20/70. He did not wear glasses previously. He is a star baseball player and is being scouted for college scholarships. His ED care should include which of the following?
- Ophthalmology consult
 - CK level
 - ECG and cardiac monitoring
 - Urinalysis for myoglobinuria
 - High-dose steroids
9. A 16-year-old Ultimate Frisbee player is brought to the ED by EMS with a report that lightning hit the national tournament playing fields. At least one player was killed. The patient reportedly had a cardiac arrest but now has a pulse. She is unresponsive to verbal stimuli. She has no external signs of injury but is wet and cold. Her lower extremities are mottled with no palpable pulses. Which of the following should be initiated in the ED?
- Begin hyperbaric oxygen therapy.
 - Induce hypothermic coma.
 - Consult surgery for fasciotomies on her lower extremities.
 - Admit to the ICU/CCU with monitoring and cardiology consults.
 - Reassure the parents that she will probably recover without sequelae since there are no signs of burns.
10. A 12-year-old was riding her bicycle home from school when a thunderstorm arose. She was found on the roadside by a passing motorist who called 911. On presentation to the ED, she is moaning softly and responds to pain but is otherwise incoherent. Her vital signs are stable and appropriate for her age. She has scattered arborescent markings on her left neck, trunk, and leg. She has multiple abrasions. How should her ED management proceed?
- Begin hyperbaric oxygen therapy.
 - Induce hypothermic coma.
 - Consult surgery for fasciotomies on her lower extremities.
 - Admit to the ICU/CCU with monitoring and cardiology consults.
 - Assess as a multiple trauma patient including head CT.

ANSWERS

- C. Debridement should be conservative and left to the plastic surgeon/oral surgeon/pediatric orthodontist. The child will not need tetanus prophylaxis if she has had her DPT series. Fluid resuscitation is not warranted for a local burn in an alert patient. A dipstick can be done for myoglobin if curious but it is highly unlikely that it will be positive.
- E. This patient can be discharged home with the parents but because of labial artery bleeding that occurs in some children with eschar separation,

- parents should be warned of this and instructed how to manage it. An ECG could be done but with a normal pulse in an alert child it is highly unlikely that it will show anything that needs to be treated. Cardiac monitoring, blood tests, and admission are not warranted.
3. A. The current in a cattle fence is insufficient to cause any problems and the child can be safely discharged.
 4. A. While the physician could elect to do any of these tests based on the presentation, it is unlikely that any tests will show anything of consequence. Since he is alert, able to communicate, has stable vital signs and no burns, he can be safely discharged with NSAID/acetaminophen for any pain control that may be necessary and follow-up with his primary care physician.
 5. A. While all of these can be done (with the exception of debridement which is not indicated in the ED), the most important care would be stabilization, fluid resuscitation and transfer to a burn unit with experience in electrical trauma and burn care. CK levels are not indicative of the degree of burn and do not guide care.
 6. B. Minor static discharges/tingling are not serious and need only follow-up if there are any signs or symptoms that develop in the next few days. Therefore, the patient could be safely discharged. On the other hand, the woman and her husband were concerned enough about this to come to the ED which probably warrants evaluation of the pregnancy by L&D to provide fetal monitoring and reassurance.
 7. A. This is a real case and it was unclear whether the boy had a lightning discharge contact injury or an electrical injury. Although the physician may elect to do some of these tests both for his curiosity as well as the parent's reassurance, it is unlikely that any will be positive. However, if they are, they certainly indicate more substantial injury. The boy developed significant learning disabilities and discoordination after the injury.
 8. A. This is another real case. This young man had bilateral macular holes and developed a cataract in one eye. His depth perception was remarkably affected and he was no longer able to play baseball. Except for muscle aches and pains that lasted for months, he had no other continuing symptoms and was able to finish high school and is now in medical school. While the ophthalmologist began high-dose steroids and subsequently reported the case in the literature, there is no evidence that the steroids had a significant impact on the outcome.
 9. D. Multiple people may be injured by the same storm. Anoxic brain injury is a real possibility with this patient. The use of hypothermic coma has been reported in only one case; the use of hyperbaric oxygen has not been reported. Keraunoparalysis (vascular spasm, cold, mottled, pulseless extremities) usually resolves over a period of hours so that fasciotomies are not indicated. Vascular flow to the extremities can be confirmed by Doppler in the ED.
 10. E. While this girl presumably suffered a lightning injury (Lichtenberg figures are pathognomonic for lightning), she might also have sustained blunt trauma and head injury when she left her bicycle or if she was hit by a car. She should be evaluated as a trauma patient.

140 HEAT AND COLD ILLNESS

Heather M. Prendergast

HEAT ILLNESS

- Spectrum self-limited (heat cramps) to life-threatening (heatstroke).
- *Predisposing factors:* poorly developed thermoregulatory systems in infants; lesser sweating capacity in exercising children and slower rate of acclimatization in children.
- *Risk factors:* Dehydration, excessive clothing and bundling, infections, mental retardation, obesity, previous episodes of heatstroke
- *Pathophysiology:* The body generates heat at 1°C/hour and heat production increases 12-fold with heavy exertion. Mechanisms of heat dissipation are evaporation, conduction, convection, and radiation
- *Heat Cramps* are involuntary skeletal muscle spasms that occur independently of weather and temperature. Suspected etiologies include alterations in the spinal neural reflex activity stimulated by fatigue in susceptible individuals and dilutional hyponatemia.
- Manifestations of heat cramps include muscle spasms often resistant to narcotics in the absence of adequate fluid rehydration that occur in the setting of normal body temperature with active sweating but in the absence of central nervous system signs.
- Management priorities involve moving the patient to a cool environment and providing rest and oral

electrolyte solutions. Avoid use of replacement fluids with caffeine or alcohol. Salt tablets cause gastrointestinal cramping and are not recommended.

- Signs and symptoms of *heat exhaustion* are dizziness, postural hypotension, nausea, vomiting, headache, weakness, and syncope. The temperature may be normal or moderately elevated (39°C–41.1°C). Heat exhaustion occurs in unacclimatized individuals and is associated with profuse sweating and normal mental status.
- Etiologies of heat exhaustion are salt depletion which occurs when fluid losses are replaced by water or hypotonic solutions and water depletion which occurs when fluid losses are unable to be replaced leading to hypernatremic dehydration.
- Management involves moving the patient to a cool environment and providing rest and intravenous rehydration with normal saline unless hypernatremic dehydration is suspected. Patients may be discharged home if all symptoms have resolved during emergency department treatment and observation.
- *Heat stroke* is the most severe form of heat illness and represents complete thermoregulatory failure. Mortality ranges from 17% to 80%. Manifestations include disorientation, seizures, and coma.
- Classic heatstroke occurs at the extremes of age (infants and the elderly). Classic heatstroke develops over a period of days. The skin is hot and dry.
- Exertional heatstroke is more likely in the pediatric population. The skin may be dry or wet. The risk is greatest in individuals performing high-intensity exercise over a short time span. Predisposing conditions include dehydration, obesity, neurologic disorders, hyperthyroidism, extremes of age, alcohol consumption, and sickle cell trait. Some medications, such as phenothiazines, anticholinergics and diuretics, interfere with heat dissipation.
- Complications of heat stroke include neurologic dysfunction (encephalopathy), acute tubular necrosis, hepatic failure, disseminated intravascular coagulation (DIC), adult respiratory distress syndrome, and multiorgan system dysfunction.
- Management priorities begin with stabilization of airway, breathing, and circulation. Cooling measures should be instituted immediately. Spraying the skin with room-temperature water and directing an electric fan onto the patient is an excellent way to begin. Ice packs can be placed in the groin and axilla, but avoid applying ice water directly to skin, since this causes vasoconstriction and impairment of heat dissipation. Monitor core temperature continuously until it reaches 39°C.
- Diazepam, 0.2 to 0.3 mg/kg/dose IV, may be required to prevent shivering. Antipyretics are ineffective.

- Intravenous fluids are given as isotonic crystalloid at a rate of 20 mL/kg over the first hour.
- Diagnostic testing may include arterial blood gases, complete blood count (CBC), serum electrolytes, urinalysis, liver enzymes, coagulation studies, CT of the brain and electrocardiogram. Findings may include hyponatremia or hypernatremia, hyperkalemia secondary to rhabdomyolysis and elevated specific gravity. Serum transaminases are usually elevated, peak in 24 to 48 hours and correlate with severity of injury. The patient is evaluated for evidence of disseminated intravascular coagulation, intracranial pathology, and myocardial infarction.

COLD ILLNESS

- Hypothermia can be separated into three categories: mild (core body temperature 35°C to 32°C), moderate (core body temperature 32°C to 30°C), and severe (core body temperature less than 30°C).
- Predisposing factors include younger age (large surface area to body mass ratio, paucity of subcutaneous tissue and poorly developed thermoregulatory systems) and adolescence (winter sports, risk-taking and inexperience).
- *Pathophysiology*: Skin receptors stimulated by cold exposure result in peripheral vasoconstriction and heat conservation. As the temperature declines, stimulation of the preoptic anterior hypothalamus occurs. Shivering increases heat production 4–5 times, but ceases at temperature of 31°C and is absent in neonates.
- Cardiovascular findings are the most noticeable and include transient tachycardia followed by bradycardia, decrease in mean arterial pressure, and decrease in the cardiac output. The J-wave (Osborn wave) is pathognomonic for hypothermia but has no prognostic or predictive value. The J-wave is an notching at junction of the QRS complex and ST segment.
- Benign atrial dysrhythmias are common at body temperature <32°C; ventricular ectopy with risk of ventricular fibrillation occurs at <30°C. Asystole typically occurs at <19°C.
- Neurologic signs and symptoms include poor judgment, apathy, confusion (<33°C), ataxia (<33°C), coma (27°C). There is a linear decrease in cerebral metabolism with decreasing temperature. Autoregulation fails at 25°C. The electroencephalogram shows a flat line at 20°C.
- Respiratory findings include a transient stimulation of respiratory drive followed by a decline in minute ventilation with decrease in temperature, bronchorrhea, and pulmonary edema.

- Gastrointestinal findings include decreased motility, gastric dilatation, ileus, constipation, poor rectal tone, and inflammatory changes in the pancreas.
- Renal effects include cold diuresis and dilution of the glomerular filtrate.
- Diagnosis is based on the core temperature but diagnostic tests are useful for monitoring and detection of complications.
- *Arterial Blood Gases* will reveal evidence of decreased tissue perfusion and metabolic acidosis. The oxyhemoglobin dissociation curve is shifted to the left.
- *CBC* is used to establish a baseline. The hematocrit increases 2% for each 1°C drop in temperature. Hemoglobin may be decreased due to blood loss or chronic illness. White blood count is reduced by sequestration and bone marrow depression.
- *Serum electrolytes* should be monitored during the rewarming process.
- *Renal function tests* establish the baseline renal function but are poor indicators of fluid status in hypothermia. Acute tubular necrosis may develop after rewarming.
- *Serum glucose* is elevated due to catecholamine effect and insulin inactivity below 30°C.
- Persistently elevated levels suggest pancreatitis or diabetic ketoacidosis. Hypoglycemia may develop due to inadequate glycogen stores in neonates and malnourished children.
- *Clotting* studies are indicated in cases of moderate-to-severe hypothermia and may demonstrate cold induced thrombocytopenia and prolonged clotting times. Persistent changes after rewarming suggest development of disseminated intravascular coagulation.
- *Amylase and lipase* may be elevated due to pancreatitis that is associated with poor outcome.
- *Urinalysis* shows low specific gravity secondary to cold diuresis.
- *Cultures of body fluid may reveal sepsis* which is a common cause of hypothermia in infants.
- *Chest radiograph* is indicated in all cases of significant hypothermia. Pulmonary edema may develop during rewarming. Aspiration is relatively common.
- Cervical spine films may be indicated if there is suspicion of trauma.
- Cranial computed tomographic scanning is indicated in the setting of trauma or to search for other etiologic factors, especially when mental status does not clear along with rewarming.
- *Electrocardiogram* is indicated for all patients with a core temperature <32°C to detect dysrhythmias or evidence of myocardial ischemia. The J-wave (Osborn wave) is seen when the temperature falls below 32°C.
- In prehospital care, the first priority is basic life support while maintaining a high index of suspicion for hypothermia. Warm resuscitation fluids are used and wet clothing is removed. Heated, humidified oxygen is used when available. Cardiac monitoring is used to detect dysrhythmias.
- Management priorities in the ED begin with airway, breathing, and circulation. In patients who are obtunded or who have absent protective airway reflexes, early endotracheal intubation should be accomplished. Fluid resuscitation is guided by vital signs, urinary output, and pulmonary status. Cardiac monitoring is continued.
- For pulseless patients, cardiopulmonary resuscitation (CPR) is maintained. If initial defibrillation attempts are unsuccessful, rewarm to 30°C before defibrillation is repeated.
- Cold myocardium may be resistant to pharmacologic defibrillation. Pharmacologic agents are ineffective in normal doses due to abnormal protein binding and agents can accumulate in the peripheral circulation, leading to toxicity with rewarming.
- Continue resuscitative efforts until warmed to 30°C.
- Continue temperature monitoring (rectal, esophageal, or tympanic).
- For moderate to severe hypothermia, active rewarming with heated, humidified oxygen; intravenous fluids heated to 40°C; radiant warmers for infants; and covering with dry blankets. Active core rewarming techniques include gastric lavage, bladder lavage, peritoneal lavage (very effective), and mediastinum (pleural cavity) lavage via thoracostomy tube (effective but invasive). Forced air rewarming is also effective and is associated with no after-drop phenomenon. Extracorporeal rewarming is the most rapid and is used for hypothermic cardiac arrest.
- For mild hypothermia (35°C–33°C), passive external rewarming is used. Techniques include maintaining a warm environment and covering with dry insulating materials.
- Most patients with hypothermia will require hospitalization.
- Core temperature of <32°C will require cardiac monitoring.
- Hypothermia patients with cardiac arrest or completely frozen extremities are candidates for extracorporeal rewarming.
- Patients with mild accidental hypothermia (35°C–32°C) may be rewarmed and discharged to a safe environment if there is no evidence of underlying disease.

FROSTBITE

- Earlobes, nose, hands, and feet are the most susceptible
- Predisposing factors include environmental, individual, behavioral, and occasion-linked factors.
- *Pathophysiology*: three distinct pathways are extracellular formation of ice crystals, hypoxia secondary to cold-induced local vasoconstriction, and release of inflammatory mediators (Prostaglandin PGF₂ and thromboxane A₂). Cold exposure increases blood viscosity, promotes vasospasm, precipitates microthrombus formation, and releases inflammatory mediators. Avoid recurrent freezing to prevent increase tissue levels of inflammatory mediators.
- The clinical signs and symptoms of frostbite differ according to the depth of injury. First-degree frostbite is limited to the superficial epidermis. Erythema and edema occur and resolve without sequelae. Second-degree frostbite is present with deeper epidermal involvement. Third-degree injury (deep) is a full thickness skin injury associated with hemorrhagic blisters on rewarming.
- Treatment is by immersion of the affected part in circulating warm water (40°C–42°C). Narcotic analgesics are often required to control pain during rewarming.
- It is difficult to determine tissue viability after significant hypothermic injury. Debridement of nonviable tissue is best delayed for several days to weeks to preserve as much tissue as possible.
- Topical aloe vera cream and ibuprofen may be used for outpatient treatment after rewarming.
- Rewarmed body parts are highly susceptible to refreezing, leading to even greater tissue loss. If exposure is anticipated, it is better not to rewarm the tissue.
- Frostbite often results in sequelae that are persistent and may be permanent.
- Prevention depends on awareness of cold risks, good nutrition, and hydration, avoiding exhaustion, excessive sweating, alcohol and tobacco.

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QUESTIONS

1. A 4-month-old male is brought in the ED with a fever to 104.2. He has a pulse of 168 and a respiratory rate of 22 and has had a recent cough and nasal congestion. Which of the following conditions is associated with the greatest risk of development of hyperthermia in the infant/neonate?
 - A. Higher metabolic rate leading to faster rates of acclimatization.
 - B. Decreased aldosterone secretion and inability to expand extracellular fluid volume.
 - C. Poorly developed thermoregulatory systems.
 - D. Inability to dissipate heat because of inefficient sweating capacity.
 - E. Overbundling.
2. A 15-year-old male football player presents to the ED with the complaint of dizziness, nausea, vomiting, generalized weakness, profuse sweating, and a temperature of 39°C after an afternoon practice session in August. He has a normal mental status. Which of the following is the most appropriate in the management of this patient?
 - A. Oral electrolyte solutions.
 - B. Salt tablets.
 - C. Intravenous rehydration.
 - D. Spraying the skin with room-temperature water.
 - E. Inpatient admission.
3. In addition to a temperature of 41.1°C to 42.2°C, which of the following is the most characteristic feature in establishing the diagnosis of heatstroke in the pediatric patient?
 - A. Fluid and electrolyte abnormalities.
 - B. History of high intensity exercise over a relatively short time span.
 - C. Profuse sweating.

- D. Seizures.
E. Failure of antipyretics to reduce core temperature.
4. A 15-year-old presents with a temperature of 42°C, dry skin and altered mental status after running in a cross country meet. Which diagnostic test carries the greatest predictive value for serious illness and complications related to this patient?
A. Arterial blood gases.
B. CBC.
C. Elevated potassium levels.
D. Renal function tests.
E. Liver enzymes.
5. Core temperature should be continuously monitored in the above patient (question #4) during treatment until the core temperature falls to what temperature?
A. 40°C
B. 39°C
C. 38°C
D. 37°C
E. 36°C
6. A 4-year-old girl is brought in after falling into a lake and being submerged from her waist down for several minutes. On arrival, she has a core temperature below 30°C. Which of the following will most likely be found in this patient?
A. Sinus tachycardia.
B. Loss of cerebral autoregulation.
C. Clear sensorium and normal mental status.
D. Ventricular ectopy.
E. Uncontrollable shivering.
7. A 10-year-old patient suffering from profound hypothermia has an Osborn or J-wave on her EKG. This pathognomonic but not prognostic Osborn or J-wave is typically seen at what core temperature?
A. 19°C
B. 20°C
C. 28°C
D. 32°C
E. 34.4°C
8. Which of the following statements is true concerning active rewarming for hypothermia?
A. Active external rewarming with hot packs and electric blankets is considered a first line therapy.
B. Heated, humidified oxygen and intravenous fluids heated to 40°C are indicated for mild to moderate cases of hypothermia.
C. Gastric irrigation is one of the most effective methods of active core rewarming
D. Forced air rewarming is an effective noninvasive technique; however, use is limited by the after-drop phenomenon.
E. For the hypothermic cardiac arrest, peritoneal lavage with heated fluid is the preferred method of rewarming.
9. A 12-year-old girl presents to the ED with the complaint of pain, redness, and blisters to multiple toes on both feet after getting them wet in a pond while sledding and walking home in the snow. Which is the preferred technique for management of this injury?
A. Vasodilators.
B. Hyperbaric oxygen.
C. Immersion in circulating warm water.
D. Debridement of nonviable tissue.
E. Topical aloe vera cream and ibuprofen.

ANSWERS

1. C. Infants are predisposed to the development of heat illness due to their poorly developed thermoregulatory systems. Whereas infants have higher metabolic rates, the rate of acclimatization tends to be slower in pediatric patients as compared to adults. Aldosterone secretions are increased once a patient is acclimatized. Increased aldosterone secretion leads to sodium retention and expansion of extracellular fluid volume. Exercising children have a reduced ability to dissipate body heat because of a lesser sweating capacity. Overbundling is one of the risk factors associated with the development of heat illness. Others include dehydration, infections, mental retardation, and obesity.
2. C. This patient is suffering from heat exhaustion and intravenous rehydration is recommended, starting with 20 mL/kg of normal saline over 30 minutes. Oral electrolyte solutions are recommended for treatment of heat cramps. Salt tablets are not recommended in the treatment of heat related illness because of the risk of causing gastrointestinal cramping. Spraying the skin with room-temperature water is part of the recommended treatment for heatstroke. Victims of heat exhaustion may require observation in the hospital. However, if all symptoms have resolved during emergency department observation, the patient may be released to continue rest and rehydration in a cool environment.
3. D. Disorientation, seizures, and coma are characteristic features of heatstroke, in addition to temperature elevation. There is no clear scientific evidence to indicate that heatstroke results from fluid or electrolyte abnormalities. Although the risk of developing exertional heatstroke appears greater in individuals

performing high-intensity exercise for a relatively short time span, it is not a characteristic feature. The skin may be dry, but profuse sweating may be seen in exertional heatstroke. Antipyretics are ineffective in the management of heatstroke but are not part of the diagnostic criteria.

4. E. This patient is suffering from heat stroke and his liver enzymes may be elevated, since the liver is very sensitive to heat stress. Transaminase levels peak in 24 to 48 hours and correlate well with severity of injury. Very high levels (aspartate transaminase >1000 IU) are predictive of severe illness and complications. Arterial blood gases are helpful in evaluating oxygenation, ventilation, and acid-base status, but have no specific prognostic value. CBC will usually show an elevated white blood cell count. Elevated band counts are more consistent with an underlying infection and should prompt a complete septic workup. Elevated potassium levels may indicate the development of rhabdomyolysis but also carry no specific prognostic value. Renal function tests may initially be elevated due to dehydration and may rise later, as renal failure develops.
5. B. Core temperature should be continuously monitored during treatment of heatstroke until the core temperature falls to 39°C.
6. D. Ventricular ectopy is seen with temperatures < 30°C and the risk of ventricular fibrillation is greatly increased. Atrial dysrhythmias commonly appear at temperatures below 32°C but are usually considered innocent. Cerebral perfusion is maintained until autoregulation fails at approximately 25°C. As the core body temperature drops below 33°C, the patient becomes confused and ataxic. Shivering ceases by the time the temperature reaches 31°C.
7. D. The J-wave (Osborn wave) is usually seen when the temperature falls below 32°C. Asystole commonly occurs at 19°C. The electroencephalogram shows a flat line at 20°C. As the core temperature drops below 33°C, the patient becomes confused and ataxic. Most thermometers for routine clinical use will record a temperature down to only 34.4°C.
8. B. Heated, humidified oxygen and intravenous fluids heated to 40°C are indicated for mild to moderately severe cases of hypothermia and have been shown to be safe and efficacious. They are used from the beginning. Active external rewarming with hot packs and electric blankets is considered dangerous and is not recommended. Rewarming

of cold extremities can result in the mobilization of cold peripheral blood to the central circulation resulting in core-temperature after-drop. Gastric, as well as bladder and colon, lavage are examples of active core rewarming techniques but heat transfer by these techniques is somewhat limited. Forced air rewarming is an effective noninvasive technique that is not associated with an after-drop phenomenon. Extracorporeal rewarming is the most rapid method and is indicated in hypothermic cardiac arrest.

9. C. The preferred technique is immersion of the affected part in circulating warm water (40°C–42°C). Vasodilators have been recommended, but firm evidence for their effectiveness is not yet available. Hyperbaric oxygen is another adjunctive therapy that has been recommended, but firm evidence for its effectiveness is not yet available. Debridement of nonviable tissue is best delayed for several days to weeks to preserve as much tissue as possible. Topical aloe vera cream and ibuprofen are recommended as outpatient treatment after rewarming.

141 HIGH ALTITUDE ILLNESS

Janis P. Tupesis

Ira J. Blumen

GENERAL

- Results from body's altered response to hypoxemia and decreased barometric pressure.
- Illness ranges from mild, self-limited disease processes, such as acute mountain sickness (AMS) to severe life threatening illness, such as high-altitude pulmonary edema (HAPE) or high-altitude cerebral edema (HACE)
- Slow, graded ascent is key to the body's ability to acclimatize to altitude.
- There are a few pharmacologic agents to help with acute altitude illness; descent to a lower altitude is key.

ATMOSPHERE AND GAS LAWS

- As altitude increases, partial pressure of oxygen decreases according to Boyle's and Dalton's laws.
- Decreasing partial pressure of oxygen causes decreased alveolar and arterial oxygenation.

PHYSIOLOGIC RESPONSES

- Respiratory
 - Decreased alveolar $P_{A}O_2$ leads to hypoxic ventilatory response.
 - Body will attempt to increase arterial P_aO_2 by increasing ventilatory rate.
 - Hyperventilation.
- Cardiovascular
 - Increased catecholamine release.
 - Result: increased heart rate, peripheral vascular resistance and systolic blood pressure, leading to increased cardiac output.
- Central nervous system
 - Due to hyperventilation, pH of both blood and CSF are slightly alkalotic.
 - Alkalosis causes chemoreceptor-mediated vasodilatation of arterioles.
 - Intracranial vasodilatation causes increased intracranial pressure.
- Renal
 - Respiratory alkalosis causes the renal excretion of bicarbonate.
 - Peripheral vasoconstriction causes increased central blood flow; increased volume to the kidneys causes moderate diuresis.

ACUTE MOUNTAIN SICKNESS

- Most common cause of altitude-related illness.
- Typical at lower altitudes.
- Clinical presentation
 - Typically in first 24 hours of being at altitude, but can be delayed by 3–4 days.
 - Most common findings include: headache, sleep disturbances, fatigue, and shortness of breath.
- Pathophysiology
 - Generalized fluid retention with fluid leaking into the extracellular space.
 - Mild cerebral edema from both cytotoxic edema (hypoxic induced capillary damage) and vasogenic edema (increased capillary blood flow and secondary hydrostatic induced capillary damage).
- Treatment
 - Since AMS is typically a self-limiting disease process, symptom management is key: acetaminophen for headache, prochlorperazine for nausea.
 - If symptoms not improving within 24 hours, descent of at least 1000 feet is indicated.
 - Acetazolamide.
- Prevention
 - Slow, graded ascent.
 - Adequate hydration.

- Avoidance of excess alcohol, sleeping medications and tobacco.

HIGH ALTITUDE CEREBRAL EDEMA

- Uncommon at low altitudes
- Affects only 1–2% of people at altitude, more common at higher altitudes
- Clinical presentation
 - Typically start out with signs and symptoms of AMS.
 - Progresses to neurologic dysfunction that heralds HACE, typically headache, nausea, and vomiting; progressing to truncal ataxia, visual disturbances, confusion, hallucinations, seizures, and eventually coma.
- Pathophysiology
 - Cytotoxic edema: endothelial damage from hypoxemia,
 - Vasogenic edema: vasodilatation and pressure mediated endothelial damage.
 - End result: cerebral edema.
- Treatment
 - Descent
 - High-flow oxygen
 - Dexamethasone, 1–2 mg/kg PO or IM initially, followed by 1–1.5 mg/kg q 4–6 h tapered over 5 days
 - If available, hyperbaric therapy (Gammow Bag) to mimic decrease in atmospheric pressure.

HIGH ALTITUDE PULMONARY EDEMA

- Form of noncardiogenic pulmonary edema.
- Thought to affect between 0.5% and 15% of those at altitude.
- Most cases at altitudes over 14,500 feet.
- Most common form death at altitude.
- Clinical presentation
 - Most commonly affects people within 1–4 days of ascent to altitude.
 - Common symptoms: cough, fatigue and dyspnea on exertion.
- Pathophysiology
 - Hypoxia-mediated vasoconstriction of pulmonary vascular bed leads to pulmonary hypertension.
 - Along with increased volume, leads to noncardiogenic pulmonary edema.
- Treatment
 - Almost all treatments aimed at decreasing pulmonary capillary pressure
 - Very limited experimental data in children.
 - Descent (1000–2000 feet).
 - High-flow oxygen.

- Nifedipine (slow release calcium channel blocker—aimed at decreasing pulmonary pressures): 10 mg initially, followed by 20 mg every 6 hours
- Newer data: β -adrenergic agonists (salmeterol) and phosphodiesterase inhibitors (sildenafil, tadalafil).

ALTITUDE RELATED SYNDROMES

- Retinal hemorrhages: due to vasodilatation of retinal blood vessels and associated papilledema.
- Chronic Mountain Sickness: due to inadequate ventilatory response, same symptom complex as AMS.
- UV keratitis: UV light exposure to eyes, severe pain, and foreign body sensation approximately 12 hours after exposure.
- High-altitude pharyngitis and bronchitis: severe dry, hacking cough caused by chronic exposure to cold and dry air at altitude.

PREEXISTING DISEASE

- Any child with preexisting pulmonary disease could be at higher risk for complications at altitude.
- Patients with sickle cell disease are at higher risk for vasoocclusive crises and complications due to hypoxemia and dehydration.

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QUESTIONS

1. A healthy 17-year-old male travels to Denver, Colorado, for a weekend hiking trip with his family.

Physiologic responses to traveling to this altitude might include which of the following?

- A. Tachycardia
 - B. Increased systolic blood pressure
 - C. Tachypnea
 - D. Peripheral vasoconstriction
 - E. All of the above
2. The day after arriving, the same young man becomes ill with a headache, nausea, and fatigue. He visits a local clinic and is diagnosed with AMS. Which of the following is characteristic of this illness?
 - A. Symptoms present at extreme altitude.
 - B. Self-limited symptoms.
 - C. Altered mental status.
 - D. Profound breathlessness and hypoxemia.
 - E. Infrequent presentation to people at altitude.
 3. Decreased partial pressure of oxygen at altitude and resulting hypoxemia result in which of the following physiologic changes?
 - A. Pulmonary vasoconstriction
 - B. Cerebral vasoconstriction
 - C. Renal vasoconstriction
 - D. Coronary artery vasodilatation
 - E. Peripheral vasodilatation
 4. Which of the following is the most common cause of death at high altitude?
 - A. Acute mountain sickness (AMS)
 - B. High altitude pulmonary edema (HAPE)
 - C. High altitude cerebral edema (HACE)
 - D. Hypothermia and thermal injury
 - E. Infection
 5. A 15-year-old male presents to your clinic after traveling to a ski resort in the mountains. Eight hours after arrival he develops headache, nausea, and difficulty sleeping. Which of the following should be recommended for this patient?
 - A. Descent to a lower altitude
 - B. Over the counter analgesics such as acetaminophen
 - C. Antiemetics
 - D. Acetazolamide
 - E. All of the above
 6. An 18-year-old female climber attempts to summit Mount Everest. She is noted by her guide to be confused, ataxic and has been complaining of a severe headache with intermittent vomiting and difficulty with her vision. Which of the following is the first line treatment for this patient?
 - A. Rest
 - B. Symptomatic treatment of symptoms
 - C. Diuresis
 - D. Steroids
 - E. Calcium channel blockers

7. Physiologic derangements at altitude include which of the following?
- Acidosis
 - Hypercarbia
 - Alkalosis
 - Volume retention
 - All of the above

ANSWERS

- E. All of the above are true. Increased catecholamine release leads to tachycardia, peripheral vasoconstriction, and increased blood pressure. The decreased partial pressure of oxygen stimulates carotid chemoreceptors that directly stimulate ventilation. Increased respiratory rate leads to respiratory alkalosis.
- B. AMS is usually a self-limited illness that affects travelers to low-moderate altitudes. It is the most common of the altitude related illnesses. Typically, there is not profound breathlessness or altered mental status as there is in HAPE or HACE.
- A. Hypoxemia at altitude leads to elevated pulmonary artery pressure by pulmonary vasoconstriction. Other physiologic findings at altitude include: cerebral vasodilatation and peripheral vasoconstriction.
- B. HAPE is the leading cause of death of travelers to intermediate and high altitudes.
- E. All of the above are true. Symptoms of headache, nausea, and vomiting can be effectively managed by acetaminophen and antiemetics. Descent to a lower altitude can help symptoms as it will change atmospheric pressure, partial pressures of oxygen and overall tissue oxygenation.
- D. These patient has HACE, which is a true medical emergency. The only treatments initially effective are steroids, supplemental oxygen and immediate descent to a lower altitude. Rest, symptom management, diuresis, and calcium channel blocker therapy are not indicated.
- C. Due to a lower partial pressure of oxygen, the body responds by increasing respiratory rate. A higher respiratory rate leads to a respiratory alkalosis.

142 DYSBARIC INJURIES

Ira J. Blumen

Lisa Rapoport

INTRODUCTION

- Dysbaric injuries* may be the result of several distinct events that expose an individual to a change in barometric pressure.
- Scuba (self-contained underwater breathing apparatus) diving was developed in the mid-1940s and currently allows the sport diver to descend to depths >100 ft.
 - In general, candidates must be 15 or 16 years old for full certification.
 - Pool-based divers may be certified at 8 years of age and some organizations will certify 10 years olds for ocean diving to 40 ft (12 m).
 - However, certification is not required to dive.
- The Divers Alert Network is a nonprofit association that provides medical expertise and education for divers around the world.
 - Between 1988 and 2002, there was an average of 16 diving injuries requiring hyperbaric recompression therapy in scuba divers aged 19 years and younger in North America. During this time period, the youngest diving fatality was 14 years old and the youngest injured diver was 11.
- The term dysbarism covers the general topic of pressure-related injuries.
- Barotrauma*, the most common diving injury, refers to the injuries that are a direct result of the mechanical effects of a pressure differential.
- The complications related to the partial pressure of gases and dissolved gases are called *decompression sickness*.

PHYSICAL GAS LAWS

- Individuals and objects under water are exposed to progressively greater pressure due to the weight of the water (Table 142-1).
- Small changes in the underwater depth result in large atmospheric pressure and volume changes, explained by *Boyle's law* (pressure and volume are inversely related) (Figure 142-1).
- Dalton's law of partial pressure* states that each gas will exert a pressure equal to its proportion of the total gaseous mixture ($P_{\text{total}} = P_1 + P_2 + P_3 \dots P_n$).
- Henry's law* states that the quantity of gas dissolved in a liquid is proportional to the partial pressure of

TABLE 142-1 Effect of Altitude or Depth on Air Pressure

ALTITUDE (FT) OR DEPTH (FT OF SEA WATER [FSW])	ABSOLUTE PRESSURE (ATA)	TORR	PSI	VOLUME RATIO
Altitude				
40,000	0.19	141	2.72	5.39
30,000	0.30	228	4.36	3.33
20,000	0.46	349	6.75	2.18
10,000	0.69	523	10.11	1.45
5000	0.83	632	12.23	1.20
1000	0.97	733	14.17	1.04
Sea level	1	760	14.7	1
Depth				
33	2	1520	29.4	0.50
66	3	2280	44.1	0.33
99	4	3040	58.8	0.25
132	5	3800	73.5	0.20
165	6	4560	88.2	0.17
198	7	5320	102.9	0.14

Courtesy of Ira Blumen, M.D., Section of Emergency Medicine, Department of Medicine, University of Chicago Medical Center.

the gas in contact with the liquid. This law will help explain the increased absorption of nitrogen during descent.

PATHOPHYSIOLOGY AND PRESENTATION

- The clinical findings of dysbaric injuries may be immediate or delayed in onset up to 36 hours. Most will occur during descent or in close proximity to ascent. A delayed presentation is possible, however, which may make diagnosis difficult.
 - There are three mechanisms for dysbaric injuries.
 - The first follows Boyle's law for trapped gas and changes in ambient pressure.
 - The second follows Henry's law when gas dissolved in blood is released.
 - The third deals with abnormal tissue concentrations of various gases.

BAROTRAUMA: DYSBARISMS FROM TRAPPED GASES

- Barotrauma is the direct result of a pressure difference between the body's air-filled cavities and the surrounding environment.
- During descent or ascent, equalization must occur between the outside pressure and internal air-filled structures. Inability to equalize the pressures within an enclosed air space could cause rupture of such

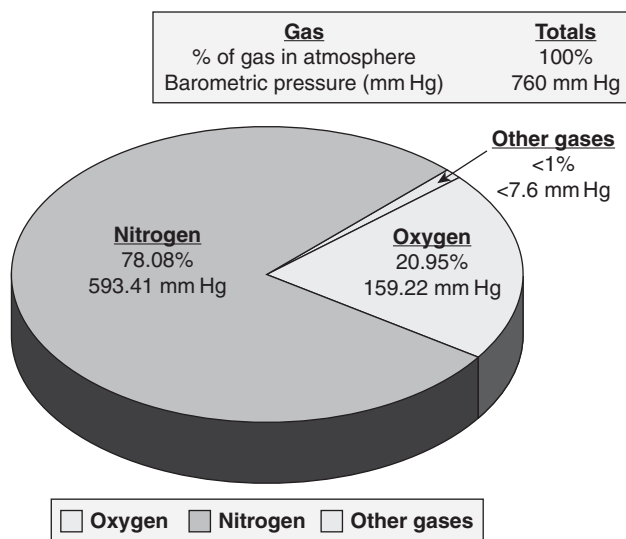


FIG. 142-1. Atmospheric composition at sea level.

spaces or damage of adjacent structures (e.g., compression, edema, or hemorrhage).

- These trapped gas disorders are differentiated by the gas-filled part of the body that is affected.
- *Barotitis media*, also known as *middle ear squeeze*, involves the middle ear, and usually begins on descent close to the surface. Symptoms include fullness in the ears, severe pain, tinnitus, vertigo, nausea, disorientation, and transient conductive hearing loss. Further descent without equalization will exacerbate symptoms and may perforate the tympanic membrane.
 - Physical examination may reveal erythema or retraction of the tympanic membrane (TM), blood behind the TM, a ruptured TM or a bloody nasal discharge.
 - Prevention is key and can be done through various maneuvers to open the Eustachian tubes, such as blowing the nose against pinched nostrils or repositioning the jaw (false yawning), while keeping a regulator in the mouth.
 - Equalization may be compromised if the eustachian tube mucosa is swollen from any cause. Pre-dive treatment of pseudoephedrine or a topical vasoconstrictor nasal spray may be helpful: oxymetazoline hydrochloride, 0.05%, applied 15 minutes before a dive, ages 6+, 2–3 sprays/nare. Not recommended for ages <6 years. Ages 6 to 12, treat with pseudoephedrine 30 mg PO. For children 12+ years old, the adult dose of 60 mg PO may be used.
 - Patients with barotitis media should not dive until all signs and symptoms have resolved.
 - For a perforated TM, prescribe a 10-day course of antibiotics (oral plus otic suspension drops) and

ENT follow-up. The TM must heal before any further diving.

- *Alternobaric vertigo* is a sudden change in middle ear pressure or asymmetrical middle ear pressure that affects vestibular function. Symptoms include transient vertigo, tinnitus, nausea, vomiting, and fullness in the affected ear. Symptoms may last minutes to several hours after the completion of a dive. Decongestants, antiemetics, and medication for vertigo are recommended.
- *Barotitis externa* or *external ear squeeze* occurs when the external auditory canal is occluded during descent. Obstruction can be caused by cerumen, ear plugs, or other foreign bodies. A diver may experience pain with or without bloody otorrhea.
- *Barotitis interna* or *inner ear squeeze* is uncommon but may result in permanent injury to the inner ear. It often follows a vigorous Valsalva maneuver. Symptoms include sudden sensorineural hearing loss, severe pain or pressure, vertigo, tinnitus, ataxia, nausea, vomiting, diaphoresis, and nystagmus. These patients must be seen emergently.
- *Altitude-related barotitis media* is the most common barotrauma of air travel. As with diving, equalization may be accomplished by maneuvers that open the Eustacian tubes. Instructing children to swallow more frequently, or giving an infant a bottle, may help with equalization.
- *Barosinusitis*: With sinus inflammation, air-filled frontal or maxillary sinuses may not equilibrate, resulting in sinus squeeze. Pain in the affected sinus may persist for hours and may be accompanied by a bloody nasal discharge. The treatment for barosinusitis also involves the use of a vasoconstrictor nasal spray before initiating a dive or before starting a descent from altitude in an airplane. Reverse sinus squeeze is felt during a diving ascent when an obstruction of the sinuses results in excessive pressure. The diver should redescend to a greater depth and then ascend at a slower rate.
- Barodontalgia or tooth squeeze is often associated with recent dental extraction, dental fillings, periodontal infection, periodontal abscess, or tooth decay. Treatment is directed toward preventative dental care and pain control. Following dental procedures, a minimum of 24 hours is advised before initiating a scuba dive.
- Facemask squeeze occurs during descent, when the increased ambient pressure exerts increasing pressure against the air-filled facemask of a scuba diver. The diver may develop facial or eye pain, subconjunctival hemorrhages, subconjunctival edema, epistaxis, or periorbital edema.
- This is prevented by using a low volume face mask that minimizes the amount of air and allows for additional small amounts of air to be blown into the mask from the nose.
- *Aerogastralgia*: Under normal circumstances, the stomach and intestines contain approximately 1 quart of gas, which may cause discomfort with expansion.
- Avoiding carbonated beverages, chewing gum (and swallowing air), large meals and may help in prevention.
- Aerogastralgia is rarely serious; however, gastric rupture has been reported and pain may occasionally cause a vaso-vagal response.
- Pulmonary barotrauma follows drowning as the second most common cause of death among scuba divers.
- Breathing from a compressed air source provides positive-end-expiratory pressure (PEEP), thereby preventing lung collapse when a diver is at depth.
- Positive-pressure barotrauma, e.g., pneumothorax or pulmonary vein rupture, can happen during ascent as the air in the lungs expands.
- To reduce the risk of pulmonary barotrauma, divers must not hold their breath. This is important not only during ascent but also in the event a diver is not aware of an unintended decrease of depth.
- Children and beginning-level divers who may not be skilled at managing depth regulation using a buoyancy device are at increased risk for barotrauma. Also at risk are divers with obstructive airway diseases, including asthma and chronic obstructive pulmonary disease.
- Air embolism.
- Specific signs and symptoms will be determined by the final destination of air emboli, such as the brain or the heart.
- The onset of symptoms can be immediately on ascent, or within 10 to 20 minutes of surfacing. Neurologic symptoms that develop later than this are more likely due to decompression sickness.
- Aggressive care includes 100% oxygen, intravenous fluids and hyperbaric treatment. Patients are placed in the trendelenburg or left lateral decubitus. A careful medical evaluation is warranted for the onset of any neurologic symptoms during or within a short time after the conclusion of a dive.
- A rapid onset and severe symptoms are suggestive of a poorer prognosis with both air embolism and decompression sickness.
- Pneumothorax and emphysema.
- The patient with a confirmed or suspected pneumothorax, pneumopericardium, pneumomediastinum, or subcutaneous emphysema should not be exposed to any further barometric pressure changes.
- Hyperbaric (recompression) treatment is avoided since it can convert a simple pneumothorax to a

tension pneumothorax. If hyperbaric treatment will be necessary, chest tubes must be placed before initiating recompression.

DECOMPRESSION SICKNESS: DYSBARISMS FROM EVOLVED GASES

- Gases coming out of solution results in decompression sickness. A diver breathing compressed air is exposed to nitrogen, oxygen, and carbon dioxide. Approximately 4/5 of the air is nitrogen. As ambient pressure increases, more nitrogen will be dissolved in the blood. As a dive progresses, the gas in the blood will equilibrate quickly with the gas in the alveoli.
- The body will absorb more nitrogen gas at a rate that depends on the depth and duration of the dive. The longer and deeper the dive, the more nitrogen gas will be accumulated within the body.
- Nitrogen is not metabolized and will remain dissolved until ascent. If the ascent is too rapid, nitrogen levels do not have the opportunity to equalize among the tissues, blood, and alveoli in order to be exhaled through the alveoli. This results in the gas coming out of solution rapidly and the formation of gas bubbles in the blood or tissue (ie, decompression sickness).
- The risk of decompression sickness is increased by increased duration and depth of a dive, particularly with inadequate surface times between dives. Other risk factors include increased physical activity during a dive, cold temperatures, obesity, alcohol ingestion, and flying within 12 hours of a dive. Diving tables are used to safely balance dive and surface times.
- Decompression sickness can be classified as follows:
 - *Type I*: Extravascular gas bubbles (causes joint pain, skin rashes, and lymphedema).
 - *Type II*: Intravascular nitrogen gas emboli. The presentation may be very similar to that of air emboli and clearly have serious sequelae. Children are more prone to Type II injuries.
- Musculoskeletal decompression sickness (also known as the bends) occurs in up to 75% of all decompression injuries and is caused by the release of nitrogen gas bubbles from the blood into the tissues surrounding the joint.
 - Symptoms usually develop within 6 to 12 hours after a dive. Sharp, throbbing, or dull achy pain is a common presentation, sometimes with paresthesias. The joints most often affected are the knees, shoulder, and elbows.
- Cutaneous decompression sickness.
 - The extravascular release of nitrogen gas bubbles from the blood into the skin usually results in benign dysbarisms, such as rashes, with or without pruritus.
 - The release of nitrogen gas bubbles can cause subcutaneous emphysema, often involving the neck and other sites.
 - Watch for alterations in a diver's voice or any subcutaneous emphysema above the collarbone, as this may be a sign of ruptured alveoli, a more serious pulmonary barotrauma. Treatment of individuals with subcutaneous emphysema begins with 100% oxygen, followed by an exam for more serious dysbarisms, and admission.
- Pulmonary decompression sickness (the chokes) is caused by arterial or venous nitrogen gas embolization that obstructs the pulmonary vasculature.
 - Symptoms may begin immediately after a dive but may also take up to 12 hours to develop. The classic triad of symptoms includes shortness of breath, cough, and substernal chest pain or chest tightness. The patient may also experience uncontrollable paroxysmal cough.
 - Minor presentations can last between 12 and 48 hours but severe presentations can progress to a rapid deterioration.
- Neurologic decompression sickness.
 - Nitrogen gas embolism is the most serious decompression sickness. Venous gas emboli can result in venous obstruction, and arterial gas emboli can cause ischemia as a result of arterial obstruction or induced vasospasm.
 - The brain is most commonly affected, but the spinal cord can also sustain ischemic injury through an embolic event blocking the venous return in the epidural vertebral venous system. Watch for back pain, numbness in the extremities, weakness, paralysis, and urinary retention.
 - Onset of symptoms will usually be delayed, with symptoms developing within 1 to 6 hours after a dive is concluded.
 - These victims require aggressive care, which includes 100% oxygen, intravenous fluids and hyperbaric treatment. Placement in the trendelenburg or left lateral decubitus position may minimize embolization to the brain.
 - Cerebral decompression injuries are more common with altitude-related decompression than with diving injuries. The symptoms are similar to those of the air embolus. Children primarily present with abnormal behavior, disorientation, and memory loss.
- Decompression shock may be secondary to hypovolemia or due to vasovagal responses. Hypovolemia is caused by fluid loss and third spacing. Watch for evidence of inadequate end-organ perfusion, at first compensated, then decompensated. The patient may become agitated, restless, cool to the touch,

tachycardic, tachypneic and, finally, hypotensive. If vasovagal symptoms dominate initially, the victim may present with diaphoresis, nausea, vomiting, bradycardia, light-headedness, and hypotension.

- Aggressive and timely management with intravenous fluids, 100% oxygen and recompression therapy should be initiated as quickly as possible.
- Treatment of decompression sickness and air embolus.
 - The survival of dysbaric injuries depends on the severity of the injury, rapid identification of illness, and timely access to appropriate medical care.
 - Treatment begins with the administration of 100% oxygen, hydration, and rewarming. The treatment of choice for most air emboli and decompression illnesses is hyperbaric (recompression) oxygen therapy (HBO). Although there is still benefit in providing delayed HBO treatment, it should be initiated as soon as possible, ideally within 6 hours of the onset of symptoms.
 - The goal of HBO is to reduce the size of the liberated gas bubbles, facilitate the resorption of these air bubbles, prevent the formation of new bubbles, and improve oxygenation. The mechanism of HBO is complex and not completely understood. It is thought that by decreasing the size of gas bubbles, hypoxia can be reduced downstream of blocked vessels, and that HBO removes the nidus for activation of the complement system. Hundred percent oxygen also helps to replace undissolved nitrogen with oxygen, which is easier for tissues to utilize and eliminate from the body. HBO may also help to deliver oxygen to tissues damaged by ischemic-reperfusion injury.
 - Before hyperbaric treatment is initiated, endotracheal tube cuffs and urinary catheter balloons should be filled with saline rather than air. Any pneumothorax must be identified and a chest tube placed prior to recompression. Plastic infusion containers are preferred over glass bottles. Consider topical anesthetic otic drops to anesthetize the TM of smaller children who may have difficulty equalizing middle ear pressure during hyperbaric treatment.
 - In HBO, patients are taken to a “depth” of 60 ft (FSW), which is equal to 2.8 atmospheres. Supplemental oxygen at an FIO_2 of 100% is provided at 20-minute intervals, alternating with room air. The hyperbaric pressure will be reduced at a rate of 1 ft per minute to equal a depth of 30 ft for a period of time and then slowly brought back to “sea level.”
 - Patients with an arterial air embolus will commonly be brought to an initial hyperbaric depth

of 165 ft (6 atmospheres). After 30 minutes, the patient will be brought slowly “up” to a depth of 60 and then 30 ft before returning to the normal ambient pressure.

- Patients should be kept dry and warm to prevent hypothermia, as well as receive adequate fluid resuscitation to a goal urine output of 1 to 2 mL/kg/hr.
- Special precautions should also be taken for victims who must be transported by helicopter or airplane in which exposure to a decreased ambient pressure may compromise the victim further. Helicopter transports should be done at as low an altitude as safely possible (less than 1000 ft). Airplane transport should be conducted in aircraft that are capable of being pressurized to sea level.
- Victims of Type I decompression sickness are advised to abstain from any further scuba diving for at least 4 to 6 weeks and Type II victims must wait at least 4 to 6 months.

DYSBARISMS CAUSED BY ABNORMAL GAS CONCENTRATION: NITROGEN NARCOSIS

- As a diver descends, the partial pressure of inhaled nitrogen gas will increase. Nitrogen narcosis occurs when an increased partial pressure of dissolved nitrogen produces a narcotic or intoxicating effect. Symptoms can include euphoria, uncontrollable laughter, impaired judgment, memory loss, light-headedness, hallucinations, loss of coordination and impaired reflexes.
- The signs of nitrogen narcosis may become evident at depths beyond 80 ft. With any suspicion of nitrogen narcosis, the dive should be terminated. The affected diver should be escorted slowly to the surface by a second diver.

CONSIDERATIONS FOR DIVING IN CHILDREN

- When screening a child for scuba-diving, physiologic, anatomic, and developmental considerations should be kept in mind. Compared with adults, children have increased surface-to-body area, higher lung volume-to-body ratio and longer sustained compensation in shock states. No concrete recommendations can be found in the medical literature for or against children participating in scuba diving.
- Children with chronic sinusitis or otitis media should be adequately prophylaxed against congestion and potential difficulty with equalization.

- Scuba diving is contraindicated in serious chronic lung conditions (e.g., cystic fibrosis, alpha-1 antitrypsin deficiency or chronic obstructive pulmonary disease), as well as cardiac conditions (e.g., ASD, VSD or congenital arrhythmias). Children with active asthma exacerbations or cold- or exercise-induced asthma should be advised against diving, as should any child with a prior history of spontaneous pneumothorax or thoracic surgery.

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QUESTIONS

1. A 12-year-old is taking diving lessons. During the pool lessons which of the following instructions are given on how pulmonary barotrauma injuries are best prevented during a dive?
 - A. Wearing a buoyancy device.
 - B. Monitoring dive temperature.
 - C. Using compressed air with a higher concentration of oxygen.
 - D. Never holding one’s breath during a dive.
 - E. Wearing a low-volume face mask.
2. A 17-year-old diver describes having pain and severe pressure in his ears during his descent. It starts whenever he gets below 7 ft. What advice can you give him?
 - A. Descend very slowly, while equalizing his ears before he feels any pressure.
 - B. Pretreat with an oral over-the-counter decongestant such as Sudafed.
 - C. If he is unable to equalize his ears, slowly reascend, rest, then try again.
 - D. Practice the maneuver to equalize ears while still on land.
 - E. All of the above.
3. A 15-year-old diver surfaces after an emergent and rapid ascent. She complains of headache and shortness of breath. What condition must be ruled out before transferring her to a hyperbaric chamber?
 - A. Pneumothorax
 - B. Air embolism
 - C. Hemiplegia
 - D. Cutaneous decompression sickness
 - E. Pulmonary decompression sickness
4. The initial treatment for any diving emergency would be which of the following?
 - A. 100% oxygen
 - B. IV fluid resuscitation
 - C. Alert the local rescue authorities for possible transport to a hyperbaric chamber
 - D. Assess and monitor for shock, exposure or any disability
 - E. All of the above
5. The risk of decompression sickness is reduced by which of the following?
 - A. Decreased depth and increased dive time.
 - B. Increased depth and decreased dive time.
 - C. Increased depth and increased dive time.
 - D. Decreased depth and decreased dive time.
 - E. Nothing can improve the risk of decompression sickness.
6. You are the expedition doctor for a dive trip in Honduras. Soon after surfacing from a dive, one of the younger participants in the group develops weakness in her legs, with paresthesias. Your biggest concern is which of the following?
 - A. The bends
 - B. Pulmonary embolism
 - C. Spinal cord decompression sickness
 - D. Pneumothorax
 - E. Musculoskeletal decompression sickness
7. A 14-year-old male with asthma requests a medical screening to participate in scuba diving lessons for his summer camp. Upon taking a history, it appears that his asthma is triggered by mold and dust, but not exercise or cold temperatures. He has evidence of chronic sinusitis on examination. Your recommendations are
 - A. It is not okay to participate in diving, ever.
 - B. Diving is okay so long as he’s not having an acute exacerbation of his asthma and he should pretreat with Sudafed.
 - C. He should pretreat with albuterol, 2 puffs q 15 minutes for 3 doses.
 - D. He should pretreat with steroids.
 - E. He should have no concerns about diving at all.

8. An 18-year-old diver has the bends and needs hyperbaric oxygen therapy (HBO). Which of the following would be the goal of this therapy?
 - A. Decrease the pain of the bends.
 - B. Facilitate reabsorption of nitrogen gas bubbles.
 - C. Improve oxygenation.
 - D. Prevent formation of new bubbles.
 - E. All of the above.
9. A diver comes to see you in the ED. He describes that during his dive, at around 80 ft, he began to feel disoriented and slightly “high.” His dive buddy helped him to shallower water and they aborted their dive safely. Afterward, he felt fine and has had no complaints since. He most likely experienced which of the following dive related conditions?
 - A. The bends (musculoskeletal decompression sickness)
 - B. Transient pulmonary embolism
 - C. Breathing unpurified air
 - D. Nitrogen narcosis
 - E. Inner ear squeeze
10. A 15-year-old diver surfaces after spending 35 minutes at 70 ft. Within 30 minutes of surfacing, he notices a “rice krispy” feeling in the skin in his neck. Then, he begins to experience changes in his voice and you notice his eyes begin to grow puffy. What are you most worried about?
 - A. Barodentalgia
 - B. Cutaneous decompression sickness
 - C. Allergic reaction to jellyfish envenomation
 - D. Ruptured alveoli leading to pneumothorax
 - E. Nitrogen narcosis

ANSWERS

1. D. Divers are taught never to hold their breath underwater. Inability to release gas and equalize the pressures between an enclosed air space and the surrounding environment can cause rupture of such spaces or the compression of adjacent structures. This is highlighted in the lungs during ascent, when the air begins to expand as the diver is exposed to less ambient pressure. In order to prevent pulmonary barotrauma, divers are trained to never hold their breath so that the lungs never become a closed air space. This is important not only during ascent but also in the event a diver is not aware of an unintended decrease of depth. Children and beginning-level divers who may not be skilled at managing depth regulation using a buoyancy device are at increased risk for barotrauma. Wearing a low-volume face mask helps to prevent face squeeze, another type of barotrauma. Monitoring dive temperature does not help prevent injuries to the lung, although it is smart practice to be aware of heat loss during time spent underwater. Higher concentrations of oxygen in compressed air tanks may be used for specialized divers going to greater depths, but requires advanced training and does not prevent barotrauma.
2. E. *Barotitis media* is the most common diving-related barotrauma and involves the middle ear. It is commonly referred to as *middle ear squeeze* or *ear block* and begins on descent close to the surface. The symptoms include fullness in the ears, severe pain, tinnitus, vertigo, nausea, disorientation, and transient conductive hearing loss. Further descent without equalization will exacerbate symptoms and may perforate the tympanic membrane. With perforation, the caloric stimulation of cold water entering the middle ear can cause vertigo, nausea, and disorientation. Prevention of middle ear squeeze is key. Scuba divers should attempt to clear their ears every 2 to 3 ft during descent by actively opening the eustachian tube, which opens and exposes the middle ear to ambient pressures. This is done through various maneuvers such as blowing the nose against pinched nostrils or repositioning the jaw (false yawning). If a diver is unable to successfully equalize the middle ear, she/he should reascend to a depth where there is no pain and then try again, descending very slowly and equalizing frequently. Occasionally practicing the equalizing maneuver before a dive can be helpful. If a diver is congested, a pre-dive treatment of Sudafed PO or oxymetazoline spray per nare may help.
3. A. A patient with a pneumothorax must have a chest tube placed before being exposed to high pressures during HBO. If the pneumothorax is not treated, there is a risk of developing a tension pneumothorax, which is urgently life-threatening. Any air-filled structures, such as an endotracheal tube cuff, should be filled with water instead of air before “diving” during HBO. Plastic IV solution bags are preferred over glass infusion bottles. All other diagnoses are indications for treatment with HBO.
4. E. As with any medical emergency, remember the emergency medicine mantra of “IV, O₂, Monitor.” In the field, it may be very difficult to distinguish pulmonary barotrauma from, for example, air embolism to the lungs (the chokes). It is important

to recognize diagnostic limitations in the field and, instead, focus on when to transfer a patient to the appropriate level of care, while keeping the patient oxygenated, warm, monitored, and always treated for shock.

5. D. A diver breathing compressed air is exposed to nitrogen, oxygen, and carbon dioxide. Approximately 4/5 of the air is nitrogen. As ambient pressure increases, the positive-pressure gradient between the alveoli and the blood will result in more nitrogen being dissolved. As a dive progresses, the gas in the blood will equilibrate quickly with the gas in the alveoli. Therefore, the body will absorb more nitrogen gas at a rate that depends on the depth and duration of the dive. The longer and deeper the dive, the more nitrogen gas will be accumulated within the body. The risk of decompression sickness is also increased by increased physical activity during a dive, cold temperatures, obesity, alcohol ingestion, previous dives with inadequate surface time to equilibrate and flying within 12 hours of a dive. Diving tables are used to safely balance dive and surface times. They help to calculate the amount of time needed on the surface between dives to prevent accumulation of too much systemic dissolved nitrogen.
6. C. Although initially it is difficult to distinguish musculoskeletal from spinal cord decompression sickness, the latter, caused by nitrogen gas embolism (ie, Type II decompression sickness, intravascular) is generally a more serious threat to life and limb. Venous gas emboli can result in venous obstruction and arterial gas emboli can cause ischemia as a result of arterial obstruction or induced vasospasm. The brain is most commonly affected, but the spinal cord can also sustain ischemic injury. The onset of symptoms, however, will usually be delayed, with symptoms developing within 1 to 6 hours after a dive is concluded. These victims require aggressive care, which includes 100% oxygen, intravenous fluids, and hyperbaric treatment. Placement in the Trendelenburg or left lateral decubitus position may minimize embolization to the brain. In spinal cord decompression injuries, an air embolism affects the spinal cord by blocking the venous return in the epidural vertebral venous system. Watch for back pain, numbness in the extremities, weakness, paralysis, and urinary retention.

Colloquially termed the *bends*, musculoskeletal decompression sickness occurs in up to 75% of all

decompression injuries and is caused by the release of nitrogen gas bubbles from the blood into the tissues surrounding the joint (ie, Type I decompression sickness, extravascular). Symptoms usually develop within 6 to 12 hours after a dive. Sharp, throbbing, or dull achy pain is a common presentation, sometimes with paresthesias. The pain may start diffusely, but becomes more localized as the intensity increases. Since this patient is not having pulmonary symptoms, pulmonary embolism, and pneumothorax are unlikely.

7. B. Scuba diving is contraindicated in serious chronic lung conditions (e.g., cystic fibrosis, alpha-1 antitrypsin deficiency or chronic obstructive pulmonary disease), as well as cardiac conditions (e.g., ASD, VSD or congenital arrhythmias). Children with active asthma exacerbations or cold- or exercise-induced asthma should be advised against diving as the cold temperatures or physical exertion during a dive may trigger an exacerbation. However, children with asthma not currently experiencing a flare may proceed to dive cautiously, without pretreating with albuterol or steroids. Children with chronic sinusitis or otitis media should be adequately prophylaxed against congestion and potential difficulty with equalization.
8. E. The mechanism by which HBO therapy improves decompression sickness is unclear. The goal of HBO treatment, however, is to reduce the size of the liberated gas bubbles, facilitate the resorption of these air bubbles, prevent the formation of new bubbles, and improve oxygenation. The mechanism of HBO is complex, but, by causing bubbles to decrease in size, hypoxia can be reduced downstream of blocked vessels. In theory, HBO removes the nidus for activation of the complement system. Giving 100% oxygen also helps to replace undissolved nitrogen with oxygen, which is easier for tissues to utilize and eliminate from the body. HBO may also help to deliver oxygen to tissues damaged by ischemic-reperfusion injury.
9. D. Scuba diving is made possible through the use of compressed air tanks. As a diver descends, ambient pressure will increase causing a proportionate increase in the partial pressure of the compressed gases (Dalton's law). As a result, the partial pressure of the inhaled gas will increase at greater depths. The increased partial pressure of nitrogen represents the greatest concern to scuba divers. Nitrogen narcosis is when an increased partial pressure of dissolved nitrogen produces

a narcotic or intoxicating effect. Symptoms can include euphoria, uncontrollable laughter, impaired judgment, memory loss, light-headedness, hallucinations, loss of coordination and impaired reflexes. The signs of nitrogen narcosis may become evident at depths beyond 80 ft. With any suspicion of nitrogen narcosis, the dive should be terminated. The affected diver should be escorted slowly to the surface by a second diver. No other treatment is required. Transient pulmonary embolism would cause shortness of breath, and possibly pleuritic chest pain, as opposed to euphoria. Inner-ear squeeze would be severe pain in the ears and would present much closer to the surface at the beginning of a dive. Although it is advised to rent compressed air tanks from a reputable source to avoid exposure to concentrated levels of toxins during a dive, breathing unpurified air would not cause nitrogen narcosis.

10. D. The extravascular release of nitrogen gas bubbles from the blood into the skin (cutaneous decompression sickness) usually results in benign dysbarisms. Rashes from cutaneous decompression sickness, with or without pruritus, can present with any of the following patterns: scarlatiniform, mottling (*cutis marmorata*), and erysipeloid. The release of nitrogen gas bubbles can cause subcutaneous emphysema, often involving the neck and other sites. However, alterations in a diver's voice or any subcutaneous emphysema above the collarbone may be a sign of ruptured alveoli, a more serious pulmonary barotrauma. Treatment of individuals with subcutaneous emphysema begins with 100% oxygen, followed by an examination for more serious dysbarisms and admission. Although an allergic reaction may cause a patient's throat to swell, he does not report a history of being stung, nor any hives or rashes, nor is he displaying the hemodynamic instability of anaphylaxis. This patient does not report any tooth pain, which makes barodentalgia, or tooth squeeze, unlikely. Barodentalgia is often associated with recent dental extraction, dental fillings, periodontal infection, periodontal abscess, or tooth decay. Treatment is directed toward preventative dental care and pain control. Following dental procedures, a minimum of 24 hours is advised before initiating a scuba dive. This presentation would not be nitrogen narcosis, which resolves when a diver surfaces.

143 RADIATION EMERGENCIES

Ira J. Blumen

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INTRODUCTION

- Children have a higher risk for internal exposure from inhaled radioactive gases, are at greater risk for developing subsequent malignancies, and are more likely to experience psychological trauma or injury after a radiation disaster.

TYPES OF RADIATION

- *Radiation* is energy emitted from a source and a *radioactive* substance, referred to as a *radioisotope* or *radionucleotide*, gives off radiation.
- People exposed to external or remote radiation have been *irradiated* but do not become radioactive. Victims give off radiation only if there was external or internal *contamination* caused by the presence of radioactive particles (α and β)
- Radiation can be classified as either *ionizing* or *nonionizing and nonparticulate (electromagnetic)* or *particulate*.
 - Electromagnetic radiation has no mass and no charge; it occurs in waveforms and is described by wavelengths; it can be ionizing or nonionizing.
 - Particulate radiation has mass and is ionizing (charged or uncharged).

IONIZING RADIATION

- Ionizing radiation interacts with matter, converting atoms to ions through gain or loss of electrons.
- Ionizing radiation is more dangerous as it can damage DNA and RNA affecting biologic functions at the cellular metabolic level.
- Ionized radiation has a high frequency, short wavelength and a billion times more energy than nonionizing radiation. Common sources are nuclear reactors, nuclear weapons, radioactive material, and radiography equipment.
- Y-rays have the highest energy content of the nonparticulate radiations and can penetrate deep into the tissue. Constant low-level exposure through cosmic radiation is a usual source. Y-radiation is a common cause of acute radiation syndrome due to radioisotope decay and radiation from linear accelerators. Lead

shields 1 to 2 inches in depth or thick concrete provide satisfactory protection from γ -rays.

- X-rays have the next highest energy content of the nonparticulate radiations and originate from outside the nucleus emitted by excited electrons. X-rays can penetrate tissue and deposit energy deep within the cells. Their usual source is medical or industrial in nature.
- α -particles are composed of two protons and two neutrons and possess a 2^+ electrical charge. They originate from the nucleus of the atom, travel only inches from their source and generally do not penetrate paper or epidermis. These are rarely harmful externally. Examples include plutonium, uranium, and radium.
- β -particles have a small mass, composed of a single electron emitted from the atom's nucleus, and possess a 1^- charge, disperse only a few feet from their source and penetrate tissues only a small amount (up to 8 mm), primarily causing thermal injuries. Clothing alone can often provide adequate protection from β -particles.
- Both α -particles and β -particles are harmful if ingested or inhaled or contaminate wounds.
- Neutrons are without electrical charge and ionize by colliding with atomic nuclei within cells and tissues. They possess strong power to penetrate and represent the only form of radiation that can make previously stable atoms within the body radioactive. They can be more damaging than x-rays or γ -rays and are responsible for radioactive fallout. Specialized concrete is necessary to provide shielding from neutron radiation. Nuclear reactors, nuclear weapons, and nuclear accelerators are common sources of neutron radiation (Fig. 143-1).

Radiation source	Amount of exposure (mrem/y)
Consumer products	111
Cosmic	26
Terrestrial	28
Nuclear medicine	14
Medical	19
Internal	40
Radon	200

NONIONIZING RADIATION

- Nonionizing radiation is relatively low energy in nature and does not result in acute radiation injuries or contamination. Adverse effects to humans are limited to local heat production. This includes ultraviolet rays, visible light, infrared radiation, microwaves, and radio waves (in order of decreasing energy).

MEASURING RADIATION

- Radiation can be detected and quantified by dosimeters or Geiger–Mueller tubes at levels below that which results in biologic consequence.
- *X-rays are measured in roentgens*, which measure ion pairs produced in a given volume of air.
- *Dose* represents the amount of energy deposited by radiation per unit of mass.
- *Rad* (roentgen absorbed dose) is the basic unit of measurement.
- *Gray (Gy)* represents the standard international (SI) unit for dose: $1 \text{ Gy} = 100 \text{ rad}$ and $1 \text{ cGy} = 1 \text{ rad}$.
- *Rem* (roentgen equivalent in man) represents a calculated radiation unit of dose equivalent. The absorbed dose (rad) is multiplied by a factor to account for the relative biologic effectiveness (RBE) of various types of radiation: $\text{rem} = \text{rad} \times \text{RBE}$.
- *Seivert (Sv)* is the SI unit for dose equivalent where: $1 \text{ Sv} = 100 \text{ rem}$ and $1 \text{ cSv} = 1 \text{ rem}$.
- For β -particles, x-rays and γ -rays, the $\text{RBE} = 1$. Therefore, for these sources of radiation, $1 \text{ rad} = 1 \text{ rem}$ and $1 \text{ Gy} = 1 \text{ Sv}$.

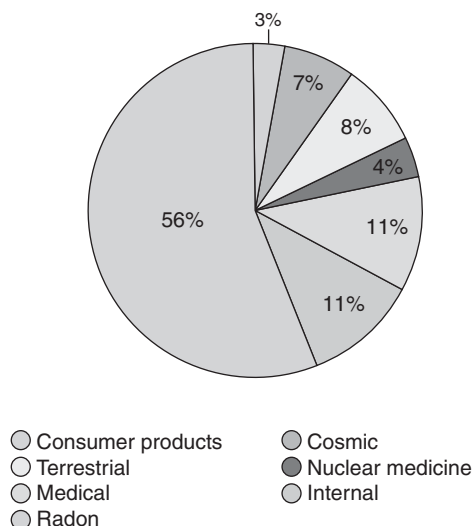


FIG. 143-1. Common sources of natural and technological radiation exposure in the United States.

TABLE 143-1 Localized Radiation Injuries

TYPE	PRESENTATION	EXPOSURE	COMMENTS
Type I	Erythema only	600–1000 rad	Similar to a first-degree thermal burn. Erythema may be delayed up to 2–3 wk. A dose of 300 rad can result in a delayed hair loss (epilation). Dry desquamation or scaling may occur.
Type II	Transepidermal injury or wet desquamation	1000–2000 rad	Similar in severity to a second-degree partial-thickness thermal burn.
Type III	Dermal radionecrosis	>2000 rad	Severe pain with or without paresthesia. Resembles a severe chemical or scalding burn. Skin grafting may be necessary.
Type IV	Chronic radiation dermatitis	Recurrent exposure over several years	Can result in an eczematous appearance of the skin. Ulcerations and carcinoma are not uncommon.

RADIATION EXPOSURE

- Clinical impact of reexposure depends on type of radiation involved, amount, and nature of exposure, total time of exposure, distance from source and the amount and type of shielding.
- Exposure over a prolonged period of time is less likely to be harmful than the same dose over a shorter period of time.
- There is an inverse square relationship between distance from a radiation source and the resultant exposure, making increased distance an effective means to reduce exposure. (Tripling the distance from the source reduces exposure by a factor of 9.)
- Shielding can reduce radiation exposure from low-energy sources like x-rays, but is impractical in the setting of medium- or high-energy radiation because of the amount of lead or concrete required.
- Background ambient radiation may represent an annual exposure of between 300 and 360 mrem. Radon accounts for the largest portion of the background radiation (~200 mrem/year). Technological sources include color television 1 mrem/y; coast-to-coast airline travel 2–5 mrem; a chest x-ray 5–10 mrem and angiography 1000 mrem.

RADIATION INJURIES

EXPOSURE

- Exposure radiation injuries can be classified as either a *localized radiation injury* or a *whole-body exposure*.
- A large dose of radiation to a small part of the body will result in a local radiation injury; these injuries often occur over months to years. Common sites include the upper extremities, buttocks, and thighs and are usually the result of an inadvertent occupational exposure. The initial clinical picture of a localized radiation injury is a thermal injury to

the skin—erythema from a local radiation injury is often delayed. Local radiation thermal injury can be painless and lead to tissue necrosis. Classification is divided into four types based on increasing epidermal and dermal injury (Table 143-1).

- Whole body exposure of 100 rad or more over a short time period may result in *acute radiation syndrome*. Organ systems with rapidly dividing cells (bone marrow and gastrointestinal tract) are the most vulnerable. With significant exposure, all organ systems may be affected including the central nervous system. It can be difficult to estimate the exposure of a whole-body radiation victim; dosimeters worn by victims during time of exposure are helpful. Biologic dosimetry via history, physical examination, and baseline laboratory testing is commonly all that is available (Table 143-2).
 - Biologic dosimetry for whole-body radiation exposure uses time of onset of symptoms and depression of absolute lymphocyte count as primary indicators of quantifying exposure. The earlier signs and symptoms develop, the higher the exposure. A biologic dosimetry calculator is available through the Radiation Event Medical Management Website (https://www.remm.nlm.gov/ars_wbd.htm). Gold standard dosimetry involves chromosomal cytogenetics and is available only to the military.
 - A progressive sequence of signs and symptoms following a whole-body exposure can be divided into four phases: prodromal, latent, manifest illness and recovery, or death.
 - Prodromal symptoms include nausea, vomiting, fatigue, and can begin within minutes to hours of exposure. Exposure to less than 100 rad rarely produces symptoms. Patients who do not exhibit symptoms within 6 hours are unlikely to have had significant whole-body exposure (>100 rad). Higher dose exposures produce symptoms more rapidly, likely due to acute tissue injury and associated release of vasoactive substances.

TABLE 143-2 Biological Dosimetry

INDICATOR	TOTAL-BODY DOSE	COMMENTS
Nausea and vomiting		
Onset within 6 h	>100 rad (1 Gy)	Prodromal stage represents a good clinical and biologic indicator to estimate whole-body exposure
Onset within 4 h	>200 rad (2 Gy)	
Onset within 2 h	>400 rad (4 Gy)	
Onset within 1 h	>1000 rad (10 Gy)	
Lymphocyte count at 48 h		
>1200/mm ³	100–200 rad (1–2 Gy)	Prognosis: good
300–1200/mm ³	200–400 rad (2–4 Gy)	Prognosis: fair
<300/mm ³	>400 rad (4 Gy)	Prognosis: poor
Diarrhea	>400 rad (4 Gy)	
Erythema of the skin	>600 rad (6 Gy)	Delayed onset
CNS symptoms (disorientation, ataxia, seizures, coma)	>1000 rad (10 Gy)	Rule out trauma Death within days

- A lower dose exposure will yield a resolution of the prodromal symptoms over days to weeks during the latent stage.
- During the manifest illness stage, specific organ symptoms develop and the patient is at greatest risk of infection and hemorrhage. Three syndromes may develop during this stage, depending on the total amount of radiation exposure: the hematopoietic syndrome (220–600 rad), the gastrointestinal syndrome (600–1000 rad) and the neurovascular syndrome (>1000 rad).
- Although the effect of radiation on the hematopoietic system is characterized by pancytopenia, the absolute lymphocyte count represents the best way to estimate exposure hematologically. Leukocyte counts may be elevated initially because of demargination, but the lymphocyte portion of the differential will quickly start to decrease. An absolute lymphocyte count >1200/mm³ indicates a 100- to 200-rad exposure and most often a good prognosis; 300 to 1200/mm³ suggests a 200- to 400-rad exposure, which promises a fair outcome. Exposure to >400 rad is marked by a poor prognosis and is expected with lymphocyte counts <300/mm³. Pancytopenia may develop after a few days to weeks; the patient will subsequently suffer from dyspnea, malaise, purpura, bleeding, and opportunistic infection.
- Gastrointestinal illness will be most evident with total-body exposures of 600 to 1000 rad; it is marked by severe vomiting and diarrhea. The radiosensitive mucosal cells of the small bowel begin to slough, which, combined with the coexistent hematopoietic abnormalities, produces severe bloody diarrhea, relentless fluid loss, fever and prostration. Even with intense supportive care, the patient rarely survives.

- Total-body irradiation with >1000 rad results in a neurovascular syndrome; ataxia and confusion quickly develop and there is direct vascular damage, with resultant circulatory collapse. The patient usually expires within hours (Table 143-3).

CONTAMINATION

- Neutrons, β -particles and α -particles are most commonly responsible for contamination. Radioactive particles, solid or liquid, may remain on the victim, resulting in external contamination. Internal contamination may be the result of inhaled, ingested, or absorbed radioactive particles.
- Unlike exposure, contamination creates potential risk to hospital and prehospital personnel. In most situations, decontamination should begin in the prehospital setting.
- Prehospital management
 - First assess and stabilize airway, breathing, and circulation. No acute, life-threatening complications of a survivable radiation injury require immediate intervention.
 - Emergency treatment is supportive and directed toward preventing complications. Quickly determine whether patients were exposed or contaminated as contamination requires that decontamination begin promptly after stabilization.
 - History obtained by prehospital personnel is of paramount importance and includes the exact type, location, and duration of exposure, route of entry, type and quantity of radioactive material.
 - Rapid decontamination limits exposure to the victim and decreases further contamination of health care equipment and personnel. At minimum, contaminated clothing should be removed before any patient, even unstable patients are transported.

TABLE 143-3 Whole-Body Exposure

WHOLE-BODY DOSE, RAD	CHARACTERISTICS
5	Asymptomatic; normal blood studies
15	Chromosome abnormality may be detectable
50–75	Asymptomatic; minor depression of platelets and white cell count may be detectable
75–100	Nausea, vomiting, fatigue in 10%–15% of victims within 2 d
100–200	Prodrome: mild nausea, vomiting, and fatigue; onset within 6 h, lasting 3–6 h Latent stage: >2 wk Manifest illness stage Lymphocyte count >1200/mm ³ at 48 h Transient sterility in men Recovery: good prognosis with only symptomatic treatment
200–600	Prodrome: nausea and vomiting within 2–4 h, last <24 h Latent stage: 1–3 wk Manifest illness stage: hematopoietic @ 200–400 rad: lymphocyte count 300–1200/mm ³ at 48 h >400 rad: lymphocyte count <300/mm ³ at 48 h Pancytopenia may develop after a latent period of up to 3 wk: the patient will subsequently suffer from dyspnea, malaise, purpura, bleeding, and opportunistic infection Requires hospitalization, protective isolation, and support Upper dose range may require bone-marrow transplantation within 7–10 d of exposure Recovery: @ 200–400 rad: fair prognosis with supportive care and if the bone-marrow damage was not irreversible @ 400 rad: poor prognosis; lethal in approximately 50% of victims
600–1000	Prodrome: severe nausea, vomiting, and diarrhea within 1–2 h, lasting >48 h Latent stage: 0–7 d Manifest illness stage: gastrointestinal Recurrence of nausea and vomiting Fever, bloody diarrhea, dehydration, electrolyte imbalance, early sepsis, hemorrhage Leukocyte count drops to zero Recovery: Overall 90%–100% mortality within 30 d Lower-dose exposure with medical care has a 50% mortality
>1000	Prodrome: nausea and vomiting within 1 h Latent stage: none Manifest illness stage: neurovascular Dehydration, hypotension Disorientation, ataxia, confusion, seizures, coma Erythema and epilation (onset may be delayed) Recovery: 99%–100% incidence of death within days
>5000	Prodrome: almost immediate onset of nausea, vomiting Latent stage: none Manifest illness stage: cardiovascular, GI, and CNS. Hypotension, ataxia, cerebral edema, seizures (rapid onset) Recovery: death within 1–4 d

- After transport, EMS personnel and their vehicles must be inspected for the presence of radioactive contamination before they leave the facility.
- Few hospitals will be treat life-threatening radiation accidents unless they are in close proximity to nuclear power plants or there is nuclear war. It is more likely that hospitals will treat minor industrial or transport accidents.

RADIATION ACCIDENT PLAN

- The Joint Commission requires each emergency department to have a radiation accident plan. A major part of a well-prepared plan facilitates the identification of *significant* versus *perceived* radiation dangers.
- The final component of an emergency department's resource plan for radiation emergencies is a list

of “additional references” including local, state, regional, and national agencies and their 24-hour telephone numbers.

- The U.S. Department of Energy is available to coordinate a federal response and provide assistance through the radiological assistance program (RAP).
- The REAC/TS in Oak Ridge, Tennessee, is also available to provide treatment and medical consultation for injuries resulting from radiation exposure and contamination (REAC/TS Telephone 865-576-1005).

GENERAL PROCEDURES

- When exposed solely to irradiation from γ -rays, x-rays, β -particles and, frequently, neutrons, patients do not become radioactive.
- Separate contaminated and clean treatment areas must be established.
- The floor of the contaminated treatment area and the ambulance receiving area must be covered with plastic or paper sheets.
- All personnel in the treatment area must wear gowns, caps, masks, shoe covers, double gloves, and personal monitoring devices (film badges). If airborne contaminants are suspected, respirators must be worn. In cases of a highly radioactive contaminant or foreign body, a lead shield or apron is required.
- Separate staff members are assigned to the clean and contaminated treatment areas.
- Ideally, several people should be assigned to each contaminated patient in order to decrease individual exposure time maintain more distance from the patient when the health worker is not involved in direct decontamination or medical management.
- A radiation safety officer should monitor the treatment area and oversee decontamination with a Geiger-Mueller counter for detecting β - and γ -radiation or a scintillation detector, which offers a higher sensitivity in detecting α -, β -, γ - and neutron particles.
- Patients enter the emergency department through a separate entrance where radiation detection equipment is in place. Patients on ambulance stretchers are transferred to clean hospital carts in the ambulance bay.
- The ideal decontamination site is an isolated room designed with a closed drainage and ventilation system and fully equipped for a major resuscitation. Following resuscitation, the radiation victim is carefully evaluated to determine if there is any surface contamination or if there is the possibility of inhaled or ingested radioactive material.
- Burns and open wounds must be evaluated for contamination, irrigated copiously with water and exam-

ined for foreign bodies. Highly contaminated foreign bodies, while rare, may represent the greatest single hazard to hospital personnel.

- Radiation burns may be delayed in their presentation. They are managed in the same way as nonradiation-induced partial- and full-thickness burns.
- Obtain a baseline complete blood count, differential, platelet count, and electrolytes and take blood for serial laboratory tests at 12 and 24 hours. Cross type patients with a decrease in absolute lymphocyte count in case a bone-marrow transplant is needed.
- Manage infections as usual and give severely neutropenic patients broad-spectrum antimicrobial agents for prophylaxis, including a fluoroquinolone with streptococcal coverage or a fluoroquinolone without streptococcal coverage plus penicillin or amoxicillin, antiviral drugs and antifungal agents.
- Not all victims will require hospitalization; exposures >100 rad may warrant inpatient care. If radiation victims exhibit severe vomiting, they should be admitted. Reverse isolation measures are used for all documented exposures of 200 to 1000 rem and for those patients with absolute lymphocyte counts $<1200/\text{mm}^3$ or 50% of the baseline value. Treatment with colony-stimulating factors should be considered for those at risk for developing neutropenia.
- Victims may develop significant fluid and electrolyte complications. Any indicated surgery must be performed without delay to avoid these additional problems. Transfusion of selected blood products is based on the individual hematologic derangement encountered and should follow the usual guidelines for their use.

EXTERNAL CONTAMINATION

- When dealing with an external contamination, it is important to prevent it from becoming an internal contamination.
- Begin decontamination as soon as possible and ensure rescue personnel wear protective clothing, including rubber gloves, shoe covers, masks, and film badges.
- Up to 70% to 90% of external contamination can be eliminated by covering open wounds and removing the patients clothing. Articles of clothing should be placed in labeled plastic bags.
- Any open wound is considered contaminated until proven otherwise, and decontamination should precede the irrigation. Wash with copious amounts of water and soap, with particular attention to skin folds, ears, and fingernails.

- The use of damp washcloths, rather than rinsing with running water, may be more practical for some emergency departments. Disposal of contaminated washcloths in plastic bags may be easier than the collection of contaminated wash water.
- All waste must be captured in sealed containers labeled “Radioactive Waste.”
- A dosimeter should be used to determine the completeness of the decontamination. The goal is to get the radiation level “as low as reasonably achievable.” This is commonly referred to as the ALARA principle.

INTERNAL CONTAMINATION

- Radioactive particles that are ingested or inhaled or that contaminate open wounds can cause significant cellular damage. These particles will continue to irradiate tissues until they are eliminated, neutralized, or blocked, or until they decay naturally.
- In general, there is a 1- to 2-hour window of time during which absorption of these particles occurs. It is crucial that any interventions be performed during this period and as soon as possible.
- It may be difficult to determine the presence of an internal contaminant, especially if an external contaminant still clouds the picture. Clues may include evidence of contamination around the mouth and nose.
- Diethylenetriaminepentaacetic acid (DTPA) administered as calcium-DTPA (Ca-DTPA) or zinc-DTPA (Zn-DTPA) as injectable chelators is approved for removal of plutonium and other transuranics (e.g., americium and curium) from the body.
- Prussian blue (ferric ferrihexacyanate) is approved for the treatment of known or suspected internal contamination with radioactive cesium and thallium.
- Potassium iodide (KI) is a blocking agent that reduces the uptake of radioactive iodine (^{131}I) by the thyroid gland, and is most effective when given soon after exposure.
- Initial stabilization and decontamination of radiation ingestions are the same as those for “routine” ingestions: to prevent absorption and enhance elimination. Gastric decontamination procedures such as gastric emptying methods and activated charcoal are used. All bodily excretions (lavage fluid, emesis, urine, and feces) should be saved and labeled for radioactive evaluation and proper disposal.
- Acute inhalation is much less common than chronic low-level exposure and can occur in the event of a radioactive accident in conjunction with a fire or explosion. A moistened cotton-tipped applicator can be used to swab the nasal passages and check for radioactivity. Bronchopulmonary lavage is performed

for removal of particulate matter. Specific blocking agents and chelating agents should be administered in this setting.

- Decontaminated wounds can be surgically closed. Wounds that remain contaminated are left open for 24 hours and may require debridement for further decontamination. Contaminated surgical instruments must be replaced to prevent further wound contamination. Amputation of contaminated extremities is rarely indicated.

EXPOSURE

- Despite the significant illness and injury that can result from either a local radiation injury or a whole-body exposure, an emergency physician can offer only limited treatment.
- Aggressive supportive care is the mainstay of treatment for these patients including fluid resuscitation for severe vomiting and diarrhea, standard trauma, and burn care. Prophylactic antimicrobial agents, administration of cytokines and stem cell transplants are other measures that may help decrease morbidity and mortality.
- For triage purposes, patients with whole-body exposure of >1000 rad will likely die within 2 to 3 weeks and should be classified as expectant or impending.

SPECIAL CONSIDERATIONS

- Nuclear explosions present unique logistic and patient care issues: medical resources may be quickly depleted depending on the number of victims and the magnitude of injuries. Routine communications equipment, electronic equipment, and computers may be rendered useless by the electromagnetic pulse generated by the nuclear blast.
- Victims of a nuclear explosion will be subject to three types of injury patterns. *Mechanical trauma* (blunt and penetrating) secondary to the blast effect of the explosion accounts for 50% of the released energy while *thermal injury* from heat dissipation represents 35% of the energy release. The remaining 15% of the thermonuclear energy release will cause *radiation injury*, 10% from radioactive fallout, and only 5% as a result of the immediate release of γ -rays and neutrons.

PROGNOSIS

- Prognosis for survival and the concern for delayed complications, while important, are not of immediate concern to the emergency physician.

- Leukemia and delayed thyroid cancer and breast cancer are of significant concern in children <10 years of age.
- In utero exposure to as little as 5 to 10 rad can be associated with mental retardation or a small head circumference.
- Based on the presenting symptoms, patients can be classified into three major prognostic classifications: *survivor probable*, *survivor possible*, and *survivor improbable*.
- Survivor probable includes individuals who are asymptomatic or who have minimal complaints that resolve within hours.
- Initial and subsequent leukocyte counts are not affected and estimated exposure is <200 rad (2 Gy).
- Following satisfactory decontamination, inpatient care is rarely needed.
- Survivor possible consists of patients with relatively brief gastrointestinal sequelae, usually lasting <48 hours. After initial presentation and the latent period, patients develop characteristic pancytopenia.
- Estimated exposure for this group is between 200 and 800 rad (2–8 Gy). An exposure of 400 rad represents the median lethal dose.
- Survival in this group is influenced by the aggressiveness of supportive therapy and hematologic intervention, antecedent health of the victim and the response to bone-marrow transplantation (when indicated).
- Survivor improbable victims have estimated whole-body exposure that exceeds 800 rad (8 Gy). The prognosis is dismal despite aggressive supportive therapy and even the implementation of bone-marrow transplantation.
- When nausea, vomiting, and diarrhea begin within 1 hour of exposure and CNS symptoms appear early, a relatively early death can be expected.

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QUESTIONS

1. A 5-year-old girl brought in by her mother presents with a chief complaint of nausea and vomiting for the past 5 days. The mother explains that she had her child evaluated in another emergency department the day before and they ordered a CT scan of the abdomen and a chest x-ray. The mother believes her child has acute radiation illness. Your best course of action is
 - A. Order a complete blood count.
 - B. Admit the child for symptomatic management.
 - C. Administer antiemetics, IV fluids, and obtain a repeat CT scan for possible acute appendicitis.
 - D. Explain to the mother that acute radiation illness is highly unlikely and discharge the patient to follow up with primary pediatrician.
 - E. Explain to the mother that acute radiation illness is highly unlikely and take a full history and perform physical examination as appropriate for routine patients with vomiting.
2. Which of the following are the most radiosensitive cells?
 - A. Muscle cells
 - B. Blood cells
 - C. Nerve cells
 - D. Skin cells
 - E. Bone cells
3. Which of the following methods is most effective in reducing external contamination with radioactive material?
 - A. Vigorous rubbing to displace contaminated debris from body surfaces.
 - B. Covering open wounds and removing the victim's clothing.
 - C. Infusing chelating agents intravenously.

- D. Continuous copious irrigation with hot water.
E. Wrapping the victim in lead blankets.
4. You are on duty in the pediatric emergency department and the charge nurse alerts you that a 12-year-old boy is being transported from a radiation accident with contact exposure. Decontamination is in progress by EMS. In preparation for the patient's arrival, you instruct the ED staff to gather which of the following for personal protective equipment?
A. Gowns and gloves
B. Eye protection
C. Prophylactic potassium iodide
D. Irrigation equipment
E. Self-contained breathing apparatus
5. As the safety officer at a radiation emergency, you are responsible for monitoring treatment areas and everyone within. You carry a Geiger–Mueller counter (“Geiger counter”) which allows you to detect which types of radiation?
A. Alpha-particles only.
B. Beta-particles only.
C. Gamma-particles only.
D. Beta-, and gamma- particles only.
E. Alpha-, beta-, gamma- and neutron particles.
6. During a radiation emergency, you are caring for a 10-year-old female who is believed to have radioactive material imbedded in her left lower extremity. Appropriate treatment includes which of the following?
A. Amputation of lower-left extremity.
B. Topical application of chelating agent to open wound.
C. Rapid surgical removal of foreign body.
D. Copious irrigation until no radiation detected.
E. Intravenous type specific chelating agent administration.
7. You are caring for an 11-year-old male who presents 90 minutes after a motor vehicle collision, passenger vehicle versus commercial truck transporting radioactive materials. The patient has bloody diarrhea and intractable nausea and vomiting. EMS approximates exposure at approximately 700 rad. This patient most likely has which of the following:
A. Gastrointestinal syndrome.
B. Hematopoietic syndrome.
C. Neurovascular syndrome.
D. Pulmonary syndrome.
E. No radiation exposure.
8. Children are at greatest risk of developing subsequent malignancies when they are exposed to radiation. Which of the following are of significant concern in children less than 10 years of age when exposed to radiation?
A. Leukemia
B. Leukemia and lymphoma
C. Leukemia and delayed thyroid and brain cancer
D. Leukemia and delayed brain and breast cancer
E. Leukemia and delayed thyroid and breast cancer
9. According to the National Council on Radiation Protection, which of the following is the generally accepted radiation exposure limit threshold, under which morbidity is unlikely?
A. 5 rem
B. 10 rem
C. 20 rem
D. 40 rem
E. 80 rem
10. A 7-year-old presents after radiation exposure and on evaluation has an absolute lymphocyte count of less than 300 per cubic millimeter. This patient has likely been exposed to which amount of radiation and has which of the following prognoses?
A. <100 rad; good prognosis
B. Between 100–200 rad; fair prognosis
C. Between 200 and 300 rad; fair prognosis
D. >400 rad; poor prognosis
E. >400 rad: good prognosis

ANSWERS

- E. Medical imaging confers a relatively small exposure to radiation, CXR 5–10 mrem, CT Angiography <1000 mrem. Acute radiation illness may develop following a whole-body exposure of 100 rad or more. Although radiation is a legitimate concern on the part of the mother, other diagnoses are much more likely in a 5-year-old with 5 days of nausea and vomiting. Answers A, B, C, D are incorrect; a complete history, physical examination, and routine evaluative testing and treatment are indicated.
- B. Cells with rapid turn over, highest mitotic activity, are most sensitive. Muscle and nerve cells are permanent. Skin and bone cells are relatively less rapid in turn over than hematopoietic cell lines.
- B. Up to 70–90% of external contamination can be eliminated by ensuring any open wounds are covered and that the victim's clothing is removed. All articles should be placed in plastic bags. Although copious irrigation is helpful, hot water should not

be utilized, therefore answer choice D is incorrect. Rubbing the skin surface may increase penetrance of radioactive debris by disrupting skin integrity and increasing contact. Chelating agents do not have a role in decontamination and wrapping the patient in lead does nothing to remove the radioactive contamination on the victim's external surfaces.

4. A. All personnel in treatment areas must wear protective clothing including gowns, caps, masks, shoe covers, double gloves, and personal monitoring devices (film badges). Eye protection alone will offer little protection. Potassium iodide may have a role in patient management, depending on the specific exposure, but has no role in personal protection for ED staff. Irrigation equipment may be important for patient decontamination, but not staff PPE. Self-contained breathing apparatus is not routinely appropriate unless radioactive material was airborne or gaseous.
5. D. This officer is given a Geiger–Mueller counter for detecting β - and γ -radiation. A scintillation detector offers a higher sensitivity by detecting α -, β -, γ -, and neutron particles.
6. C. Highly contaminated foreign bodies, while rare, may represent the greatest single hazard to hospital personnel. These contaminants must be removed from the victim as safely and quickly as possible. Amputation is rarely indicated unless foreign body cannot be removed. Topical or intravenous chelating agents and copious irrigation will not prevent ongoing radiation exposure from the embedded radioactive foreign body; it must be removed.
7. A. Gastrointestinal illness will be most evident with total-body exposures of 600 to 1000 rad. Often within hours of exposure it presents with severe vomiting and diarrhea. The radiosensitive mucosal cells of the small bowel begin to slough. Total-body irradiation with >1000 rad results in a neurovascular syndrome; ataxia and confusion quickly develop and there is direct vascular damage, with resultant circulatory collapse. Although the effect of radiation on the hematopoietic system is characterized by pancytopenia, the absolute lymphocyte count represents the best way to estimate exposure hematologically. Leukocyte counts may be elevated initially because of demargination, but the lymphocyte portion of the differential will quickly start to decrease. No specific pulmonary syndrome has been described, although pulmonary symptoms may be associated with the other syndromes. This scenario describes illness from radiation, not just exposure.
8. E. Leukemia and delayed thyroid cancer and breast cancer are of significant concern in children <10 years of age. Prognosis for survival and the concern for delayed complications, while important, are not of immediate concern to the emergency physician. In utero exposure to as little as 5 to 10 rad can be associated with mental retardation or a small head circumference.
9. A. A threshold of 5000 mrem (5 rem) should be the exposure limit, except to save a life. A once-in-a-lifetime exposure to 100,000 mrem (100 rem) to save a life has been established by the National Council on Radiation Protection as acceptable and will not result in any undue morbidity.
10. D. Although the effect of radiation on the hematopoietic system is characterized by pancytopenia, the absolute lymphocyte count represents the best way to estimate exposure hematologically. Leukocyte counts may be elevated initially because of demargination, but the lymphocyte portion of the differential will quickly start to decrease. An absolute lymphocyte count >1200/mm³ indicates a 100- to 200-rad exposure and most often a good prognosis; 300 to 1200/mm³ suggests a 200- to 400-rad exposure, which promises a fair outcome. Exposure to >400 rad is marked by a poor prognosis and is expected with lymphocyte counts <300/mm³. Pancytopenia may develop after a few days to weeks. The patient will subsequently suffer from dyspnea, malaise, purpura, bleeding, and opportunistic infection.

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144 **SEXUAL ABUSE**

Sara L. Beers
Matthew Cox

ETIOLOGY

- Child sexual abuse is defined as contact or interaction between a child and an individual when a child is being used for the sexual stimulation of that individual.
- Children are most often abused by adults or older children who are known to them, with the victim knowing the offender in 8 out of 10 reported cases.
- The offender is more frequently male.
- Risk factors associated with child sexual abuse include:
 - Poor parent–child relationships.
 - Poor relationships between parents.
 - Absence of a protective parent.
 - Presence of a nonbiologically related male in the house.

EPIDEMIOLOGY

- Sexual abuse affects approximately 100,000 children each year in the United States.
- Most abuse goes unreported during childhood.
- An estimated 20% of girls and 9% of boys are the victims of sexual abuse during childhood.
- Children are most likely to be abused sexually during preadolescence, around the age of 8 to 12 years.
- Sexual abuse of children crosses all socioeconomic status and ethnicities.

PATHOGENESIS

- The vast majority of children who are victims of sexual abuse will have normal exams.

- Factors that contribute to exams being found normal include the following.
 - Most sexual abuse of children occurs without the use of physical force or restraint.
 - Anogenital tissue heals very quickly.
 - Genital tissues are elastic in nature making them less prone to injury.
 - Delayed disclosure gives injuries time to heal.
- When injuries do occur, they can involve the genitalia, anus, oral cavity, extragenital sites, or any combination of the above.
- Injuries may include superficial abrasions, bruises, tearing of the hymen, or deeper genital injury.
- Findings of the hymenal tissue from 3- to 9-o'clock positions (with the child in a supine position) are particularly noteworthy when assessing for injuries from abuse.
- Key issues in discerning accidental injuries such as straddle injuries from sexual abuse:
 - Straddle injuries typically include injury to the clitoris, clitoral hood, mons pubis, and labia.
 - Straddle injuries are usually asymmetric.
 - Straddle injuries do not involve the hymen.

RECOGNITION

- Concern for child sexual abuse often arises when:
 - A child discloses abuse.
 - There is abnormal appearance of a child's anus or genitals.
 - There are behavior changes in a child.
- Sexual abuse may present with a variety of nonspecific symptoms ranging from anogenital pain, itching, bleeding or discharge, abdominal pain, dysuria, constipation, and/or painful defecation.
- A detailed external genital examination of the female child should include noting the appearance of the labia majora, labia minora, urethral meatus, posterior fourchette, fossa navicularis, and the hymen. See Fig. 144-1 for normal anatomy.

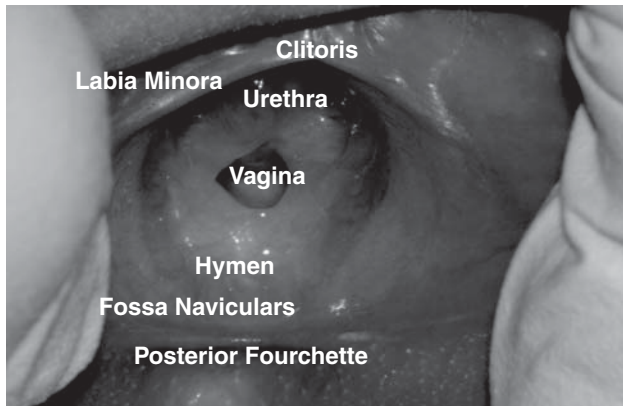


FIG. 144-1. Normal female anatomy.

- Physician and medical providers must be able to identify other medical conditions in children that may present as suspected sexual abuse.
 - Various soaps, shampoos, and lotions may cause chemical irritation.
 - Restrictive clothing may lead to mechanical trauma.
 - Poor hygiene can lead to fecal contamination and cause pruritus.
 - Pinworms and scabies can present as pruritus of the anogenital area.
 - Perianal streptococcal dermatitis is a bright red, sharply demarcated tender rash.
 - Candidiasis is uncommon in healthy prepubertal girls, but can be associated with oral antibiotics, diabetes mellitus, and immunodeficiency.
 - Peripubertal and pubertal girls can develop vaginal candidal infections that present with pruritus, inflammation, and a thick white discharge.

MANAGEMENT

- Management of the child with suspected sexual abuse involves both medical management and legal management and is required by law to be reported to child protective services (CPS) and law enforcement.
- The history taken from the child is often the most important part of the overall evaluation, and thus great detail should be taken when documenting the history provided, with actual quotes from the child when possible.
- The child should be interviewed away from the parent if possible, using open-ended nonleading questions. “W” words (who, what, where, when, and how) are recommended.
- The child should not be repeatedly questioned or interviewed by multiple examiners whenever possible.

- Children should have an immediate medical evaluation if the abuse was within the previous 72 hours or there is bleeding or concern of acute injury. The AAP recommends forensic evidence collection if the evaluation is within 72 hours of penetrating sexual abuse; however, the yield from such evidence collection significantly decreases after 24 hours.
- Prepubertal children should *not* have a speculum examination. A detailed external genital examination is sufficient. In cases with suspected intravaginal injuries or active bleeding without an obvious external source, one must consult a pediatric general surgeon who can perform an exam and repair if needed in the operating room under anesthesia.
- Universal screening for sexually transmitted diseases (STDs) is not necessary because the incidence of STDs among children who have been sexually abused is low. When cultures are needed, swabs taken from the external genitalia are sufficient in prepubertal female patients. In adolescent female patients with history of rape, a speculum examination with cervical cultures is recommended.
- Empiric treatment for STDs is usually not necessary, except in cases of stranger assaults and in adolescent rape victims.
- Blood tests for syphilis, HIV, hepatitis B and C should also be considered in high-risk cases.

ANCILLARY STUDIES

- Colposcopy provides a noninvasive method for visualizing the anogenital structures. It provides magnification and a light source, both of which can be helpful in identifying injury. It also allows video or still photography for documentation.
- Alternative light sources, including the Blue Max 6000 or a Wood’s lamp, can be used during forensic evidence collection to guide collection of possible body fluids on victims. It is important to note though that material other than semen may also fluoresce with a Wood’s lamp.

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QUESTIONS

- A 10-year-old girl is brought into the ED by her mother after disclosing that about a week prior the 18-year-old neighbor put his penis in her vagina and in her bottom. The girl told her mom that it also happened several times in the past but he told her not to tell. Her exam is normal. Which of the following explains the lack of physical findings?

 - Genital tissues are elastic in nature making them more prone to injury.
 - Genital tissues are well vascularized and injuries heal slowly.
 - There may have been injuries that have already healed.
 - Most sexual abuse occurs with the use of physical force.
 - The anus can enlarge to large diameters to pass bowel movements, and thus penetrating injuries to the anus are common.
- A 6-year-old girl is brought into to the ED by her parents after coming into the house crying and holding her genital area. Mom found some blood in her underpants. She had been playing with some neighbor kids, one of whom tells the girl’s parents that he saw her stop suddenly on her bike and straddle the bar on the bike. Which if the following are commonly found with accidental straddle injuries?

 - Hymenal tears in the 9 o’clock to 3 o’clock position.
 - Hymenal tears in the 3 o’clock to 9 o’clock position.
 - Bruising to the posterior forchette and fossa navicularis.
 - Bruising to the posterior forchette.
 - Bruising to the clitoris, clitoral hood, mons, and labia.
- A 5-year-old boy is brought to the ED by his mother after telling her that his 17-year-old brother put his penis in his bottom. The mother tells you that she wants the boy checked but does not want to report anything because she does not think the older brother would really do this. As the emergency department physician, you are required by law to report the alleged abuse to the following:

 - Police department.
 - CPS.
 - Police department and CPS.
 - Leave the reporting to the discretion of the ED social worker.
 - Since mom is requesting no reports to be made, you do not have to make any reports.
- A 12-year-old female was sexually assaulted by mother’s boyfriend. This happened about 36 hours prior to coming to the ED. She has bathed and changed clothes since the assault. She is brought to the ED to be evaluated. With regard to forensic evidence collection, you should do which of the following:

 - Since she has bathed and changed clothes, there is unlikely to be any evidence yielded from a forensic collection and therefore it should not be done.
 - Since it has been greater than 24 hours, no forensic evidence needs to be collected.
 - Forensic evidence collection should be done only if the family requests it.
 - Forensic evidence collection should be done only if the patient requests it.
 - She should have forensic evidence collection.
- An 8-year-old girl is brought in to the ED by her mother after the babysitter’s boyfriend sexually assaulted her. The girl is complaining of vaginal pain and bleeding. Upon your external genital examination, you find moderate active bleeding from the vaginal opening. You are not able to visualize where the bleeding is coming from. You should:

 - Consult a pediatric surgeon to perform an exam and repair if needed in the operating room under anesthesia.
 - Get child life involved and use a speculum to get a better internal exam.
 - Provide midazolam anxyolysis and use a speculum to get a better internal exam.
 - Admit the patient until the bleeding resolves.
 - Check a CBC and if it is normal, discharge the patient and have her follow up for a repeat exam in 24 hours.
- A 7-year-old boy is brought in to the ED after disclosing to his mother that a male teacher put his

finger in his bottom at school today. Mom brought the child to the ED to be evaluated. Which of the following is true when interviewing a child about the alleged sexual abuse:

- A. The child should be interviewed many times to make sure there are no variations in his story.
 - B. Open ended questions including the use of “why?” should be used.
 - C. The child’s history should be carefully documented using actual quotes from the child when possible.
 - D. The child should always be interviewed with the parent at the bedside.
 - E. None of the above.
7. A 10-year-old girl is brought into the ED by her mother after disclosing that her older stepbrother has been fondling her genitalia. She has no specific complaints. Her GU exam is normal without evidence of infection. In regards to screening for STDs, you as the ED physician should:
- A. Screen for HIV, *Chlamydia trachomatis*, *Neisseria gonorrhoeae*, syphilis, and trichomonas.
 - B. Screen for *Chlamydia trachomatis*, *Neisseria gonorrhoeae*, syphilis, and trichomonas.
 - C. Screen for *Neisseria gonorrhoeae*, *Chlamydia trachomatis*, and trichomonas.
 - D. Screen for *Neisseria gonorrhoeae* and *Chlamydia trachomatis*.
 - E. No need to screen for STDs.

ANSWERS

1. C. With frequent delays in disclosure of sexual abuse, injuries that may have been present at the time of the abuse will often be healed by the time the child undergoes a physical examination. Genital tissues are elastic in nature making them less prone, not more prone to injury. Genital tissues are well vascularized so injuries heal quickly, not slowly. Most sexual abuse occurs without the use of physical force. The anus can enlarge to large diameters to pass bowel movements, and thus penetrating injuries to the anus are rare, not common.
2. E. Accidental straddle injuries on playground equipment, toys, furniture, etc often result in physical injuries. Key in discerning such injuries from sexual abuse is that straddle injuries typically include injury to the clitoris, clitoral hood, mons pubis, and labia. Also important to note is that straddle injuries are usually asymmetric and do not involve the hymen. Conversely, the posterior fourchette, fossa navicularis, and posterior hymen are the structures/area that are injured with penetrating sexual abuse.
3. C. All cases of suspected sexual abuse in children are required by law to be reported to CPS and law enforcement. Often a hospital’s social worker is involved and helps with reporting the suspected abuse to law enforcement and CPS, but it would never be left to the discretion of the social worker as to whether or not the reports need to be made. While parents will often not want reports filed, it is the law and must be done.
4. E. The AAP recommends forensic evidence collection if the evaluation is within 72 hours of the sexual abuse. The yield from such evidence collection does significantly drop off when greater than 24 hours has passed and when the patient has bathed or changed clothes, but it should still be done with penetrating sexual abuse. The requests of the family and patient should not factor into the decision to collect forensic evidence.
5. A. When there is concern for internal injuries a pediatric surgeon should be consulted to perform an exam and repair if needed in the operating room under anesthesia. Prepubertal children should never have a speculum exam. Admitting the child until the bleeding resolves or sending the child home to be rechecked in 24 hours are not acceptable as there may be significant ongoing blood loss and/or injuries that need immediate surgical repair.
6. C. The history taken from the child is often the most important part of the overall evaluation. The child’s history should always be carefully documented using actual quotes from the child when possible. If a skilled social worker has already obtained a detailed history from the child, the physician’s interview can be abbreviated. Further questioning of a child can be deleterious. The child may find repetitive questioning unpleasant or threatening, may infer that he or she is not believed, or may modify his or her history in response to repetitive questioning. Ideally, the interviewer should use opened-ended nonleading questions. “W” words (who, what, where, when, and how) are recommended. However, “Why” questions should be avoided because they may imply blame to the child. If possible, it is recommended that children with a developmental age of 3 or older should be interviewed alone. Having the family leave the room for the interview will often allow the child to open up more without the fear of upsetting the family or getting into trouble.
7. E. Universal screening for STDs is not necessary because the incidence of STD’s among children who have been sexually abused is low. The Centers for Disease Control and Prevention recommend testing for STD in the following situations: when the child has had symptoms or signs of an STD, when

a suspected assailant is known to have an STD or to be at high risk for STD's, when a sibling or another child or adult in the household or child's immediate environment has an STD, when the patient or parent requests testing, or when evidence of genital, oral, or anal penetration or ejaculation is present. In nonacute evaluations, careful examinations without STD screening may be acceptable in asymptomatic, prepubertal children who lack clear history or physical examination findings indicative of penetrating sexual abuse.

145 ABUSE AND NEGLECT

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INTRODUCTION

Child abuse is a serious cause of morbidity and mortality affecting young children in the United States and throughout the world. The identification of an abused child is challenging because the history provided is typically inaccurate, physical findings are nonspecific, and the physician fails to include abuse in the differential diagnosis. A careful and complete medical exam and detailed documentation is important in cases of suspected abuse. This chapter delineates the types of abuse most commonly seen in the emergency department, describes historical and physical indicators to help differentiate inflicted from noninflicted injuries, describes diagnostic studies useful in the medical evaluation, and discusses the legal obligations to report suspected abuse and neglect. It is vital that emergency medical care providers recognize, evaluate, and report suspected child abuse and neglect in order to facilitate the safety and well-being of children.

EPIDEMIOLOGY

- Child physical abuse is physical harm to a child at the hands of a caregiver that may encompass a single incident or repeated incidents.
- Examples include abusive head trauma, immersion burns, skeletal injuries, and inflicted, patterned bruises.
- 3.3 million referrals to child welfare agencies in the United States in 2006, with abuse documented in 30% of cases.

- More than 1500 deaths in the United States annually are related to abuse and neglect.
- Abuse is ranked as the third leading cause of homicide in children older than 1 year of age.
- The youngest children are most at risk for being abused.
- Children commonly present initially to an emergency department with injuries or medical problems caused by abuse and neglect.
- The spectrum of child abuse and neglect is broad and includes physical abuse (16%), sexual abuse (9%), emotional abuse (6.6%), and neglect (60%).
- The diagnosis of child abuse depends on information obtained from the medical history, physical examination, and injuries identified by ancillary studies.
- A detailed medical record must be kept in cases of suspected abuse, since this information will be frequently used by investigating agencies such as the police and child protection services.

ETIOLOGY

- Parental risk factors that are associated with child abuse:
 - Maternal age less than 19 years.
 - Single marital status.
 - Late or no prenatal care.
 - Parental depression.
 - Lack of maternal education.
 - Parental substance abuse.
 - Parental mental illness.
 - Domestic violence.
- Risk factors for physical abuse involving children include:
 - Male gender and young age.
 - Prematurity .
 - Chronic illness.
 - Congenital abnormalities, children with physical disabilities and behavioral problems.
 - Large family size and low family income.
- Fatal child abuse is most common among children in the first year of life.
- Children who live in poverty are overrepresented in the child welfare and foster care systems.
- Screening for domestic violence is an important aspect of the evaluation of child abuse.

MEDICAL HISTORY

- Obtain history from the caregiver and the child whenever possible.

- Note discrepancies between the clinical findings and historical data supplied by the parents.
- History is often inaccurate because the adult is either unaware of what happened to the child or is the perpetrator of the abuse.
- Victims are often too young or too ill to provide a history of their assault.
- Older victims may be too scared or intimidated to provide information.
- Distinguish accidental from inflicted injury.
- Medical conditions that mimic abuse need to be considered in the differential diagnosis; watch for:
 - Lack of history of trauma.
 - History of low-impact trauma in patients with persistent neurologic deficits.
 - Changing histories.
 - Trauma blamed on home resuscitative efforts.
- Consider inflicted injury in differential diagnosis for all pediatric injuries (Table 145-1).
- Include the location, time, and mechanism of any injury.
- Identify the caretakers at the time of the injury.
- Denial of trauma should be carefully documented.

PHYSICAL EXAMINATION

- Findings indicative of abuse may be found incidentally during a routine physical examination or during an examination specifically intended to look for injuries.
- Physical examination of a child with suspected inflicted injuries must be complete and all identified injuries should be carefully documented.
 - The child should always be completely undressed.
 - In older children, allow them to undress and put on a hospital gown while you step out of the room.
- In infants, subtle external injuries are often a clue to a more serious internal injury.
- 50% of children intentionally injured will have injuries to the head and neck.
- Bruises on the face and ears are highly concerning and warrant a detailed medical examination and evaluation.

TABLE 145-1 Historical and Examination Findings Suggestive of Abuse

FEATURES SUGGESTIVE OF ABUSE

- History inconsistent with injuries
- History incompatible with child's development
- History that changes with time
- Contradictory histories
- Delay in seeking treatment
- Pathognomonic injuries (such as forced immersion burn, multiple fractures in different stages of healing)

- Oral injuries might include torn frenula, lacerations to the mucosal surfaces or palate, and dental trauma.
 - Tears of the frenulum are highly suspicious in children who are not yet ambulatory.
 - May occur from a blow to the face or from an object such as a pacifier, spoon, or bottle being forced into an infant's mouth.
- Bruises, burns, and scars should be measured, and their size, shape, location, and color carefully documented.
- Photographs are important adjuncts to the recorded physical examination and are not a substitute for accurate medical documentation.

CUTANEOUS INJURIES

- The skin has the highest number of inflicted injuries.
- Burns are the most serious form of inflicted skin injury because they can be quite deep and involve large areas of a child's skin.
- The most important aspect of evaluating suspected abusive burns is correlating the history with the physical examination findings.
 - Does the mechanism make sense?
 - What was the temperature of the substance that caused the burn?
 - Duration of exposure?
- Water temperatures in excess of 120°F can result in burns within a few seconds, depending on the age of the patient and the location on the body (Table 145-2).
- Scald burns with hot tap water are the most frequent type of inflicted burns. The history of the injury must be carefully correlated with the observed pattern of injury, burn depth, and wound appearance.
- The immersion burn is a pathognomonic injury with involvement of the buttocks, posterior thighs, and feet, with relative sparing of the inguinal area.
- Immersion burns characteristically have uniform depth, an unvaried appearance, and distinct wound borders.

TABLE 145-2 Temperatures Required to Cause Full-Thickness Burns

TEMPERATURE OF WATER IN DEGREES CELSIUS (DEGREES FAHRENHEIT)	DURATION OF EXPOSURE
44 (111)	360 min
45 (113)	180 min
47 (116)	40 min
48 (118)	15 min
49 (120)	10 min
51 (124)	6 min
53 (127)	30 s
55 (131)	30 s
60 (140)	3 s

- Bruises in an unusual distribution or location are a cause for concern.
- The distribution of normal bruises varies by age and motor development.
 - Bruising is uncommon in nonambulatory children.
 - Bruises to the extremities and over other bony prominences are common in normal children.
 - Bruises centrally located, such as on the buttocks, chest, and abdomen, are less common.
- Estimating the age of a bruise is not recommended as it is fraught with multiple variables that affect accuracy.
- Patterned skin injuries, such as slap marks, loop marks, and bites, can be identified with careful examination.
- May present with signs and symptoms, such as hypovolemic shock, peritonitis, or nonspecific complaints, such as abdominal pain or vomiting.
- Some children have asymptomatic injuries.
- Most abusive abdominal injury is caused by blunt trauma that results in solid organ injury, perforation of a hollow viscous, or shearing of mesenteric vessels.
- Most victims are between the ages of 6 months and 3 years.
- Asymptomatic injuries may be discovered by routine screening of liver function tests and pancreatic enzymes or on abdominal computed tomography.

MUSCULOSKELETAL TRAUMA

- The physical examination may not always reveal skeletal deformity or tenderness.
- All children younger than 2 years with suspected abuse should have a skeletal survey.
- In battered infants, it is not uncommon to identify occult healing fractures indicating a pattern of repeated trauma.
- Skeletal injuries with a moderate to high specificity for child abuse include:
 - Posterior rib fractures, especially bilateral or multiple.
 - Metaphyseal fractures of the long bones.
 - Scapular fractures.
 - Fractures of the digits.
 - Sternal fractures.
- Skeletal injuries in nonambulatory children should prompt a medical evaluation for additional injuries.
- Skeletal injuries with a low specificity for abuse include:
 - Clavicle fractures
 - Long bone fractures
 - Linear skull fractures
- A follow-up skeletal survey approximately 2 weeks after the initial study increases the diagnostic yield, and should be considered when abuse is strongly suspected.

VISCERAL TRAUMA

- Accounts for less than 1% of identified cases of child maltreatment.
- Second most common form of fatal inflicted injury, after neurotrauma.
- May go unrecognized as it commonly results in nonspecific symptoms, and because external indicators of abdominal trauma are often absent, even with severe injury.

ABUSIVE HEAD INJURY

- The leading cause of morbidity and mortality in physically abused children.
- Classic triad associated with inflicted neurotrauma:
 - Subdural hemorrhages
 - Retinal hemorrhages
 - Metaphyseal fractures
- Refer to the injury with an inclusive term that does not specify the exact mechanism of injury, such as inflicted neurotrauma or abusive head trauma (AHT).
- The etiology of AHT is rarely clear because an accurate history is almost always lacking, and the mechanisms of injury varies among patients.
- Victims are generally younger than 3 years; most are infants
- Perpetrators tend to be men—fathers or a maternal boyfriend—although mothers and babysitters are frequently implicated.
- Symptoms vary from mild lethargy, vomiting, or irritability to apnea and coma.
- Seizures are common in victims of abusive head injury and are reported in up to 80%.
- Factors associated with missed diagnosis include age less than 6 months, caucasian race, both parents living in the home, and presentation with mild, nonspecific symptoms such as vomiting, fever, and irritability.
- Features of AHT seen on physical examination include irritability, lethargy, soft tissue swelling of the scalp, full fontanelle, opisthotonic posturing, or coma.
- The hallmark feature of AHT is subdural hemorrhage.
- Injuries associated with AHT include retinal hemorrhages, skeletal injuries, cutaneous injuries, and visceral injuries.
 - 80% of children with AHT have retinal hemorrhages.
 - May be unilateral or bilateral.

- Dilated, indirect ophthalmoscopy performed by an ophthalmologist is preferred in the evaluation of suspected head injury to identify and document the extent of retinal involvement.
- All victims with AHT require a skeletal survey to evaluate for further injuries.
- Extracranial abnormalities are detected in 30% to 70% of abused children with AHT

DIFFERENTIAL DIAGNOSIS OF CHILD ABUSE

- The differential diagnosis of child abuse can be reviewed in (Table 145-3).
- Dermatologic findings that can be mistaken for bruises including “mongolian” spots, cultural practices such as coining, phytodermatitis, and connective tissue diseases such as Ehlers–Danlos syndrome.
- Conditions such as bullous impetigo, epidermolysis bullosa, and folk treatments may be confused with burns.
- Conditions that may mimic inflicted neurotrauma include accidental or birth-related trauma, hemorrhagic disease of the newborn, vascular malformations, and glutaric aciduria type I.
- With excessive or unusual bruising consider isoimmune thrombocytopenic purpura (ITP), hemophilia, infection such as meningococemia, Henoch–Schonlein purpura (HSP), and vitamin K deficiency.

ANCILLARY STUDIES

- Laboratory or radiographic testing is guided by the age of the child, the injury pattern, the clinical condition of the child, and the consideration of differential diagnosis.
 - Coagulopathy screen is indicated for children who present with isolated bruising.
 - Abdominal enzymes, including liver function enzymes, amylase, and lipase, are indicated for children with suspected abdominal trauma.
 - For injury patterns that are pathognomonic for inflicted injury, no search for an alternative diagnosis is warranted.
- The skeletal survey is indicated for all children younger than 2 years with any suspicious injury.
 - Skeletal surveys are generally not indicated for children older than 5 years of age since older children rarely have occult fractures.
 - Patients between the ages of 2 and 5 years, there should be a high index of suspicion for abuse in order to justify a skeletal survey.
- Additional radiographic studies when indicated include radionuclide bone scans, ultrasound, CT scans, and MRI scans.

- Neuroimaging may be positive in up to 30% of infants with physical injuries.

NEGLECT

- Neglect is the inattention or omission on the part of the care-giver to provide for the needs of a child.
- Most common type of child maltreatment.

TABLE 145-3 Differential Diagnosis of Injuries Associated with Child Abuse

Bruises

1. Accidental injury
2. Dermatologic disorders
 - Mongolian spots
 - Erythema multiforme
 - Phytodermatitis
3. Hematologic disorders
 - Idiopathic thrombocytopenic purpura (ITP)
 - Leukemia
 - Hemophilia
 - Vitamin K deficiency
 - Disseminated intravascular coagulopathy (DIC)
4. Cultural practices
 - Cao gio (coining)
 - multiforme
 - Quat sha (spoon rubbing)
5. Genetic diseases
 - Ehlers–Danlos
 - Familial dysautonomia (with congenital indifference to pain)
6. Vasculitis
 - Henoch–Schonlein purpura

Burns

1. Accidental burns
2. Infection
 - Staphylococcal scalded skin syndrome
 - Impetigo
3. Dermatologic
 - Phytodermatitis
 - Stevens–Johnson reaction
 - Fixed drug eruption
 - Epidermolysis bullosa
 - Severe diaper dermatitis
4. Cultural practices
 - Cupping

Fractures

1. Accidental injury
2. Birth trauma
3. Metabolic bone disease
 - Osteogenesis imperfecta
 - Copper deficiency
 - Rickets
4. Infection
 - Congenital syphilis
 - Osteomyelitis

Head Trauma

1. Accidental head injury
2. Hematologic disorders
 - Vitamin K deficiency (hemorrhagic disease of the newborn)
 - Hemophilia
3. Intracranial vascular abnormalities
4. Infection
5. Metabolic diseases
 - Glutaric aciduria type I

- Core needs include access to health care, appropriate shelter, proper nutrition, education, and emotional support.
- Neglect also includes failure to properly supervise and protect children from harm.
 - Refers to omissions that are within the parent's control.
- Failure to thrive (FTT), medical neglect, drug-exposed newborns, and child abandonment are all examples of neglect.
- Medical neglect can range from a caretaker who refuses, denies, or fails to provide prescribed treatment for serious acute illness to the caretaker who fails to seek basic medical care for the child.
 - An example is the child whose care provider is non-compliant with medications and medical follow-up for a readily treatable disease, which results in an increase in the severity of the disease that requires escalating medical care.
- Supervisional neglect includes child abandonment and lack of appropriate supervision.
- The physician must serve as an advocate for the safety and well-being of the child.

MANAGEMENT

- Child protection laws mandate all professionals to notify their local child protection agency when there is a suspicion on child abuse or neglect.
 - “Suspicion” is defined as having a reasonable cause/concern that a child may be or has been harmed or neglected.
 - Failure to report suspected abuse can result in criminal charges and loss of medical licensure.
 - Medical reporters are typically protected from retaliation when their report to child welfare agencies is made in good faith.
- Maintain communication with the family or care provider even when there is a concern of abuse or neglect.
- Maintain objectivity and refrain from confrontational or accusatory statements when talking with the family.
- Specific discussions regarding mechanisms of injury and timing of injury should be avoided until law enforcement and child welfare have met with the family.
- Investigators may request written medical opinions or interviews with the medical care provider to assist with their investigation.
- A physician may be asked to testify in both family and civil court proceedings
 - The family court proceeding help determine who has custody of the child.

- The criminal court proceeding deals with the criminal prosecution of cases.
- In either case, the emergency physician may be requested to formulate an opinion within a reasonable degree of medical certainty if a child has been abused.
- When available, the emergency physician should consult with a child abuse pediatrics specialist to aid in this assessment.

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QUESTIONS

1. A 3-year-old female child presents with bruising suspicious for abuse. She is the product of a term delivery but has cerebral palsy and developmental delays. Both parents work. Which of the following is a risk factor for abuse in this patient?
 - A. Age >1 year
 - B. Female gender
 - C. Physical disabilities
 - D. Term gestation
 - E. Working parents

2. You are giving a lecture to first year emergency medicine residents about child abuse. You try to provide case scenarios that should prompt a higher level of suspicion. Which of the following cutaneous injuries would be most consistent with abuse?
 - A. A two year old with scald burns to the chest from spilled coffee.
 - B. A 14-month old with bruises on the anterior surfaces of both legs.
 - C. A 2-year old with a forehead hematoma and small laceration from a fall.
 - D. A 4-month old with linear bruises to both legs.
 - E. A 2-year old recently arrived from Southeast Asia who presents with fever and linear ecchymoses on back.
3. In addition to describing historical and physical indicators to help differentiate inflicted from non-inflicted injuries, you discuss the legal obligations to report suspected abuse and neglect. Which of the following case scenarios would most likely need to be reported to child protective services (CPS)?
 - A. A 2-month old with bleeding from the mouth secondary to a frenulum tear.
 - B. A 3-year old with a clavicle fracture after a fall from a slide.
 - C. A 5-month old with a linear skull fracture after a fall from mother's arms.
 - D. A 6-month old with type 1 glutaric academia and retinal hemorrhages.
 - E. A 2-year old with a buckle fracture of the distal radius and no history of injury.
4. A 2-year old child presents with pattern bruises over the chest, abdomen, back, and buttocks consistent with being beaten with a cord loop. A skeletal survey is performed and there are multiple fractures of the ribs and long bones in different stages of healing. You arrange to have the patient transferred to a pediatric specialty hospital for further evaluation. Before transfer, you must
 - A. Report the case to CPS only if your state requires reporting.
 - B. Report the case to CPS immediately and notify them of the transfer to the pediatric center.
 - C. Hold any reporting until a comprehensive social services evaluation can be completed at the receiving hospital.
 - D. Contact CPS only if there are other children at risk, since this patient is being admitted and is in no danger.
 - E. Only report to CPS if you are certain that there is no fear of retribution.
5. A 5-year old child with asthma on multiple maintenance and rescue drugs is being seen in the ED for an acute asthma exacerbation. Mother states that the patient "ran out of medications" and has had no treatment for 3 days. Review of history shows three admissions for status asthmaticus in the past year, and six additional ED visits for treatment of asthma. An appropriate management plan would include
 - A. Immediate removal of the patient from the caretakers due to medical neglect
 - B. Discharge home with follow up if medications can be obtained, and acute asthma is cleared.
 - C. Hospitalize the patient until the social situation can be resolved.
 - D. If acute exacerbation is resolved and medication can be obtained, report to CPS for family home follow up due to medical neglect.
 - E. You have no responsibility for a difficult social situation.

ANSWERS

1. C. Risk factors in patients for abuse include young age (<1 year), male gender, prematurity, chronic illness and congenital anomalies. Our patient had cerebral palsy and developmental delays. Parental risk factors include poverty, depression, single marital status, depression, substance abuse, and lack of education.
2. D. A nonambulatory patient with any bruising, especially under 6 months of age should always make one suspicious of abuse. Bruising on the anterior surfaces of the legs in toddlers is common and expected. Likewise, a forehead hematoma and/or laceration from a fall in a toddler are common. Linear ecchymoses on a recent immigrant from Southeast Asia is likely from "coining" a common folk practice for treatment of fever. Splash burns from scalds are common and rarely abuse. Pattern burns should make one suspicious of abuse.
3. A. Tears of the frenulum are highly suspicious of abuse in nonambulatory infants, and usually results from a bottle being forced into the infant's mouth. Clavicle fractures and distal radial fractures are common in ambulatory children and are rarely a result of abuse. A linear skull fracture in a 5-month old does warrant a careful history and examination, but could be consistent with a fall from mothers arms onto a hard surface. Glutaric academia type 1 is a rare metabolic disorder that often has retinal hemorrhages not from abuse.
4. B. Child protection laws mandate all professions in all 50 states to notify local CPS agencies whenever there

is reasonable concern that a child has been abused and/or neglected. All medical professionals are mandated to assure reporting is done, and failure to report can result in criminal charges and loss of medical license. Even if the patient is no longer at risk, or is being transferred to another facility, reporting still must be done.

5. D. This is an example of the most common form of medical neglect; a caretaker who fails to provide prescribed treatment for a chronic serious illness. This should be reported to CPS. It may not be necessary to remove the patient from the home, but reporting is necessary so follow-up support and education can be arranged for the caretaker.

146 PSYCHIATRIC EMERGENCIES

Catherine P. Moore

INTRODUCTION

- The first priority in evaluating and treating psychiatric patients in the emergency department (ED) is to determine the risk the patients pose to themselves and others. Safe rooms that have no equipment and are highly visible to staff are optimal for psychiatric patients. Some patients may need one-on-one supervision by staff; others may need physical or chemical restraint.
- Examination of the psychiatric patient includes a full physical examination as well as complete neuro and mental status examination. When performing a mental status examination, particular attention should be paid to patient and caretakers. Assessment of the mental status of the caretaker can reveal much about the parent–child relationship and function.

THE SUICIDAL PATIENT

ETIOLOGY/PATHOGENESIS

- Most suicidal patients have psychiatric pathology.
 - Most common being major depressive disorder (MDD)/dysthymia.
 - Disruptive behavior disorder.
 - Drug/alcohol abuse and dependence.
 - Anxiety disorder.

RECOGNITION

- Chief complaint of a suicide attempt.
- It is important to assess risk of suicide while in the ED:

- Are you here because you tried to hurt yourself?
- In the past week, have you been having thoughts about killing yourself?
- Have you ever tried to hurt yourself in the past *other than this time*?
- Has something very stressful happened to you in the past few weeks?
- The more “yes” answers, the greater the risk of the patient posing a threat to themselves.

MANAGEMENT

- The interviewer should ascertain.
 - Reason the patients desire to hurt themselves.
 - Plan to do so.
 - Whether the patient has the means necessary to carry out the plan.
 - Timing of the plan’s execution.
- Admission is warranted if there are any medical issues or the patient is not deemed safe for discharge.
- Concerns for patient safety include:
 - Inability to maintain a safety (no-suicide) contract.
 - Active suicidal ideation (plan and intent).
 - High intent or lethality of attempt.
 - Psychosis.
 - Volatile/unsafe family and home environment.
- Treatment of the patient with suicidal tendency may include both medical therapy and counseling.
- Multiple studies suggest that combining medical therapy, antidepressants such as a selective serotonin reuptake inhibitor (SSRI), and cognitive behavioral therapy have the highest success rates.
- In 2004, the Food and Drug Administration (FDA) issued a black box warning for all SSRIs, warning that “antidepressants increased the risk compared to placebo of suicidal thinking and behavior (suicidality) in children, adolescents, and young adults in short-term studies of major depressive disorder (MDD) and other psychiatric disorders.”
- Bridge and colleagues published a meta-analysis that showed antidepressant use in children, adolescents, and young adults is of greater benefit than harm.
 - This study suggested close follow-up and evaluation on a case-by-case basis and that the patient on SSRIs is not at a greater risk in the ED than the one who is not on medications.

ANCILLARY STUDIES

- If the patient’s mental status is altered, a toxicology screen and ethanol level is warranted.
- Many intoxicated suicidal patients are no longer suicidal once the intoxicant wears off.

- Organic disease must be ruled out.
 - Thyroid disease, autoimmune disease, and other organic disease states can cause or augment a patient's depression and tendency to commit suicide.

THE PSYCHOTIC PATIENT

ETIOLOGY/PATHOGENESIS

- Pediatric schizophrenia can present with psychosis at or before 12 years of age.
- DSM-IV-TR criteria include hallucinations (79–82%), delusions (54–63%), and thought disorder (40–100%).
- Disorganized behavior, negative symptoms, and impaired functioning.
- Symptoms of later onset disease are not particularly diagnostic in children.
- Psychosis can be a feature of many other psychiatric diagnoses including MDD, bipolar disorder, and schiziform disorder.
- Most pediatric patients present with a chief complaint of auditory and/or visual hallucinations.
- Etiology can be basic psychiatric illness, but again it is essential to rule out organic disease such as infection, rheumatic disease (especially lupus), cerebral blood flow changes/hypoxia, temporal lobe epilepsy, toxicological, vitamin deficiencies, metabolic and endocrine disorders, Reye syndrome, Wilson's disease, and encephalopathy.

MANAGEMENT

- Mainly pharmacological: olanzapine, risperidone, and haloperidol have been used in pediatric ED settings.

ANCILLARY STUDIES

- Workup for the patient with psychosis/altered mental status should include toxicology, electrolytes, kidney and liver functions, and ammonia level.
- Imaging to rule out organic causes including CT scan, MRI, and EEG may be necessary.
- A lumbar puncture may be warranted to evaluate for meningitis and encephalitis.

POSTTRAUMATIC STRESS DISORDER (PTSD)

ETIOLOGY/PATHOGENESIS

- Posttraumatic stress disorder (PTSD) emerges after traumatic events that evoke intense fear, helplessness, or horror.

- The hypothalamic–pituitary–adrenal axis has been noted to be hyperactive in children and adolescents diagnosed with PTSD.
- Overall, these patients have elevated cortisol levels in urine and saliva.

RECOGNITION

- PTSD should be recognized by its symptoms of intrusive memories and/or dreams of the traumatizing event, avoidance of stimuli connected to the traumatic event, symptoms of excessive arousal/anxiety including inability to concentrate on tasks, exaggerated startle response, hypervigilance, insomnia, and inappropriate outbursts.
- Any trauma that warrants emergency evaluation can provoke a case of PTSD.
- PTSD is distinguished from acute stress disorder by duration of symptoms being 4 weeks or longer.
- Children with burn injuries have over a 50% chance of having symptoms of PTSD.

MANAGEMENT

- Treatment with counseling and pharmacotherapy should be determined on a case-by-case basis.
- There is no laboratory or radiographic test to confirm or deny the existence of PTSD.

THE PSYCHOPATHIC PATIENT: WHEN JUNIOR KILLS FIDO

- *American Heritage Dictionary* defines psychopath as “a person with an antisocial personality disorder (APD), manifested in aggressive, perverted, criminal, or amoral behavior without empathy or remorse.”
- APD can only be diagnosed at 18 years of age or older. Children and adolescents often are diagnosed with oppositional defiant disorder (ODD) and conduct disorder (CD) prior to their diagnosis of APD.
- ODD is characterized by a pattern of negativity, hostility, and defiance lasting for at least 6 months during which four DSMIV criteria are met.
- Diagnosis of CD is based on persistent history of a child who seriously misbehaves with aggressive or nonaggressive behaviors against people, animals or property that may be characterized as belligerent, destructive, threatening, physically cruel, deceitful,

disobedient, or dishonest. This may include stealing, intentional injury, and forced sexual activity.

ETIOLOGY/PATHOGENESIS

- It has been suggested that APD, CD, and ODD originate in poor bonding with the mother in the first five years of life.
- These patients have deficits in fear recognition similar to patients with amygdala damage.

CONVERSION/SOMATIZATION DISORDER

ETIOLOGY/PATHOGENESIS

- Thought to be triggered by psychosocial stressors. It is not understood how the stressor turns into somatic complaints.
- These complaints are not intentional, unlike factitious disorder and malingering.
- In one study, it was noted that boys with somatoform disorders had significantly poorer interpersonal relations and communications, whereas girls had higher rates of conflicts with family members.

RECOGNITION

- Once the possibility of organic disease has been excluded, psychiatric evaluation can make the diagnosis.
- In paradoxical vocal cord dysfunction, the patient presents in refractory respiratory distress. Direct visualization of abnormal vocal cord movement (closing of the cords on inhalation) is the definite means of establishing the diagnosis.

TREATMENT/MANAGEMENT

- After organic disease is ruled out, patients require psychotherapy and often medical management (SSRIs) to help cope with the underlying stressor(s).

THE AGGRESSIVE PATIENT

- Look for the etiology of the patient's aggression/discontent. If it can be easily removed from the patient's environment, it should be.
- Need to be well supervised and placed where they do not have the means to harm themselves or others.
- If verbal reassurance is ineffective in diffusing the aggression, escalation in care proceeds to seclusion

and therapeutic holding. Therapeutic holding refers to at least two people physically restraining the patient.

- Recommendations of the American Academy of Pediatrics are
 - Explain the necessity of restraint to the patient.
 - Have specific physician orders including indication for and duration of restraint.
 - Explain everything to the family.
 - Perform and document ongoing assessment of correct application of restraints, skin and neurovascular integrity, as well as efficacy of the restraints in meeting the indication for application.
- If care requires escalation from physical restraint/therapeutic holding, chemical restraint may be warranted, with goals of
 - Decreasing the patient's anxiety and discomfort.
 - Minimizing disruptive behavior.
 - Preventing escalation of behavior.
 - Reversing the underlying cause.
- All chemical restraint use requires careful monitoring of the patient on cardiac apnea monitor and pulse oximetry.

CHEMICAL RESTRAINT AGENTS IN THE AGGRESSIVE PATIENT

- Several of the most common chemical restraints are detailed in Table 146-1.
- Lorazepam, midazolam, and diazepam are benzodiazepines that work to sedate patients by activating GABA receptors and can, therefore, be helpful in aggressive behavior modification. The major side effect is respiratory depression.
- Neuroleptic drugs, such as haloperidol, have been utilized extensively in treating the aggressive patient. The value of these medications in the acute care setting is due to their sedating effects rather than their antipsychotic effect.
- The rare incidence (1%) of extrapyramidal symptoms (EPS) can occur after one dose of neuroleptic drugs.
 - Most commonly seen EPS is dystonic reaction involving eyes, neck, and/or back.
 - Rarely does EPS affect the airway.
 - Treatment for EPS is diphenhydramine (IV or IM) and/or benzotropine (IV or IM).
- One also must be aware of potentially fatal neuroleptic malignant syndrome characterized by fever, sweating, hypertension, severe muscle rigidity, and delirium sometimes progressing to coma.
 - Treatment of neuroleptic malignant syndrome is with dantrolene and supportive care.

TABLE 146-1 Medication Options for Acute Agitation

GENERIC NAME	BRAND NAME(S)	DRUG CLASS	DELIVERY	ONSET OF ACTION	MAJOR/COMMON SIDE EFFECTS
Lorazepam	Ativan	Benzodiazapine	IV/IM/po	5–10 min IV 20–40 min IM 20–30 min po	Respiratory depression
Haloperidol	Haldol	Typical antipsychotic	IM/po	5–10 min	EPS, Qtc ↑, NMS, hypotension
Chlorpromazine	Thorazine	Typical antipsychotic	IV/IM/po	15 min IM 30–60 min po	Altered cardiac conduction, esophageal dysmotility, EPS, NMS, hypotension
Droperidol	Inapsine	Typical antipsychotic	IV/IM	3–10 min	EPS, Qtc ↑, NMS, hypotension
Olanzapine	Zyprexa, Zydis	Atypical antipsychotic	IM/ODT/po	15–30 min IM 65 hours po	Bradycardia, few EPS, prolactin elevation, anticholinergic SE, orthostatic hypotension
Ziprastine	Geodon	Atypical antipsychotic	IM/po	30–43 min IM	Nausea/HA/dizziness few EPS, Qtc ↑, orthostatic hypotension, prolactin elevation
Hydroxyzine	Vistaril Atarax	Antihistamine (H1 receptor antagonist)	IM/po	15–30 min	Hypotension, SVT, hallucination, seizure, tremor

- Atypical psychotics, such as ziprasidone and olanzapine, are gaining favor in the acute management of aggressive patients.
 - With a lower incidence of EPS, multiple routes of administration, and better tolerance, they are commonly used in the pediatric ED.
 - Ziprasidone and olanzapine are recommended for the agitated schizophrenic patients.
 - Olanzapine is also indicated for aggression/agitation associated with bipolar disorder.
- Hydroxyzine is an antihistamine that has been used as an anxiolytic, can be administered IM, and has an onset of action comparable to that of lorazepam.

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QUESTIONS

1. The clinical psychologist on call is evaluating a 14-year-old boy with suicidal ideation in the ED’s “safe room.” You observe the patient becoming aggressive and yells at the psychologist. Your first intervention is
 - A. Ask psychologist to leave room
 - B. Call security
 - C. Get help for therapeutic holding
 - D. Order haloperidol IM
 - E. Talk to patient

2. Once you call for help, talk to the patient and excuse the psychologist the patient settles down, but only temporarily. When the nurse asks the patient if he would like some water the patient spits on him and attempts to bite him. Your next action is to
 - A. Call security
 - B. Execute therapeutic holding
 - C. Order IM ativan to bedside
 - D. Order IM haldol to bedside
 - E. B and C
3. A 5-year-old female involved in a high-speed motor vehicle collision in which her mother and brother were killed. She has burns over 25% of her body. She was hospitalized for 2 weeks including a 6-day ICU course. One-week post discharge she presents to the ED with inappropriate outbursts at school. Her aunt who is her full time caretaker reports that she has been suffering with anxiety since she was discharged from the hospital. The most appropriate diagnosis is
 - A. Acute stress disorder
 - B. Anxiety disorder
 - C. Attention deficit hyperactivity disorder
 - D. Major depressive disorder
 - E. Post-traumatic stress disorder
4. The best course of action for this patient is
 - A. Physical and mental status exam
 - B. Psychiatric referral
 - C. Prescribe a benzodiazepine
 - D. Prescribe a SSRI
 - E. Screen for early predictors of PTSD
5. A 15-year-old girl presents in mild respiratory distress that began 2 hours ago. Her medical history is significant for schizophrenia that was diagnosed 2 years prior and she was started on haldol 2 days ago. She is afebrile, her heart rate is 127, respiratory rate is 26, O₂ sats 94% on room air, and her blood pressure is 121/73. Physical exam reveals wheezing throughout her chest. Your first intervention is
 - A. Albuterol
 - B. Benadryl
 - C. Epinephrine
 - D. Steroids
 - E. Oxygen
6. Several rounds of beta agonist later, you get a chest x-ray which is normal but the patient is still appears anxious. You have given steroids. On her repeat examination, the wheezing has resolved. Your colleague suggests you try a benzodiazepine. The mechanism of action of a benzodiazepine in this patient
 - A. Combat EPS associated with haloperidol use.
 - B. Increase the density of type 2 pneumocytes in the aveoli.
 - C. Relax the patient's airway.
 - D. Treat anxiety associated with bronchospasm.
 - E. Treat seizure.
7. A 16-year-old boy presents with acute onset of psychosis in which he complains, "Bugs are crawling all over me." He is diaphoretic, tachycardic, and tachypenic and appears apprehensive when you go to approach him. You order an IM dose of haloperidol, which works beautifully to calm this patient. Neuroleptic drugs are utilized in aggressive patients because they
 - A. Intoxicate the patient acting at GABA receptors.
 - B. Decrease the hallucinations experienced by the patient through antidopenergic effects.
 - C. Decrease sensory irritation by antihistamine quality.
 - D. Produce alpha-adrenergic mediated anxiolysis.
 - E. Sedate by its action on the limbic system.
8. As the patient arouses, he complains of neck pain and has some abnormal posturing on movement. Your most appropriate intervention is
 - A. Diphenhydramine
 - B. Lorazepam
 - C. Prednisolone
 - D. Psychiatric consult
 - E. Repeat haloperidol dose
9. An 8-year-old boy presents to the ED after being found by his mother trying to hang himself. He has some marks around his neck, but otherwise his physical exam is benign. After careful evaluation you decide the patient requires admission. The most important reason this patient requires admission is
 - A. Age
 - B. Lethality of the attempt
 - C. Safety concerns
 - D. Psychosis
 - E. Flat affect of mother.

ANSWERS

1. E. Talk to the patient. If it is the psychologist that is insighting the aggression, he should be removed. However, before one attempts to make an intervention with an aggressive patient it is essential to have backup, so getting help should be done *before* the intervention. Chemical restraints, such as haloperidol are appropriate after failure to talk to the patient and therapeutic holding.
2. E. while you are executing therapeutic holding is always the best idea. In this instance. It is important to plan ahead with the aggressive patient, and having a chemical restraint en route ativan is the more widely popular choice of pharmacological agent (likely due to

the smaller side effect profile), but remember that olanzapine would also be an appropriate choice for this child if it were given that he suffered with bipolar disease. In addition, when use of a chemical restraint is being entertained, it is important to explain the action to the patient and family as well as to appropriately document an assessment of the patient.

3. A. The diagnosis of acute stress disorder is one based on the time from the incident. Although the symptoms described fit a classical scenario of PTSD, the time since the accident has been only 3 weeks that is not enough time to make a diagnosis of PTSD as it takes at least 1 month from the time of the inciting incident.
4. A. Physical and mental status exam. This patient had extensive injuries as a result of the original crash and her behavior must be ruled inorganic before psychiatric treatment can commence.
5. A. Albuterol is a beta-agonist used to treat bronchospasm, which seems evident from the vignette.
6. D. Manage the anxiety associated with bronchospasm. EPS rarely presents with airway complications, most often will present with dystonia involving eyes neck and back.
7. E. Neuroleptics are antidopaminergic agents, and that is how they treat psychosis. However, these effects take 7–10 days of treatment. In the acute setting, it is their sedating action that is of use in treating the aggressive patient.
8. A. This is likely a dystonic reaction and diphenhydramine is the rescue drug of choice. A benztropine would also be appropriate.
9. C. Although all of the above are concerning, the patient's safety is your utmost concern. If there is any reason the patient is not safe to go home admission to the hospital is warranted.

147 DEATH OF A CHILD IN THE EMERGENCY DEPARTMENT

William R. Ahrens

- Informing family members and friends of a loved one's death is a fact of life for the emergency physician. Such deaths are often sudden and unexpected, and survivors are confronted with the loss of a loved one with no prior psychological preparation. The situation is particularly difficult when the dead patient is a child. The interaction between the emergency physician and the Emergency department (ED) staff

and the dead child's family can be an important first step in recovery.

- The majority of emergency physicians feel that managing the death of a child is far more difficult than managing the death of an adult; some consider it the most difficult aspect of their job. Many feel guilty or inadequate after a failed pediatric resuscitation. Many feel impaired for the remainder of their shift. Few have had any formal training in how to tell parents that their child is dead.
- The immediate reaction of family members to the sudden loss of a child is disbelief, even though many say that they knew before being told that their child had died. On the part of parents, a sense of failure or guilt is probably universal.
- Many describe the experience in the ED as one that is replayed in their minds "like a tape," thousands of times. They can often recall minute details of their experience and can remember verbatim exactly what they were told, and by whom.

THE INTERVIEW

- One of the most common complaints of families whose loved one died in an ED is that they were not kept informed of the progress of events. Given the reality that the vast majority of pediatric patients who arrive pulseless and apneic will die in the ED, the process of dealing with the patient's death should be considered part of the resuscitation; the family is the patient. Parents should be placed in a private, quiet room, with adequate seating. A staff member is designated to communicate with the family; ideally this is an individual who is experienced in delivering bad news.
- There are some families who will want to be present during the resuscitation. The decision to allow or invite family members into the resuscitation room depends largely on the comfort level of the ED staff. Recent publications have suggested that family members be at a minimum offered the opportunity to be present during the resuscitation.
- It is the responsibility of the attending physician to tell the parents that their child is dead. If possible, the physician should be seated during the interview. The language used should be direct and nonjudgmental; it cannot be overemphasized that the dead child should be referred to by his or her name. Euphemisms for death are to be avoided; phrases like "the little guy did not make it," or "the baby expired" can be deeply resented by parents, who perceive them as depersonalizing their child. "I am very sorry, but Brendon is dead," or "I am sorry to have to tell you that Becky has died," are two examples of acceptable terminology.

- Parents want to know why their child died, and that the staff did everything that could be done to save the child's life. The physician must reassure the family that they are not responsible for their child's death.
- It is important that parents believe that the emergency physician and staff experience sorrow for the loss of the child.
- If an interpreter is involved in the interview, they should stand behind the physician so that the physician can maintain eye contact with the parents during the discussion.

AFTER THE INTERVIEW

- Family members should be offered an opportunity to spend time with the dead child after the interview. Before this is done, resuscitation equipment is removed, the body is cleaned, and preferably wrapped in a blanket. Most but not all parents find that spending time with the child is helpful; some will want to hold the body, others will not. A reluctance to do so does not signify child abuse. Parents should be allowed to stay until they are ready to leave.
- The vast majority of family members who have a child die in an ED would like a physical memento of the patient. These include a lock of hair and/or an ink-print or plaster mold of a hand or foot. Clothing and other personal items should be returned. Mementos are concrete objects that allow survivors to maintain a sense of contact with their dead loved one. Unless one has lost a child, it is probably difficult to imagine how important they are.
- Because most pediatric arrests involve prolonged tissue asphyxia, organ donation is usually not possible. However, many patients are eligible to donate heart valves, skin, and corneas. While approaching family members about tissue donation immediately after the death of their child is extremely difficult, there are at least some family members who retrospectively wish they had been asked.
- Most cases of sudden death require an autopsy. It is important that this process be explained to family members. They must be told how to obtain the results. In some cases the family's personal physician may be available to discuss the findings; in many cases the local coroner will do so. In cases of suspected sudden infant death syndrome (SIDS), parents should not be told they will feel better if indeed the autopsy confirms SIDS as the cause of death; in many cases this is not true.
- It is important to realize that in situations where the cause of death of a previously healthy baby is unknown, as is the case with babies who die of SIDS, the local police will investigate the family's home as a potential crime scene.
- The long-term effects of the death of a child on family members are not well known and are difficult to study. Spouses, grandparents, and surviving siblings will each experience the loss of the child in their own way. For children, the understanding of death largely depends on their developmental stage; young children in particular may have no concept of death, or may not perceive it as permanent. Older children develop an understanding that death is permanent, but can perceive it as evil or an entity that punishes people. Parents will of course feel profound grief that is most intense during the first year after the loss of the child.
- Mothers may have a higher risk of pathologic grief responses than fathers. There may be an increase in psychiatric hospitalizations in parents who have lost a child. It is often assumed that divorce is more common among spouses who have lost a child; there is no evidence to support this assumption, which is bitterly resented by many bereaved parents.
- A child's death can shatter the relationship of people with the everyday world, as well as their concepts of spirituality and their relationship with religion. An important aspect of recovery involves finding meaning and value in the dead child's life, however short it may have been. One mother is quoted, "I wanted her gown because it was the last thing she wore. I wanted the sheet from the bed. I wanted her bracelet from the hospital." They said they could not give us anything. Sensing compassion on the part of the medical staff who attended the patient and family is important; parents want to know that their child was valued as an individual.
- Offering parents the opportunity to return to the hospital and discuss the child's death may in some cases be helpful.
- Bereaved parents will receive support from other family members, friends, and in many cases, clergy. There are also organized grief-support groups that have extensive experience and profound commitment to help parents cope with the death of a child. Parents should be made aware of their existence (see Table 147-1).

TABLE 147-1 Grief-Support Groups

Sudden Infant Death Syndrome Alliance: www.sidsalliance.org (800-221-7437)
The Compassionate Friends: www.compassionatefriends.org (877-969-0010)
Survivors of Suicide: www.survivorsof suicide.com
National Organization of Parents of Murdered Children: www.pomc.com (513-721-5683)
Share Pregnancy and Infant Loss: www.nationalshareoffice.com (800-821-6819)

MISCARRIAGE IN THE EMERGENCY DEPARTMENT

- Miscarriage, or spontaneous abortion, is defined as the loss of pregnancy up to the 20th week of gestation. Between 10% and 20% of pregnancies will end in miscarriage. How many patients are diagnosed as having miscarried in an ED is unknown. Ironically, there is a large body of literature evaluating the effect of miscarriage on both mothers and fathers compared to that evaluating the effect of the loss of a born child. It is clear that miscarriage has profound psychological effects on both parents. Many people suffer from depression and anxiety after a miscarriage. Relationships between spouses are affected, as well as attitudes toward future pregnancies.
- Many people feel that the medical staff trivialized their loss; for most parents, the miscarriage was the loss of a baby. For some people, follow-up intervention can be helpful.
- While the death of a child is clearly different from a miscarriage, it is important for the emergency physician and the ED staff to acknowledge to patients who miscarry that a baby has been lost.

CARING FOR THE ED STAFF

- Managing the death of a child in the ED will always be difficult and will affect all staff members. At least some emergency physicians and presumably other staff use counseling services after suffering traumatic events in the department, but they are by no means universally available.
- While critical incident stress debriefing is commonly used, there is limited data documenting its efficacy. It is likely that educating physicians and other staff members in the best way to communicate the death of a child or other bad news can benefit both staff and patients. At the very least, it creates a mutually supportive culture within the department regarding the management of death. There are multiple modalities that have been successfully used, including role-playing, hearing from parents whose child was pronounced dead in an ED, and incorporating “death-telling” into advanced life support courses.
- Some comments and suggestions from parents whose child died in an ED are as follows
 - Just let the parents know you are sorry and that everything possible was done.
 - Unless it has happened to you, do not say “I know how you feel,” because you do not.

- Just be honest; say everything possible was done to save the child.
- Look the parents in the eyes. Explain that all attempts at resuscitation were performed.
- Tell them how sorry you are, that life is not fair, and most importantly that it was not their fault.
- Allow your humanity to show. Our physician was very compassionate and he had tears in his eyes.
- Make sure you express how sorry you are and tell the loved ones how it affects the staff.
- It is okay to show emotion; do not be overly clinical.
- The doctor should say, “I am sorry, we did all we could.”
- As you well know, once your child is dead, you can never have enough mementos.

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QUESTIONS

1. An unresponsive 2-month old is brought in by EMS. The infant was full term, vaginal delivery with no complications. You suspect SIDS. From the emergency physician’s point of view managing the death of a child
 - A. Is rarely problematic.
 - B. Is something they are well prepared to do.
 - C. Can cause feelings of guilt or inadequacy.
 - D. Does not affect their job performance.
2. A 4-year old with severe cerebral palsy and congenital heart abnormalities is brought to the ED for decreased responsiveness. Over the last several

weeks, the child has been deteriorating and was made DNR 3 days ago. Shortly after arrival to the ED the child dies. Which of the following statement is correct regarding holding their dead child in the ED?

- A. Is something all parents find helpful.
 - B. Is something all parents want to do.
 - C. Is normal for parents and failure to want to do so should raise the possibility of child abuse.
 - D. Is something all parents should be allowed the opportunity to do.
3. A 3-year-old girl is brought by EMS following being struck by a automobile while playing. Despite resuscitation in the ED, the patient expires. When telling parents their child has died
- A. Euphemisms such as “the little guy did not make it” soften the blow.
 - B. Using straightforward, direct language is important.

- C. Referring to the dead child by name is not important.
- D. Eye contact can be threatening to parents.

ANSWERS

1. C. Most emergency physicians are not well prepared to manage the death of a child. Most feel guilt or inadequacy after a failed pediatric resuscitation, and many at least feel impaired for the remainder of their shift.
2. D. Although most parents will want to hold their dead child, some do not, and some do not find doing so helpful. A reluctance to hold the child does not signify that the child was abused. All parents should be allowed the opportunity to hold the child.
3. B. Making eye contact and using direct, straightforward language is important, as is referring to the child by name. Euphemisms are to be avoided.

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Section 22

EMERGENCY MEDICAL SERVICES AND MASS CASUALTY INCIDENTS

148

PEDIATRIC PREHOSPITAL CARE

*Craig J. Huang
Maeve Sheehan*

STATE OF PREHOSPITAL PEDIATRIC EMERGENCY CARE IN THE UNITED STATES

- Regional EMS systems and training courses for emergency care providers started in the 1960s.
- The Emergency Medical Services Act of 1973 provided funding and specific guidance on establishing EMS systems and personnel.
- The Emergency Medical Services for Children (EMSC) program is the only federal program whose purpose is to improve the quality of pediatric emergency care.
- In 2007, the Pediatric Emergency Care Subcommittee of the IOM analyzed the challenges of providing integrated emergency services to pediatric patients, specifically examining prehospital EMS services and systems.
- Several resource centers, grants, national and federal partnerships, and allied organizations help integrate EMSC into regional and state EMS systems.

ANTICIPATORY GUIDANCE AND OFFICE PREPAREDNESS

- Out-of-hospital cardiac arrest is common and family members usually do not initiate basic CPR.
- Prehospital care begins in the pediatrician/primary care provider's office and is the first link in "the chain of survival" in the emergency care system.

- Caregivers and triage personnel should know when to call 911.
- Office preparedness, in the form of personnel training and available equipment, is dependent on the usual response time for EMS systems.
- Primary care providers should routinely provide caregivers with their child's problem list, vaccinations, required medications, and allergies especially for children with special health care needs.

PREHOSPITAL PROFESSIONALS

- The NAEMSE, with the cooperation of NHTSA and the MCHB, have developed the National EMS Education Standards that replaces the current national standard curricula outlining the comprehensive educational guide and instructional materials for EMT basics, intermediates, and paramedics.
- TRIPP (<http://cpem.med.nyu.edu/teaching-materials>) and PEPP (www.PEPPsite.com) are supplemental educational resources for prehospital pediatric emergency medicine training that augment BLS/ALS information, offers resources for child abuse and neglect and pediatric disaster preparedness.

EQUIPMENT AND SUPPLIES/MEDICAL DIRECTION AND PROTOCOLS/CARE IN THE COMMUNITY ED

- The EMSC National Resource Center (www.child-rensnational.org/EMSC/Pubres/PediatricEquipment.aspx) lists a number of resources that can provide recommended pediatric BLS/ALS equipment and supply lists.
- EMS medical oversight is divided into direct/online medical control which is "real-time" supervision directly on-scene or remotely, and indirect/offline

medical control that usually involves delegated orders and predefined patient care protocols.

- Community EDs can appropriately stabilize and provide pediatric emergency care by identifying physician and nursing representatives to advocate for pediatric preparedness at their hospital.
- Regional pediatric centers and pediatric specific educational programs (e.g., PALS, APLS, and ENPC) are resources community EDs can use to measure their pediatric preparedness.

LEGAL ISSUES/TRANSFER CONSIDERATIONS

- Minor patients cannot refuse treatment and transport in an emergency situation.
- Legal guardians should be informed of the potential risks and asked to sign a waiver releasing the EMS service from responsibility when refusing treatment.
- Transport must occur, regardless of consent, in situations involving life-threatening emergencies and suspected abuse.
- Policies and protocols regarding DNR documents and orders should comply with state regulations and can always be rescinded based on the wishes of the legal guardian.
- ED providers should understand the pediatric capabilities of the institution where they are working in order to know when to transfer a patient and to whom.
- EMTALA mandates that the pediatric patient be stabilized to the full extent capable by the referring institution, prior to the transfer.

SPECIAL CONSIDERATIONS/DISASTER PREPAREDNESS/CHILDREN WITH SPECIAL HEALTH CARE NEEDS

- Management of pediatric traumatic injuries should prioritize the ABCDs with a focus on differences in airway anatomy, cardiovascular and respiratory physiology, and particular attention to traumatic brain and cervical spine injuries.
- The issues necessitating pediatric specific resources, equipment, and supplies are magnified when considering mass casualty incidents/disaster situations.
- Pediatric-specific considerations need to be addressed in any disaster-planning situation and be brought to the forefront of discussions by health care advocates for children.
- The AAP and ACEP have published a joint policy on emergency preparedness for Children with Special Health Care Needs (CSHCN), use of the Emergency

Information Form, and other available resources. (www.aap.org/advocacy/emergprep.htm)

- EMS personnel, as mandated reporters of child abuse in almost every state, should remember their primary responsibility is the stabilization and safe transport of those patients.
- EMS personnel must adequately protect themselves from harm when stabilizing and transporting psychotic and/or aggressive pediatric patients.
- Law enforcement officials, in addition to physical and pharmaceutical restraints may be necessary.

AREAS FOR RESEARCH/ CONTROVERSIES/ PECARN/NEDARC

- Pediatric prehospital research is important but difficult to accomplish due to multiple factors.
- A number of controversial clinical topics that require high-quality research include: advanced airway management, pain management, out-of-hospital resuscitation, and proper spinal immobilization.
- PECARN is a pediatric emergency care research network (consisting of a data coordination center, research node centers, and multiple emergency departments) whose goal is to promote and facilitate high-priority, multi-institutional, collaborative research amongst EMSC investigators.
- NEDARC is a national resource center whose mission is to help provide technical, administrative, and research support to EMS agencies and EMSC grantees.

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QUESTIONS

1. A 15-year-old boy is involved in a motor vehicle collision (MVC). He was driving his parents' automobile without permission. While at a stop light, his vehicle was rear-ended. There is no damage to either vehicle. EMS arrives on the scene. The boy has no complaints but has a 2 cm laceration to scalp. He appears visible "shakened" by the accident. Which of the following statements are true?
 - A. Since there is no damage to vehicles, the patient can legally refuse transport.
 - B. EMS is required to evaluate and transport the patient to the nearest facility.
 - C. EMS should provide local wound care and if the patient remains asymptomatic then allow the patient to sign a waiver of transport.
 - D. EMS should attempt to contact his parents.
 - E. EMS should defer management of the patient to the law enforcement officials.
2. A pediatrician has a child in his office suffering from an acute asthma exacerbation. The child does not appear to be responding to the nebulizer treatment he has been administered. What is the proper course of action for the pediatrician?
 - A. He/she should call 911.
 - B. He/she should send the patient in his parent's car to the closest emergency department.
 - C. He/she should send the patient in his parent's car to the closest children's hospital.

- D. He/she should call a regional pediatric specialty hospital and try to get their specialized pediatric transport service.
 - E. He/she should continue giving nebulizer treatments until the patient's asthma is better controlled.
3. Paramedics are called to the group home of a 14-year-old aggressive bipolar patient. The patient became upset at staff and began smashing windows. The paramedics arrive prior to local law enforcement and the scene is not secure. Which of the following statements is correct?
 - A. The paramedics should attempt to physically restrain the patient until the police arrive.
 - B. The paramedics should attempt to chemically restrain the patient until the police arrive.
 - C. The paramedics should have the caregivers sign a refusal of transfer on behalf of the patient with the option of returning once the scene is secure.
 - D. The paramedics should transport the patient once law enforcement is present.
 - E. The paramedics should take control of the scene in order to protect the patients and staff of the group home.

ANSWERS

1. B. Minor patients cannot refuse treatment and transport in an emergency situation. Since the patient has sustained a head injury and appears "shakened" by the MVC, transport must occur, regardless of consent, in situations involving life-threatening emergencies and suspected abuse.
2. A. Prehospital care begins in the pediatrician/primary care provider's office and is the first link in "the chain of survival" in the emergency care system. Caregivers and triage personnel should know when to call 911. In this case, the pediatrician is still responsible for the patient until they arrive at a higher level of care. It would be unsafe to transport this patient by family car.
3. D. EMS personnel must adequately protect themselves from harm when stabilizing and transporting psychotic and/or aggressive pediatric patients. Law enforcement officials, in addition to physical and pharmaceutical restraints may be necessary.

149 INTERFACILITY TRANSPORT

Maeve Sheehan

Craig J. Huang

HISTORICAL PERSPECTIVES

- Specialized medical transport systems have evolved from military conflicts.
- Earliest references of specialized transport systems date from the Napoleonic wars.
- The first reported fixed wing medical transport was in 1915 and rotor wing in 1944.
- Specialized pediatric transport teams began in the 1970s.
- Outcomes for critically ill and injured children improve when care is provided by skilled pediatric specialists.

LEGAL CONSIDERATIONS

- All efforts to obtain consent for treatment and transfer of a pediatric patient with an urgent or emergent medical condition should be made, but appropriate medical care, including transport, should never be withheld or delayed because of problems obtaining consent.
- Referring providers are responsible for stabilizing a patient's emergency medical condition, within the capabilities of their institution, before the patient is transferred to another institution for definitive care.
- Transport teams should be aware of and discuss with the medical control physician, any DNR and other limitations in resuscitation orders, realizing that those orders may be revoked at any time by the legal guardian.
- Although in reality there is some shared responsibility/liability for the patient between the transport team and the referring physician, the referring physician remains legally responsible for the patient while at their institution, chooses the particular transport service, mode of transport, and has the right to intervene or cancel the transport.

TEAM COMPOSITION AND TRAINING

- Transport teams are comprised of a variety of medical professionals, each of whom has their own potential advantages and disadvantages, with the ideal team composition determined by the patient's needs.

- Most adult EMS providers and transports have limited pediatric exposure, especially with critically ill and injured children.
- Specialized neonatal and pediatric transport teams can help fill this knowledge and skill gap and help to reduce transport-related morbidity.
- All transport team members should have the opportunity to develop and maintain advanced procedural skills on an ongoing basis.

COMMUNICATION

- A communication center is vital for the safety of patients and transport personnel.
- An ideal transfer center receives the initial intake call, facilitates the dialogue between the referring and receiving physician, notifies and dispatches the appropriate team, and tracks their location.
- A standard set of basic questions can help transfer center coordinators identify the most appropriate receiving hospital unit.
- It is essential to record any communication with outside facilities pertaining to transport.

EQUIPMENT AND MEDICATION/ PROTOCOLS/MEDICAL DIRECTION/ STABILIZATION FOR TRANSPORT

- Special equipment consideration is needed for basic life support and advanced life support transports of neonates, children, and adult-size patients.
- Lists of supplies, medication, and equipment should be checked regularly to ensure proper compliance with regulatory standards, expiration dates and storage requirements including refrigeration.
- Regularly updated, written protocols to direct patient care serve as a guideline for transport teams until the medical control physician can be contacted.
- Familiarity with the protocols is essential to guide each team member's actions within their specific profession's scope of practice, and should clearly define when to seek help from their medical control physician.
- The medical director of the transport service is involved in all aspects of care provided by the teams including the hiring and education of the team members and transport medical control physicians, development of policies, procedures, and transport protocols, infection control principles, quality assurance, and overall safety of the program for the patients and staff.
- Transport medical control physicians provide "on-line" medical advice and may give recommendations

- to the referring physician regarding the most appropriate mode of transport, initial patient stabilization prior to the team arrival, and on-going interventions.
- One of the goals of interfacility transport is to ensure that the child is as stable as possible for the transport that may require multiple interventions such as establishing a definitive airway, treating a pneumothorax prior to air transport, and having adequate vascular access.
 - The medical control physician should have the appropriate skill level regarding the patient population served by the transport service.

REGULATORY REQUIREMENTS/ CERTIFICATION STANDARDS

- Interfacility transport services must meet all hospital standards and city/state requirements for ambulance services.
- Air medical programs must also comply with FAA regulations.
- Programs may choose to be accredited by the Commission on Accreditation of Medical Transport Systems (CAMTS) a voluntary body that sets minimum standards for quality of patient care and safety in the medical environment.
- The Guidelines for Air and Ground Transport of Neonatal and Pediatric Patients manual published in 1993 by the AAP serves as a resource for the interfacility transport of infants, children, and adolescents.

MODES OF TRANSPORT

- Each mode of transport has its own particular advantage and disadvantage.
- Ground ambulance advantages include being the safest mode of transport, having the ability to stop the vehicle to perform procedures, space to permit transport of a family member, no requirement for a landing zone, and a lessened impact from inclement weather.
- Ground transport's major disadvantage is the travel time needed to cover long distances, particularly if the child is critically ill.
- Rotor wing transport is fast and available for remote and difficult terrain scene transports.
- The disadvantages of rotor wing transport include the stresses of flight, lack of space to perform emergency interventions, need for a specific landing zone, and unavailability in inclement weather.
- Fixed wing aircrafts are usually reserved for long-distance interfacility transports and often have enough room to perform emergency interventions and to permit transport of a family member.

- Disadvantages of fixed wing transport include altitude considerations, weather limitations, and the need for an airport and arrangement for ground transportation for pick up and return at each end of the transport.

SAFETY/ALTITUDE PHYSIOLOGY AND AIR MEDICAL CONSIDERATIONS

- Air and ground transports carry real risks, especially with a substantially increasing accident rate and number of flight hours with air transports.
- Safety considerations must be an essential and ongoing part of a transport team's training and policies, including; proper use of restraints, management of violent or psychotic patients, use of lights and sirens on the ambulance, weight limitations for flight, nighttime air transport, and transport during inclement weather.
- In the case of hazardous weather the final decision as to safety of travel should reside with the pilot or driver of the transport vehicle although any member of the transport team should have the ability to abort the mission due to safety concerns.
- Personal protective equipment is important to reduce or prevent injury in the event of unforeseen circumstances and to provide adherence to universal infectious disease precautions.
- Guidelines should mirror institutional-based policies/procedures in the event of accidental needle sticks or exposure to blood and/or body fluids.
- Although changes in barometric pressure and hypoxia are the most significant stressors to the patient and crew during air transport, temperature, dehydration, noise, vibration, g-forces, third-spacing, and fatigue should also be taken into consideration.
- Laws governing the behavior of gases under conditions of changing pressure have significant clinical implications such as barotraumas and volutrauma from rapid gas expansion or decreased oxygen saturations with increase in the altitude.
- For each 1000-ft gain in altitude there is a 2°C drop in temperature along with a decrease in ambient humidity in a pressurized aircraft.

SPECIFIC CLINICAL TRANSPORT ISSUES

- Neonates, especially if they are premature, have particular relevant physiologic demands and specialized equipment needs that should be considered when they are transported.

- Transport isolettes should be utilized for infants weighing less than 10 kg because they provide protection from environmental hazards.
- The medical control physician should be actively involved in the situation when an interfacility transport team is asked to transport a child in active cardiac arrest that occurs at the referring institution.
- It is important to try and identify the cause of the arrest prior to transport, so necessary interventions may be implemented to prevent further arrest.

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QUESTIONS

1. A 3-year old who was struck by a car is at a community hospital emergency department (ED), they are not a designated trauma center. Who determines by what mode the patient should be transported?
 - A. The administrator on call at the receiving facility.
 - B. The administrator on call at the referring facility.
 - C. The nurse at the referring facility taking care of the patient.
 - D. The pediatric hospitalist at the receiving facility.
 - E. The ED physician at the referring facility taking care of the patient.
2. A pediatric transport team is picking up an 8-year-old patient at a community hospital ED 10 miles away, who has an angulated radius and ulna fracture. He has no evidence of neurovascular compromise and his pain has been well controlled with ibuprofen. The ideal composition of the transport team consists of:
 - A. A nurse, respiratory care practitioner, paramedic, and helicopter pilot.
 - B. A physician and helicopter pilot.
 - C. A physician, respiratory care practitioner, and ground ambulance driver.
 - D. A physician and fixed wing pilot.
 - E. A nurse, respiratory care practitioner, and ground ambulance driver.
3. A 9 kg infant is being transferred to the local children's hospital with a femur fracture. The most appropriate way to transport this child in an ambulance is
 - A. In her mother's arms so she can console the child.
 - B. In an infant car seat with five point restraints.
 - C. In a transport isolette.
 - D. Restrained in a stretcher in the back of the ambulance.
 - E. In a seatbelt in the front of the ambulance.
4. A 5-year-old child injured in a motor vehicle collision needs to be emergently transferred to a pediatric trauma center. Her parents have been injured and are unavailable to give consent. The physician should
 - A. Wait for her grandparents who are on their way to give consent.
 - B. Transfer the child without delay as the parents are unavailable.
 - C. Call the local judge and get a court order to transfer the patient.
 - D. Obtain the signature of two physicians to give consent.
 - E. Keep the child in the ED until the parents can be found to give consent.
5. The ICU physician in a pediatric referral center receives a call from a community hospital that does not have a transport service. They wish to arrange

transfer of a child to the ICU for emergent dialysis. The transport team cannot fly due to bad weather. The physician should

- A. Ask to speak to the pilot and insist the team fly
- B. Call the referring doctor, tell him they cannot fly and ask him to call back tomorrow.
- C. Turn away the transfer.
- D. Call the hospital administrator and complain about the transport team not facilitating the transfer.
- E. Work with the transfer center and see if alternative transport arrangements can be made.

ANSWERS

1. E. Referring providers are responsible for stabilizing a patient's emergency medical condition, within the capabilities of their institution, before the patient is transferred to another institution for definitive care. The referring physician remains legally responsible for the patient while at their institution, chooses the particular transport service, mode of transport, and has the right to intervene or cancel the transport.
2. E. Transport teams are comprised of a variety of medical professionals, each of whom has their own potential advantages and disadvantages, with the ideal team composition determined by the patient's needs. This patient is hemodynamically stable. A nurse, respiratory care practitioner, and ground ambulance driver nurse is sufficient.
3. C. Transport isolettes should be utilized for infants weighing less than 10 kg because they provide protection from environmental hazards.
4. B. All efforts to obtain consent for treatment and transfer of a pediatric patient with an urgent or emergent medical condition should be made, but appropriate medical care, including transport, should never be withheld or delayed because of problems obtaining consent.
5. E. In the case of hazardous weather the final decision as to safety of travel should reside with the pilot or driver of the transport vehicle although any member of the transport team should have the ability to abort the mission due to safety concerns. A communication center is vital for the safety of patients and transport personnel.

An ideal transfer center receives the initial intake call, facilitates the dialogue between the referring and receiving physician, notifies and dispatches the appropriate team, and tracks their location.

150 MASS CASUALTY MANAGEMENT

Janet Lin

EPIDEMIOLOGY

- The effect of a disaster is nondiscriminatory; however, certain populations are more vulnerable than others.
- Unique issues surround the care and clinical management of the child during mass casualty events (MCEs). Children are not simply small adults.

MASS CASUALTY EVENTS

- A MCE is an event that is characterized by an imbalance between the needs and resources available within a health care system.
- The inciting event may be due to natural disasters, transportation-related failures, civil disturbances, war, or terrorist-related activities.
- Pediatric patients have physiologic, developmental, and behavioral differences from adults that influence their management in an MCE.

UNIQUE PEDIATRIC CONSIDERATIONS AGE, SIZE, AND WEIGHT

- A MCE involving pediatric victims demands a wide range of skills and the availability of equipment to accommodate a range of sizes and weights.
- Age is often considered a surrogate for size and weight in clinical MCE management.
- Medications like antibiotics, vaccines, or antidotes need to be dosed according to the age, size, or weight of a child.
- Response to medication treatment may vary because of the metabolic differences in children when compared to adults.

DEVELOPMENTAL AND BEHAVIORAL CONSIDERATIONS

- The stage of motor and cognitive development of a child will influence a responder's ability to communicate with and care for a victim.
- Nonverbal children cannot voice their complaints or injuries and may not be able to cognitively distinguish between one who is trying to help them from one who may be trying to hurt them.

- Nonambulatory children will not be able to flee a dangerous situation and need to be carried and transported away.
- Depending on the age and cognitive development of a child, he/she may refuse to move or even run toward a threat. Children may lack self-preservation skills that tell them to run away from a dangerous situation or lack decisional capacity to follow directions from strangers who are trying to help.
- Children may suffer anxiety due to separation from their family or primary caretakers during a disaster. They are also susceptible to the reactions and mental state of their caretakers.
- Children rarely carry personal identification, which makes the process of identifying victims and later reuniting family with their children more difficult.

PHYSIOLOGIC DIFFERENCES

- Children have increased respiratory rates, which can lead to increased absorption of aerosolized chemicals, and thus more severe illness.
- Children are generally closer to the ground because of their small size. They are potentially more vulnerable to agents that either settle on the ground or do not become airborne.
- Children have an increased body surface area and thinner skin relative to adults, which can lead to increased absorption of toxic agents.
- Children are more susceptible to hypothermia, especially when skin is exposed.
- Children have smaller circulating blood volumes.
- Children tend to preserve their hemodynamic function despite a relatively large volume loss; cardiovascular collapse can occur suddenly.

CONSIDERATIONS FOR THE DELIVERY OF CARE TO THE PEDIATRIC POPULATION

PROVISION OF CARE

- Mass casualty disaster plans should include recommendations for care of pediatric patients.
- These include: use of a pediatric-specific mass casualty triage protocol, availability of pediatric-sized equipment and supplies, proper decontamination guidelines, plans for reunification of children with their family, and plans to address postevent mental health needs of the children and family.
- From a prehospital perspective, it is best to transport pediatric MCE victims to either a regional pediatric hospital or one that has extensive pediatric expertise.

- Hospitals need to think about providing care as a family unit. That is, general hospitals need to be able to care for pediatric patients and pediatric hospitals need to be prepared to care for parent/adult victims as well.

TRIAGE

- Adult mass casualty triage protocols cannot be universally used for pediatric patients, as adult protocols often rely on a victim's ability to verbalize and ambulate.
- Physiological differences between adults and children make the adult MCE triage protocols inappropriate for infants and children.
- The JumpSTART method focuses on pediatric-specific parameters and places an emphasis on early airway intervention because respiratory failure usually precedes circulatory failure in children (Fig. 150-1).
- The pediatric assessment triangle (PAT) relies on visual cues of appearance, work of breathing, and skin circulation to rapidly assess the acuity of a patient illness (Fig. 150-2).

EQUIPMENT

- A pediatric emergency measuring tape, like a Broselow tape, is useful in estimating the proper equipment size or medication dose without having to perform any time-consuming calculations or measurements.
- Establishing intravenous access may be difficult and time consuming. Alternative access sites (scalp, umbilical, central venous sites) and means [ultrasound aided, intraosseous (IO) access] must be considered.
- MCE plans should include provisions for a minimum supply of appropriate pediatric-sized equipment and extra supplies at all times.

DECONTAMINATION

- Decontamination needs to be performed in any MCE that involves suspected biological, chemical, or radiation exposures.
- Children are at higher risk for hypothermia than adults.
- It takes longer to decontaminate children due to hesitancy of children to disrobe, inability to disrobe themselves, fear or anxiety about being separated from parents or known caretakers.

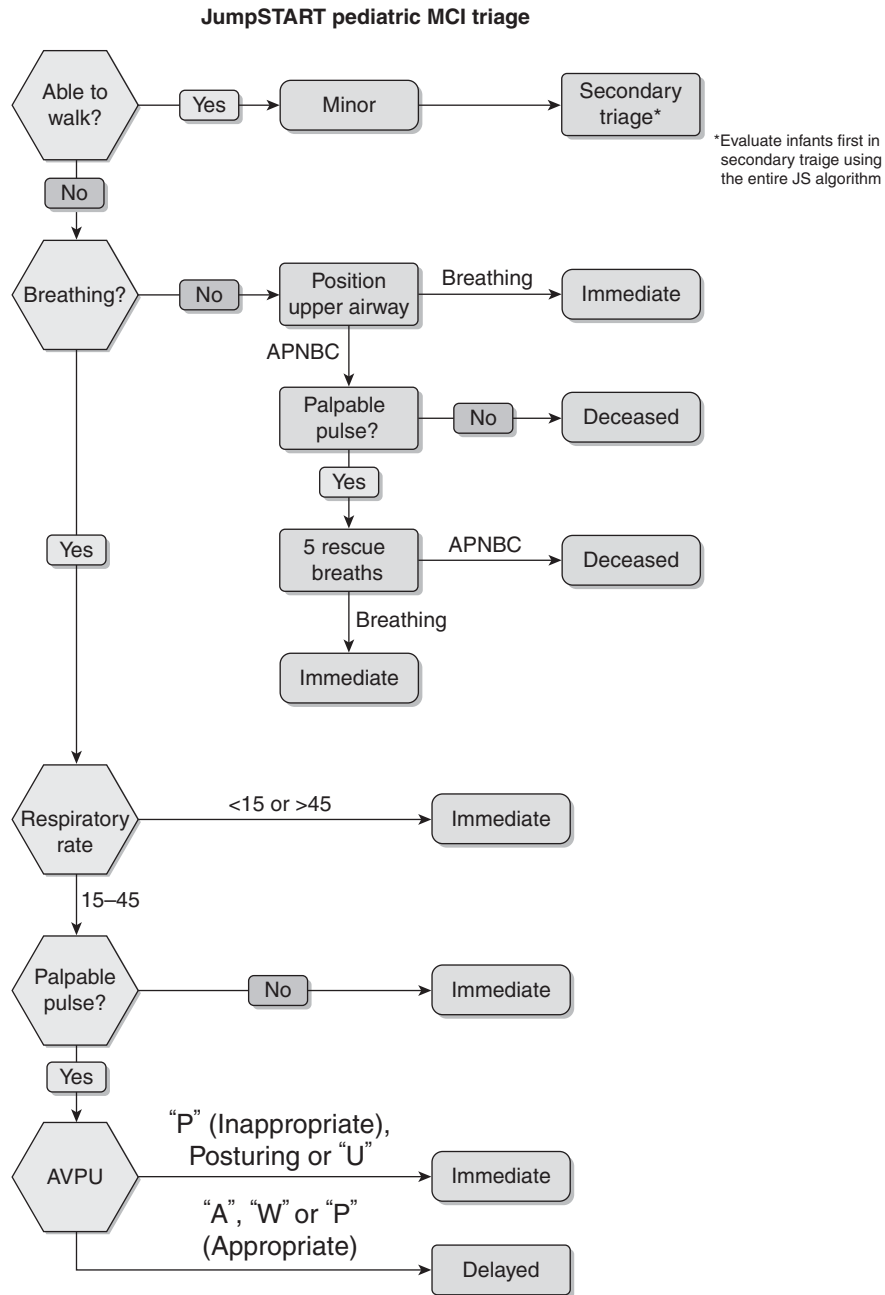


FIG. 150-1. JumpSTART Triage algorithm. (With permission from developer, Lou E. Romig MD, FAAP, FACEP.)

- Personal protective equipment limits the dexterity of health care personnel. Handling small children and infants requires a higher degree of caution.

MEDICATIONS

- Newer recommendations support the use of medications that historically have not been used in children

- because of their potential toxic effects during a disaster or a MCE because the risks outweigh the benefits (eg, ciprofloxacin and doxycycline).
- The FDA has approved pediatric-specific atropine autoinjectors. Autoinjectors have the advantage of being prefilled, easy-to-use devices that do not require intravenous access.
- The Broselow tape system has recently made available a new color-coded pediatric tape that contains

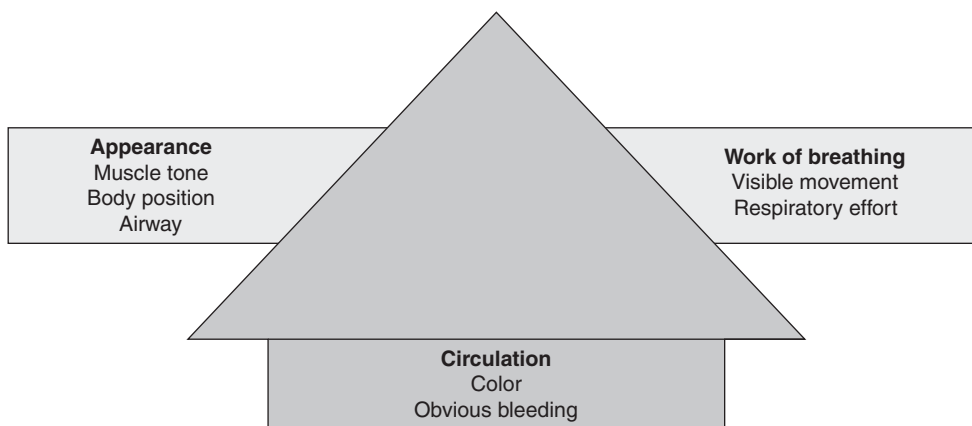


FIG. 150-2. Pediatric assessment triangle.

doses for chemical treatment agents, including adult autoinjectors, thus decreasing the amount of time required to determine doses for victims.

RECOVERY

- Every effort should be made to reunite family members with the child as soon as possible. Keeping and treating parent and child victims of a MCE together in the same facility is a logical strategy.
- Children suffer varying degrees of psychological disturbances after an MCE.
- Somatization is common and parents may not identify symptoms as psychological in origin.
- Try to establish a safe environment. This may be accomplished simply by reuniting a child with his/her family.
- Children need reassurance that their reactions are not a result of something right or wrong, but a normal reaction to an abnormal event.

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QUESTIONS

1. It is important for health care centers to create emergency management plans that specifically address the pediatric population because
 - A. Plans that specifically address adult patients are easily adapted to pediatric patients.
 - B. When pediatric patients present with the same injury or exposure as adults, their symptoms or complaints are completely different.
 - C. Pediatric patients in a MCE have unique issues that influence care and management of the child.
 - D. Pediatric patients are a vulnerable population that is singularly affected during MCEs.
 - E. Accrediting bodies require a specific pediatric plan for health care centers to function.

2. A cross-country passenger train derailment occurred on the railroad that runs near your hospital. Victims are being brought to several of the area hospitals in your region. You are receiving several pediatric victims. What is a treatment priority for this type of event?
 - A. Extra help should be available to assist in the more complicated decontamination process for pediatric patients.
 - B. Pediatric patients should be reunited with parents or known caretakers as soon as possible.
 - C. Calls should be made immediately to the closest pediatric hospital to arrange transfer of pediatric patients to that facility.
 - D. Pediatric patients should be given priority in the initiation of treatment over adults because they are more likely to respond to earlier interventions.
 - E. The most important parameter to take into account in determining doses of medications for children is age.
3. A 3-year-old child arrives at the ambulance bay of your hospital after a mass casualty plan is called to the area medical centers. The child is awake but visibly scared. Why is a specific pediatric triage protocol preferable to an adult triage protocol for assessing this 3-year old child?
 - A. Adult triage protocols place more emphasis on circulation/perfusion than respiratory status.
 - B. Pediatric triage protocols are based on and contain the entire range of normal vital signs expected in all pediatric age groups.
 - C. Pediatric triage protocols incorporate more life saving interventions in the triage process than an adult triage protocol would.
 - D. Adult triage protocols often rely on the ability of a patient to verbalize their complaints.
 - E. Adult triage protocols underestimate the fluid loss and, therefore, underestimate the fluid requirements of pediatric patients.
4. A mass casualty plan has been called after an aerosolized agent was released in a public park during a summer festival. Most patients are not presenting with physical injuries. What might you expect to find in the pediatric patients that present to your hospital?
 - A. More severe illness because children have increased respiratory rates, which can lead to increased absorption of aerosolized chemicals.
 - B. As the aerosolized agent dissipates, children may be more vulnerable to the agent because they are generally closer to the ground because of their small size.
 - C. An increased absorption of the toxic agents because children have an increased body surface area and thinner skin relative to adults.
 - D. Because there is more skin exposed, they will be more susceptible to hypothermia.
 - E. Sudden cardiovascular collapse because children have smaller circulating blood volumes and they can preserve their hemodynamic function despite a relatively large volume loss.
5. The stage of development of children can increase the risk of exposure during a disaster. Developmental stage may also introduce challenges for healthcare providers to provide appropriate care. Behaviors that may influence a healthcare providers ability care for or assess children include
 - A. Children who are nonverbal will voice their complaints or injuries through sounds. Health care providers can differentiate whether these sounds are from those children who are scared versus those who are in pain.
 - B. Children will always need to be carried out of dangerous situations because they will not flee on their own, requiring more resources and time to evacuate pediatric patients.
 - C. Children that carry personal identification will readily show them to responders or care providers, which makes the process of identifying victims and later reuniting family with their children easier.
 - D. Children will run from a dangerous situation because they have an innate sense of danger telling them to flee and not seek care, but find shelter and hide.
 - E. Children may not follow directions from strangers (ie, responders) who are trying to help because they lack decisional capacity or are scared.

ANSWERS

1. C. Unique issues are highlighted in the text and varied. A is incorrect. Pediatric patients cannot simply be treated as adults. Many plans address and assume certain cognitive and behavioral capabilities that may or may not be present in children. B is incorrect. While presentations or complaints may be different, oftentimes they are not. D is incorrect. Vulnerable populations are definitely affected more in disasters or MCEs, but pediatric population is not the only vulnerable population that will be affected. E is incorrect. While this would be ideal, there is no specific requirement.

2. B. Every effort should be made to either reunite or keep families together as much as possible. A is incorrect. Mechanical disaster involving a passenger train, no concern for possible exposure, decontamination is not required. C is incorrect. Depending on location, this may or may not be possible. In addition, the immediate need is to care for patients, so nonpediatric facilities should be prepared to care for pediatric patients. Transfer to a pediatric facility if it is far away, may not be practical or safe. D is incorrect. Priority should be based on severity of injury and likelihood of survival. While age may play into, by no means is that a guarantee of improved outcome or survival. E is incorrect. Age can be used as a surrogate for estimating medication dose, but weight and height is probably more ideal, or using a device like Broselow's tape.
3. D. Given that pediatric patients cannot often talk, visual clues are incorporated into pediatric triage protocols. A is incorrect. Adult triage protocols generally emphasize respiratory status over circulation, as does pediatrics. B is incorrect. While pediatric populations do have a wider range of vital signs, the emphasis in pediatric triage protocols is based on visual clues rather than actual numbers. C is incorrect. Quick and immediate life-saving maneuvers are employed in both pediatric and adult triage protocols, ie, airway maneuvers, however, not any more than in adult protocols. E is incorrect. Fluid resuscitation does not really play into the triage protocols.
4. A is correct. B is incorrect. Aerosolized agents may not necessarily fall to the ground, but rather dissipate in the air, so just because children may be smaller and closer to the ground does not make them more vulnerable. C is incorrect. Again, because this is an aerosolized agent, the more likely mode of exposure is through inhalation. Skin absorption plays a lesser role. D is incorrect. Same as above, this question does not focus on skin exposure. E is incorrect. While this may play a role in general, this is not a trauma related disaster where you would expect blood loss, therefore volume loss.
5. E is correct. A is incorrect. Nonverbal children may be a challenge because they cannot voice complaints or injuries. It is often difficult to differentiate scared from pain, especially if a child is not familiar with the stranger/responder. B is incorrect. If children are non-ambulatory, they may need to be carried out, but not all children will have to be. C is incorrect. Children oftentimes do not carry identification. In addition, they are unlikely to volunteer such information, either they do not know or are scared. D is incorrect. Children sometimes will not realize that they are in danger and may stay in place or even run towards a dangerous situation.

Section 23

MEDICOLEGAL AND ADMINISTRATIVE ISSUES

151 MEDICO-LEGAL CONSIDERATIONS

William R. Ahrens

- The emergency medicine physician must work within the context of a society's laws as they apply to the emergency department (ED). Laws in the United States can be divided into three categories: statutory, common, and administrative.
- Statutory law is codified by legislative bodies; an example is the requirement to report child abuse to authorities.
- Common law is based on the case law; it has evolved over centuries, and is interpreted by judges as it applies to litigated cases. Common law applies to medical malpractice.
- Administrative laws are enabled by legislation and are issued by agencies in the form of regulations; an example is the regulations by Drug Enforcement Agencies regarding the prescription of narcotics.
- U.S. law is also divided into state and federal law. The federal statutes that apply most directly to an ED are the Emergency Medical Treatment and Labor Act (EMTALA) and the Health Insurance Portability and Accountability Act (HIPAA).

EMTALA

- EMTALA was enacted by congress in 1986 as part of the Consolidated Omnibus Reconciliation Act of 1985 (COBRA) as a response to the refusal of some EDs to provide care to uninsured patients. One of the key provisions of EMALA is that when a patient requests care at an ED, "the hospital must provide an appropriate medical screening examination within the capability of the hospital's ED, including ancillary

services routinely available to the ED, to determine whether an emergency medical condition (EMC) exists."

- If an EMC exists, the hospital must provide "with the staff and facilities available, for such further medical examination and such treatment as may be required to stabilize the medical condition or transfer the patient to another facility." The medical screening examination must be performed by a physician or someone accredited to do so by the medical staff.
- An EMC is a condition "manifesting itself by acute symptoms of sufficient severity, including severe pain, psychiatric disturbances, or symptoms of substance abuse, such that the absence of immediate medical attention could reasonably be expected to result in (a) placing the health of the individual in serious jeopardy, and (b) serious dysfunction of any bodily organ or part."
- The broad definition of an EMC gives the emergency physician performing a legally required medical screening examination wide discretion to err on the side of caution, especially in the case of unaccompanied minors.
- Stabilization of the patient means that within reasonable clinical confidence, no material deterioration of the condition is likely to result from or occur during transfer. The receiving facility must agree to accept the transfer; all pertinent medical records must accompany the patient. The condition of the patient as well as the risks and potential benefit of the transfer must be documented.

HIPPA

- The Health Insurance Portability and Accountability Act (HIPAA) and the finalized version "Standards for Privacy of Individually Identifiable Health Information" (HIPAA Privacy Rule) require health care providers

and institutions to provide a set of minimum standards to protect medical record privacy and confidentiality.

- The law reflects the fundamental ethical paradigm that all patients are entitled to privacy. Protecting privacy is within the code of ethics for emergency physicians, part of which states that “sensitive information may only be disclosed when such disclosure is necessary to carry out a stronger conflicting duty, such as a duty to protect an identifiable third party or to comply with a just law.”
- Confidentiality is closely related to privacy; in medicine, it refers to the duty not to disclose information conveyed to a health care provider without the patient’s approval. Generally, HIPPA requires patient permission for information disclosure; consent is not required for disclosure of personal health information for purposes of treatment, payment, and health care operations. Patient’s consent is also not required for communication between consultants when threats to patient of public safety are involved, such as suicidal or homicidal ideation or terrorist threats, certain instances of abuse and neglect, and certain issues involving law enforcement.
- HIPPA confers a high degree of confidentiality or records pertaining to psychiatric treatment. A patient must approve the release of psychotherapy notes, unless they are being used for the purposes of treatment by the physician who originated them, or if they are necessary to avert an imminent threat to public health or safety.
- Confidentiality issues regarding minors, especially adolescents, are problematic. There needs to be a balance between the adolescent patient’s inherent right of privacy and parents’ right of access to information pertaining to the child for whom they are legally and morally responsible.
- The HIPPA Privacy Rule states that in most cases a parent is the “personal representative” of the child and thus has legal power to control the release of the minor’s medical records. If the parent is not the personal representative of the child, state or other regulations apply. Examples where the parent is not the personal representative of the minor include situations where the minor can consent for their own medical care, when the parent agrees to a confidential relationship between the minor and physician, when the physician believes the child has been abused or that the parent is not acting in the child’s best interest, and when a court-appointed guardian is the minor’s personal representative.
- Laws regarding parents’ right to access the medical records of adolescents who have received treatment for which they were able to consent vary from state to state.

CONSENT AND REFUSAL OF CARE

- Informed consent is predicated on a patient or patient’s representative demonstrating the intellectual capacity and emotional maturity to fully understand the information presented and the consequences of both acceding to medical treatment and refusing care. In most states, the age at which a patient can consent for or refuse medical care is 18.
- States have increasingly recognized that a “bright-line rule” regarding who is an adult may not be appropriate, and many have adopted “mature minor” statutes. Such statute presume that minors, in most cases beginning at age 14, have the cognitive and developmental capacity to comprehend the nature of the medical treatment offered and, therefore, the decisional capacity to accept or decline care. These statutes are case sensitive; consent relates to specific situations in which adolescents demonstrate the capacity to make decision related to their care.
- Almost all states have doctrines involving “emancipated minors”; laws vary, but most commonly include minors who are married, serving in the armed forces, are pregnant or parents, or who are self-sufficient as capable of making their own medical decisions.
- Most states have provisions that allow adolescents to seek psychiatric care without the consent of their parents; the age at which they may do so varies from state to state.
- Other conditions that may preclude the need for consent in minors include treatment of sexually transmitted diseases, including HIV, contraceptive services, and treatment for substance abuse.
- Increasingly, minors who are not emancipated seek care in EDs. While every effort should be made to contact their parent or personal representative, EMTALA specifically requires a medical screening examination: this in essence obviates the issue of consent. The American College of Emergency Physicians states: “Under federal law, a minor can be examined, treated, stabilized, and even transferred to another hospital without consent ever being obtained from a parent or legal guardian.” This applies to cases where the minor is brought to the ED with an EMC by a personal representative who for whatever reason may be impaired, and therefore incapable of informed consent, and who may not act in the best interests of the patient.
- Refusal of care is an issue that often involves a complex interaction between what is the law and what is perceived the patients’ best interest, either by the patients themselves or their personal representative(s). There is no question that adults with adequate decisional capacity have the right to refuse medical care.

When a conflict involving refusal of care for a minor arises, it is the best interest of the minor that is the priority. In cases involving a nonemancipated minor with an EMC, the physician has the obligation to use the legal system to protect the patient.

FORENSIC DOCUMENTATION

- Because most assaults entail some form of criminal activity, the examining physician has a high likelihood of becoming involved in the legal aftermath of the case.
- In the pediatric patient who is a victim of maltreatment, careful and correct documentation can serve to protect the child from further harm; poor documentation can result in the child being returned to a potentially fatal environment.
- In cases that go to trial, the emergency physician may be asked to provide expert testimony.
- Most emergency physicians have no forensic training. The extent of documentation is correlated with the filing of charges, and the conviction and sentencing of perpetrators.
- In the case of child maltreatment, elements such as the explanation provided, whether witness were present, the developmental stage of the child, and a clear description of injuries are often absent from the documentation.

MEDICAL MALPRACTICE

- The four requirements for malpractice are duty, a breach of duty, damages, and causation. A physician treating a patient in an ED has a “duty” to treat the patient. A breach of duty occurs when the physician does not treat the patient according to “the standard of care,” or in a way that a reasonable physician practicing in a comparable setting would have done in a similar situation. Damages occur when a patient suffers an adverse outcome as a result of the care rendered. Causation means that the breach in care is shown to have caused the damages (some jurisdictions vary).
- A recent review of medical malpractice suits in the United States involving pediatric patients found that 49% of the cases involved children younger than 2 years. Diagnostic error was a factor in 39% of cases. The most common diagnoses involved were meningitis, fracture, appendicitis, and testicular torsion. Other factors cited were improper performance of procedure, failure to supervise staff, delay in treatment, failure to consult, failure to admit, and medication error. Most cases were settled out of court.

- Preventing, or at least minimizing, errors in the ED, and therefore medical malpractice suits, involves a combination of systems analysis, quality improvement, education, and teamwork.
- There is a rapidly growing and most likely unstoppable movement to inform patients when they have been the victim of a medical error. Studies have found that patients want to know when a mistake has happened; physicians are less likely to want to disclose an error. There is little information regarding error disclosure in the pediatric population. How error disclosure affects medical malpractice is as yet unknown—it may decrease the likelihood of a lawsuit.
- Many states have requirements for the standardized reporting of medical error. A few states require patient notification of medical error. Some states protect health care providers with “apology statutes,” where expressing remorse or apologizing for an error is not admissible in court as an admission of liability. Legislation in this area is constantly changing and will, no doubt, develop rapidly as public sentiment demanding full error disclosure grows.
- The standards set by Joint Commission on Accreditation of Healthcare Organizations require that patients and families be informed about medical errors. The American College of Emergency Physicians policy on error disclosure is summarized in Table 151-1.

END-OF-LIFE CARE

- It is likely that an emergency physician will someday be confronted with a pediatric patient suffering from a terminal illness who presents to the ED in a near-death situation. In most of these situations medical decision making is the responsibility of the parents of

TABLE 151-1 ACEP Policy on Disclosure of Medical Errors

Health care institutions should develop policies and procedures for identifying and responding to medical errors and procedures for disclosing significant errors to patients.
Medical educators should develop and incorporate into their curricula programs on identifying and preventing medical errors and on communicating truthfully and sensitively with patients and their representatives about errors.
Emergency department directors . . . and other leaders in emergency medicine should play a leading role in developing institutional and ED policies for prompt error identification, responsible reporting, and proper remediation.
Society should adopt tort reforms and system changes that improve patient safety by encouraging disclosure of medical errors.

the patients, who are expected to act in the best interest of their child.

- As the concept of the emancipated minor expands and develops a stronger legal footing, more young patients will be able to make their own medical decisions, including refusing resuscitation.
- Legally and ethically, emergency physicians are not required to provide treatment that would be ineffective. However, futility is very difficult to define; situations involving resuscitative efforts are best guided by the parents, guardian, or patient, rather than the physician.
- Parents who initially request aggressive treatment may later change their mind; withdrawing life support, including ventilation, is ethically and legally permissible even in the ED when it is no longer in the best interest of the patient.

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QUESTIONS

1. The charge nurse tells you there is a 14-year-old female in the waiting room complaining of a sore

throat. Her parents cannot be reached. Which of the following is true?

- A. To look in her throat constitutes assault.
 - B. You can only examine her with the approval of hospital administration.
 - C. She should be told to come back later with her parents.
 - D. She can be triaged by the admissions clerk and discharged.
 - E. She must be examined by a qualified individual.
2. A 15-year-old male presents to the ED and in the course of the interaction tells you he plans on killing his girlfriend because she is seeing someone else. The patient refuses to allow you to call his psychiatrist. HIPPA
- A. Has no applicability in this situation.
 - B. Mandates confidentiality even in this scenario.
 - C. Provides criminal penalties to physicians who share information even in a case like this.
 - D. Allows communication between health care providers when there is a threat to an individual or public safety.
 - E. Is written in language such that it would be best to ignore the patient's threat as he is unlikely to carry it out.
3. A 17-year-old female who is a Jehova's witness presents with vaginal bleeding. She is pale and tachycardic, with a normal blood pressure. Her mental status is normal. Her hemoglobin is 5 mg/dl. She refuses a transfusion. Her parents agree with her decision. Which of the following is true?
- A. Under no circumstances can she refuse a transfusion since she is under 18 years of age.
 - B. She should be immediately transfused, even if she is restrained.
 - C. Hospital administration should take decision regarding a potential transfusion.
 - D. Her parents have the absolute right to decide whether to transfuse or not.
 - E. The patient may have the right to refuse a transfusion, even if it means she will die.
4. A 14-year-old female presents to the ED with the complaint of being raped by her mother's boyfriend. You
- A. Send the intern in with a nursing student to do the exam and rape kit.
 - B. Avoid "overdocumenting" as it may complicate future testimony.
 - C. Document all details of the history and physical meticulously and nonjudgementally.
 - D. Do a quick, superficial exam to avoid further traumatizing the patient.
 - E. Avoid a detailed history of the assault because it is embarrassing for both of you.

5. You order 10 units of insulin on a diabetic child; the nurse accidentally gives 100 units. You successfully manage the complication and the child does well. The following is true:
- Under no circumstances should you tell the family about the mistake.
 - Patients do not want to know about medical errors unless the plan to sue.
 - You may live in a state where the law requires you to inform a patient about a medical error.
 - JACHO advises physicians to avoid discussing medical errors with patients.
 - Disclosing medical errors definitely decreases litigation.

ANSWERS

- E. EMTALA requires a screening examination of all patients to rule out a medical emergency by an individual qualified by the hospital. Administrative consent is not necessary. The law designed to protect patients, including minors, and gives the examining physician or other health care provider wide discretion.
- D. HIPPA is designed to provide patient confidentiality; it is especially protective of psychiatric information. However, when there is a threat to an individual or to public safety communication between health care providers is permissible.
- Discussion. This very real scenario is a nightmare. Given relatively stable vital signs there is time to consider options so restraint and forced transfusion is inadvisable. Hospital administration should probably be aware of such a scenario, but the decision to transfuse rests ultimately with the patient and her parents. A mentally competent 17-year old may be considered a mature minor, and in some states may be allowed to refuse a transfusion despite her parents wishes. Such a situation usually requires legal adjudication.
- C. In a scenario like this that goes to trial the examining physician is likely to be asked to testify as an expert. It is vital that the history and physical be documented accurately and nonjudgementally by an experienced health care provider. Good documentation can mean the difference between a conviction and a perpetrator that is set free.
- C. At least many patients want to know if they or a family member is a victim of a medical mistake. JACHO standards require that patients be informed about medical errors. Such disclosure may decrease litigation. Some states require that patients be informed about medical errors.

152 ETHICAL CONSIDERATIONS

Alan Johnson

- When caring for an ill or injured child, patients, parents, guardians, and physicians share a common goal: to act in the best interests of the child.
- Ethical conflicts may arise if there are differences of opinion about those best interests.
- Sometimes disagreements may arise which, if unresolved, could jeopardize the health and well-being of the child. Significant effort may be required to resolve conflicts in order to provide for the best possible outcome for the child.
- A core ethical value of emergency medicine is that emergency departments (EDs) serve as an essential medical safety net.
- An emergency medicine physician needs to be familiar with current recommendations, policy statements, principles, and controversies that guide the practice of emergency medicine as they apply to the care of children.

TRIAGE AND OVERCROWDING

- Pediatric patients make about 30 million visits to EDs in the United States each year, accounting for about 25% of all ED visits. Although overcrowding of EDs is difficult to quantify, a reasonable definition is “the need for medical services exceeds available resources.”
- One solution to overcrowding is to triage low-acuity patients away from the ED to some other source of medical care, but no pediatric triage protocol identifies all patients with a serious illness. It is a good practice to develop systems within the department and institution to provide a place to care for lower-acuity patients and to triage such patients to this area.
- No child should be turned away or denied care, based on an initial triage assessment that the complaint is low acuity.

MEDICALLY UNDERSERVED CHILDREN

- Poor, immigrant, homeless, uninsured, and migrant children often do not have access to outpatient medical care.
- Poor children have higher rates of under immunization, acute illnesses, asthma, injury, malnutrition, and mental health issues than the general population.

- The Emergency Medicaid program was established in 1986 and provides coverage for uninsured documented and undocumented children with an emergency medical condition; eligibility and benefits are set by each state.
- These vulnerable children not only require excellent emergency medical care, but also need to be connected to any federal, state, and community resources to help them access the Emergency Medicaid program and other support services to facilitate their overall health and well-being.
- Care should be provided for the presenting complaints and, whenever possible, a referral should be made to an accessible source of ongoing primary care in the community.
- The American College of Emergency Physicians opposes federal and state initiatives that would require refusal of care to undocumented persons or reporting suspected undocumented persons to authorities.

CHILD ABUSE

- When treating a patient that may have been abused, the physician must consider the best interests of the patient as the primary focus of the evaluation.
- A parent or caretaker may have conflicting interests, wanting what is best for the child while possibly wanting to avoid investigation and prosecution of themselves or another parent or family member.
- The treating physician not only needs to explain the process of investigation to the parent, assure the parent that they are dedicated to the child's well being, but also explain the legal mandate to report a suspected abusive situation to the local child protection agency.
- Even when a child is in protective custody, the parent may still remain in charge of medical decision making for the child.
- In the event that a parent does not seem to be acting in the child's best interests, a court proceeding to establish a guardian for medical decision making should be pursued.

INFORMED CONSENT, PARENTAL PERMISSION, AND ASSENT

- The informed consent process is an ethical cornerstone of providing care to patients, as it balances the physician's desire to do what is best from a medical and scientific perspective with the patient's right to comprehensive information in order to decide what is best from their personal perspective.
- Informed consent is the appropriate term for an adult or adolescent with decision-making capacity, informed

permission is the preferred term when a parent or other surrogate makes decisions for a patient lacking decision-making capacity, and assent (or agreement) of a patient lacking decision-making capacity should be sought whenever possible and appropriate.

- In an emergency, a societal standard of presumed or implied consent allows medical treatment to prevent harm based on the assumption that a person in danger would want to be saved.
- Treatment for an emergency medical condition should never be delayed despite the inability of the patient to provide informed consent or the presence of a parent to provide permission.
- Parents are almost always strong advocates for what they believe to be best for their children, but children have rights independent of their parents.
- Children have an evolving decision-making capacity that is dependent on psychological, emotional, and intellectual development and maturity that the physician must consider and should therefore participate in decision making as appropriate for their developmental status.
- Adolescents may seek confidential and independent health care for conditions defined by state law, including sexually transmitted infections, pregnancy, psychiatric complaints, and substance abuse problems.
- Minors may also be legally "emancipated" or "mature" and able to consent for their own health care if they meet one of several conditions defined by the state as signifying independence, including marriage, pregnancy, parenthood, participation in military service, or financial independence. This varies by state.
- Adolescents, 14 and older, usually can make informed health care decisions as well as adults, so for conditions not covered by statute, a "mature minor" approach allows low-risk, high-benefit treatments to be provided if the physician believes that the minor is as capable to consent for the treatment as an adult.

PARENTAL AND PATIENT REFUSAL

- An adult patient with decision-making capacity may refuse medical treatment, but refusal of recommended diagnostic testing and treatment for children is more problematic.
- In almost every circumstance, the child, the child's parents, and the child's physicians have the child's best interests in common, but there may be disagreement about what those interests are.
- A parent refusing an invasive or painful diagnostic procedure may believe that they are acting in the best interests of the child—and the child may very well agree.
- Such a conflict can arise during the evaluation of a common complaint in a well-appearing patient with a

symptom that suggests the possibility of a very serious disease, such as a fever in a neonate.

- Physicians consider painful and invasive tests to be routine, but they are often anything but routine from the perspective of the child and the parent.
- It is unusual for parents to refuse any testing at all, but sometimes they are concerned about a particular test, such as a lumbar puncture or a urethral catheterization.
- In the event that a parent refuses a diagnostic test, the physician needs to review the remaining options and make therapeutic decisions based on the available data in a similar manner to what must be done if a test is attempted but not successful.
- Therapy for a potentially dangerous or progressive condition should not be delayed or forgone for lack of a complete diagnostic evaluation.
- Providing emergency care to children requires a delicate balance of obtaining parental permission and the assent of the patient for care, while at the same time acknowledging the societal mandate that in a life-threatening situation the parent and patient cannot refuse life-saving therapy.
- The most commonly discussed example is a child of the Jehovah's witness faith needing a blood transfusion, and courts have consistently ruled that the child's welfare supercedes the religious practice.
- In the case of an exsanguinating injury, the child should be transfused; but if the child can be stabilized and is likely to need a transfusion in the future, the local child protection agency should be involved and a judicial order mandating transfusion should be obtained.
- Adolescent refusal of care is more complicated, but should be respected if the refusal would not subject them to a great risk of harm.

FAMILY PRESENCE FOR PROCEDURES AND RESUSCITATION

- Traditionally, parents and families were excluded from the room for invasive procedures or when a critically ill or injured child was being resuscitated due to concern that their presence might interfere with optimal care and outcome.
- Physicians are concerned that a parent might interfere with the procedure or resuscitation, might distract the caregivers, might be traumatized by the procedure or resuscitation, and might subject the physicians to physical harm or a higher degree of malpractice risk in the event of a bad outcome.
- Survey research reveals that families want to be given the choice about being present for an invasive procedure or resuscitation and most choose to be present, and most feel that their presence is helpful to the child and to the health care team.

- Physician opinions vary and are influenced by institution, specialty, age, experience, and geographic area but there is a growing trend to support the parental presence.
- Family presence should be considered for all procedures and resuscitations, but assurance of safety for the patient and health care team is essential and may require exclusion of family members who are combative, threatening, intoxicated, mentally ill, or emotionally overwhelmed.

PRACTICING PROCEDURES ON THE NEWLY DEAD

- Teaching and practicing procedures on the newly dead has a long history in medicine and is practiced in about half of emergency medicine training programs, with endotracheal intubation being the most common procedure practiced.
- It has been argued that practicing certain procedures is an invaluable learning experience for the trainee, and the trainee can learn an invasive but not disfiguring procedure on a newly dead body in order to better serve the next living patient.
- A more problematic approach is adopting the pretense of therapeutic intent prior to pronouncing a patient dead in order to practice invasive procedures that have no hope of benefiting the patient.
- Even if practicing procedures on the newly dead is an excellent way for a medical trainee to learn and others will benefit from the physician's procedural skill in the future, it should only be done with permission from a parent.
- If teaching procedures on the newly dead is to be done, the teaching institution and program should have a policy covering this practice.
- Only those trainees requiring the skill should practice, and only after mastering the procedure on artificial models or donated cadavers, and only nonmutilating procedures should be performed.
- Most families will agree to allow procedures to be practiced, but they want to be asked.

ADOLESCENT ISSUES

CONFIDENTIALITY

- Adolescents must be assured that communications will be kept confidential, but the limits of confidentiality must be discussed as confidentiality may be breached in the event of a disclosure of abuse or homicidal or suicidal intent.
- Although every effort should be made to protect confidentiality, the adolescent needs to be aware that

their parents may get information from a medical bill or statement from an insurer.

- An acceptable and confidential method to provide the patient with test results should be established, include instructions for the adolescent to obtain their test results, discussing the situation with the patient's primary care physician, or calling the patient on their personal phone.

CHAPERONES AND PHYSICAL EXAMINATION

- A problem-focused physical examination of an adolescent may often be performed without discomforting or embarrassing the patient.
- When a breast, anorectal, genital, or pelvic examination is indicated, the examination should be clearly explained.
- The patient, parent, or physician will want to have a chaperone present for the more sensitive parts of the examination. The presence of the third party may protect the interests of both the patient and physician.

EMERGENCY CONTRACEPTION

- Although teen birth rates have been on the decline, 74% to 95% of adolescent pregnancies are unintended.
- Emergency contraception—the use of hormonal therapy up to 120 hours after unprotected sexual intercourse—may decrease the rate of unintended adolescent pregnancies.
- Education about and access to emergency contraception does not increase the frequency of protected or unprotected intercourse and should be offered to all adolescent women when sexuality issues are discussed.
- The American Academy of Pediatrics supports improved availability of emergency contraception, including full support of over-the-counter access.

ABORTION

- One of the most ethically challenging and controversial issues is providing confidential care to the pregnant adolescent considering an abortion.
- Minors have the legal right to obtain an abortion without parental involvement or consent unless state law requires such involvement.
- Most adolescents considering abortion actively involve their parents or other trusted adults in their decision-making process.

- Parental involvement in an adolescent's important medical decisions might be desirable or even ideal, but legally required involvement may cause a significant obstruction to seeking care.
- The American Academy of Pediatrics recommends that an adolescent should involve a parent or other trusted adult in the decision-making process, but the adolescent's right to confidential care must be respected.

DRUG USE AND DRUG SCREENING

- There is a clear medical indication to perform testing for illicit drugs as part of the diagnostic evaluation of a patient with an altered level of consciousness.
- Drug testing may also be required if a patient is in need of mental health services and needs to be referred to the most appropriate treatment program or facility, but such testing should only be done with the consent of the patient, unless the patient is under an involuntary psychiatric hold.
- Sometimes drug testing may be required by law enforcement, but physicians should only participate in criminal investigations as required by law or court order.
- A parent may request drug testing if they suspect their child has been using drugs, but there may be a conflict between the interests of the patient from the patient's and parent's perspective.
- It might be appropriate to test a young child without their assent but with a parent's permission, but an older adolescent should only be tested with fully informed consent.
- All tests have false-positives and false-negatives, and issues related to sensitivity, specificity, and pretest probability require careful interpretation of a test result that might lead to misinterpretation of reality.
- The American Academy of Pediatrics opposes involuntary screening of older adolescents on the basis of parental request and also opposes commercially available home drug testing or testing as a condition for participation in sports or any other school function.

CONCLUSION

- Providing emergency medical care to children embodies many of the core ethical values of medicine, including providing care to anyone in need; despite their perceived need at presentation, immigration status, insurance, or ability to pay.
- Ethical challenges unique to pediatric emergency medicine present an opportunity and need for ongoing

exploration and discussion with emergency medicine physicians and pediatricians.

- Children have an evolving capacity to take part in health care decisions, and by adolescence, they may be able to make decisions as well as adults.
- Keeping the best interests of the child as the central focus should help guide physicians through the most challenging cases.

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QUESTIONS

1. A mother brings a 6-month old to the ED with a chief complaint of a fever. The baby had ibuprofen at home and is nontoxic, afebrile, and has normal vital signs in triage. The most appropriate action is to
 - A. Reassure the mother that the baby is fine and send the baby home from triage.
 - B. Offer the mother the use of the phone to call the baby's doctor.

- C. Tell the mother to take the baby to the baby's doctor.
 - D. Triage the baby to be seen in the ED following standard triage protocol.
 - E. Provide the mother with a list of community urgent care centers and recommend that she take the baby to one of them.
2. A child of migrant laborers presents to the ED with an asthma exacerbation. The child has no local physician and is out of his asthma medications. The most appropriate action is to
 - A. Refer the patient from triage to a nearby clinic with a large immigrant patient population.
 - B. Provide the family with a list of services and clinics available to immigrant families.
 - C. Triage the baby to be seen in the ED following standard triage protocol.
 - D. Ask for documentation of legal immigrant status.
 - E. Report the family to the Department of Homeland Security, Immigration Health Services.
3. A 2-year old is brought to the ED by emergency medical services. The police were investigating a domestic violence report and noted that the child had multiple bruises. You are worried that the child may have serious injuries. A parent accompanying the child refuses all medical treatment and diagnostic testing. The most appropriate initial action is to
 - A. Contact the hospital's legal department for permission to treat the patient.
 - B. Perform an appropriate diagnostic evaluation and any necessary therapeutic interventions needed to assure the well-being of the child while simultaneously reporting the case to the local child protection agency.
 - C. Report the case to the local child protection agency and wait for authorization of treatment.
 - D. Obtain a court order to allow treatment.
 - E. Try to contact the other parent to get permission for treatment.
4. A child who appears to be about 10 years of age is brought in by emergency medical services after being hit by a car. He is unconscious, tachycardic, and hypotensive. His identity is unknown. The appropriate initial action is to
 - A. Perform any appropriate diagnostic and therapeutic procedures necessary to stabilize the child, including blood transfusion, invasive procedures, and surgery.
 - B. Obtain a court order to allow treatment.
 - C. Perform any appropriate diagnostic and therapeutic procedures necessary to stabilize the

- child, but do not perform blood transfusion, invasive procedures, or surgery until a parent can provide permission to treat.
- D. Maintain close contact with the investigating police to obtain permission from a parent to treat.
 - E. Contact the hospital's legal department for advice about what to do.
5. An adolescent presents with a disfiguring facial laceration that should be repaired. The patient has a severe needle phobia and refuses local anesthetic administration and suturing. The most appropriate approach is to
 - A. Honor the patient's right to refuse treatment and discharge her.
 - B. Give the patient a mild sedative and then obtain consent for repair.
 - C. Offer the patient procedural sedation or general anesthesia for repair.
 - D. Restrain the patient and perform the repair.
 - E. Give the patient a strong sedative and perform the repair.
 6. A 3-week old presents with a high fever. Your standard management would be to obtain blood for a complete blood count and culture, urine for urinalysis and culture by bladder catheterization, and cerebrospinal fluid for analysis and culture by lumbar puncture followed by intravenous antibiotics and admission. The parents refuse to allow a lumbar puncture to be performed but give permission for the other procedures, antibiotics, and admission. The most appropriate action is to
 - A. Obtain a court order to perform the lumbar puncture.
 - B. Take protective custody of the baby, report the parents to the local child, protection agency, perform the lumbar puncture, and then relinquish custody back to the parents.
 - C. Perform all diagnostic tests except the lumbar puncture and hold antibiotics until you can convince the parents to allow the lumbar puncture.
 - D. Ask the parents to leave the room while the blood and urine samples are obtained. Perform the lumbar puncture at the same time.
 - E. Perform all diagnostic tests except the lumbar puncture, administer antibiotics, admit the baby to the hospital, and ask the parents to continue to consider the lumbar puncture and discuss it with the inpatient service.
 7. A 2-year old is brought in pulseless and not breathing by emergency medical services after being found on the bottom of a swimming pool. The child was intubated and received four rounds of epinephrine via an intraosseous line. The child has been without a pulse with a flat electrocardiogram for 50 minutes. You decide to terminate resuscitation but there are several interns in the room with minimal procedural experience. The most appropriate action is to
 - A. Supervise an intern to check the endotracheal tube placement, remove the endotracheal tube, and replace it prior to terminating resuscitation.
 - B. Supervise two interns in performing a simple needle thoracostomy, once on each side prior to terminating the resuscitation.
 - C. Terminate the resuscitation and after breaking the news to the parents ask permission to allow the interns the opportunity to reinsert the endotracheal tube.
 - D. Dislodge the endotracheal tube and supervise an intern replacing it prior to terminating the resuscitation.
 - E. Supervise an intern in placement of an internal jugular intravenous line prior to terminating the resuscitation.
 8. A 15-year-old female presents with a complaint of dysuria and mild abdominal pain. When interviewed alone, she states she is sexually active with a single partner, but she does not want her mother to know about her sexual activity. Her urinalysis is normal, urine tests are sent for Chlamydia and gonorrhea, and she is treated presumptively with azithromycin and cefixime. The next day, her urine test is positive for Chlamydia. The most appropriate action is to
 - A. Recheck to make sure the patient was treated, with no further follow up.
 - B. Contact the patient via her cell phone and inform her of the test result and need for partner notification and testing.
 - C. Leave a message on the family's phone giving the information about the positive Chlamydia test and need for partner notification and testing.
 - D. Call the mother to tell her about the test result.
 - E. Report the case to the local child protection agency to investigate for possible sexual abuse.
 9. A 16-year-old female presents 12 hours after consensual intercourse. The condom broke and she is worried that she might become pregnant. The most appropriate action is to
 - A. Offer her testing and empiric treatment for sexually transmitted infections and emergency contraception.
 - B. Notify her parents to get permission for testing and empiric treatment for sexually transmitted infections and emergency contraception.

- C. Report the case to the local child protection agency to investigate for possible sexual abuse.
- D. Offer her testing and empiric treatment for sexually transmitted infections only.
- E. Refer her to her primary physician for follow up.
10. A 17-year-old male presents with his parents with a concern that he is acting sullen and defiant. They believe he may be using drugs and want him tested. When interviewed alone, he admits to drinking alcohol 3 weeks before and smoking marijuana 4 weeks before. The most appropriate action is to
- A. Advise the patient to consent for testing, as it will be negative and prove that he does not have any problems.
- B. Recommend that the parents obtain home drug testing kits available online.
- C. Tell the patient that you are worried about a medical cause of his behavior and that he needs tests to be sure he is OK. Do urine and blood testing for illicit drugs and let the parents know the results?
- D. Explain to the parents that teen boys are often sullen and defiant, but it is not usually simply due to illicit drug use. Refer them to their pediatrician or community family counseling services.
- E. Obtain a urine sample from the patient, do no testing, and tell the parents it was negative.
5. C. Adolescent refusal of care should be respected if the refusal would not subject them to a great risk of harm. Although sedation and general anesthesia may not be routine for an adolescent with a disfiguring facial wound, it would be appropriate in a case of severe needle phobia. This choice is the only one that allows for an optimal cosmetic outcome without violating the practice of informed consent.
6. E. In the event that a parent refuses a diagnostic test, the physician needs to review the remaining options and make therapeutic decisions based on the available data in a similar manner to what must be done if a test is attempted but not successful. Therapy for a potentially dangerous or progressive condition should not be delayed or forgone for lack of a complete diagnostic evaluation.
7. C. Even if practicing procedures on the newly dead is an excellent way for a medical trainee to learn and others will benefit from the physician's procedural skill in the future, it should only be done with permission from a parent.
8. B. Adolescents may seek confidential and independent health care for conditions defined by state law, including sexually transmitted infections, pregnancy, psychiatric complaints, and substance abuse problems. An acceptable and confidential method to provide the patient with test results should be established, include instructions for the adolescent to phone back for their test results, discussing the situation with the patient's primary care physician, or calling the patient on their personal phone.
9. A. Adolescents may seek confidential and independent health care for conditions defined by state law, including sexually transmitted infections, pregnancy, psychiatric complaints, and substance abuse problems. The American Academy of Pediatrics supports improved availability of emergency contraception, including full support of over-the-counter access.
10. D. A parent may request drug testing if they suspect their child has been using drugs, but there may be a conflict between the interests of the patient from the patient's and parent's perspective. The American Academy of Pediatrics opposes involuntary screening of older adolescents on the basis of parental request.

ANSWERS

1. D. Although the child appears well, no pediatric triage protocol has been demonstrated to identify all patients with a serious illness. No child should be turned away or denied care based on an initial triage assessment that the complaint is low acuity.
2. C. Care should be provided for the presenting complaints and, whenever possible, a referral should be made to an accessible source of ongoing primary care in the community. The American College of Emergency Physicians opposes federal and state initiatives that would require refusal of care to undocumented persons or reporting suspected undocumented persons to authorities.
3. B. When treating a patient that may have been abused, the physician must consider the best interests of the patient as the primary focus of the evaluation. A child with potentially serious injuries must be evaluated and treated immediately.
4. A. In an emergency, a societal standard of presumed or implied consent allows medical treatment to prevent harm based on the assumption that a per-

153 WITHHOLDING OR TERMINATING RESUSCITATION AND BRAIN DEATH

Howard Hast

WITHHOLDING OR TERMINATING RESUSCITATION

- There are certain circumstances when resuscitation in the emergency department (ED) may be withheld.
- A terminally ill patient who presents to the ED with an advance directive such as a DNR order may be treated but not resuscitated. The order must be signed by the patient's attending physician and legal guardian and should be reviewed by the attending physician in the ED with the patient's guardian. The parents or legal guardian may withdraw the DNR order at any time, including shortly after arrival in the ED.
- A terminally ill patient with no advance directives must be provided with advanced life support and resuscitation. Time and resources in the ED are insufficient to initiate the conversations that might result in a DNR order.
- Termination of resuscitation and declaration of death is appropriate when there is no return of spontaneous circulation. There are no absolute standards of care that dictate the duration of resuscitation in the pediatric population.
- Survival after out-of-hospital arrest is poor and has been correlated to the number of doses of Epinephrine administered. In several studies, there were no survivors if more than one dose of epinephrine was administered, and a larger number of investigations revealed no survivors if more than two doses of epinephrine were administered.
- In terms of duration of resuscitation, multiple studies have demonstrated few survivors in resuscitations lasting more than 20 to 30 minutes, while other investigations have demonstrated intact survival only in patients who received less than 15 minutes of resuscitation. Lacking universally accepted guidelines, the physician must determine the duration of resuscitation based upon the individual case.
- Hypothermic patients should have resuscitation continued until they have a body temperature of at least 35°C.
- A number of studies have demonstrated that family members typically prefer to be present during resuscitation, and that their grieving is less problematic if they were present during the resuscitation. There should be one staff member assigned to be present

with the family to answer questions and explain what is being done; the ED staff should be sensitive to the presence of family members during the resuscitation.

BRAIN DEATH

- Brain death cannot be determined in the ED because of the observation period required.
- In some cases, particularly in normothermic children who have the return of spontaneous circulation after a prolonged resuscitation, the children will subsequently progress to brain death. Brain death is defined as a total and irreversible loss of cerebral function, including the brain stem function.
- The most recent, widely accepted guidelines were penned in 1987 by The Task Force for the determination of brain death in children. These guidelines were a consensus opinion regarding the necessary clinical history, physical examination criteria, observation periods, and confirmatory laboratory tests required to determine brain death in children.
- The guidelines are summarized below
 - The *clinical history* must be consistent with the diagnosis of brain death.
 - The cause of coma should be determined whenever possible.
 - There must be no remediable or reversible conditions.
 - Confounding factors such as toxic and metabolic disorders, the presence of sedative-hypnotic drugs, paralytic agents, hypothermia, hypotension, and surgically remediable conditions must be eliminated prior to establishing a diagnosis of brain death.
- The *physical examination* is necessary to determine the failure of the brain function. The physical examination criteria must consider the following:
 - Coma and apnea must coexist.
 - A complete loss of consciousness, vocalization, and volitional activity.
 - Absent brain stem function.
 - Fixed and dilated or midposition pupils.
 - Absence of spontaneous eye movements.
 - Absent oculocephalic and oculovestibular reflexes.
 - Absent corneal reflexes.
 - No cough or gag; no sucking or rooting reflexes.
 - Respiratory movements absent without the ventilator.
 - Flaccid tone and absence of spontaneous or induced movements, excluding spinal cord events such as reflex withdrawal, or spinal myoclonus.
 - After meeting these criteria, apnea testing may be performed.

- The examination results should remain consistent with brain death throughout the *observation and testing period*.
- The observation period varies according to age:
 - For children aged 7 days to 2 months, two examinations, and EEGs separated by at least 48 hours.
 - For children aged 2 months to 1 year, two examinations, and EEGs separated by at least 24 hours. Repeat examination and EEG are not necessary if a concomitant cerebral radionuclide angiographic study demonstrates no visualization of cerebral arteries.
 - For children older than 1 year, two examinations should be performed at least 12 hours apart. No corroborative laboratory studies are necessary.
 - If hypoxic encephalopathy is present, observation for at least 24 hours is recommended. This may be reduced if an EEG shows electrocerebral silence or a radionuclide study is negative for cerebral blood flow.
 - Although it is beyond the scope of this chapter, there are occasions when supportive studies are useful if not potentially essential, for example, in fulminant cases of acute peripheral neuropathies, such as acute inflammatory demyelinating polyradiculopathy or infant botulism, where the physical examination may be consistent with brain death.
 - In many institutions, two examiners—typically from the pediatric neurology, pediatric neurosurgery, or pediatric critical care specialties—are necessary to complete a brain death determination.
 - Corroborative studies such as EEG or a radionuclide study are not mandatory in children older than 1 year of age when the physical examination and apnea testing are consistent with brain death.
 - Following the 1987 guidelines, different institutions may vary in minor degree as to the specifics of how brain death is determined. Within each institution it is essential that the same protocol be followed for each infant or child to avoid uncertainty or error in this most important determination.
 - Clinicians must devote the necessary time to explain the concept of brain death to the patient's family. Typically, multiple conversations during the observation and testing period are needed by most families to grasp and accept the concept that brain death is equivalent to cardiac death within our society.
 - During this time, extended family and institutional support should be organized to assist the child's family in coping with this tragic diagnosis and their profound loss.

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QUESTIONS

1. A 4-yo girl is brought to the ED in full cardiac arrest. She was an unbelted rear seat passenger in a rollover motor vehicle collision. She was ejected from the vehicle and sustained massive head and chest trauma. Which of the following statements is correct regarding terminating the resuscitation?
 - A. The patients should ideally receive a minimum of three rounds of Epinephrine and two rounds of atropine prior to termination the resuscitation.
 - B. It is standard of care to attempt resuscitation for a minimum of 30 minutes on a pediatric patient.
 - C. Termination of resuscitation efforts is appropriate when there is no return of spontaneous circulation.
 - D. The chances of survival after traumatic arrest is poor therefore resuscitation should be terminated upon arrival to the ED.
 - E. The length of the resuscitation should take into consideration whether family members are present in the ED.
2. A 15-yo boy sustained massive head injuries while snowboarding at a nearby ski resort. He was intubated in the field and transported by EMS to your ED. Upon arrival to the ED, he has a Glasgow Coma Scale of 3. He has no corneal reflexes and his pupils are fixed and dilated. Which of the following statements would apply to this patient?
 - A. It is appropriate to declare the patient brain dead.
 - B. Contact the pediatric neurosurgeon to complete a brain death determination in the ED.
 - C. Obtain a stat bedside EEG to corroborate your suspicion of brain death.
 - D. Brain death cannot be determined in the ED because of the observation period required.
 - E. Since the cause of the coma is easily determined, the consensus guidelines allow for the brain death determination to be made at the bedside.

ANSWERS

1. C. Termination of resuscitation and declaration of death is appropriate when there is no return of spontaneous circulation. There are no absolute standards of care that dictate the duration of resuscitation in the pediatric population. Survival after out-of-hospital arrest is poor and has been correlated to the number of doses of Epinephrine administered.
2. D. Brain death cannot be determined in the ED because of the observation period required. The task force for the determination of brain death in children. Provide a consensus opinion regarding the necessary clinical history, physical examination criteria, observation periods, and confirmatory laboratory tests required to determine brain death in children. The *clinical history* must be consistent with the diagnosis of brain death, the cause of coma should be determined whenever possible, and there must be no remediable or reversible conditions.

INDEX

Page numbers followed by *f* or *t* indicate figures or tables, respectively.

- A**
- Abdominal pain
 - diagnostic test, 437
 - gynecologic causes of
 - ectopic pregnancy, 439
 - ovarian cysts, 439
 - ovarian torsion, 439
 - pelvic inflammatory disease, 439
 - history, 437
 - inflammation, 437
 - nonsurgical causes of
 - acute gastroenteritis, 438
 - bleeding and pain, 438
 - colic, 438
 - constipation, 438
 - urinary tract infections, 438
 - peritoneal irritation, 437
 - Abdominal trauma, 193
 - Abscess, 25*f*, 109
 - Abuse, drugs of, 692
 - Acetaminophen
 - clinical presentation, 643
 - laboratory studies, 643
 - management
 - antidote, 644
 - gastrointestinal decontamination, 644
 - pathophysiology, 643
 - pharmacology, 643
 - Achilles reflexes, 22
 - Acquired coagulopathies
 - laboratory findings, 595
 - Acute abdominal conditions
 - obstructions
 - acute appendicitis with perforation, 43
 - incarcerated hernias, 43
 - intraabdominal sepsis, 43
 - intussusception, 42
 - malrotation with midgut volvulus, 42
 - necrotizing enterocolitis, 44
 - pyloric stenosis, 42
 - spontaneous peritonitis, 43
 - Acute ankle injury, 237*f*
 - Acute appendicitis
 - with perforation, 43
 - without perforation, 43
 - Acute bilirubin encephalopathy, 57
 - Acute chest syndrome
 - laboratory evaluation, 588
 - treatment, 588
 - Acute hemarthrosis
 - treatment, 593
 - Acute hemorrhage, 365
 - Acute inflammatory demyelinating polyradiculoneuropathy (AIDP), 355
 - Acute pelvic pain
 - etiology, 563
 - management, 563
 - Acute septic arthritis
 - clinical presentation, 613
 - diagnostic evaluation, 613
 - differential diagnosis, 613
 - physical examination, 613
 - radiographic imaging, 613
 - treatment, 613
 - Acute splenic sequestration crisis (ASSC)
 - laboratory evaluation, 589
 - treatment, 589
 - Acute suppurative tenosynovitis
 - management, 616
 - Acyclovir, 8, 9
 - Adjunctive therapy, 51
 - Adolescent pregnant patient
 - deep vein thrombosis, 555
 - eclampsia, 553
 - ectopic pregnancy, 555
 - emergency contraception, 556
 - HELLP syndrome, 553
 - hydatidiform mole, 556
 - preeclampsia, 553
 - trauma in pregnancy, 556
 - vaginal bleeding, 554
 - Adrenal insufficiency
 - clinical presentation, 462
 - differential diagnosis, 463
 - disposition, 463
 - epidemiology, 462
 - etiology, 462
 - laboratory findings, 462
 - management, 463
 - pathophysiology, 462
 - Afebrile seizure, 29, 36
 - Airway management, 119
 - Alanine aminotransferase (ALT), 198
 - Albumin, 600
 - Altered mental status
 - diagnostic testing, 23
 - disposition, 25
 - history, 22
 - pathophysiology, 22
 - physical examination, 22
 - special consideration
 - congenital adrenal hyperplasia, 26
 - hypoglycemia, 26
 - inborn errors of metabolism, 26
 - intussusception, 26
 - lead encephalopathy, 26
 - reye's syndrome, 26
 - therapy, 23
 - Ambient temperature, 9
 - American Academy of Pediatrics, 1
 - American College of Emergency Physicians, 1
 - Amoxicillin, 6
 - Amphetamines
 - clinical manifestations, 700
 - diagnostic testing, 700
 - disposition, 701
 - hyperthermia, 701
 - hyponatremia, 701
 - management, 701
 - pharmacology, 700
 - Analgesia, 99
 - Anaphylaxis
 - allergens, 435
 - ancillary tests, 435
 - disposition, 436
 - presentation, 435
 - treatment, 435
 - Anatomy, 84
 - Ancillary testing, 89
 - Anemia, 581
 - Anesthesia, 211
 - Aneurysmal bone cysts (ABCS), 631
 - Anorexia, 43
 - Antibiotic coverage, 4
 - Antibiotic therapy, 5
 - Antiepilepsy Drugs (AEDs)
 - carbamazepine, 32*t*
 - levetiracetam, 32*t*
 - primidone, 32*t*
 - valproic acid, 32*t*
 - Antipyretic analgesics, 643
 - Antipyretics, 9
 - Anxiolysis, 101
 - Aplastic crisis
 - laboratory evaluation, 589
 - Apparent life-threatening event (ALTE)
 - disposition, 18
 - epidemiology, 17
 - etiology, 17
 - evaluation, 18
 - initial assessment and stabilization, 17
 - management, 18
 - physical examination, 18
 - Arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD), 345

- Arterial blood gas (ABG)
 acid–base balance of the body, 13
 blood gas exchange in the lungs, 13
 electrolyte levels, 13
- Arterial ischemic strokes (AIS), 372
- Arterial thromboembolism (ATE), 335–336
- Arteriovenous malformation (AVM), 372
- As low as reasonably achievable (ALARA), 2
- Aspirin
 clinical presentation, 646
 laboratory studies, 646
 management, 647
 pathophysiology, 646
 pharmacokinetics, 646
- Asthma
 clinical presentation, 258–260
 differential diagnosis, 260–261
 disposition/outcome, 264
 etiology/pathophysiology, 257
 laboratory and radiographic findings, 260
 treatment
 anticholinergics, 263
 corticosteroids, 263
 heliox, 264
 intubation, 264
 magnesium, 264
 mechanical ventilation, 264
- Asystole, 139
- Ataxia
 acute ataxia, 349
 acute cerebellar ataxia, 349
 chronic nonprogressive ataxia, 351
 chronic progressive ataxia, 350
 evaluation, 348
 intermittent/episodic ataxia, 350
 pathophysiology, 347
 physical examination, 348
 tumors, 349
- Atelectasis, 14
- Atlanto-axial rotatory subluxation (AARS),
 181–182
- Atlantoaxial injuries, 178
- Atlantooccipital dislocation, 178
- Atonic seizures, 28
- Atypical antipsychotics
 aripiprazole, 684
 clozapine, 684
 olanzapine, 684
 paliperidone, 684
 quetiapine, 684
 risperidone, 684
 ziprasidone, 684
- Autoimmune hepatitis
 chronic progressive inflammatory
 disorder, 62
- Avulsion injuries, 220
- Axis fractures, 179
- B**
- Bacteremia, 10
- Bag-valve-mask (BVM) ventilation, 128
- Barbiturates, 103
- Bartholin abscess
 clinical presentation, 563
 etiology, 563
 management, 563
- Behavioral pediatric pain management, 111
- Bell's palsy, 357
- Benign childhood epilepsy, 28, 37
- Benign familial neonatal convulsions, 31f
- Benign myoclonic epilepsy, 31f
- Bilious emesis, 47, 67
- Bimodal positive airway pressure (BiPAP), 129
- Bioterrorism
 anthrax, 426
 botulism, 427
 plague, 426
 smallpox, 427
- Bladder injuries, 203
- Bleeding disorders, 592
- Blood component therapy, 599
- Blood culture, 5t
- Blood urea nitrogen (BUN), 501
- Blunt abdominal trauma, 196
- Bone, nonmalignant tumors of, 628
- Brachioplexus injuries, 221
- Brain death, 832
- Brain Dysfunction, 24t
- Breast milk jaundice, 57
- Bronchiolitis
 clinical presentation, 269
 differential diagnosis, 270
 disposition/outcome, 272
 etiology, 269
 pathophysiology, 269
 radiographic findings, 269
 treatment, 271
- Bronchopulmonary dysplasia (BPD),
 differential diagnosis, 289
 disposition, 290
 etiology/pathophysiology
 infection, 288
 inflammation, 288
 malnutrition, 288
 mechanical ventilation, 288
 oxygen therapy, 288
 laboratory and radiographic findings, 289
 treatment, 289
- Broselow tape and carts, 2
- Brugada syndrome, 344
- Burns
 clinical evaluation, 750
 diagnostic studies, 751
 disposition, 752
 etiology, 750
 management, 750
 pathophysiology, 750
- C**
- C-reactive protein (CRP) level, 70
- Calcium channel blockers
 laboratory evaluation, 673
 Management, 673
 pathophysiology, 672
 pharmacokinetics, 672
 pharmacology, 672
- Carbon monoxide poisoning
 clinical presentation, 688
 disposition, 689
 laboratory studies, 688
 pathophysiology, 688
 treatment, 688
- Cardiac syncope, 344
- Cardiac tamponade, 188
- Cardiac transplant patients, 305
- Cardiopulmonary resuscitation, 18, 136
- Cardiotoxins
 β-adrenergic blocking agents
 clinical presentation, 671
 disposition, 672
 laboratory evaluation, 671
 management, 671
 pathophysiology, 671
 pharmacokinetics, 671
 pharmacology, 671
- Cardiovascular deterioration, 16
- Cardiovascular emergencies, 297
- Cardiovascular injuries, 188
- Catheterized specimen, 5t
- Causes of neonatal seizures
 drug withdrawal, 33t
 hemorrhage, 33t
 hereditary disorders, 33t
 infection, 33t
 structural anomalies, 33t
- Caustics
 alkali burns, 660
 diagnosis, 660
 hydrofluoroic acid, 661
 laboratory and radiology studies, 660
 management, 660
 pathophysiology, 660
 presentation, 660
 special concerns
 button batteries, 661
 caustic eye injuries, 661
- Central nervous system emergencies, 608
- Cephalohematoma, 58
- Cerebral edema, 169
- Cerebral palsy (CP)
 clinical presentation, 370
 complications, 370
- Cerebral sinovenous thrombosis (CSVT), 372
- Cerebrospinal fluid shunts, 11t
- Cerebrovascular syndromes
 diagnosis
 diagnostic evaluation, 374
 history, 372
 physical examination, 374
 treatment, 375
- Cervical lymphadenopathy, 89
- Cervical spine, 162
- Chemical weapons
 nerve agents, 724
 sulfur mustard, 725
- Chest pain
 clinical presentation, 38
 diagnostic evaluation, 38
 differential diagnosis
 cardiac, 38
 gastrointestinal, 39
 idiopathic, 39

- musculoskeletal, 39
 - psychogenic, 39
 - respiratory, 39
 - management, 38
 - Chest radiograph, 5*t*
 - Child abuse, 193
 - Child death in emergency department
 - after interview, 805
 - caring for ED staff, 806
 - interview, 804
 - miscarriage, 806
 - Childhood cancer, common complications of
 - hypercalcemia, 605
 - tumor lysis syndrome (TLS), 605
 - Children with special health care needs (CSHCN), 105
 - Children, mild head injury in, 74
 - Chlamydia infection
 - topical erythromycin ointment, 6
 - Chloroquine, 722
 - Cholinergic crisis, 356
 - Chronic bilirubin encephalopathy (Kernicterus), 57
 - Chronic toxicity, 687
 - Clavicle fractures
 - medial clavicle fractures, 223
 - Clinical pharmacologist, 1
 - Clostridium difficile*, 50
 - Clostridium perfringens*, 49
 - Cocaine toxicity
 - clinical manifestations, 696
 - diagnosis, 696
 - disposition, 696
 - management, 696
 - pathophysiology, 696
 - pharmacology, 695
 - coelenterates. *See under* Marine envenomations
 - Cold illness
 - diagnosis, 764
 - hypothermia, 763
 - pathophysiology, 763
 - Coma, 21–27
 - Common allergic presentations
 - allergic rhinitis, 433
 - clinical presentation
 - allergic conjunctivitis, 431
 - differential diagnosis, 432
 - pathophysiology, 431
 - treatment, 432
 - Common parasitic infections
 - arthropods
 - pediculosis, 415
 - scabies, 415
 - cestodes
 - flatworms, 414
 - tapeworms, 414
 - nematodes
 - ascariasis, 411
 - enterobiasis, 411
 - hookworms, 412
 - trichinosis, 411
 - trichuris trichiura, 411
 - protozoa
 - entamoeba histolytica*, 414
 - giardia lamblia*, 414
 - pneumocystis, 414
 - trematodes (flukes), 412
 - Common pediatric malignancies, 604
 - acute leukemias, 602
 - anaplastic large cell lymphoma (ALCL), 603
 - central nervous system tumors, 603
 - hodgkin disease, 603
 - non-hodgkin lymphomas (NHL), 603
 - Wilms' tumor, 604
 - Complete blood count (CBC), 5*t*
 - Complete metabolic panel, 5*t*
 - Computed tomography (CT), 23, 87, 183
 - Concussion, 220
 - Congenital adrenal hyperplasia (CAH), 26, 93
 - Congenital defects, 16
 - Congenital heart disease
 - epidemiology, 297
 - evaluation, 297
 - hperoxia test, 298
 - other ancillary tests, 298
 - physical examination, 298
 - physiology, 297
 - fetal circulation, 297
 - neonatal circulation, 297
 - Congenital long QT syndrome (LQTS), 326
 - Congenital masses, 88
 - Congenital short QT syndrome, 344
 - Congestive heart failure (CHF)
 - laboratory testing, 310
 - management, 310
 - physical examination, 310
 - signs and symptoms, 310
 - Conjugated hyperbilirubinemia, 59
 - Continuous positive airway pressure (CPAP), 129
 - Corneal reflex, 24*t*
 - Croup, 10
 - Crying infant
 - differential diagnosis, 65
 - physical examination, 64
 - Cryoprecipitate, 599
 - Cyanide poisoning
 - clinical presentation, 710
 - disposition, 711
 - laboratory evaluation, 710
 - pharmacopathology, 710
 - treatment, 710
 - Cyanotic breath-holding spells, 29
 - Cyanotic cardiac lesions, 91
 - Cyclosporine, 97
 - Cystic fibrosis
 - clinical presentation, 292
 - differential diagnosis, 293
 - etiology/pathophysiology, 292
 - laboratory and radiologic findings, 292
- D**
- Deep sedation, 103
 - Deep vein thrombosis (DVT), 335
 - Delirium
 - delusions, 22
 - disorientation, 22
 - fearful responses, 22
 - hallucinations, 22
 - irritability, 22
 - sensory misperception, 22
 - Dengue fever, 420
 - Dermacentor andersoni*, 408
 - Dermatologic emergencies, 507
 - Diabetic ketoacidosis (DKA), 23*t*
 - Diagnostic evaluation, 39
 - Diaphoresis, 9
 - Diaphragmatic hernia, 149
 - Diaphyseal clavicle fractures, 223
 - Diarrheal dehydration, 13
 - Digoxin
 - antidotal therapy, 675
 - clinical presentaion, 674
 - diagnosis, 675
 - disposition, 675
 - management, 675
 - pathophysiology, 674
 - pharmacology, 674
 - Disability, 159
 - Dislocations
 - proximal interphalangeal (PIP) joint, 220
 - Disorder of glucose metabolism
 - diabetic ketoacidosis, 457
 - clinical manifestations, 457
 - epidemiology, 457
 - laboratory studies, 457
 - management
 - fluid resuscitation, 458
 - insulin therapy, 458
 - phosphate, 458
 - potassium, 458
 - sodium, 458
 - pathophysiology, 457
 - Disrhythmias in children
 - fast rates
 - atrial flutter and fibrillation, 324
 - paroxysmal supraventricular tachycardia, 323
 - premature ventricular contractions, 324
 - ventricular fibrillation, 325
 - ventricular tachycardia, 324
 - slow rates
 - atrioventricular blocks, 323
 - pacemakers in children, 323
 - sinus bradycardia, 322
 - Disseminated intravascular coagulation (DIC), 583, 595
 - Distal radius and ulna fracture, 228
 - Drowning
 - epidemiology, 747
 - management, 748
 - pathophysiology, 747
 - prognosis, 748
 - Dysbaric injuries
 - barotrauma, 771
 - decompression sickness, 773
 - dysbarisms, 774
 - pathophysiology, 771
 - physical gas laws, 770-771
 - presentation, 771
 - Dysfunctional uterine bleeding
 - clinical presentation, 575

- Dysfunctional uterine bleeding (*Cont'd.*)
 diagnostic evaluation, 576
 etiology, 575
 management, 577
 pathophysiology, 574
 physical examination, 576
- Dysmenorrhea
 clinical presentation, 573
 diagnostic evaluation, 573
 etiology, 572
 management, 573
 pathophysiology, 572
 treatment, 573
- E**
- Ear and nose emergencies
 acute otitis externa, 527
 acute otitis media, 527
 epistaxis, 528
 foreign body of nose and ear, 528
 mastoiditis, 527
 sinusitis, 529
- Echinoderms. *See under* Marine envenomations
- Effective screening tools
 infrared, 9
 liquid crystal, 9
- Eisenmenger syndrome, 304
- Elbow, 224
- Electrical injuries
 anatomic sites of injury, 756
 electrophysiology, 755
 management
 emergency department care, 756
 prehospital care, 756
 mechanisms of injury, 756
 prevention, 757
- Electrocardiograms (ECGs), 40
- Electroencephalogram, 20
- Electrolytes, 5*t*
- Elliptocytosis (HE), 583
- Emergency department (ED), 1, 9, 38, 55
- Emergency medical services for children (EMSC), 809
- Encephalopathy, 26
- Enchondromas, 630
- End-tidal capnometer (ETCO₂), 100
- Endotracheal intubation, 151
- Entameba histolytica*, 50
- Enteroinvasive bacteria, 49
- Enteroviruses
 clinical findings, 521
- Enthesitis-related-arthritis, 623
- Environmental emergencies, 727
- Environmental poisons, 710
- Eosinophilia, significance of, 419
- Eosinophilic esophagitis (EE), 447
- Epidemiology, 4
- Epiglottitis, 254
- Epilepsy, 23*t*
- Epistaxis. *See under* Ear and nose Emergencies
- Erythema multiforme, 81*f*
- Erythrocyte sedimentation rate (ESR), 70
- Erythromycin, 5
- Escherichia coli*, 4, 10, 49
- Esophagitis, 609
- Ethanol. *See under* Household chemicals
- Ethical considerations
 adolescent issues
 abortion, 828
 chaperones and physical examination, 828
 confidentiality, 827
 drug screening, 828
 emergency contraception, 828
 child abuse, 826
 medically underserved children, 825
 parental and patient refusal, 826
 practicing procedures on
 newly dead, 827
 triage and overcrowding, 825
- Ethylene glycol. *See under*
 Household chemicals
- Eutectic mixture of local anesthetics (EMLA), 107
- Excisional biopsy, 90
- Extra-hepatic biliary obstruction, 61
- Eye emergencies
 physical examination
 anterior segment, 545
 cellulitis, 546
 common eye complaint, 545
 conjunctivitis, 548
 episcleritis, 546
 intraocular pressure, 545
 lids and orbit, 544
 nontraumatic eye disorders, 545
 ophthalmia neonatorum, 546
 posterior segment, 545
 pupils and extraocular movements, 545
 scleritis, 546
 vision, 544
- Eye trauma
 epidemiology, 539
 history, 539
 medications, 541
 physical examination, 539
 specific injuries, 541
- F**
- Facial laceration, 209
- Febrile or septic appearing neonate
 epidemiology, 4
 evaluation, 5
 fever and sepsis, 4
 sepsis workup, 5
 treatment, 5
- Febrile seizures, 31*f*, 33
- Fecal leukocytes, 52
- Feeding disorders
 gastroesophageal reflux
 treatment, 54
- Fever
 bacteremia, 10
 management, 11
 physical examination, 10
 presentation and history, 10
 sepsis, 10
- Fifth disease (erythema infectiosum)
 clinical findings, 518
 complications, 518
 epidemiology, 518
- Flail chest, 188
- Flexion, abduction, and external rotation (FABER) test, 73, 74
- Fluid and electrolyte disorders
 calcium
 hypercalcemia, 484
 hypocalcemia, 484
 potassium
 hyperkalemia, 480
 hypokalemia, 482
 sodium
 hyponatremia, 478
 hyponatremia, 478
- Fluoroscopy, 116–117
- Focused abdominal sonography for trauma (FAST), 113, 163, 203
- Foot fractures, 237
- Forensic documentation, 823
- Fresh frozen plasma, 599
- Frontal sinus, 211
- Frostbite
 treatment, 765
- G**
- Gamma-hydroxybutyrate
 antidotal therapy, 704
 decontamination, 703
 diagnosis, 703
 disposition, 704
 enhanced elimination, 704
 pharmacology, 703
 treatment, 703
- Gastric decontamination
 activated charcoal, 639
 antidotes, 640
 cathartics, 639
 enhanced elimination, 640
 gastric lavage, 638
 hemodialysis and hemoperfusion, 640
 induction of emesis, 638
 whole bowel irrigation, 639
- Gastroenteritis
 diagnostic evaluation
 diagnostic studies, 50
 physical assessment, 50
 etiology, 49
 pathophysiology, 50
 treatment
 adjunctive therapy, 51
 oral rehydration therapy, 50
 prevention, 52
- Gastroenteritis, prevention of, 52
- Gastroesophageal reflux (GER)
 complications, 447
 differential diagnosis, 446
 evaluation, 446
 treatment, 447
- Gastrointestinal (GI) bleeding
 diagnostic testing, 441
 factitious bleeding, 441
 historical features, 440
 lower gastrointestinal bleeding, 442
 physical examination, 441
 upper gastrointestinal bleeding, 441
- Gastrointestinal emergencies, 437

- Gastrointestinal foreign bodies
 anatomic sites for obstruction, 450
 colonic/rectal foreign bodies, 451
 diagnosis, 450
 etiology, 450
 small bowel, 451
 surgical approach, 451
 therapeutic options, 451
- Gastrointestinal foreign bodies, 44
- Gastroschisis, 149
- Genitourinary emergencies, 493
- Genitourinary trauma, 202
- Glasgow coma scale (GCS), 22
- Group A streptococci (GAS), 392
- Group B *Streptococcus*, 4, 7
- Grunting
 atelectasis, 13
 pneumonia, 13, 18
 pulmonary edema, 13
- Guillain–barré syndrome (GBS), 355
- Gynecologic disorders
 of infancy, childhood and adolescence
 evaluation, 560
 physical examinations, 561
- H**
- H influenzae*, 10
- Hand and wrist injuries, 229
- Head trauma
 anatomy, 168
 pathophysiology, 168
 specific injuries, 168
- Head trauma, 119
- Headache
 Cluster headaches, 364
 evaluation, 362
 laboratory studies, 362
 Migraine headaches, 363
 neuroimaging, 362
 primary headaches, 363
- Heart rate, 8
- Heat and cold illness, 762
- Heat illness
 diagnostic testing, 763
 heat stroke, 763
 management, 762
 manifestations, 762
 pathophysiology, 762
 risk factors, 762
 signs and symptoms, 763
- Hematologic and oncologic emergencies, 581
- Hematologic complications
 anemia, 606
 hemorrhage, 606
 hyperleukocytosis, 606
- Hematoma, 25*f*, 184
- Hemolytic uremic syndrome (HUS), 438, 596
- Hemophilia, 592–593
- Hemothorax, 185
- Hepatosplenomegaly, 24*t*
- Hereditary spherocytosis (HS), 583
- Herpes simplex
 ancillary tests, 520
 clinical findings, 520
 complications, 520
 epidemiology, 520
 management, 520
- Herpes simplex, 4
- Herpes zoster (shingles)
 clinical findings, 519
 complications, 519
 treatment, 519
- High altitude illness
 acute mountain sickness, 768
 altitude related syndromes, 769
 atmosphere and gas laws, 767
 high altitude cerebral edema, 768
 high altitude pulmonary edema, 768
 physiologic responses, 768
 preexisting disease, 769
- High anion gap metabolic acidosis, 13
- Highly active antiretroviral therapy (HAART), 400
- Hip joints, 69
- Hirschsprung disease, 46*f*, 44, 48
- Hollow organs, 199
- Household chemicals
 ethanol
 clinical presentation, 652
 disposition, 652
 laboratory, 652
 management, 652
 pathophysiology, 652
 pharmacokinetics, 652
- ethylene glycol
 clinical presentation, 654
 laboratory, 654
 management, 654
 pathophysiology, 654
 pharmacokinetics, 654
- isopropanol
 clinical presentaiton, 655
 disposition, 656
 laboratory, 655
 management, 656
 pathophysiology, 655
 pharmacokinetics, 655
- methanol
 clinical presentaiton, 653
 disposition, 654
 laboratory, 653
 management, 653
 pathophysiology, 653
 pharmacokinetics, 653
- Human and animal bites
 antibiotics, 728
 epidemiology, 727
 history, 727
 physical examination, 727
 rabies prophylaxis, 728
 tetanus prophylaxis, 729
 wound care, 727
- Human immunodeficiency virus (HIV)
 infection, 86
- Humerus fractures
 proximal humerus fracture, 224
- Hydrocarbons
 classification, 662
 clinical presentation, 662
 disposition, 662
 management, 662
 pathophysiology, 662
 properties, 662
- Hydrocephalus, 368
 clinical presentation, 368
 management, 368
- Hyperbilirubinemia, 58
- Hyperpyrexia, 9, 13
- Hypersensitivity, 54
- Hypertensive encephalopathy, 365
- Hyperthyroidism
 clinical presentation, 466
 differential diagnosis, 466
 disposition, 467
 epidemiology, 465
 etiology, 465
 management, 466
 neonatal thyrotoxicosis, treatment of, 467
 pathophysiology, 465
- Hypertrophic cardiomyopathy (HCM), 303
 hypertrophic obstructive cardiomyopathy (HOCM), 303
 idiopathic hypertrophic subaortic stenosis (IHSS), 303
- Hypocalcemia. *See under* Fluid and electrolyte disorders
- Hypoglycemia
 diagnostic evaluation, 459
 disposition, 460
 pathophysiology, 459
 signs and symptoms, 459
- Hypoplastic left heart syndrome (HLHS), 302
- Hypothermia *See under* Cold illness, 18
- I**
- Idiopathic, 29, 39
- Idiopathic central apnea, 18
- Idiopathic intracranial hypertension, 365
- Idiopathic neonatal hepatitis, 60
- Imaging, 99, 112
- Imidazoline decongestants, 723
- Immune globulins, 600
- Immune thrombocytopenic purpura (ITP), 596
- Immunologic emergencies, 431
- Imported diseases in travelling child
 chemoprophylaxis, 417
 immunizations, 417
 incubation periods, 417
 physical signs and symptoms, 417
 possible pathogens, 417
 travel locations, 417
- Inborn errors of metabolism (IEM)
 diagnostic testing, 487
 newborn screening, 489
 presentation, 486
 treatment, 489
- Incarcerated hernias, 43
- Infancy and childhood, disorders of
 congenital vaginal obstruction, 561
 labial adhesions, 561
 neonatal physiologic variants, 561

- Infant
febrile, 9
Septic appearing, 9
- Infant rashes
benign infant rashes, 523
disorders of pigmentation, 524
vascular lesions, 524
- Infection
hospitalization, 588
laboratory evaluation, 588
treatment, 588
- Infection prophylaxis, 98
- Infectious emergencies, 379
- Infectious musculoskeletal disorders, 613
- Inflammatory and infectious heart disease
acute rheumatic fever, 319
endocarditis, 319
myocarditis
clinical presentation, 317
diagnosis, 318
etiology, 317
treatment, 318
pericarditis, 317
- Inflammatory bowel disease related
arthropathies, 624
- Inflammatory musculoskeletal disorders, 619
- Influenza, 10t
clinical presentation, 379
epidemiology and transmission, 379
laboratory and radiographic findings, 382
pathophysiology, 379
prevention, 385
treatment, 383
- Insidious meningitis, 388
- Interfacility transport
altitude physiology, 813
communication, 812
equipment, 812
legal considerations, 812
medication, 812
modes of transport, 813
regulatory requirements, 813
safety, 813
specific clinical transport issues, 813
stabilization, 812
- Intervertebral diskitis
clinical manifestations, 616
diagnostic testing, 616
treatment, 616
- Intraabdominal sepsis, 43
- Intracranial hemorrhage, 593
- Intracranial pressure (ICP), 23, 103
- Intramuscular bleeding, 593
- Intubation, 123
- Iron
clinical manifestations, 707
diagnosis, 707
management, 707
pathophysiology, 707
pharmacology, 707
- Iron deficiency, 581
- Irritability, 5
- Isoniazid toxicity
clinical presentation, 686
diagnosis, 686
disposition, 687
pathophysiology, 685
pharmacology, 685
treatment
antidotal therapy, 686
decontamination, 686
enhanced elimination, 686
stabilization, 686
- Isopropanol. *See under* Household chemicals
- J**
- Jaundice
pathophysiology, 56
- Jefferson fracture, 178
- Juvenile idiopathic arthritis, 624
- Juvenile myoclonic epilepsy
(Janz syndrome), 28
- K**
- Kawasaki disease (KD)
ancillary data, 398
cardiovascular complications, 398
differential diagnosis, 398
etiology and pathogenesis, 396
management, 398
phases of
acute or febrile phase, 396
subacute phase, 397
prognosis, 399
- Ketamine, 103
- Knee and patella dislocations, 235
- L**
- L monocytogenes* infection, 4
- Laceration, 108, 184
- Laryngeal mask airways (LMAs), 146
- Lateral condyle (capitellum), 226
- Lead poisoning, 582
lead sources, 705
management, 706
pathophysiology, 705
pharmacokinetics, 705
- Legg–Calvé–perthes disease (LCPD), 72
- Lennon gastaut syndrome, 36
- Leptomeningeal cyst, 174
- Lesions
diagnostic features of, 80
morphology of, 80
- Lethal toxins
benzocaine exposure
clinical signs and symptoms, 722
treatment, 722
camphor ingestion
clinical symptoms, 721
management of camphor ingestion, 721
lomotil
initial manifestations, 722
management, 722
- Lethargy, 21
- Leukocyte count, 5t
- Ligamentous injuries
anterior cruciate ligaments (ACL), 220
anterior talofibular ligaments (ATFL), 220
calcaneofibular ligaments (CFL), 220
- Lightning injuries
management
disposition, 759
emergency department care, 758
prehospital care, 758
prevention, 759
sequelae, 759
mechanisms of injury, 757
types of injuries, 757
- Limping child
evaluation, 70
microbiology, 71
physical examination
ankle/foot, 70
hip joints, 69
knee, 70
sacroiliac joints, 69
spine, 69
radiographic analysis, 70
bone scan, 71
CT scan, 71
MRI, 71
ultrasound, 71
- Listeria monocytogenes*, 4
- Live attenuated (LAIV) influenza, 385
- Liver, 198
- Liver and gallbladder disease
biliary atresia, 454
biliary tract disease, 454
cholecystitis, 454
choledochal cysts, 454
fulminant hepatic failure, 453
gallbladder hydrops, 454
hepatitis, 453
hyperbilirubinemia, 452
neonatal hyperbilirubinemia conjugated, 453
neonatal hyperbilirubinemia unconjugated, 452
- Liver function tests, 5t
- Long QT syndrome, 344
- Low reticulocyte counts, 584
- Lumbar puncture, 5t, 7
- Lymphadenitis, 10t
- M**
- Magnetic resonance imaging (MRI), 89, 188
- Malaria, 420
- Male genitourinary problems
balanitis and balanoposthitis, 495
epididymitis, 493
Henoch–schönlein purpura, 495
hydrocele, 495
inguinal hernia, 494
paraphimosis, 495
phimosi, 495
priapism, 496
scrotal swelling, 495
scrotal trauma, 494
testicular torsion, 493
testicular tumors, 494
varicocele, 495
- Marine envenomations
coelenterates
disposition, 743

- management, 743
 - pathophysiology, 743
 - echinoderms
 - clinical presentation, 745
 - disposition, 745
 - management, 745
 - pathophysiology, 745
 - sea snakes
 - clinical presentation, 745
 - disposition, 745
 - management, 745
 - venomous fish
 - clinical presentation, 744
 - disposition, 745
 - management, 744
 - pathophysiology, 744
 - Mass casualty management
 - epidemiology, 815
 - mass casualty events, 815
 - Massive Transfusions, complications of
 - infectious complications, 601
 - Maxillofacial trauma
 - associated injuries, 208
 - emergency management, 208
 - etiology, 208
 - history, 208
 - incidence, 208
 - physical examination, 208
 - Meconium, 148
 - Medial epicondylar fracture, 226
 - Medical malpractice, 823
 - Medico-legal considerations
 - emergency medical treatment and labor act (EMTALA), 821
 - health insurance portability and accountability act (HIPAA), 821
 - Megacolon, 45
 - Meningitis, 609
 - antibiotic treatment, 389
 - corticosteroid treatment, 390
 - differential diagnosis, 389
 - management of stable patients, 389
 - management of unstable patients, 389
 - pathophysiology, 388
 - prevention, 388
 - sequelae, 390
 - Meningitis, 10*t*, 365
 - Meningococemia, 82*f*
 - Metabolism, inborn error of, 26
 - Metacarpal fractures, 230
 - Methanol. *See under* Household chemicals
 - Methicillin resistant Staph aureus (MRSA), 12, 13
 - Methyl salicylate, 723
 - Microcytic anemia, 581
 - Mild head injury in children
 - anatomic considerations, 74
 - assessment, 74
 - biomechanical considerations, 74
 - disposition, 75
 - history, 74
 - pathophysiology, 74
 - physical examination, 75
 - Moderate sedation, 103
 - Mononuclear pleocytosis, 5
 - Motor response, 22*t*
 - Moyamoya, 375
 - Multiorgan dysfunction syndrome (MODS), 10
 - Multiple-trauma patient, 153
 - Muscular dystrophies
 - progressive degeneration of muscle, 357
 - Musculoskeletal tumor, 604
 - Mushroom poisoning
 - allenic norleucine group, 713
 - classification, 712
 - coprine group, 712
 - cyclopeptide group, 712
 - gastrointestinal irritant group, 713
 - gyromitrin group, 712
 - ibotenic acid and muscimol group, 712
 - muscarine group, 712
 - myotoxin group, 713
 - orellanine group, 713
 - psilocybin group, 713
 - Myalgias, 11*t*
 - Myasthenia crisis, 356
 - Myasthenia gravis, 356
 - Mycophenolate, 97
 - Myocardial contusion, 188
 - Myoclonic seizures, 28
 - Myopathies, 357
- N**
- Nasal fractures, 209
 - Nasopharyngeal specimen, 5
 - Neck masses
 - anatomy, 84
 - ancillary testing, 89
 - assessment, 85
 - congenital masses, 88
 - differential diagnosis, 86
 - disposition, 89
 - inflammatory conditions, 86
 - neoplastic masses, 89
 - traumatic conditions, 88
 - Neck trauma, 119
 - Necrotizing enterocolitis (NEC), 44, 48
 - Needle cricothyrotomy with transtracheal jet ventilation (TTJV)*, 158
 - Needle thoracostomy, 189
 - Neisseria gonorrhoeae*, 6
 - Neonatal emergencies, 91
 - Neonatal resuscitation, 147
 - pharmacologic agents, 147
 - vascular access, 147
 - volume expansion, 148
 - Neonatal seizures, 30, 31*f*
 - Neonate
 - febrile, 4, 5
 - septic appearing, 4
 - Neoplastic masses, 89
 - Nephrotoxicity, 98
 - Neuroblastoma, 604
 - Neuroleptic malignant syndrome
 - clinical presentation, 683
 - disposition, 683
 - management, 683
 - Neuroleptics
 - clinical presentation, 682
 - management, 682
 - pathophysiology, 681
 - Neurologic emergencies, 341
 - Nitrous oxide, 101
 - Noncardiac syncope, 345
 - Noncritical care setting, 9
 - Noninvasive mechanical ventilation (NIPPV), 129
 - Nonpolio enterovirus, 4
 - Nonpulmonary diseases
 - DKA, 13
 - shock, 13
 - Nonspecific vulvovaginitis.
 - clinical presentation, 565
 - etiology, 565
 - management, 565
 - Nonsteroidal anti-inflammatory drugs
 - clinical presentation, 649
 - laboratory studies, 649
 - management, 649
 - Nontraumatic bone and joint disorders, 613
 - Normal anion gap metabolic acidosis
 - hypernatremic dehydration, 14
 - rapid volume expansion, 14
 - renal tubular acidosis, 14
 - Normal Cerebrospinal Fluid (CSF), 389*t*
 - Normal peripheral perfusion
 - capillary refill, 10
 - Normal saline (NS), 160
 - Normocytic anemia, 583
- O**
- Oligoarticular arthritis, 625
 - Omphalitis, 6, 9
 - Omphalocele, 149
 - Oncologic emergencies, 602
 - Ophthalmologic emergencies, 539
 - Opioids
 - clinical presentation, 693
 - disposition, 694
 - laboratory and diagnostic testing, 693
 - management, 694
 - pathophysiology, 692
 - pharmacology, 692
 - toxicokinetics, 692
 - Oral antibiotics, 9
 - Oral cavity and neck, emergencies of
 - dentoalveolar infections, 532
 - dentoalveolar trauma, 531
 - oral piercings, 534
 - oral soft tissue, infections of, 532
 - oropharyngeal trauma, 531
 - peritonsillar abscess, 533
 - pharyngitis, 532
 - retropharyngeal abscess, 534
 - Oral rehydration therapy (ORT), 50
 - Orbital fractures, 210
 - Organ donation, 191
 - Organophosphates and carbamates
 - clinical presentation, 658
 - laboratory studies, 658
 - pathophysiology, 657
 - treatment, 658
 - Orthopedic injuries, 213
 - Osteochondromas (Cartilaginous exostoses), 629

- Osteoid osteomas, 628
- Osteomyelitis
- chronic recurrent multifocal osteomyelitis, 615
 - clinical manifestations, 615
 - diagnostic testing, 615
 - etiology, 614
 - sequelae, 615
 - treatment, 615
- Otitis media, 10*r*
- Otolaryngologic emergencies, 527
- Ovarian torsion
- clinical presentation, 563
 - etiology, 562
 - management, 563
- Overuse syndromes, 220
- Oxygen administration, 120
- P**
- Packed red blood cells, 599
- Pain assessment, 107
- Pain management, 211
- procedural pain management, 107
- Painful stimulus, 22
- Pancreas, 198
- Pancreatitis, 609
- Paradoxical irritability, 68
- Paraphimosis reduction, 109
- Paratyphoid enteric fevers, 423
- Paroxysmal supraventricular tachycardia (PSVT), 139, 143
- Patent ductus arteriosus (PDA), 135
- Pediatric Advanced Life Support (PALS), 1
- Pediatric airway anatomy, 119
- Pediatric Assessment Triangle (PAT)
- appearance, 1
 - circulation to skin, 1
 - effort of breathing, 1
- Pediatric cervical spine injury
- anatomy and physiology, 175
 - epidemiology, 175
 - evaluation and management, 175
- Pediatric HIV patient in ED
- clinical presentations, 401
 - complications, 402
 - diagnosis, 401
 - epidemiology, 400
 - postexposure prophylaxis, 404
- Pediatric hypertension
- assessment
 - history, 329
 - physical examination, 330
 - testing, 330 - treatment, 331
- Pediatric patient, 1
- Pediatric prehospital care, 809
- anticipatory guidance, 809
 - legal issues, 810
 - office preparedness, 809
 - prehospital professional, 809
 - transfer considerations, 810
- Pediatric procedures
- papoose board, 4
- Pediatric trauma systems, 154
- Pelvis and lower extremities, injuries of
- osgood-schlatter apophysitis, 235
 - femoral shaft fractures, 234
 - hip dislocations, 234
 - Knee and patella, 234
 - pelvic fracture, 233
 - proximal femur fractures, 234
- Penile injuries, 205
- Pericardiocentesis, 190
- Periodic paralysis, 357
- Peripheral vascular resistance, 9
- Pertussis
- clinical presentation, 283
 - diagnostic evaluation
 - differential diagnosis 283
 - laboratory diagnosis, 283 - pathophysiology, 282
 - treatment, 283
- Petechiae and purpura
- by sepsis, 507
 - clinical course, 507
 - complications, 507
 - treatment, 507 - henoch-schonlein purpura, 508
 - idiopathic thrombocytopenic purpura, 509
- Phencyclidine & ketamine
- clinical presentation, 699
 - diagnosis, 699
 - disposition, 699
 - pharmacology, 699
- Phenobarbital, 35, 36
- Physical injuries, 214
- Physiologic heat loss
- conduction, 9
 - convection, 9
 - evaporation, 9
 - radiation, 9
- Pityriasis rosea
- clinical presentation, 521
- Platelet concentrate, 599
- Platelet disorders, 595
- Platelet dysfunctions, 595
- Pneumonia
- causes of, 276
 - clinical presentation, 276
- Pneumothorax, 14, 149, 184
- Poisonous plants
- epidemiology, 714
 - identification of plants, 714
 - psychoactive plants, 718
 - specific toxic plants
 - gastrointestinal irritants, 715
 - mucosal irritants, 715
 - systemic toxins, 715
 - ackee (*blighia sapida*), 718
 - anticholinergic, 715
 - cardiac glycosides, 716
 - colchicine, 717
 - poison hemlock (*Conium maculatum*), 717
 - sodium channel, 716
 - solanine, 716
 - tobacco (*nicotiana sp.*), 717
 - toxalbumins, 717
 - water hemlock (*Cicuta sp.*), 717
 - yew (*taxus sp.*), 716
- Poliomyelitis, 358
- Polyarticular arthritis, 624
- Polymorphisms, 16
- Positive end-expiratory pressure (PEEP), 121, 184
- Positive pressure ventilation (PPV), 158
- Postconcussion syndrome, 75
- Postnatal age, 59*f*
- Posttransplant lymphoproliferative disease (PTLD), 97
- Preadarrest dysrhythmias, 138
- Prescription drugs
- antidepressants
 - atypical antidepressants
 - clinical manifestations, 679
 - diagnosis, 679
 - pathophysiology, 679
 - pharmacology, 679
 - treatment, 679 - selective serotonin reuptake inhibitors
 - diagnosis, 678
 - disposition, 679
 - management, 679
 - pathophysiology, 678
 - pharmacology, 678 - tricyclic antidepressants
 - clinical manifestations, 678
 - diagnosis, 678
 - disposition, 678
 - management, 678
 - pathophysiology, 678
 - pharmacology, 678
- Priapism, 589
- Procalcitonin, 5*r*
- Procedural sedation, 101
- Prolonged QT syndrome, 17
- Prothrombin time (PT), 593
- Pruritic rashes
- atopic dermatitis
 - diagnostics, 511
 - treatment, 511 - contact dermatitis
 - treatment, 512 - erythema multiforme, 513
 - papular urticaria, 512
 - pediculosis
 - treatment, 512 - scabies
 - treatment, 512 - urticaria
 - Acute urticaria, 513
 - Chronic urticaria, 513
- Pseudoseizure, 36
- Psoriatic arthritis, 624
- Psychiatric emergencies
- aggressive patient
 - chemical restraint agents, 801
 - conversion/somatization disorder
 - etiology, 801
 - management, 801
 - pathogenesis, 801
 - recognition, 801
 - treatment, 801

- posttraumatic stress disorder (PTSD)
 etiology, 800
 management, 800
 pathogenesis, 800
 recognition, 800
 psychopathic patient
 etiology, 801
 pathogenesis, 801
 psychotic patient
 ancillary studies, 800
 etiology, 800
 management, 800
 pathogenesis, 800
 suicidal patient
 ancillary studies, 799
 etiology, 799
 management, 799
 pathogenesis, 799
 recognition, 799
 Psychogenic headaches, 365
 Psychosocial emergencies
 abuse and neglect
 abusive head injury, 795
 ancillary studies, 796
 cutaneous injuries, 794
 differential diagnosis, 796
 epidemiology, 793
 etiology, 793
 management, 797
 medical history, 793
 musculoskeletal trauma, 795
 physical examination, 794
 visceral trauma, 795
 sexual abuse
 ancillary studies, 790
 epidemiology, 789
 etiology, 789
 management, 790
 pathogenesis, 789
 recognition, 789
 Pubertal vaginal bleeding
 precocious puberty, 562
 urethral prolapse, 562
 vaginal foreign body, 561
 Pulmonary contusion, 184
 Pulmonary embolism (PE), 335
 Pulmonary lesions, 91
 Pulse oximetry reading, 3
 Purified protein derivative (PPD)
 tuberculin test, 89
 Pyloric stenosis, 42
 Pyomyositis, 358
Q
 Quetiapine, 682
R
 Radial head subluxation, 227
 Radiation emergencies
 External contamination, 783
 general procedures, 783
 internal contamination, 784
 prognosis, 784
 radiation accident
 plant, 782
 radiation injuries
 exposure, 780
 special considerations, 784
 types of radiation
 ionizing radiation, 778
 nonionizing radiation, 779
 Rapid antigen test, 11*f*
 Rapid sequence intubation (RSI) method, 121
 Rash, 79
 Reactive arthritis, 624
 Renal injuries, 203
 Rescue breathing, 120
 Respiratory, 39
 Respiratory distress
 clinical presentation, 13
 differential diagnosis, 14
 disposition, 14
 laboratory and radiographic findings, 13
 pathophysiology, 13
 treatment, 14
 Respiratory emergencies, 253
 Respiratory failure
 anatomy, 127
 assisted ventilation
 advanced airway management, 128
 bag-valve-mask ventilation, 128
 noninvasive mechanical
 ventilation, 128
 laboratory studies, 128
 physical examination, 128
 physiology, 127
 Respiratory syncytial virus, 11*t*
 Retinal hemorrhages, 29
 Retinoblastoma, 604
 Retrograde urethrogram (RUG), 161
 Retropharyngeal abscess, 255
 Return to play (RT), 76
 Rheumatic fever
 carditis, 622
 clinical features, 621
 diagnostic evaluation, 622
 differential diagnosis, 622
 erythema marginatum, 622
 joint, 621
 nodules, 622
 treatment, 623
 Rhinovirus, 11*t*
 Rib fractures, 187
 Rickets
 breast-feeding, 469
 clinical manifestations, 470
 decreased sunlight exposure, 468
 diagnosis, 470
 etiology, 468
 imaging, 472
 laboratory evaluation, 471
 monitoring, 472
 pathophysiology, 469
 prevention, 473
 treatment, 472
 vitamin D deficiency, 468
 Ringer's lactate (LR), 160
 Risk factor for thromboembolism, 335
 Rodenticides
 clinical findings, 664
 developmental considerations, 664
 disposition, 667
 laboratory and diagnostic testing, 664
 management, 665
 pathophysiology, 664
 pharmacology, 664
 toxicology, 664
 Roseola (exanthem subitum)
 clinical findings, 517
 complications, 517
 epidemiology, 517
 Rotavirus, 11*t*
 Rubella (German measles)
 clinical findings, 517
 complications, 517
 epidemiology, 517
 management, 517
 Rubeola (measles)
 clinical findings, 516
 complications, 516
 epidemiology, 516
 management, 516
S
 S pneumoniae, 10
 Sacroiliac joints, 69
 Scarlet fever
 ancillary tests, 521
 clinical findings, 521
 complications, 521
 management, 521
 pathophysiology, 521
 Scrotal trauma, 205
 Sea snakes. *See under* Marine envenomations
 Second impact syndrome (SIS), 75
 Secondary headaches
 brain tumors, 364
 hydrocephalus, 364
 Sedation agent, 123
 Seizure
 classification
 febrile seizures
 disposition, 34
 laboratory evaluation, 33
 therapy, 34
 first afebrile seizure
 differential diagnosis, 29
 history, 29
 laboratory evaluation, 30
 physical examination, 29
 radiologic evaluation, 29
 neonatal seizures
 laboratory evaluation, 31
 treatment, 33
 differential diagnosis, 29
 Selected injuries, management of
 abrasions, 246
 consultation guidelines, 249
 ear lacerations, 247
 fingertip injuries, 247
 forehead lacerations, 247
 lip lacerations, 247
 puncture wounds, 248
 scalp lacerations, 247
 Sepsis evaluation, 4

- Septic appearance, 4
 Septic arthritis, 10*t*
 Septic shock, 12
 Serious bacterial infection (SBI), 4, 10, 13
 Serum aspartate aminotransferase (AST), 198
 Sexually transmitted diseases (STD)
 cervicitis, 570
 chlamydia
 clinical presentation, 567
 diagnosis, 567
 etiology, 567
 treatment, 567
 Epididymitis
 clinical presentation, 570
 diagnosis, 570
 etiology, 570
 treatment, 570
 genital warts
 clinical presentation, 567
 diagnosis, 567
 etiology, 567
 treatment, 567
 gonorrhea
 clinical presentation, 569
 diagnosis, 569
 etiology, 567
 treatment, 569
 herpes simplex
 clinical presentation, 570
 diagnosis, 570
 treatment, 570
 HIV and AIDS
 clinical presentation, 570
 diagnosis, 570
 etiology, 570
 treatment, 570
 pelvic inflammatory disease (PID)
 clinical presentation, 570
 diagnosis, 570
 etiology, 570
 management, 571
 treatment
 inpatient, 570
 outpatient, 570
 syphilis
 clinical presentation, 569
 diagnosis, 570
 etiology, 569
 treatment, 570
 trichomoniasis
 clinical presentation, 569
 diagnosis, 569
 etiology, 569
 treatment, 569
 urethritis, 570
 Shock
 cardiogenic shock, 132
 distributive shock, 130
 hypovolemic shock, 130
 obstructive shock, 132
 Sickle cell disease (SCD), 587
 laboratory evaluation, 587
 treatment, 587
 vasoocclusive crisis, 587
 Sickle cell pain, 110
 Skin closure
 mattress stitches, 244
 running stitch, 244
 simple interrupted stitch, 244
 Skin mottling, 5
 Snake envenomations
 coral snakes
 clinical presentation, 734
 disposition, 734
 management, 734
 epidemiology, 730
 exotic snakes, 734
 pit vipers
 anatomy, 731
 antivenom therapy, 732
 clinical presentation, 731
 diagnostic studies, 732
 disposition, 733
 hospital management, 732
 pathophysiology, 731
 prehospital management, 731
 Soft tissue injury and wound repair
 history and physical examination, 241
 management
 antiseptic and scrubbing, 242
 debridement, 243
 foreign body evaluation, 243
 hemostasis, 242
 irrigation, 243
 local anesthesia, 242
 nerve block, 242
 sedation, 242
 Specific renal syndromes
 acute glomerulonephritis, 500
 acute renal failure, 503
 etiology, 503
 laboratory evaluation, 503
 hemolytic uremic syndrome
 (HUS), 502
 nephrotic syndrome, 501
 Spider and arthropod bites
 bees and vespids
 dispositions, 740
 management, 739
 pathophysiology, 739
 black widow spiders
 anatomy, 736
 clinical presentation, 736
 disposition, 736
 management, 736
 pathophysiology, 736
 brown recluse spiders
 anatomy, 737
 clinical presentation, 737
 disposition, 738
 management, 737
 pathophysiology, 737
 hymenoptera, 738
 imported fire ants
 clinical presentation, 740
 management, 741
 pathophysiology, 740
 scorpions
 anatomy, 738
 clinical presentation, 738
 management, 738
 pathophysiology, 738
 tarantulas, 738
 Spinal cord injury, 176*t*
 Spinal cord injury without radiographic
 abnormality (SCIWORA), 162, 180
 Spondyloarthropathies, 623
 Spontaneous peritonitis, 43
Staphylococcus aureus, 6, 10
 Status epilepticus, 34
 Sternocleidomastoid muscle, 84
 Stevens–Johnson syndrome, 83
 Stomatitis, 10*t*
 Stool culture, 5*t*
 Stroke (Cerebrovascular accidents)
 initial management, 588
 treatment, 588
 Subaxial cervical spine injuries, 179
 Subcutaneous hemorrhage, 593
 Sudden infant death syndrome (SIDS)
 disposition, 17
 epidemiology, 16
 evaluation, 16
 management, 16
 pathology, 16
 pathophysiology, 16
 Superficial skin infections
 fungal infections
 tinea capitis infection, 515
 tinea corporis infections, 515
 impetigo, 515
 staphylococcal scalded skin syndrome, 515
 Superior mediastinal syndrome (SMS), 607
 Superior vena cava syndrome (SVCS), 607
 Supracondylar humerus fractures, 225
 Supraventricular tachycardia (SVT), 65
 Symptomatic bradycardia, 139
 Syncope
 history, 341
 laboratory studies, 342
 pathophysiology, 341
 physical examination, 341
 Systemic immune response syndrome (SIRS),
 10, 12
 Systemic lupus erythematosus (SLE)
 clinical presentation, 620
 diagnostic evaluation, 620
 differential diagnosis, 620
 prognosis, 621
 treatment, 621
 Systemic onset disease
 complications, 625
 diagnosis, 625
 diagnostic evaluation, 626
 differential diagnosis, 626
 prognosis, 625
 treatment, 626
- T**
 Tachycardia, 9, 12
 Tachypnea, 9, 12
 Tacrolimus (FK506), 97
 Telecanthus, 213
 Tension-type headaches, 364
 Thalassemia, 581

- The phalanges, fracture of, 229
 Therapy, 34
 Thermoregulation, 9
 Thoracic trauma, 182
 Thromboembolic disease, 335
 Thyrotropin receptor-stimulating antibodies (TRSAbs), 465
 Tibia and fibula fractures, 236
 Tick paralysis, 355
 Tick-borne infections
 babesiosis, 408
 colorado tick fever, 408
 human granulocytic anaplasmosis, 408
 human monocytic ehrlichiosis (HME), 408
 lyme disease, 407
 rocky mountain spotted fever, 407
 tularemia, 408
 Total body water (TBW), 475
 Toxic appearing infants, 65
 Toxic megacolon, 48
 Toxic shock syndrome
 clinical manifestations, 392
 differential diagnosis, 393
 epidemiology, 392
 etiology and pathogenesis, 392
 management, 393
 recurrences, 393
 Toxicologic emergencies
 diagnostic aids and laboratory, 635
 epidemiology, 635
 history, 635
 management
 stabilization, 636
 physical examination, 635
 Transcutaneous devices (TcB), 58
 Transfusion, complications of
 acute hemolytic transfusion reaction (AHTR), 600
 allergic transfusion reaction, 600
 delayed hemolytic transfusion reactions (DHTR), 600
 febrile nonhemolytic transfusion reactions (FNHTR), 600
 Transient erythroblastopenia of childhood (TEC), 583
 Transient synovitis
 clinical presentation, 619
 diagnostic evaluation, 619
 differential diagnosis, 619
 physical examination, 619
 prognosis, 619
 radiographic imaging, 619
 treatment, 619
 Transient synovitis (TS), 69
 Transplant patient, 96
 transverse myelitis, 355
 Trauma, 153
 Trauma scores, 165
 Traumatic asphyxia, 186
 Traumatic diaphragmatic hernia, 187
 Traumatic esophageal rupture, 187
 Travellers' diarrhea (TD)
 foodborne infections, 422
 waterborne infections, 422
 Treatment for pneumothorax, 293
 Trichinosis, 358
 Trivalent inactivated (TIV) Influenza, 385
 Tube thoracostomy, 189
 Tuberculosis, 86
 Tularemia, 428
 Tympanic, 9
 Typhlitis, 609
 Typhoid fevers (TF), 423
- U**
 Uncomplicated vaginal delivery, 7
 Unconjugated hyperbilirubinemia, 56
 Unicameral bone cysts, 630
 Upper airway emergencies
 pathophysiology
 lower airway considerations, 253
 upper airway considerations, 253
 signs of respiratory distress, 253
 specific clinical scenarios
 bacterial tracheitis, 255
 croup, 254
 epiglottitis, 254
 foreign body aspiration, 255
 peritonsillar abscess, 255
 retropharyngeal abscess, 255
 Upper extremities, injuries of
 carpal fractures, 231
 diaphyseal clavicle fractures, 223
 diaphyseal radius and ulnar fractures, 228
 dislocations
 carpometacarpal joint, 231
 distal interphalangeal joint, 231
 metacarpophalangeal joint, 231
 proximal interphalangeal joint, 231
 medial clavicle fractures, 223
 Uremia, 231
 Ureteral injuries, 203
 Urethral catheterization, 109
 Urinalysis, 51, 7
 Urinary tract diseases
 urinary tract infection, 498
 urolithiasis, 498
 Urinary tract infection, 4
- V**
 Vaginitis
 candidal vulvovaginitis, 566
 garnereella vaginitis, 566
 nonspecific vulvovaginitis, 565
 parasitic vulvovaginitis, 566
 shigella vaginitis, 566
 Varicella, 101
 Varicella (chicken pox)
 complications, 518
 epidemiology, 518
 management, 519
 Venocclusive disease, 609
 Venomous fish. *See under* Marine envenomations
 Venous thromboembolism
 clinical presentation, 336
 complications, 337
 imaging, 337
 laboratory testing, 337
 treatment, 337
 Venous thrombotic events (VTE), 335
 Ventilation bag, 120
 Vesicular rashes, 5
 Viral hemorrhagic fever, 428
 Viral infections
 herpes simplex virus (HSV) infections, 607
 varicella zoster virus (VZV) infections, 607
 Viral myositis, 358
 Vomiting, life threatening cause of
 diabetic ketoacidosis, 491
 inborn errors of metabolism, 491
 increased intracranial pressure, 491
 reye's syndrome, 491
 toxic ingestions, 491
 Von willebrand factor (vWf)
 clinical manifestations, 594
 laboratory findings, 594
- W**
 Weakness
 diagnosis
 history, 354
 laboratory evaluation, 355
 physical examination, 354
 West Nile virus, 358
 West syndrome (infantile spasms), 28
 Wheezing represents airway obstruction, 13
 Whole blood, 599
 World Health Organization (WHO), 50
- X**
 X-ray detectors, 114
- Y**
Yersinia pestis, 427
 Yew (*Taxus* sp.), 716
- Z**
 Ziprasidone, 105