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PREP & Pediatric in Review (PIR) Content Specifications

The PREP covers all Content Specifications over a 5-year period. Thus, the material presented in PREP The Curriculum covers approximately 20% of the Content Specifications each year in either the PREP Self-Assessment or PIR. Therefore, in any 5-year continuous cycle, PREP The Curriculum covers the vast majority of these knowledge statements and provides participants with an educational program that is ideal for achieving lifelong learning.

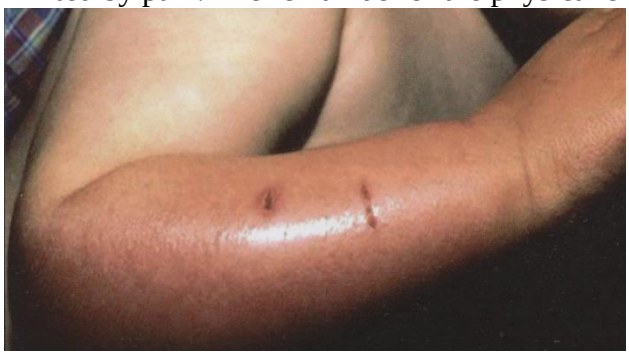
Core Competency Icons

Six core competencies considered to be the foundation of high-quality medical care.

1. **I-C:** Interpersonal and Communication Skills result in effective information exchange and teaming with patients, families, and other health professionals
2. **P:** Professionalism manifested through a commitment to professional responsibilities, adherence to ethical principles, and sensitivity to a diverse patient population
3. **PBLI:** Practice-Based Learning and Improvement involves investigation and evaluation of one's own patient care, appraisal, and assimilation of scientific evidence, and improvements of patient care
4. **SBP:** Systems-Based Practice demonstrates an awareness of and responsiveness to the larger context and system of health care and effectively calls on system resources to provide care that is of optimal value
5. **S:** Safety
6. **TE:** Interdisciplinary Teams

Item 1

A 1-year-old boy with no drug allergies has pain, swelling, and erythema 10 hours after being bitten on the forearm by his pet cat. His temperature is 38°C and vital signs are normal for his age. On physical examination, there is an area of erythema, swelling, and tenderness on the posterior aspect of the right forearm (Item Q1). You note 2 puncture sites in the center of this area. Flexion and extension of the elbow and wrist are mildly limited by pain. The remainder of the physical examination findings is normal.



ITEM Q1: Findings as described for the boy in the vignette.

Of the following, the MOST appropriate antimicrobial agent for this patient is

A. amoxicillin-clavulanate
B. azithromycin
C. clindamycin
D. dicloxacillin
E. levofloxacin

Item 1 Preferred Response: A

The patient described in the vignette has rapid onset of cellulitis and low-grade fever after a cat bite. The most appropriate antimicrobial agent for this patient is amoxicillin-clavulanate because it has excellent activity against Pasteurella multocida, a likely pathogen that causes infection after dog bites, cat scratches, or cat bites. The organism is present as part of the normal oral flora in up to 90% of cats and 50% of dogs. Pasteurella can also colonize the upper respiratory tract of livestock and poultry. Transmission from animals to humans has occurred in the absence of a bite or scratch through respiratory tract spread. Vertical transmission from mother to neonate is also possible, as is horizontal spread from colonized persons.

The onset of symptoms in cases of Pasteurella infection is very rapid, usually within 24 hours and sometimes as early as 3 hours after a cat bite. This rapid onset may distinguish Pasteurella infection from that caused by other pathogens, such as group A Streptococcus or Staphylococcus aureus, which often take 1 to 2 days to develop. For patients with cellulitis due to Pasteurella, significant pain and swelling at the site of the bite are common, and sanguinopurulent drainage occurs in approximately 40% of cases. Lymphangitis (20%) and regional lymphadenopathy (10%) can also occur. Osteomyelitis and septic arthritis commonly occur from direct inoculation of the organism; however, necrotizing fasciitis is rare. Pasteurella is a rare cause of bacteremia, meningitis, pneumonia, urinary tract infection, intraabdominal infection, endocarditis, or ocular infection.

Penicillin is the treatment of choice for Pasteurella, but rare isolates with β-lactamase activity have been reported. Amoxicillin-clavulanate has activity against β-lactamase-positive organisms and also provides coverage against oral anaerobes which may be of potential concern for the patient in the vignette. Azithromycin is an alternative for treating Pasteurella infection in patients with a serious penicillin allergy, but there are few clinical data regarding its use. Treatment failures with erythromycin, semisynthetic penicillins (eg, dicloxacillin), and clindamycin can occur because of their poor activity against Pasteurella multocida. Levofloxacin has activity against Pasteurella but is not approved for use in patients younger than 18 years with cellulitis.

PREP Pearls

- Skin and soft tissue infections due to Pasteurella multocida can occur after a dog or cat bite.
- Symptoms due to P multocida infection usually present within 24 hours of a bite.
- Penicillin is the treatment of choice for P multocida infection; amoxicillin–clavulanate adds additional coverage against possible anaerobes.

American Board of Pediatrics Content Specification (s):

- Know the mode of transmission of Pasteurella multocida
- Know the most common clinical manifestation of a Pasteurella multocida infection (ie, cellulitis at the site of an animal bite that develops within 24 hours of injury)

Suggested Reading:

- American Academy of Pediatrics. Pasteurella infections. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:542-543
- Stechenberg BW. Pasteurella multocida. In: Feigin RD, Cherry JD, Demmler-Harrison GJ, Kaplan SL, eds. Feigin and Cherry's Textbook of Pediatric Infectious Diseases. 6th ed. Philadelphia, PA: Saunders Elsevier; 2009:1615-1619

Item 2

You are seeing a 16-year-old girl for her annual health supervision visit. The girl has become a vegan, and her mother is concerned about possible nutritional deficiencies. The girl has no symptoms and has not lost any weight in the last 6 months. Her menses are regular and her physical examination findings are unremarkable.

Of the following, the girl's diet puts her at GREATEST risk of a deficiency of

A. calcium
B. iron
C. vitamin A
D. vitamin B12
E. vitamin D

Item 2 Preferred Response: D

As a vegan, the girl described in the vignette is most likely to develop a deficiency in B12, a vitamin almost exclusively found in animal products. Vegans require supplements or fortified food products to meet their need for this vitamin. Because the high intake of folic acid in vegan diets may mask the hematologic aspects of vitamin B12 deficiency, diagnosis may be delayed until neurologic symptoms occur. Other potential deficiencies associated with a vegan diet include calcium, iron, zinc, vitamin A and D, and perhaps other trace elements, but adequate intake can be ensured by following recommended vegan food group allowances and serving sizes.

Adolescents may be vegetarian to restrict their caloric intake. Although there are nutritional advantages to a vegetarian diet, such as decreased intake of protein, saturated fat, and cholesterol, along with higher amounts of fiber, magnesium, and vitamins C and E; vegans (those who eat no animal products, ie, meat, poultry, fish, seafood, eggs, and milk) are at risk for energy deficits and nutrient deficiencies.

Adolescents' nutritional needs are influenced by a number of factors. Biologic considerations include enhanced needs related to an increased growth rate and change in body composition; males have a greater blood volume and leaner body mass than females, who need a minimum fat mass for menstruation and reproduction. In addition to increased energy needs, growth in lean body mass will require increased intake of calcium, iron, nitrogen, zinc, magnesium, fluoride, and vitamins A, C, and D. Acute and chronic illness, trauma, and stress will add to the needs imposed by growth and physical activity.

Behavioral considerations during the adolescent years include changes in food habits related to levels of activity, changes in schedules, and a desire for independence, resulting in skipped meals, more frequent snacking, and more meals outside the home (including fast food). Last, dissatisfaction with body image may be associated with frequent dieting. Because boys usually eat more food than girls, their risk of deficiencies is reduced.

PREP Pearls

- Adolescents' eating habits put them at risk for nutritional deficiencies.
- Vegan diets pose potentially more serious health risks (deficiency of vitamin B12 but to a lesser extent vitamins A and D and calcium, iron, and zinc) compared with vegetarian diets.
- Vegans/vegetarians may have an underlying eating disorder.

American Board of Pediatrics Content Specification(s):

- Understand the potential nutritional deficiencies in adolescents

Suggested Reading:

- Adolescent nutrition. In: Pediatric Nutrition Handbook. 6th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2009:175-181
- Nutritional aspects of vegetarian diets. In: Pediatric Nutrition Handbook. 6th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2009:201-224
- Larson N, Neumark-Sztainer D. Adolescent nutrition. Pediatr Rev. 2009;30:494-496. doi:10.1542/pir.30-12-494
- Renda M, Fischer P. Vegetarian diets in children and adolescents. Pediatr Rev. 2009;30:el-e8. doi:10.1542/pir.30-1-el
- Messina V, Melina V, Mangels AR. A new food guide for North American vegetarians. J Am Diet Assoc. 2003;103:771-775. doi:10.1053/jada.2003.50141

Item 3

A 12-year-old Chinese-American boy complains of gas, bloating, and diarrhea for the past 3 months. He has no nocturnal symptoms and denies weight loss. His recent history is significant only for a family trip to mainland China to visit his grandparents 2 months ago. Physical examination demonstrates a well-developed, well-nourished adolescent. The remainder of the examination is unremarkable, except for a nontender, tympanitic, slightly distended abdomen with active bowel sounds. His stool is watery, however occult blood test results are negative and the pH is 5.0.

Of the following, the test that you are MOST likely to recommend is a

A. 72-hour fecal fat
B. lactose breath test
C. lactulose breath test
D. small intestinal biopsy for disaccharidase levels
E. stool for ova and parasites

Item 3 Preferred Response: B

The boy described in the vignette has of a 3-month history of watery diarrhea in the absence of weight loss or nocturnal or other constitutional symptoms. Although a recent trip abroad suggests the possibility of an acquired enteric infection, his symptoms began before he left the United States. Examination of a stool specimen demonstrates an acidic pH of 5.0 and no occult blood loss. In this scenario, a diarrheal state occurring as a consequence of dietary causes, especially carbohydrate malabsorption, is highly likely. Considering the young man's ethnic background (Item C3A), a diagnosis of hypolactasia (lactase deficiency) is suspected

Item C3A. Prevalence of Lactase Deficiency in Healthy North American Populations

Nationality	Population, %
Filipino	95
Asian (Far East)	85
African American	75
Mexican	75
American Indian	55
Puerto Rican	25
Whites	20

To confirm this diagnosis, the most effective and least invasive approach would be to perform a lactose breath hydrogen test.

Disaccharidases are glycoproteins that are abundant on the apical portion of the small intestinal enterocyte microvillus (brush-border) membrane. These enzymes cleave α-glycosidic linkages between component monosaccharides, thus permitting absorption via carrier-mediated brush-border transport. Four human disaccharidases have been identified: sucrase-isomaltase (hydrolytic products = glucose + fructose), lactase-phlorizin hydrolase (hydrolytic products = glucose + galactose), maltase-glucoamylase (hydrolytic products = glucose + glucose oligosaccharides), and trehalase (hydrolytic product = glucose). Disaccharidase deficiencies, either primary or secondary, represent the most common causes of carbohydrate malabsorption (Item C3B).

Item C3B. Major Causes of Carbohydrate Malabsorption

Primary Causes	Secondary Causes
• Lactase deficiency (hypolactasia) • Congenital sucrase-isomaltase deficiency • Glucoamylase deficiency • Congenital glucose-galactose malabsorption	• Diffuse vinous injury – Celiac disease - Malnutrition – Infection (rotavirus) – Allergic enteropathy • Short bowel syndrome • Antibiotics • Bacterial overgrowth • Iron deficiency

Symptomatic disaccharidase deficiency usually occurs within a few hours of ingesting the offending carbohydrate and may present clinically with nausea, abdominal pain, vomiting (particularly in young children), diarrhea, flatulence, and abdominal distension. The most common of these disorders is lactase deficiency (hypolactasia), which may be classified as acquired (secondary to intestinal injury) or genetically programmed, late primary deficiency. The latter classification is extremely common in individuals of non-Northern European origin, occurring in 75% of the world's population. Its prevalence in subjects of Asian (Far-Eastern) extraction is at least 85% (Item C3A). Late primary deficiency usually begins in the second half of the first decade of life but may not present until adolescence. Importantly, congenital lactase deficiency (an autosomal recessive disorder) is extremely rare.

The hydrogen breath test relies on the ability of colonic bacteria to hydrolyze unabsorbed carbohydrate, producing volatile organic acids, carbon dioxide, and hydrogen. An increase from the baseline breath hydrogen content of greater than 20 ppm within 2 hours of ingesting 2 g/kg (up to 50 g) of a disaccharide indicates malabsorption. A small percentage of healthy individuals (<5%) do not have a resident hydrogen-producing bacterial flora. In these cases, when lactase deficiency is suspected, direct enzyme measurement from an endoscopically obtained duodenal biopsy may be required to reach a definitive diagnosis.

Of the known disaccharidase deficiency states, sucrase-isomaltase deficiency represents the most common congenital disorder. It has a variable age of clinical presentation. Symptoms develop during early infancy in patients whose diet comprises formulas with carbohydrates other than lactose, especially soy and protein hydrolysate products containing Sucrose and/or glucose oligosaccharides

In infants who are either breastfed or consume cow milkbased formulas, symptoms are delayed until carbohydrates other than lactose are introduced to the diet. Sucrase-isomaltase deficiency is a rare disorder, with an overall prevalence of less than 0.2% in North America (2%-10% in Greenland Eskimos). As in suspected lactase deficiency, a hydrogen breath test (using sucrose as the testing substrate) represents the first-line diagnostic study.

Travel abroad should alert the clinician to suspect an acquired enteric infection (especially parasitic infestation) in a patient who presents with chronic diarrhea. However, because this patient's symptoms began before his trip to China, a stool test for ova and parasites would not be indicated as a first-line diagnostic study. Lactulose is a nonabsorbable disaccharide composed of galactose and fructose. A lactulose breath hydrogen test is useful in the evaluation of suspected bacterial overgrowth, particularly in patients who have undergone prior bowel surgery and present with symptoms that suggest a blind or stagnant intestinal loop. An early breath hydrogen peak, usually occurring within 30 minutes of lactulose ingestion, suggests overgrowth. Finally, in a patient whose symptoms are not accompanied by weight loss or other signs of nutritional compromise, a stool fat analysis would not be indicated.

PREP Pearls

- Watery, acidic stools are characteristic of carbohydrate malabsorption.
- Sucrase-isomaltase deficiency is the most common congenital disaccharidase deficiency.
- In the evaluation of suspected malabsorption, absence of weight loss suggests carbohydrate loss only.

American Board of Pediatrics Content Specification(s):

- Recognize the incidence of lactase and sucrase isomaltase deficiency in different ethnic groups

Suggested Reading:

- Gupta SK, Chong SKF, Fitzgerald JF. Disaccharidase activities in children: normal values and comparison based on symptoms and histologic changes. J Pediatr Gastroenterol Nutr. 1999;28:246-251
- Heyman MB. Lactose intolerance in infants, children and adolescents. Pediatrics. 2006;118:1279-1286. doi:10.1542/peds.2006-1721
- Lomer MC, Parkes GC, Sanderson JD. Review article: lactose intolerance in clinical practice—myths and realities. Aliment Pharmacol Ther.
- 2008;27:93-103. doi:10.1111/j.1365-2036.2007.03557.x
- Matthews SB, Waud JP, Roberts AG, Campbell AK. Systemic lactose intolerance: a new perspective on an old problem. Postgrad Med J. 2005;81:167-173. doi:10.1136/pgmj.2004.025551
- Robayo-Torres CC. Quezada-Calvillo R, Nichols BL. Disaccharidase digestion: clinical and molecular aspects. Clin Gastroenterol Hepatol. 2006;4:276-287. doi:10.1016/j.cgh.2005.12.023

Item 4

A newborn is determined to have Leber congenital amaurosis, which causes congenital blindness. There are no other abnormalities and no indication of central nervous system involvement. The parents ask how this diagnosis will affect their infant's development. Of the following, the MOST appropriate statement about development in this child is

A. language development will be persistently delayed
B. preserved hearing will allow this child to develop typically
C. reaching for objects is likely to be delayed
D. sitting independently is likely to be delayed
E. social development will be typical

Item 4 TE **Preferred Response: C**

Vision can be viewed as the coordinating sensory input for early development, and the ability to perceive people and objects in the environment through sight helps stimulate development not only in motor skills but also in the verbal and social realms. Congenital visual impairment, regardless of cause, influences when and how a child achieves normal milestones. Because blind children must use auditory or tac-tile input to explore their environment, they may have difficulty conceptualizing objects, may need to use trial and error to achieve milestones, and may not receive feedback from parents or others for social and verbal development. These differences in how they encounter the world are likely to contribute to the general developmental delay seen in children with visual impairment. These developmental delays may be compounded in the presence of neurologic deficits that are frequent comorbidities when visual impairment is associated with chromosomal, infectious, prematurity-related, or hypoxic-ischemic causes.
The child in the vignette has Leber amaurosis, which causes 10% to 18% of childhood blindness (visual acuity <20/200). It is the result of mutations in at least 12 different genes and is almost always inherited as an autosomal recessive condition. Some affected children have other neurologic abnormalities, but like the child in the vignette, many are otherwise neurologically normal.

Gross and fine motor development is one of the most studied and perhaps the most affected realm of development in visually impaired children. Although sighted children on average intentionally reach for objects in the environment at age 4 to 5 months, visually impaired children do not do so until 8 to 9 months of age. Reaching activity occurs at this age if there is continuous touch contact with the object or there is a continuous sound stimulus from the object. Reaching for silent objects does not occur until 4 or 5 months later even if there was prior (but not continuous) touch contact with the object. Activities of daily living that require movement through space are of particular concern if they require use of a tool (eg, a cup or other object external to the body). Activities that relate to the body, such as pulling on clothes, are more easily performed.

Postural development, ie, the ability to sit or stand, is also delayed in blind infants compared with sighted infants, but both groups develop the ability to sit independently at about the time when they are able to reach in a coordinated manner. Language acquisition is slightly delayed in visually impaired children with a 1- or 2-month difference in achieving first words. However, visually impaired children reach the 10- to 50-word developmental level at approximately the same age as sighted children. The words are qualitatively different however, with blind children producing more naming words but having difficulty learning to use personal and possessive pronouns and spatial prepositions (on, under, above, etc.). The inability to participate with parents in reciprocal interaction and visual joint attention can lead to problems in social relatedness and social interaction.

Programs that support early and intensive intervention with visually impaired children have been developed to compensate for these early developmental lags. The data thus far to determine the effectiveness of these programs are limited, but it is hoped that they will

provide a framework these children need to achieve their maximum developmental potential

PREP Pearls

- Congenital visual impairment, even in the absence of other neurologic problems, contributes to a lag in reaching normal developmental milestones.
- Motor development that requires movement through space (eg, reaching for an object) is often delayed by several months.
- Initial verbal development can be mildly delayed, but catch-up development is fairly rapid. A qualitative difference in the types of words used, however, may persist.
- Leber amaurosis causes 10% to 18% of childhood blindness.

American Board of Pediatrics Content Specification(s):

- Understand the variations in the developmental sequence that are associated with congenital visual impairment

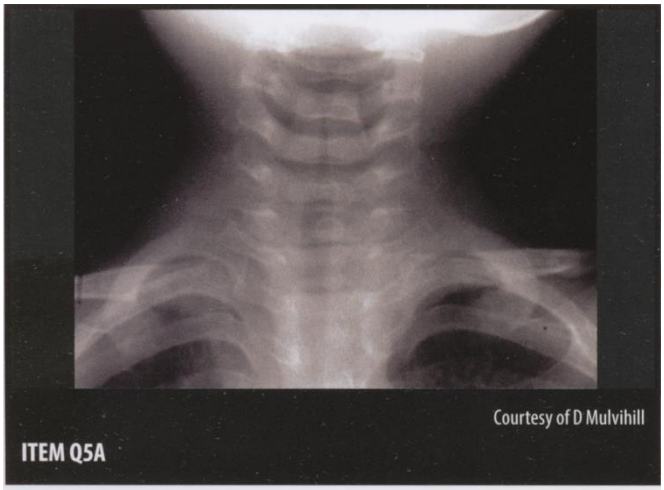
Suggested Reading:

- Brambring M. Divergent development of manual skills in children who are blind or sighted. J Visual Impairment Blindness. 2007
- Brambring M. Divergent development of verbal skills in children who are blind or sighted. J Visual Impairment Blindness. 2007
- Dale N, Salt A. Early support developmental journal for children with visual impairment: the case for a new developmental framework for early intervention. Child Care Health Dev. 2007;33:684-690
- Ihsen E, Troester H, Brambrin M. The role of sound in encouraging infants with congenital blindness to reach for objects. J Visual Impairment Blindness. 2010;104:478-488
- Kelly DP, Teplin SW. Sensory impairments: hearing and vision. In: Voigt RG, ed. Developmental and Behavioral. Pediatrics. Elk Grove Village, IL: American Academy of Pediatrics. 2011:22:476-489
- Roizen N, Kasza K, Karrison T, et al; Toxoplasmosis Study Group. Impact of visual impairment on measures of cognitive function for children with congenital toxoplasmosis: implications for compensatory intervention strategies. Pediatrics. 2006;118:e379-390. doi: 10.1542/peds.2005-1530

Item 5

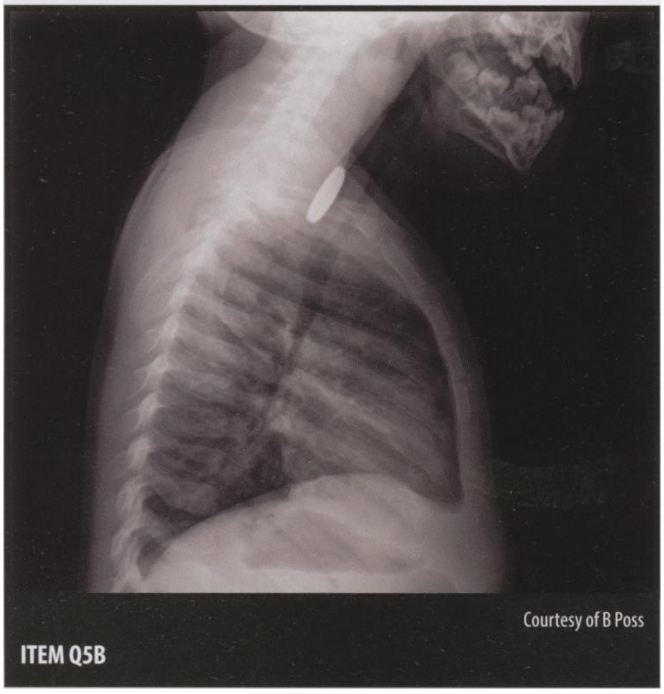
A previously healthy, 3-year-old girl developed a high fever and has trouble breathing since this morning. On physical examination, the child appears "toxic" and is leaning forward in her mother's lap. Her temperature is 40.0°C, heart rate is 138 beats/min, respiratory rate is 37 breaths/min, blood pressure is 96/62 mm Hg, and oxygen saturation on room air is 94% by pulse oximetry. You arrange rapid transfer to the local hospital and consultation with a pediatric anesthesiologist and otolaryngologist. The otolaryngologist updates you on the condition of your patient and lets you know that a large "cherry red" epiglottis was seen on visualization in the operating room.
Of the following, the image MOST consistent with your patient's diagnosis is

A. Item Q5A
B. Item Q513
C. Item Q5C
D. Item Q5D
E. Item Q5E



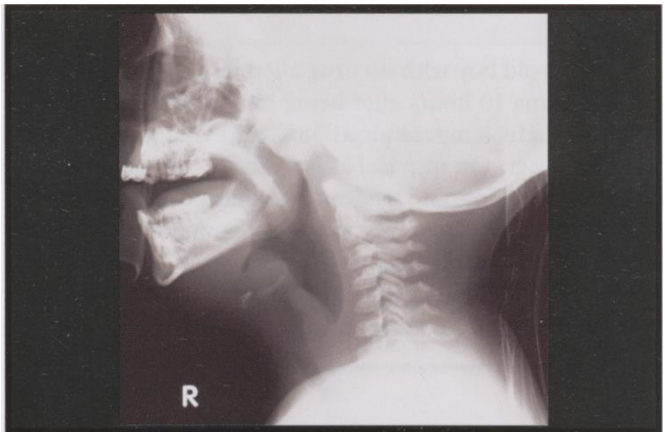
Courtesy of D Mulvihill

ITEM Q5A



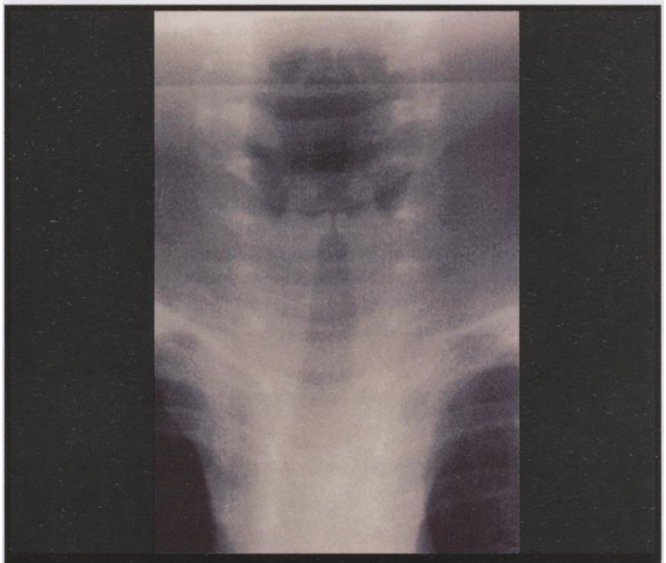
Courtesy of B Poss

ITEM Q5B



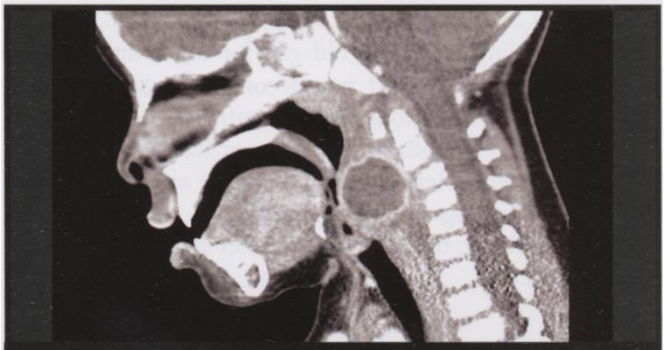
Courtesy of B Poss

ITEM Q5C



Courtesy of D Mulvihill

ITEM Q5D



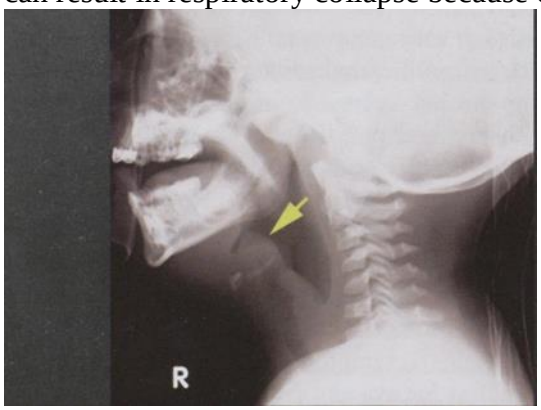
Courtesy of B Poss

ITEM Q5E

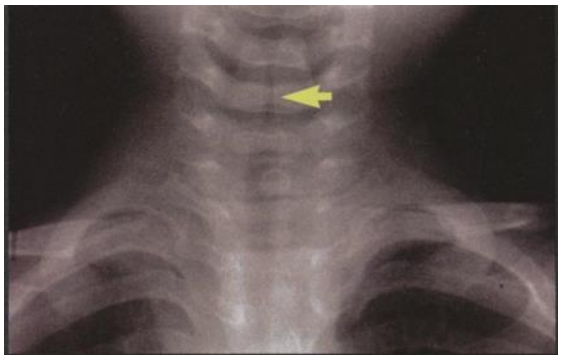
Item 5 TE S Preferred Response: C

The clinical presentation of the child described in the vignette is consistent with epiglottitis, which is a medical emergency. Lateral radiographs are often diagnostic but should be deferred in children strongly suspected of having epiglottitis until the proper personnel and equipment are available to secure the airway if needed. If obtained, the radiograph will demonstrate an enlarged epiglottis (the "thumb sign") (option C, Item C5A). Direct visualization of the airway as was done in the operating room in this child will demonstrate a red and swollen epiglottis. Other diseases of the upper airway can produce symptoms of respiratory obstruction, including bacterial tracheitis, foreign bodies, retropharyngeal abscesses, and viral laryngotracheobronchitis (croup). Option A is an anterior-posterior radiograph of the neck that shows narrowing of the subglottic trachea ("steeple sign") (Item C5B) consistent with viral laryngotracheobronchitis (croup); this finding is in contrast with the normal airway illustrated in option D (Item C5C). Option B (Item C5D, page C-5) demonstrates a foreign body that is clearly seen in the esophagus, whereas option E (Item C5E, page C-5) is a lateral neck computed tomographic image of a patient who has a retropharyngeal abscess. Radiographs obtained in bacterial tracheitis may appear indistinguishable from those obtained in viral laryngotracheobronchitis, but haziness or irregularity of the tracheal lumen may be seen Although the incidence of epiglottis has dramatically decreased since the introduction of the Haemophilus influenzae type B vaccine, prompt recognition of this disease remains imperative. Affected patients typically present between the ages of 2 and 8 years with the rapid onset of fever, sore throat, and the "four Ds" (drooling, dysphagia, dysphonia, and dyspnea). Patients often assume a position of comfort by sitting upright, leaning forward, and bracing themselves with their arms, known as the tripod position. In general, patients who have viral laryngotracheobronchitis tend to have a slower onset of disease that is associated with prodromal symptoms (eg, rhinorrhea, cough), lower grades of fever, are less ill-appearing, and have a prominent barking cough. In addition, most patients who present for medical attention are less than 2 years of age.

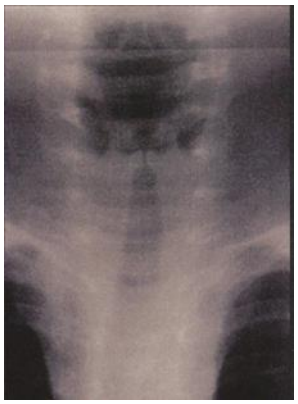
Direct examination of the airway under anesthesia (with the availability of personnel who can perform a tracheostomy if needed) is the preferred management for suspected cases of epiglottis. Airway management is critical and, therefore, invasive procedures such as blood draws, throat culture, obtaining vascular access, or intramuscular administration of medications should be deferred until the airway is properly secured; agitation of the child can result in respiratory collapse because of airway obstruction.



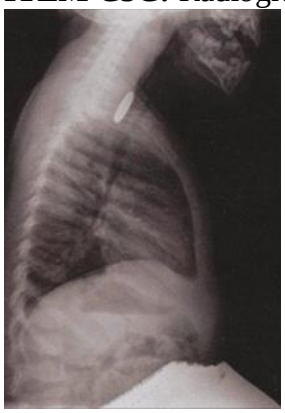
ITEM C5A: Lateral radiograph of the neck in epiglottitis: there is enlargement of the epiglottis (arrow) giving the "thumb sign.



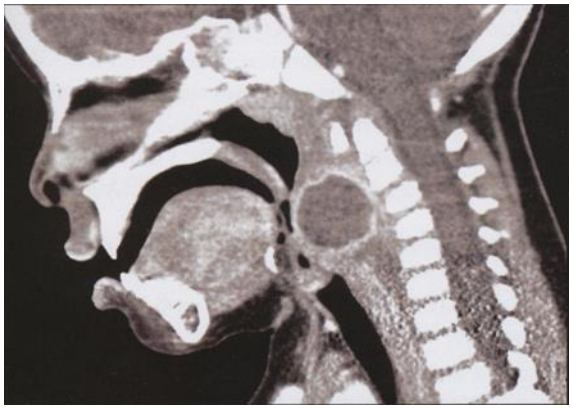
ITEM C5B: Anterior-posterior radiograph of the neck shows narrowing of the subglottic portion of the trachea (arrow) giving the "steeple sign."



ITEM C5C: Radiograph depicts a normal airway.



ITEM C5D: Radiograph depicts a foreign body lodged in the esophagus.



ITEM C5E: Lateral computed tomographic image of the neck demonstrating a retropharyngeal swelling consistent with a retropharyngeal abscess. The epiglottis is of normal size.

PREP Pearls

- Although the incidence of epiglottitis has decreased, providers must recognize it when it presents.
- Epiglottitis is characterized by the "four Ds" (drooling, dysphagia, dysphonia, and dyspnea).
- Direct visualization of the airway under anesthesia is the preferred initial management of epiglottitis.

American Board of Pediatrics Content Specification(s):

- Differentiate the clinical and radiographic findings of viral croup from those of epiglottitis and bacterial tracheitis
- Know the risks of examination of patients with suspected epiglottitis

Suggested Reading:

- Mehta R, Hariprakash SP, Cox PN, Wheeler DS. Diseases of the upper respiratory tract. In: Wheeler DS, Wong HR, Shanley TP, eds. Pediatric Critical Care Medicine: Basic Science and Clinical Evidence. New York, NY: Springer-Verlag; 2007:485-505
- Roosevelt GE. Acute inflammatory upper airway obstruction (croup, epiglottitis, laryngitis, and bacterial tracheitis. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:1445-1449
- Woods CR. Epiglottis (supraglottis): clinical features and diagnosis. UptoDate. Available online only for subscription
- Woods CR. Epiglottis (supraglottis): treatment and prevention. UptoDate. Available online only for subscription

Item 6

A 15-year-old female patient calls to request that you call in a prescription for emergency contraception. Her last sexual activity was 4 days ago, and her partner did not use a condom.

Of the following, the BEST choice for emergency contraception for this girl is

A. intrauterine device placement
B. levonorgestrel
C. mifepristone
D. ulipristal acetate
E. Yuzpe regimen

Item 6 Preferred Response: D

Many treatment options are now available for prevention of pregnancy within 72 to 120 hours after unprotected sexual intercourse. Given that newer methods do not have to be given within 12 hours of an episode of unprotected sexual activity, the terms morning-after pill and postcoital contraception are no longer appropriate. Emergency contraception (EC) is the term currently in use. The available methods can be grouped as either hormonal or intrauterine. The most effective hormonal method currently available is ulipristal acetate and it was approved by the US Food and Drug Administration for this purpose in August 2010. It is a selective progesterone receptor modulator (SPRM) and is available by prescription only. The 30-mg pill has been found to be effective up to 5 days (120 hours) after unprotected intercourse and has a favorable safety profile.
Yuzpe and his group were the first to introduce hormonal EC. Their method included the use of 100 lig of ethinyl estradiol plus 500 jig of levonorgestrel taken twice at a 12-hour interval. The rate of unwanted pregnancy was reduced by two-thirds, but this method was not effective if used more than 72 hours after unprotected intercourse.

Progesterone methods were developed next and were more effective, with fewer adverse effects. A single 1.5-mg tablet of levonorgestrel is currently recommended. The overall reduction in pregnancy rate is 60% to 93%, with the effectiveness decreasing if taken more than 72 hours after intercourse. This method is available over the counter. The World Health Organization developed the third hormonal method (ie, SPRM). The first drug in this category was mifepristone, which is a known abortifacient but is used for EC in some countries. Clinical studies comparing levonorgestrel and mifepristone in different dose regimens concluded that all regimens possess similar effectiveness. Ulipristal acetate is a second-generation SPRM and works as a contraceptive by suppressing follicular development, delaying the surge of luteinizing hormone, retarding endometrial maturation, and promoting endometrial bleeding. At the single dose of 30 mg, it has been found to be a more potent inhibitor of ovulation than levonorgestrel and can be effectively taken up to 120 hours after unprotected intercourse. Intrauterine methods include postcoital insertion of a copper-bearing intrauterine device. This method requires an office visit to a trained health care professional for insertion. Concerns about an increased incidence of infections in the presence of an intrauterine device have been discounted.

Female rape victims are usually seen in an emergency department setting, and surveys indicate that a significant number are not provided EC. Clinicians should consider giving an on-hand prescription for a hormonal EC method to all girls younger than 17 years of age because of the temporal effectiveness of treatment.

PREP Pearls

- Options for EC are now available for up to 5 days after unprotected intercourse.
- A single 1.5-mg tablet of levonorgestrel is effective for EC up to 72 hours after intercourse and is available over the counter.
- Ulipristal acetate is effective for EC up to 5 days after intercourse and requires a prescription.

American Board of Pediatrics Content Specification(s):

- Know the options for postcoital contraception for the female rape victim and recognize when they should be used

Suggested Reading:

- AGOG Committee on Practice Bulletins—Gynecology. AGOG Practice Bulletin No. 112: emergency contraception. Obstet Gynecol. 2010;115:1100119. doi:10.1097/A0G.0b013e3181deff2a
- Fine PM. Update on emergency contraception. Adv Then 2011;28:87-90. doi:10.1007/s12325-010-0090-x
- Fortin K, Jenny C. Sexual abuse. Pediatr Rev. 2012;33:19. doi: 10.1542/ pir.33-1-19
- Kaufman M; American Academy of Pediatrics Committee on Adolescence. Care of the adolescent sexual assault victim. Pediatrics. 2008;122:462. doi: 10.1542/peds.2008-1581
- Langston A. Emergency contraception: update and review. Semin Reprod Med. 2010;28:95-102. doi:10.1055/s-0030-1248133
- Patel A, Panchal H, Piotrowski ZH, Patel D. Comprehensive medical care for victims of sexual assault: a survey of Illinois hospital emergency departments. Contraception. 2008;77:426-430. doi:10.1016/j. contraception.2008.01.018
- World Health Organization. Guidelines for Medico-Legal Care for Victims of Sexual Violence. Geneva, Switzerland: World Health Organization; 2003

Item 7

A 4-year-old boy is being evaluated for a 6-cm abscess on his buttocks with surrounding erythema. An incision and drainage is performed that yields approximately 15 mL of purulent material. Therapy with clindamycin is recommended, pending culture and sensitivity results, because there is a high incidence of community-acquired methicillin-resistant Staphylococcus aureus in the area.
Of the following, the adverse reaction MOST often associated with this treatment is

A. anemia
B. diarrhea
C. glomerulonephritis
D. hepatitis
E. scalded skin syndrome

Item 7 S Preferred Response: B

Clindamycin is associated with diarrhea in up to 20% of recipients. The diarrhea is often mild and self-limited but use of clindamycin, ampicillin, amoxicillin, and the cephalosporins, can be associated with antibiotic-associated colitis due to Clostridium difficile. Individuals with chronic, debilitating underlying disease are at increased risk of developing antibiotic-associated colitis. C difficile colitis typically occurs between days 4 and 9 of therapy, but may occur up to 10 weeks after completion of a course of antibiotics.

Up to 10% of patients receiving clindamycin develop a skin rash, which is typically maculopapular and not the diffuse erythroderma of scalded skin syndrome. The latter is associated with staphylococcal toxin, not the antibiotic. The eruption of Stevens-Johnson syndrome, which may resemble scalded skin syndrome, is an uncommon reaction associated with the use of clindamycin.

An increase in liver transaminases (hepatitis) is an uncommon reaction to clindamycin. Transient neutropenia and thrombocytopenia, but not anemia, have been rarely described with the administration of clindamycin. Glomerulonephritis has not been associated with the use of clindamycin.

PREP Pearls

- Clindamycin is associated with diarrhea in up to 20% of recipients but is generally mild.
- Clindamycin, amoxicillin, ampicillin, and the cephalosporins are the antibiotics most associated with Clostridium difficile colitis.

American Board of Pediatrics Content Specification(s):

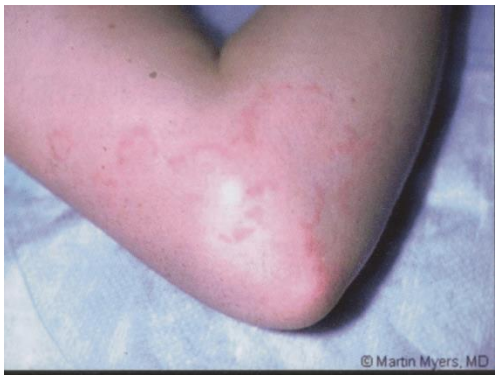
- Recognize the adverse reactions to clindamycin (eg, diarrhea, including Clostridium difficile enterocolitis)

Suggested Reading:

- Michelow IC, McCracken GH, Jr. Antibacterial therapeutic agents. In: Feigin RD, Cherry JD, Demmler-Harrison GJ, Kaplan SL, eds. Feigin and Cherry's Textbook of Pediatric Infectious Disease. 6th ed. Philadelphia, PA: Saunders Elsevier; 2009:3206-3208
- Thomas C, Stevenson M, Riley TV. Antibiotic and hospital-acquired Clostridum difficile-associated diarrhea: a systematic review. JAntimirob Chemother. 2003;51(6):1339-1350. doi: 10.1093/jac/dkg254
- MacDougall C, Chambers HF. Clindamycin. In: Brunton LL, Chabner, BA, Knollmann BC, eds. Goodman and Gliman's The Pharmacological Basis of Therapeutics. 12th ed. New York, NY: McGraw-Hill Co; 2011:1534-1535

Item 8

A 7-year-old boy presents to your office with a 3 -day history of fever and joint pain. His parents recall a sore throat 3 weeks ago. Findings on physical examination reveal swelling and erythema of his right knee and ankle. Yesterday, he had swelling and erythema of his right wrist and left knee that have since resolved. His temperature is 38.7°C, and the remainder of his vital signs is normal. Examination of his skin reveals a rash (Item Q8). His erythrocyte sedimentation rate is 30 mm/h and a rapid streptococcal test result is negative.



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ITEM Q8: Rash as described for the boy in the vignette.

Of the following, the MOST appropriate therapy for this patient is

A. azithromycin
B. ceftriaxone
C. naproxen
D. penicillin G
E. prednisone

Item 8 Preferred Response: D

The patient described in the vignette has fever, erythema marginatum, and a polyarticular migratory arthritis. This constellation of features is consistent with acute rheumatic fever (ARF). Acute rheumatic fever is a major cause of acquired heart disease worldwide; however the incidence of acute rheumatic fever has been declining in the United States. ARF is an autoimmune disease that follows an infection with group A streptococci (GAS) and can lead to rheumatic heart disease in patients that are not treated for streptococcal pharyngitis. ARF is diagnosed by the Jones criteria (Item C8A).

The most common features of ARF in the United States are carditis and arthritis. The cardiac manifestations of ARF present as mitral or aortic insufficiency, pericarditis, pericardial or myocardial involvement, and valvulitis. The arthritis associated with ARF is classically described as an early polyarticular arthritis that migrates. Often, the arthritis will completely resolve in one joint and appear in another joint. The arthritis is nondeforming and most commonly affects large joints such as the elbows, wrists, knees, and ankles. The arthritis is exquisitely painful and responds well to aspirin and other anti-inflammatory medications. Often, throat culture is negative by the time the patient presents with symptoms; however, it is recommended that a patient with active symptoms of ARF be treated with Penicillin G or 10 days of oral penicillin to eliminate residual GAS infection, even if the throat culture is negative. Treatment of ARF also includes anti-inflammatory medications, restriction of activity, and antibiotic prophylaxis to prevent progression of cardiac disease (Item C8B) Item C8C

Penicillin G is the drug of choice to treat any residual GAS infection. Azithromycin can treat GAS infections but is a second-line therapy. Ceftriaxone does not provide good coverage for a GAS infection. While nonsteroidal anti-inflammatory drugs such as naproxen can be used to treat the arthritis associated with ARF, they are second-line agents. Aspirin would be the first-line treatment choice. Glucocorticoids such as prednisone are not recommended in the treatment of ARF except in cases of severe carditis, and even then the evidence for effectiveness is weak.

Item C8A. Jones Criteria

Confirmation of preceding Group A Streptococcus by positive throat culture, positive rapid antigen test, elevated or rising antistreptolysin O titer
AND
2 major manifestations
OR
1 major and 2 minor manifestation

Major Manifestation	Minor Manifestations
• Polyarthritis • Carditis • Subcutaneous nodules • Sydenham chorea • Erythema marginatum	• Fever • Arthralgia • Prolonged PR interval on electrocardiogram • Elevated acute phase reactants (erythrocyte sedimentation rate, C-reactive protein)

Item C8B. Antibiotic Prophylaxis Recommended to Prevent Recurrence of Rheumatic Fever

Agent	Dose	Mode
Benzathine penicillin G	27 kg (60 Ibs), 600,000 U, >27 kg (60 Ibs) 1,200,000 U	Intramuscular every 4 weeks
Penicillin V	250 mg twice daily	Oral
Sulfadiazine	27 kg (60 Ibs), 0.5 g once daily >27 kg (60 lbs) 1.0 g once daily	Oral
Macrolide or **azalide***	Varies	Oral

* If allergic to penicillin and sulfadiazine.

Reprinted with permission from Gerber MA, Baltimore RS, Eaton CB, et al. Prevention of rheumatic fever and diagnosis and treatment of acute streptococcal pharyngitis: a scientific statement from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee of the Council on Cardiovascular Disease in the Young, the Interdisciplinary Council on Functional Genomics and Translational Biology, and the Interdisciplinary Council on Quality of Care and Outcomes Research, endorsed by the American Academy of Pediatrics. *Circulation*. 2009;119(11):1541-1551

Item C8C. Duration of Secondary Rheumatic Fever Prophylaxis

Category	Duration after last attack
Rheumatic fever with carditis and residual heart disease (persistent valvular disease by echocardiogram or clinical evidence)	10 years or until 40 years of age (whichever is longer), sometimes lifelong prophylaxis
Rheumatic fever with carditis but no residual heart disease (no valvular disease on echocardiogram or clinical evidence)	10 years or until 21 years of age (whichever is longer)
Rheumatic fever without carditis	5 years or until 21 years of age (whichever is longer)

Reprinted with permission from Gerber MA, Baltimore RS, Eaton CB, et al. Prevention of rheumatic fever and diagnosis and treatment of acute streptococcal pharyngitis: a scientific statement from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee of the Council on Cardiovascular Disease in the Young, the Interdisciplinary Council on Functional Genomics and Translational Biology, and the Interdisciplinary Council on Quality of Care and Outcomes Research, endorsed by the American Academy of Pediatrics. *Circulation*. 2009;119(11):1541-1551

PREP Pearls

- Throat cultures are often negative by the time symptoms of ARF are present.
- The arthritis of ARF is a nondeforming, migratory polyarthritis that involves the large joints.
- Long-term antibiotic prophylaxis is used in ARF patients to prevent worsening cardiac disease.

American Board of Pediatrics Content Specification(s):

- Know the characteristics of arthritis associated with rheumatic fever

Suggested Reading:

- Gerber MA, Baltimore RS, Eaton CB, et al. Prevention of rheumatic fever and diagnosis and treatment of acute streptococcal pharyngitis: a scientific statement from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee of the Council on Cardiovascular Disease in the Young, the Interdisciplinary Council on Functional Genomics and Translational Biology, and the Interdisciplinary Council on Quality of Care and Outcomes Research, endorsed by the American Academy of Pediatrics. Circulation. 2009;119(11):1541-1551. doi:10.1161/CIRCULATIONAHA
- John J, Chandran L. Arthritis in children and adolescents. Pediatr Rev. 2011;32(11):470-480. doi:10.1542/pir.32-11-470
- Steer AC, Carapetis JR. Acute rheumatic fever and rheumatic heart disease in indigenous populations. Pediatr Clin N Am. 2009;56(6):1401-1419. doi:10.1016/j.pc1.2009.09.011

Item 9

A 2 year-old boy is brought to your office by his parents. The child is having difficulties both at home and at his child care center. His parents report that he often makes noises, but rarely does this seem like he is making an attempt to communicate. The parents feel that he is able to hear them, but he either does not understand them or chooses not to respond. They feel he is not as interactive with them as his older sibling had been at the same age. The boy usually plays by himself and does not appear to be interested in joining group activities at child care. He becomes very frustrated when he does not get his way. He typically makes poor eye contact. The staff at his child care center is having difficulties managing his behaviors.

Of the following, the MOST appropriate intervention at this time is to perform

A. an electroencephalogram
B. a fragile X syndrome genetic test
C. a mercury level test
D. a modified checklist for autism in toddlers (M-CHAT)
E. thyroid function tests

Item 9 Preferred Response: D

The presentation of the child in the vignette suggests an autism spectrum disorder (ASD), which can be further evaluated by asking the child's parent to complete the Modified Checklist for Autism in Toddlers (M-CHAT). The M-CHAT is a free-to-use checklist that has been validated for use between 16 and 30 months of age. It is designed to identify children who would benefit from a more thorough developmental autism evaluation, and can be part of a well-child check or specialist's assessment. This patient has demonstrated several areas of impairment in social interaction and communication that are typical of a young child who may have autism, but the information provided is not sufficient to make a definitive diagnosis. The M-CHAT can be completed along with an assessment of his hearing and vision (which could very easily cause similar symptoms), inquiring about any sudden changes in developmental progress (which could indicate a specific brain abnormality), and examination for dysmorphic features that might be suggestive of a developmental syndrome. Regardless of the ultimate diagnosis, the child's evident developmental impairment requires further evaluation.

Electroencephalography (EEG) would be indicated for a child with intermittent episodes of altered mental status or altered movements that could be suggestive of a seizure disorder. EEG is not a routine part of an autism evaluation. Genetic testing for fragile X syndrome can be indicated for children with mental retardation, but in this case further assessment is needed to determine if this patient has mental retardation. If the child were found to have both autism and mental retardation, fragile X testing could be considered. Elevated mercury levels, suggested to result from vaccine preservatives like thimerosal, have been extensively researched and found not to be associated with autism. Mercury toxicity is very uncommon, and testing for it is not part of the autism evaluation. Hypothyroidism is an uncommon cause of mental retardation. Thyroid function tests would be appropriate for developmentally impaired children who were not appropriately screened for hypothyroidism at birth or for children with unexplained linear growth failure; however, such tests are not a routine part of an autism evaluation.

The symptoms of autism spectrum disorder should be observable before age 3 years, with some signs noted usually by age 1 year. Autism should be suspected in any child with impairment of both social interaction and communication that is out of sync with the child's overall developmental level. Though autism is often associated with intellectual impairment, many patients with autism have intelligence in the normal range.

PREP Pearls

- The M-CHAT is a useful screening tool for identifying autism in children under age 3 years.
- Autism can typically be diagnosed before age 3 years.
- Biological testing for autism-associated syndromes like fragile X is neither required for every patient, nor necessary to diagnose autism.

AAP Mental Health Competency:

- Identify how to intervene for a case of autism in a less than 3 year old child

Suggested Reading:

- Myers SM, Plauche Johnson C; Council on Children With Disabilities. Management of children with autism spectrum disorders. Pediatrics. 2007;120:5:1162-1182. doi:10.1542/peds.2007-2362
- Taguay PE. Autism spectrum disorders. In: Dulcan MK, ed. Dulcan's Textbook of Child and Adolescent Psychiatry. Arlington, VA: American Psychiatric Publishing; 2010:173-189

Item 10

A neonate was born at 35 weeks of gestation to a 23-year-old primigravida by spontaneous vaginal delivery that was complicated by prolonged rupture of the membranes and no prenatal care. The mother presented completely dilated and rapidly delivered a vigorous neonate. Tachypnea is noted 2 hours after birth. The mother described leaking copious clear fluid 36 hours before delivery. Her prenatal screens, including group B streptococcus status, were unknown at the time of delivery, and no antibiotics were given to the mother. Physical examination reveals a neonate with a temperature of 36.3°C, heart rate of 120 beats/min, respiratory rate of 76 breaths/min, right-arm blood pressure of 41/23 mm Hg (mean blood pressure, 29 mm Hg), and right-hand oxygen saturation of 95% on room air. The remainder of the evaluation is remarkable for a capillary refill of 4 seconds, a soft 2/6 systolic murmur at the left lower sternal border, and mild retractions. After obtaining blood and cerebrospinal fluid cultures, antibiotic therapy is initiated with ampicillin and gentamicin.

Of the following, the MOST appropriate next step in management is to

A. administer 10 mL/kg albumin bolus
B. continue to monitor cardiorespiratory status
C. initiate dopamine
D. provide supplemental oxygen
E. start prostaglandin El

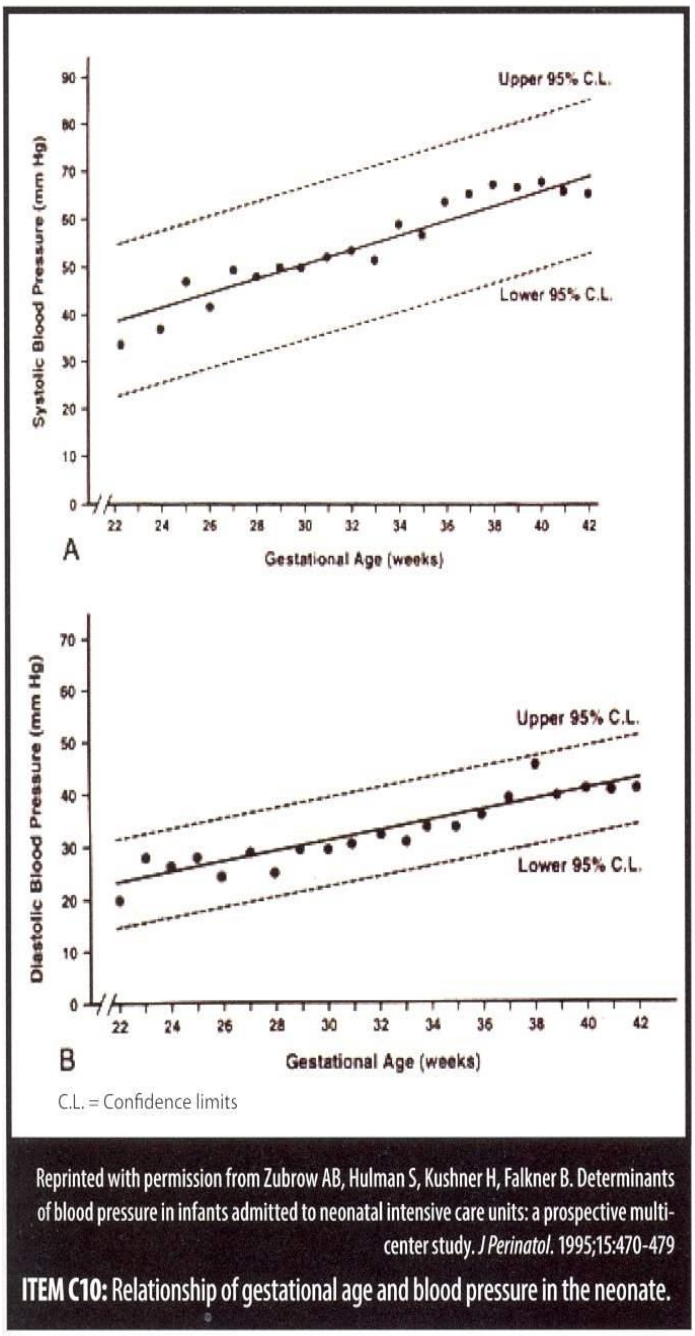
Item 10 Preferred Response: C

The infant in the vignette is demonstrating the clinical findings of septic shock, including hypotension, and would benefit from the initiation of dopamine for cardiovascular support. Septic shock in the neonatal population is often associated with increased capillary leak and vasodilation, which lead to relative hypovolemia and decreased peripheral vascular resistance. Cardiovascular support with volume replacement and vasopressor drug administration may be required to prevent circulatory collapse. Dopamine is a catecholamine that stimulates dopaminergic, α-adrenergic,β-adrenergic, and serotonin receptors in a dose-dependent manner. Systemic blood pressure is improved by the effect of dopamine on peripheral vascular resistance and myocardial contractility.

Hypotension in the neonatal population has been loosely defined as any value that falls below the fifth to tenth percentile for gestational age. The most commonly used guide for hypotension in the neonatal period is that the mean blood pressure should be equal to or greater than the gestational age of the infant. Treatment of hypotension, as presently Defined, has not been demonstrated to improve long-term outcome. Therefore some experts suggest that hypotension should be defined as the blood pressure at which cerebral blood flow regulation fails and brain tissue hypoperfusion occurs. No simple technique to assess this presently exists, but near-infrared spectroscopy imaging may allow estimation of cerebral blood flow and could be used as a surrogate measurement in the future. The mean blood pressure of 29 mm Hg for the 35-week gestation infant in the vignette supports the diagnosis of hypotension (Item C10).

The role of volume replacement in septic shock remains unclear in the immediate neonatal period. It is required in hypovolemic shock, with the preferred agents being packed red blood cells, normal saline, or lactated Ringers solution. Although albumin is as effective as normal saline in the immediate period of administration, it is not recommended in the 2010 American Academy of Pediatrics and American Heart Association Guidelines for Neonatal Resuscitation because of its increased cost and theoretical risks such as increased fluid retention. Volume replacement should be avoided in infants weighing less than 1,500 g in the first week after birth because of the potentially increased risks of intraventricular hemorrhage and chronic lung disease, unless there is evidence of blood loss from events such as maternal abruption, maternal-fetal transfusion, or subgaleal hemorrhage.

The treatment of hypotension of the infant in the vignette should be the next step in the management. If volume expansion is considered, normal saline should be used rather than albumin. The preductal saturation of 95% does not warrant supplemental oxygen and makes congenital cyanotic heart disease unlikely. If congenital heart disease remains a consideration, an echocardiogram should be obtained to determine the need for prostaglandin El.



Reprinted with permission from Zubrow AB, Hulman S, Kushner H, Falkner B. Determinants of blood pressure in infants admitted to neonatal intensive care units: a prospective multi-center study. *J Perinatol.* 1995;15:470-479

ITEM C10: Relationship of gestational age and blood pressure in the neonate.

PREP Pearls

- Newborns with hypotension due to septic shock may benefit from administration of dopamine to prevent circulatory relapse.
- The mean blood pressure should be equal to or greater than the gestational age of the newborn.

American Board of Pediatrics Content Specification(s):

- Know that blood pressure values vary directly with gestational age

Suggested Reading:

- Cayabyab R, McLean CW, Seri I. Definition of hypotension and assessment of hemodynamics in the preterm neonate. I Perinatol. 2009;29:s58-s62. doi: 10.1038/jp.2009.29
- LeFlore JL, Engle WD. Clinical factors influencing blood pressure in the neonate. NeoReviews. 2002;3:e145-e150. doi: 10.1542/neo.3-8-e145
- Seri I, Noori S. Diagnosis and treatment of neonatal hypotension outside the transitional period. Early Hum Dev. 2005;81:405-411. doi: 10.1016/j. earlhumdev.2005.03.008

Item 11

A 5-year-old girl presents to the emergency department with pallor and jaundice. Her mother states that the girl was in her usual state of health until 3 days ago, when she began to have fever, cough, and decreased activity. On physical examination, her oral temperature is 37.5°C, pulse rate is 120 beats/ min, respiratory rate is 24 breaths/min, and blood pressure is 100/60 mm Hg. The child is pale with icteric sclerae. Her breath sounds are normal, and there is no hepatosplenomegaly or lymphadenopathy. The remainder of the examination is normal. Laboratory test results reveal:

- White blood cell count, 13,500/µL (13.5 x 10^9/L), with 40% polymorphonuclear leukocytes, 53% lymphocytes, 5% monocytes, and 2% eosinophils
- Hemoglobin, 4.0 g/dL (40 g/L)
- Mean corpuscular volume, 85/μm^3 (85 fL)
- Platelet count, 402 x 10^3/µL (402 x 10^9/L)
- Reticulocyte count, 12% (0.12)

Of the following, the MOST likely cause of this child's anemia is

A. autoimmune hemolytic anemia
B. congenital erythrocyte hypoplasia (Blackfan-Diamond anemia)
C. glucose-6-phosphate dehydrogenase deficiency
D. parvovirus B19 infection
E. transient erythroblastopenia of childhood

Item 11 Preferred Response: A

Hemolysis can be divided into intrinsic and extrinsic hemolytic anemias. Intrinsic hemolytic anemias are due to defects within the erythrocyte that affect the red blood cell membrane, hemoglobin, or cytosolic components (enzymes). These defects include gene mutations and nutritional deficiencies, which can lead to abnormal production of heme, globin chains, membrane proteins, or intracellular enzymes. Examples of intrinsic hemolytic anemias are sickle cell disease (globin chain), thalassemia (globin chain), hereditary spherocytosis (membrane defect), and glucose-6-phosphate dehydrogenase deficiency (enzyme). Extrinsic hemolytic anemias are due to immunologic, chemical, or physical factors that damage the erythrocyte. Examples of extrinsic hemolytic anemias include autoimmune hemolytic anemia, hypersplenism, toxins, heat, disseminated intravascular coagulation (DIC), hemolytic-uremic syndrome, and thrombotic thrombocytopenic purpura. In some cases, both intrinsic and extrinsic mechanisms may be involved.

Pallor, jaundice, and reticulocytosis are typical features of hemolytic anemia. Reticulocytes are young erythrocytes released from the bone marrow. The reticulocyte count is useful in distinguishing between anemias of decreased production vs those of increased destruction. Normally, reticulocytes account for 1% to 2% of all circulating red blood cells. When erythrocytes have a shortened lifespan (eg, from hemolysis), the erythropoietin level increases, which stimulates the bone marrow to increase its production of erythrocyte precursors. The percentage of reticulocytes can increase significantly above the steady state of 1% to 2%, as seen in this vignette, where the child is found to have a reticulocyte count of 12%.

The girl in the vignette most likely has autoimmune hemolytic anemia (AIHA). In infants and young children, AIHA usually occurs after an infection, whereas in adolescents it is more likely to be associated with an underlying systemic disease. A positive direct antiglobulin test result (also known as the direct Coombs test), indicating the presence of antibodies to the red blood cells, would also support the diagnosis of AIHA. Glucose-6-phosphate dehydrogenase deficiency is another cause of hemolytic anemia, but its X-linked recessive inheritance pattern makes it unlikely in girls.

In contrast to the anemias of increased destruction, the anemias of decreased production will typically present with a low or normal reticulocyte count. Iron deficiency is the most common cause of anemia in the world and is due to decreased formation of the heme component of hemoglobin. A low reticulocyte count and low mean corpuscular volume are seen in iron deficiency anemia, both of which are not found in the patient in this vignette. Low reticulocyte count would be seen in congenital erythroid hypoplastic anemia (Blackfan-Diamond anemia), which usually presents in the first year of life, and transient erythroblastopenia of child-hood, which usually presents between 6 months and 4 years of life. Parvovirus B19 preferentially infects the red blood cell precursors in the bone marrow, which can lead to a transient red blood cell aplasia. This condition does not cause a significant anemia in individuals with a normal erythrocyte life span. However, in patients who have chronic hemolytic conditions and rely on reticulocytosis to compensate

for their anemia (as in patients with sickle cell disease or hereditary spherocytosis), infection with parvovirus B19 can lead to a severe aplastic crisis.

PREP Pearls

- The reticulocyte count is useful in distinguishing between anemias of decreased production vs those of increased destruction.
- Pallor, jaundice, and reticulocytosis are typical features of hemolytic anemia.
- In infants and young children, All-IA usually occurs after an infection, whereas in adolescents it is more likely to be associated with an underlying systemic disease.

American Board of Pediatrics Content Specification(s):

- Know that the reticulocyte count usually distinguishes between disorders of erythrocyte production and those of erythrocyte destruction

Suggested Reading:

- Ware RE. Autoimmune hemolytic anemia. In: Nathan and Oski's Hematology of Infancy and Childhood. 7th ed. Philadelphia, PA: Saunders Elsevier; 2009:613-658
- Recht M, Mahoney DH. Overview of hemolytic anemias in children. UptoDate. Available online only for subscription

Item 12

A 10-year-old boy is at school when his teacher notices that he is staring out the window. She can't get him to stop staring or respond to her, so he is brought to the emergency department. No other children had similar symptoms. On arrival, his physical examination reveals a temperature of 37.2°C, blood pressure of 100/60 mm Hg, heart rate of 85 beats/min, and a respiratory rate of 20 breaths/min. The boy is awake and seems restless. He follows one-step commands (eg, "take off your shoes"), but does not follow two-step commands. He knows his name, but not where he is. The remainder of the physical examination findings is unremarkable. Results of computed tomography of the head without contrast, serum sodium and glucose, and serum and urine toxicology testing are normal. As you are completing your examination, the boy's parents arrive and report no known ingestions at home, no history of seizures or headaches, and no similar prior events. The boy is adopted and no family history is known. After 2 hours of observation, he is alert and responding normally to commands, but complains of a headache and vomits.

Of the following, the MOST likely diagnosis is

A. acute psychosis
B. carbon monoxide poisoning
C. confusional migraine
D. postictal state
E. pseudotumor cerebri

Item 12 Preferred Response: C

The boy described in the vignette has a confusional migraine. Confusional migraine is a subtype of migraine headache characterized by the abrupt onset of an altered level of consciousness. The child appears disoriented and is sometimes agitated or combative; the symptoms last hours before recovery. Often, the episode is followed by a headache. A prior history of headaches or a family history of migraine headaches supports this diagnosis, but other causes of altered mental status need to be considered in the acute setting.

The boy in the vignette did not have a history of a stressful trigger that may have precipitated an acute reactive psychosis. Other causes of acute psychosis in a 10-year-old child include rare inborn errors of metabolism, porphyria, or toxin/medication exposures, but the boy in the vignette had no history consistent with this. Acute psychosis in this age rarely occurs because of childhood-onset schizophrenia.

The child also had no history of carbon monoxide poisoning and no other children from the classroom were affected. Pseudotumor cerebri presents with headache but not an altered level of consciousness. He had no history of seizures that would have caused a postictal state. Other causes of altered level of consciousness such as head injury, meningitis, or encephalitis were not suggested by the clinical presentation.

The initial evaluation of altered level of consciousness depends mostly on the history and physical examination findings. Once alternate causes have been ruled out, and especially with a history of prior headaches and a family history of migraines, confusional migraine can be considered. Confusional migraines recur, and on subsequent presentations, an extensive evaluation for alternate causes is not always necessary.

Confusional migraine is treated with the same approach as other migraine headaches. During the episode, acetaminophen, ibuprofen, fluids, and caffeine can help speed recovery. If episodes recur frequently or impair functioning, a prophylactic medication such as cyproheptadine can decrease the frequency and severity of both confusional migraine and migraine headache.

PREP Pearls

- Confusional migraine is characterized by an abrupt onset of altered consciousness.
- Initial evaluation of altered level of consciousness could include serum chemistries, toxicology, infectious evaluation, or central nervous system imaging based on the history and physical examination.

American Board of Pediatrics Content Specification(s):

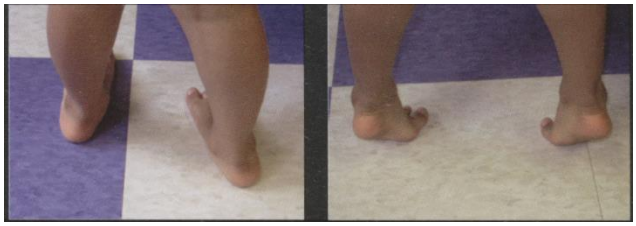
- Know the common causes of an altered level of consciousness
- Plan the initial phase of evaluation for an altered level of consciousness

Suggested Reading:

- Blume HK. Pediatric headache. Pediatr Rev. 2012;33(12):562-576. doi: 10.1542/pir.33-12-562
- Bechtel K. Acute mental status change due to acute confusional migraine. Pediatr Emerg Care. 2004;20(4):238-241

Item 13

A 4-year-old girl presents to your office for a health super-vision visit. Her parents are very worried about the appearance of her ankles. When she is sitting, the feet and ankles appear normal. However, when she walks, the parents observe that her ankles protrude inward and she seems to have "very flat feet" The child does not report any pain and has not had difficulty keeping up with her peers at play. The appearance of the girl's feet and ankles when standing on her toes is shown (Item Q13). The remainder of the examination is normal.



ITEM Q13: Feet as described for the girl in the vignette.

Of the following, the MOST appropriate management is

A. physical therapy for ankle strengthening
B. reassure the family regarding the physical findings
C. refer the girl to orthopedic surgery for possible surgical correction
D. use of rigid 3/ length custom shoe inserts
E. use of soft, full-length custom shoe inserts

Item 13 Preferred Response: B

Physiologic flat foot, also referred to as flexible flat foot or pes planus, is present in approximately 45% of preschool-age children. The child described in the vignette has asymptomatic physiologic flat foot and does not require any treatment. With physiologic flat foot, the arch seems to disappear when the child stands and becomes visible when the child goes up on his or her toes. Often, the heels deviate outward with standing ("hindfoot valgus") and the ankles appear to protrude medially. For most children, the arch develops spontaneously with growth. Braces, shoes, or shoe inserts have not been shown to facilitate arch development. As children get older, the fatty tissue over the arch disappears and the ligaments of the foot and ankle become less lax, making the arch more easily visible.

In about 1 in 7 people, the arch never develops. Factors that increase the likelihood of having persistent flat foot include obesity, ligamentous laxity, tight calf muscles, and a positive family history. The majority of children and adults with flexible flat foot are asymptomatic.

Children with flexible flat foot generally come to medical attention owing to parents' concerns about the appearance of the foot. Custom shoe inserts are not indicated for asymptomatic or minimally symptomatic patients; inserts are costly and may make children less comfortable. For children and teenagers reporting activity-related foot pain, shoe inserts may help alleviate symptoms; however, arch supports do not mold or alter the shape of the foot. Active children and teenagers often find very rigid inserts uncomfortable; soft, full-length inserts generally are tolerated better. Physical therapy for ankle strengthening may be helpful for children with ankle hypermobility and pain or with symptomatic flat foot associated with poor flexibility; however, physical therapy is not indicated for children with asymptomatic flat foot.

With pathologic, or rigid, flat foot, the ankles tend to be stiff and standing on one's toes does not make the arches visible. Individuals with rigid flat feet are more likely to become symptomatic and may require surgical treatment if pain does not resolve with a conservative approach. Tarsal coalition, an abnormal fusion between two of the tarsal bones, is one cause of rigid flat feet. Children with tarsal coalition typically present between the ages of 9 and 13 years with recurrent ankle sprains and activity-related foot pain.

PREP Pearls

- Flexible flat foot is associated with hindfoot valgus (often referred to as ankle pronation).
- Asymptomatic flexible flat foot is very common and does not require treatment.
- Accommodative shoe inserts ("orthotics") may help alleviate symptoms in children with flexible flat foot but will not change the structure of the foot.

American Board of Pediatrics Content Specifications:

- Know that no treatment for piano valgus is required in childhood
- Know that a longitudinal arch support may be helpful if piano valgus is painful for adolescents

Suggested Reading:

- Evans AM, Rome K. A Cochrane review of the evidence for nonsurgical interventions for flexible pediatric flat feet. Eur I Phys Rehabil Med. 2011;47(1):69-89
- Pfeiffer M, Kotz R, Ledl T, Hauser G, Sluga M. Prevalence of flat foot in preschool-aged children. Pediatrics. 2006;118(2):634-639. doi:10.1542/peds.2005-2126
- Rao UB, Joseph B. The influence of footwear on the prevalence of flat foot: a survey of 2,300 children. J Bone Joint Surg Br. 1992;74(4):525-527

Item 14

You are seeing a 2-year-old girl for a routine health super-vision visit. Her mother tells you that she is thinking about becoming pregnant, but she was diagnosed last year with bipolar disorder and is currently maintained on lithium. Her psychiatrist feels that lithium is her best option for medical management and has discouraged her from stopping this medication. She asks your advice about another pregnancy.
Of the following, you are MOST likely to tell her that

A. the decision whether to discontinue lithium requires a risk-benefit assessment since the risk of fetal anomaly is relatively low
B. lithium is a potent teratogen and she should not consider a pregnancy while on this medication
C. lithium use is primarily associated with cardiac and limb defects, which can be detected on second trimester ultrasonography
D. she can resume lithium after delivery and may breastfeed without concern because little is excreted into breast milk
E. she should discontinue lithium throughout the first trimester of pregnancy to avoid any adverse fetal effects

Item 14 TE Preferred Response: A

Although the risks for birth defects associated with lithium use in pregnancy are well-documented, the actual risk for birth defects, including Ebstein anomaly, are relatively low. The estimated risk for cardiac defects in fetuses with prenatal exposure to lithium is between 1% and 5%, compared to 0.5% to 1% for the general population. Therefore, the decision whether to discontinue this medication in pregnancy requires a risk-benefit decision in consultation with the woman's psychiatrist and obstetrician.

Lithium, like many potential teratogens, appears to have a dose-response curve that correlates with embryonic risks in all 3 trimesters. Hence, the lowest effective dose of medication is recommended. No teratogen can cause every type of malformation, and teratogenic effects often mimic single-gene disorders or known Mendelian syndromes.

Although controversy exists about the precise risk for birth defects, lithium has been classified as a Category-D (positive evidence of human fetal risk) teratogen. Lithium use in pregnancy continues to be advisable under certain circumstances. Second-trimester, level II ultrasonography along with a fetal echocardiogram may help identifying infants with major congenital anomalies but cannot detect all problems or defects associated with this teratogen. Infants with lithium levels greater than 1 mEq/L at birth seem to be at greatest risk for congenital goiter (which may begin to develop in the second trimester) and nephrogenic diabetes insipidus as well as transient hypothermia, cyanosis, bradycardia, shallow respirations, poor suck, hypotonia, and altered T waves on electrocardiography immediately after birth. Therefore, it is reasonable for a mother to discontinue lithium 24 to 48 hours before delivery (when feasible) and restart this medication after childbirth. Lithium is excreted into breast milk, and breastfed infants with lithium levels approaching 0.6 mEq/L have been noted to also experience cyanosis, lethargy, hypotonia, and poor feeding. As a result of these findings, the American Academy of Pediatrics has indicated that lithium use by a mother is a contraindication to breastfeeding.

In light of the above, lithium use in pregnancy poses some risks but is not completely contraindicated during pregnancy. It is associated with an increased risk for a few birth defects as well as other problems that may develop in the second trimester, and may also cause transient feeding and respiratory problems at birth. Mothers who require lithium to address a serious psychiatric disorder should not breastfeed while taking this medication.

PREP Pearls

- Lithium can be used with caution during pregnancy for patients who need this medication but should be used at the lowest effective dose to reduce fetal risks.
- Lithium use is not compatible with breastfeeding.

American Board of Pediatrics Content Specification(s):

- Know the effect on a fetus of lithium use during pregnancy

Suggested Reading:

- Brent RL. Environmental causes of human congenital malformations: the pediatrician's role in dealing with these complex clinical problems caused by a multiplicity of environmental and genetic factors. Pediatrics. 2004;113(suppl 3):957-968
- Mone SM, Gillman MW, Miller TL, Herman EH, Lipshultz SE. Effects of environmental exposures on the cardiovascular system: prenatal period through adolescence. Pediatrics. 2004;113 (suppl 3):1058-1069

Item 15

A 2-year-old boy is brought to the emergency department by ambulance. Thirty minutes ago, his mother discovered him eating pills from a pillbox at his grandparents' house and called 911. The boy's mother states that the pillbox contained a 1-week supply of his grandparents' daily medications. She did not count how many tablets remained in the box before the ambulance arrived, but she states, "I think only a couple were missing:' Both grandparents take "blood pressure medicine" and that the grandfather takes "a pill for his nerves"

The boy is well-appearing and playful. His vital signs are normal for his age, and you note no abnormalities on physical examination. The mother states that she now feels "silly for panicking over nothing. ' She asks you how soon she can take her son home.

Of the following, the BEST next step in managing this patient is

A. administration of activated charcoal at 1 g/kg
B. administration of intravenous normal saline at 20 mL/kg
C. discharge the boy after educating his mother about signs and symptoms to observe for at home
D. observe the boy in the emergency department for development of symptoms over the next 6 hours
E. perform gastric lavage to remove ingested pill fragments from the stomach

Item 15 S Preferred Response: A

The child described in this vignette was seen ingesting the contents of a pillbox thought to contain an antihypertensive agent and an antidepressant, therefore, administration of activated charcoal is warranted to decrease absorption of these toxins. Activated charcoal minimizes absorption of drugs by binding them onto its surface; it has become the gastrointestinal (GI) decontamination strategy of choice in pediatric patients and is most effective when administered within the first hour after a toxic ingestion. The dose of activated charcoal is 1 g/kg. Activated charcoal is contraindicated in patients with an unprotected airway, patients with a disrupted GI tract, or patients in whom charcoal therapy may increase the risk and severity of aspiration, such as in those ingesting a hydrocarbon. Substances that are poorly adsorbed by activated charcoal include common electrolytes, heavy metals such as iron, alcohols, cyanide, most solvents, and most water-insoluble compounds. For the asymptomatic boy described in the vignette, no contraindications for activated charcoal administration exist.

Poisoning represents one of the most common medical emergencies encountered by young children and is responsible for a significant proportion of emergency department visits in the adolescent population. More than 2 million toxic exposures are reported to the American Association of Poison Control Centers Toxic Exposure Surveillance System each year. Two-thirds of these exposures occur in individuals younger than 20 years of age, with half occurring in children younger than 6 years. All physicians caring for children must be familiar with the evaluation and management of poisoning. While poisonings in young children are usually unintentional, poisonings in adolescents and young adults generally result from substance abuse, experimental risk-taking behaviors, and depression or suicidal intent.

Numerous factors place young children at risk for unintentional poisonings. Between 1 and 2 years of age, most children learn to walk and develop the dexterity to use a pincer grasp; a common way that children within this age group explore their environments is by placing objects in their mouths. Young children like to mimic actions that they have seen their family members perform, such as using household products and taking medications. Furthermore, a number of household products and medications are brightly colored and may even resemble candy, making them particularly attractive to children.

Most substances that young children are exposed to within their home environments, such as cosmetics and personal care items, are nontoxic. Even among the significant percentage of toxic exposures involving drugs, small quantities of most agents ingested by a child require little treatment beyond reassurance. A few medications, however, can be lethal to small children in quantities of only 1 or 2 pills or teaspoon-sized swallows.

Pediatric practitioners need to be familiar with the drug classes from which "one pill can kill" when ingested by a toddler. These classes include cardio-vascular drugs (eg, -blockers and calcium-channel antagonists), antidepressants, antipsychotics, anticonvulsants, antiarrhythmic agents, salicylates, oral hypoglycemics, and opioids, all of which are widely prescribed for adults. When drugs from these classes are involved,

proper evaluation and intervention are essential for preventing severe toxic effects and even death in small children.

The first priority in managing any child who has ingested a toxic substance is to ensure stability of the airway and take any necessary steps to maintain adequate ventilation and circulation. The asymptomatic child who may have ingested only a few pills or swallows of an unknown toxin presents a clinical dilemma. A careful history, physical examination, and laboratory findings may narrow the differential diagnosis and facilitate an educated assessment of the potential severity of the exposure. In situations in which a child may have ingested a medication with potentially lethal effects, the most appropriate course of management is to decontaminate the child if no contraindications exist and to monitor closely for a period of time, depending on the poison that may have been ingested.

The boy described in the vignette is asymptomatic with normal vital signs; therefore, administration of intravenous normal saline is not warranted. Since, he may have ingested drugs with the potential to produce significant toxic effects within a few hours, GI decontamination and a period of observation are required before discharge. Although observation of the child for a period of several hours is warranted in this case, administration of activated charcoal would be the best initial step in management because activated char-coal is most efficacious within the first hour after ingestion. The clinical benefit of gastric lavage has not been confirmed in controlled studies, and its routine use in the management of poisoned patients is no longer recommended.

PREP Pearls

- Pediatric practitioners must recognize the drug classes from which "one pill can kill" when ingested by a toddler.
- The first priority in managing a possible toxic ingestion is to ensure stability of the airway and maintenance of adequate ventilation and circulation.
- Activated charcoal is the GI decontamination strategy of choice in pediatric patients with possible toxic ingestions and should be given as soon as possible provided there are no contraindications.

American Board of Pediatrics Content Specification(s):

- Understand the management of childhood poisonings
- Understand the management of poisonings by an unknown agent or by multiple agents

Suggested Reading:

- Braitberg G, Oakley E. Small dose ... big poison. Aust Fam Physician. 2010;39:826-833
- Osterhoudt KC. The toxic toddler: drugs that can kill in small doses. Contemp Pediatr. 2000;17:73
- Osterhoudt KC, Ewald MB, Shannon M, Henretig FM. Toxicologic emergencies. In: Fleisher GR, Ludwig S, eds. Textbook of Pediatric Emergency Medicine. 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2010:1171-1223
- Velez LI, Shepherd JG, Goto CS. Approach to the child with occult toxic exposure. UpToDate. Available online only for subscription

Item 16

A 19-month-old boy is seen in your office for follow-up after a diagnosis of anemia. One month ago, iron supplementation was initiated for treatment. At that time, his hemoglobin was 9.1 g/dL (91 g/L), hematocrit was 28% (0.28), and the mean corpuscular volume was 67 fL. Today, his hemoglobin is 11 g/dL (110 g/L), hematocrit is 33% (0.33), and the mean corpuscular volume is 71 fL. He eats a varied diet and continues to breastfeed well. His growth is normal.

Of the following, the BEST plan for management today is to

A. continue iron supplementation for 3 months
B. encourage more meat in the child's diet
C. have the mother take a multivitamin with iron tablet daily
D. offer reassurance and discontinue the iron supplementation
E. suggest weaning the child from breastfeeding

Item 16 Preferred Response: A

The most common nutritional deficiency in the United States is iron deficiency. It primarily affects older infants, young children, adolescent girls after menarche, and women during their childbearing years. Iron deficiency is the most likely cause of microcytic hypochromic anemia in a healthy breastfeeding toddler. The American Academy of Pediatrics Committee on Nutrition recommends that presumptive iron deficiency anemia be treated with oral (elemental) iron at a dose of 3 to 6 mg/kg per day for 4 weeks. Most commonly, ferrous sulfate (5 mg of ferrous sulfate = 1 mg of elemental iron) is used. After 4 weeks of supplemental iron, an increase in the hemoglobin concentration of more than 1 g/dL (10 g/L) or in the hematocrit of more than 3%, as seen in the boy described in the vignette, confirms the diagnosis of iron deficiency anemia. If this improvement does not occur, affirmation of adherence with medication recommendations, or further evaluation for possible blood loss should occur. Histamine 2 blockers and proton-pump inhibitors may decrease iron absorption by raising gastric pH.

If improvement occurs, as in this case, iron supplementation should continue for at least 2 months after the anemia has corrected to replenish the iron stores.
Anemia represents the most severe end of the iron-deficiency spectrum, which is a limitation of screening for iron deficiency by routine hemoglobin testing alone. Many studies have documented that early iron deficiency can lead to cognitive and motor deficits. Some of these deficits are not fully reversible with iron treatment. The potential for irreversible developmental delay, resulting from a temporary nutritional deficiency, underscores the importance of prevention, early diagnosis by screening, and complete treatment of iron deficiency.

Increasing iron intake by encouraging more meat in the patient's diet or through daily multivitamin with iron supplementation in his breastfeeding mother will not supply adequate additional iron above the daily requirement to replenish iron stores and correct deficiency. Discontinuing iron supplementation as soon as the anemia corrects may leave the patient iron deficient and at risk for suboptimal neurocognitive outcomes. Although breastfed infants need a supplemental source of iron (as either liquid supplement or high-iron food) introduced as early as 4 to 6 months of age, discontinuing breastfeeding is not recommended for the treatment of iron deficiency anemia.

PREP Pearls

- Iron deficiency is the most common nutritional deficiency in the United States.
- Presumptive iron deficiency anemia may be treated with oral (elemental) iron at a dose of 3 to 6 mg/kg per day for 4 weeks.
- If improvement in the hemoglobin concentration and hematocrit occurs after 4 weeks of treatment with supplemental iron, iron deficiency is confirmed as the cause of anemia.
- Iron supplementation should continue for at least 2 months after the anemia has corrected to replenish the iron stores.

American Board of Pediatrics Content Specification:

- Know that treatment of iron deficiency with ferrous sulfate should continue after the hemoglobin concentration has returned to normal

Suggested Reading:

- American Academy of Pediatrics. Iron. In: Pediatric Nutrition Handbook. 6th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2009:403-422
- Kett J. Anemia in infancy. Pediatr Rev. 2012;33:186-187. doi:10.1542/pir. 33-4-186
- Segel G, Hirsh M, Feig S. Managing anemia in pediatric office practice: part 1. Pediatr Rev. 2002;23:75-84. doi:10.1542/pir.23-3-75
- Wu A, Lesperance L, Bernstein H. Screening for iron deficiency. Pediatr Rev. 2002;23:171-178. doi:10.1542/pir.23-5-171

Item 17

The parents of a 4-year-old boy have questions about their son's food allergies and what foods he will need to avoid when he starts kindergarten next year. They were advised that he was allergic to milk, egg, and soy products on blood testing (serum specific IgE) performed in infancy as part of an evaluation of atopic dermatitis. His eczema worsened when he consumed milk, but he has never eaten egg or soy. He has been strictly avoiding all these foods, and he outgrew his atopic dermatitis 2 years ago. You discuss your plan to reevaluate the status of the boy's food allergies with his parents.

Of the following and according to the natural history of food allergies, the MOST appropriate response is to advise the parents that in kindergarten their son may

A. be able to tolerate eggs but not milk and soy products
B. be able to tolerate milk and soy products but not egg products
C. be able to tolerate milk, egg, and soy products
D. need to avoid all milk, egg, and soy products
E. need to avoid egg and soy products but not milk products

Item 17 TE S Preferred Response: C

The child described in the vignette may be able to tolerate milk, egg, and soy products by the time he starts kindergarten. While studies are difficult to compare because of inconsistencies in the definition of food allergy and design limitations, review of the natural history indicates that most children with food allergies will eventually tolerate milk, egg, soy, and wheat. The time course of resolution of the food allergy varies by the individual food. Most milk, egg, and soy allergies are outgrown in the first decade of life, and many by the age of 5 years. Previous data suggested that almost all infants with milk allergy developed it in infancy and that clinical tolerance developed in 80% by age 5 years. On the other hand, a more recent study hinted at a lower rate of development of clinical tolerance to milk, with only 5% being tolerant at age 4 and 21% at age 8. Approximately 50% of children with soy allergy outgrow it by 6 years and 69% by age 10.

Prior studies had indicated that most infants with egg allergy become tolerant to egg at a young age with an estimated two-thirds of children becoming tolerant by age 7. Recent data show a trend toward slower resolution, with a retrospective review of a university allergy practice showing half of patients resolving by age 10 years and 80% by age 16 years. On the basis of these reassuring data, there is a high probability that the child in the vignette will be able to tolerate milk, egg, and soy products. In addition, serum-specific IgE (sIgE) testing in the absence of clinical correlation indicates sensitization but not necessarily allergy. Because the child has never ingested the suspected foods, it is possible that he was only sensitized but not allergic, suggesting a high likelihood of being able to tolerate all 3 foods

Although a high initial level of sIgE is associated with a lower rate of resolution of clinical allergy, a drop in sIgE levels over time is often a marker for the onset of tolerance to the food. Periodic reevaluation of food sIgE to monitor the trend in levels may help discriminate between children with persistent allergy (sustained elevation in levels) vs those who have a good chance of outgrowing the allergy (levels drop below predictive cut-off values). A skin prick test response to a food can remain misleadingly positive long after tolerance to the food has developed, although a reduction in the size of the skin prick test wheal, if present, may also be a marker for the onset of tolerance to the food. If the results suggest the possibility of having outgrown the allergy, a supervised double-blind placebo-controlled oral challenge (the gold standard test) will need to be performed in order to demonstrate achievement of tolerance.

PREP Pearls

- Most milk, egg, and soy allergies are outgrown by the first decade of life.
- Half of children with soy allergy outgrow it by 5 years.
- Half of children with egg allergy outgrow it by 10 years.

American Board of Pediatrics Content Specification(s):

- Know that most milk, egg, and soy allergies are outgrown

Suggested Reading:

- NIAID-Sponsored Expert Panel; Boyce JA, Assa'ad A, Burks AW, et al. Guidelines for the diagnosis and management of food allergy in the United States: report of the NrAID-Sponsored Expert Panel. J Allergy Clin Immunol. 2010;126(6 suppl):S1-S58. doi:10.1016/j.jaci.2010.10.007
- Savage JH, Kaeding AL Matsui EC, Wood RA. The natural history of soy allergy. JAllergy Clin Immunol. 2010;125(3):683-686. doi:10.1016/j. jaci.2009.12.994
- Savage JH, Matsui EC, Skripak JM, Wood RA. The natural history of egg allergy. JAllergy Clin Immunol. 2007;120(6):1413-1417. doi:10.1016/j. jaci.2007.09.040
- Shek LP, Soderstrom L, Ahlstedt S, Beyer K, Sampson HA. Determination of food specific IgE levels over time can predict the development of tolerance in cow's milk and hen's egg allergy. I Allergy Clin Immunol. 2004;114(2):387-391. doi:10.1016/j.jaci.2004.04.032

Item 18

A 15-year-old boy is seen for a health supervision visit. The patient is healthy with normal growth parameters and development. He has no significant past medical or past surgical history. Physical examination and vital signs are unremarkable. His urine analysis in the office reveals a specific gravity of 1.010, a pH of 6.0, 4+ protein, and no blood, leukocyte esterase, or nitrites. His parents recall that their son had protein in his urine during a sports physical last year.

Of the following, the BEST next step in management is

A. evaluation by a nephrologist
B. evaluation of a first morning urine sample
C. quantitative urine protein estimation in a 24-hour urine sample
D. recheck at the next health supervision visit
E. urine microalbumin estimation in a 24-hour urine sample

Item 18 PBLI Preferred Response: B

Differentiation between benign and pathologic causes of proteinuria is important because proteinuria may be the only indication of renal disease in asymptomatic patients. Persistent dipstick-positive proteinuria or a urine protein-creatinine ratio higher than 0.2 is considered abnormal.

Twenty-four hour urine collection, though still performed in adult patients with proteinuria, is not routinely performed in children with asymptomatic proteinuria. Besides the obvious issues related with a 24-hour urine collection especially in children, studies have shown that urine protein-creatinine ratio in a first-morning urine sample is as sensitive as 24-hour urine collection for detecting pathologic proteinuria. According to the Kidney Disease Outcome Quality Initiative guidelines from the National Kidney Foundation; it is not usually necessary to obtain timed/24-hour urine collections for evaluating proteinuria in adults or children.

The boy described in the vignette is asymptomatic and has normal growth parameters, which decrease his risk for an underlying chronic kidney disease. Given the patient's presentation, he most likely has orthostatic proteinuria (OP). OP occurs during the day when the patient is active and disappears when the patient is supine/asleep for at least 2 hours. Therefore to confirm OP, a first-morning urine sample is needed. It is important that the patient collect the first urine sample immediately on waking because even a small amount of activity can lead to proteinuria. A urine protein-creatinine ratio higher than 0.2 in a first-morning sample is abnormal and indicates renal pathology, which requires evaluation by a pediatric nephrologist.

Urine dipsticks become positive when urine albumin secretion is more than 300 mg/day. Normal urine albumin secretion is less than 30 mg/day. Thus, microalbuminuria (more recently termed high albuminuria) detects urine albumin secretion in the range of 30 to 330 mg/day. Studies on patients with diabetes have shown that microalbuminuria is the earliest manifestation of diabetic nephropathy. In addition to diabetes, microalbuminuria is known to be an early marker of cardiovascular disease-associated renal injury. Urine microalbuminuria testing should be restricted and used for early detection of renal injury secondary to conditions associated with chronic renal failure.

PREP Pearls

- Persistent dipstick-positive proteinuria or a urine protein-creatinine ratio higher than 0.2 indicates renal pathology, which requires evaluation by a pediatric nephrologist.
- A first-morning urine sample is necessary to evaluate for orthostatic proteinuria.
- It is not usually necessary to obtain timed/24-hour urine collections for evaluating.
- Microalbuminuria testing should be restricted and used for early detection of renal injury secondary to chronic conditions.

American Board of Pediatrics Content Specification(s):

- Recognize the limitations of 24-hour urine collections in pediatric patients

Suggested Reading:

- Hogg RJ, Portman RJ, Milliner D, Lemley KV, Eddy A, Ingelfinger J. Evaluation and management of proteinuria and nephrotic syndrome in children: recommendations from a pediatric nephrology panel established at the National Kidney Foundation conference on proteinuria, albuminuria, risk, assessment, detection, and elimination (PARADE). Pediatrics. 2000;105(6):1242-1249. doi: 10.1542/peds.105.6.1242
- Houser MT. Assessment of proteinuria using random urine samples. Pediatr. 1984;104: 845-848. doi: 10.1016/S0022-3476(84)80478-3
- KDOQI Clinical practice guidelines for chronic kidney disease: evaluation, classification, and stratification: part 5—evaluation of laboratory measurements for clinical assessment of kidney disease
- National Kidney Foundation Web site www.kidney.org
- Sebestyen JF, Alon US. The teenager with asymptomatic proteinuria: think orthostatic first. Clin Pediatr. 2011;50:179-182. doi:10.1177/0009922810380904

Item 19

A family living in the United States comes to your office for the initial medical evaluation of their 2-year-old daughter who they recently adopted from Southeast Asia. The limited past medical history is unremarkable. The parents report that the child is eating, voiding, and stooling normally, and they have no specific concerns about her health or behavior.

On physical examination, vital signs are normal for age, and growth parameters are in the 25th percentile for age. Complete physical examination findings are normal. The peripheral white blood cell count is 15,000/µL (15.0 x 10^9/L), with 40% polymorphonuclear neutrophils, 20% lymphocytes, 10% monocytes, and 30% eosinophils. Hemoglobin is 10 g/dL (100 g/L), and platelet count is 260 x 10^3/µL (260 x 10^9/L).

Of the following, the test that is MOST likely to yield an abnormal result is

A. hepatitis B antigen test
B. human immunodeficiency virus antibody test
C. stool culture
D. stool for ova and parasites
E. tuberculin skin test

Item 19 Preferred Response: D

Internationally adopted children should have a stool ova and parasites (O&P) test and a complete blood cell count with differential performed as a part of their routine screening process. The asymptomatic child described in the vignette has hypereosinophilia (eosinophil count >1,500/4). There are varying degrees of eosinophilia (Item C19A) with many possible associated disorders (Item C19B). The eosinophilia in this child is most likely due to a helminthic parasite infection acquired in Southeast Asia. Therefore, 3 stool specimens should be examined for O&P. Pathogens such as Ascaris and hookworm can also be detected by stool O&P. Strongyloides stercoralis is particularly prevalent in Southeast Asia, and diagnosis often requires serologic testing if the fecal microscopy result is negative. Internationally adopted children should have Strongyloides and Schistosoma serologic testing performed if they have an eosinophil count greater than 450/4 and 3 negative stool O&P examination results. Giardia intestinalis (formerly G lamblia) and Cryptosporidium species can also be detected with stool O&P, but neither is associated with peripheral eosinophilia. A stool culture for bacterial pathogens would not be helpful in evaluating this child with asymptomatic eosinophilia and is not routinely recommended for internationally adopted children.

Item C19A. Degrees of Eosinophilia

Severity	Eosinophil Level (/µL)
Mild	500 – 1500
Moderate	1501 – 5000
Severe	>5000

Item C19B. Disorders Associated With Eosinophilia

Category	Examples
Allergic/atopy	Asthma, atopic dermatitis, allergic rhinitis, medication reaction
Endocrine	Hypoadrenalism
Hematologic/neoplastic	Hypereosinophilic syndrome, leukemia, lymphoma, tumor
Immunologic	Specific immunodeficiency, graft-versus-host disease
Infectious	Parasitic (especially helminths) infection, certain fungi (eg, *Aspergillus*), human immunodeficiency, virus, mycobacterial disease
Specific diseases with organ involvement	Churg-Strauss, inflammatory bowel disease, systemic lupus erythematosus

Hepatitis B (hepatitis B surface antigen test) and hepatitis C virus serologic testing are recommended for all international adoptees, however infections with these virus is not associated with peripheral eosinophilia. Although human immunodeficiency virus (HIV) infection can be associated with eosinophilia and HIV testing is recommended for all international adoptees, HIV would be unlikely in this asymptomatic child because most HIV-associated eosinophilia occurs with advanced infection. Likewise, peripheral eosinophilia may be associated with advanced, not latent, tuberculosis infection, so a tuberculin skin test would not likely explain the results of this patient's complete blood cell count.

However, all internationally adopted children should be tested for tuberculosis regardless of the results of their complete blood cell count. Additional recommended testing of international adoptees includes syphilis serologic testing (both nontreponemal [eg, rapid plasma reagin or VDRL test] and treponemal [eg, Treponema pallidum particle agglutination assays or microhemagglutination assay or fluorescent treponemal antibody absorption] tests), serologic testing for Trypanosoma cruzi (Chagas disease) in children from Mexico and Central and South America, and

serologic testing for lymphatic filariasis (Wuchereria bancrofti, Brugia malayi, and Brugia timori) in children older than 2 years with eosinophilia and from countries of origin in the tropics and subtropics of Africa, Asia, the Western Pacific, and parts of South America and the Caribbean.

PREP Pearls

- All internationally adopted children should be screened for the following infectious diseases:
  - Hepatitis B
  - Hepatitis C
  - Syphilis
  - Human immunodeficiency virus
  - Giardia intestinalis
  - Cryptosporidium species
  - Tuberculosis
  - Trypanosome cruzi (if from endemic area)
  - Lymphatic filariasis (if from endemic area)
  - Strongyloides species (if eosinophilia present)
  - Schistosoma species (if eosinophilia present and from endemic area)
- Internationally adopted children with peripheral eosinophilia should have 3 stool specimens examined for O&P.

American Board of Pediatrics Content Specification (s):

- Know the recommendations for screening for infectious diseases in internationally adopted children

Suggested Reading:

- American Academy of Pediatrics. Medical evaluation of internationally adopted children for infectious diseases, In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012: 191-197
- Centers for Disease Control and Prevention. Intestinal parasite guidelines for domestic medical examination for newly arrived refugees

Item 20

A 17-year-old, sexually active boy has complaints of intermittent burning with urination for the last 2 weeks. He says he sometimes sees some staining on his underwear but has not noticed any penile discharge or genital lesions. He reports that he has never had a sexually- transmitted infection and that he always uses condoms. He is otherwise healthy and has no systemic complaints, hematuria, or urgency.
On physical examination, he is at sexual maturity rating 5 for pubertal development. Other than some moistness at the urethral meatus, his genital examination findings are normal.

Of the following, the organism MOST likely responsible for this boy's symptoms is

A. Chlamydia trachomatis
B. Escherichia coli
C. Mycoplasma genitalium
D. Neisseria gonorrhoeae
E. Ureaplasma urealyticum

Item 20 Preferred Response: A

Dysuria and discharge experienced by the boy described in the vignette is indicative of urethritis. Occasionally, penile itching and tingling may occur without discharge; urinary frequency, urgency, or rarely, hematuria may also be present. The organisms responsible for urethritis include those that cause urinary tract infections and those that may be sexually transmitted. Almost half of all cases are caused by Chlamydia trachomatis, the organism most often responsible for nongonococcal urethritis. Usually, C trachomatis urethritis presents with a scant clear discharge; however, infection may be asymptomatic or cause a profuse purulent discharge, as is seen in disease caused by Neisseria gonorrhoeae. The frequency of C trachomatis urethritis is almost 3 times that of N gonorrhoeae. Because both Chlamydia and gonorrhea are reportable diseases and partner notification and treatment may be improved with a specific diagnosis, testing for these organisms should be performed in all patients. This is best accomplished using a nucleic acid amplification test performed on a urine specimen.

Treatment goals include symptom relief, preventing transmission to sexual partners, and reducing complications; untreated C trachomatis may lead to epididymitis and, rarely, prostatitis or reactive arthritis. Empiric treatment is with azithromycin, 1 g orally, as a single dose for C trachomatis and ceftriaxone, 250 mg intramuscularly, for N. gonorrhoeae. Patients should be instructed to abstain from sexual intercourse for a week after single-dose therapy (to minimize sexual transmission) and until all partners are treated (to prevent reinfection). Testing the patient for other infections, including syphilis and human immunodeficiency virus, should be undertaken.

In addition to C trachomatis and N gonorrhoeae, other agents causing nongonococcal urethritis include Mycoplasma genitalium, Ureaplasma urealyticum, Trichomonas vaginalis, herpes simplex virus (HSV), and adenovirus. Diagnostic testing for these organisms is not recommended in the setting of urethritis; however, testing for HSV is recommended if genital ulcers are present. Urinary tract infections are less common in men than women and are not associated with a penile discharge. Escherichia coli is the most common urinary tract pathogen and is an occasional cause of urethritis. In addition to poor hygiene, sexual practices like unprotected insertive anal intercourse may predispose patients to acquisition of other organisms. Rarely, there may be noninfectious causes of urethritis, which are less likely to affect men. These causes include chemical irritation caused by soaps, lotions, and colognes, which may cause temporary urethral pain. Contraceptive jelly, cream, or foam, as well as spermicide in condoms, can also cause irritation. Mechanical manipulation of the penis with vigorous sexually activity or masturbation can cause a temporary irritation of the urethra.

PREP Pearls

- Dysuria and discharge in males are the most common symptoms of urethritis.
- Chlamydia trachomatis is the most common etiologic agent of urethritis. Treatment includes azithromycin or doxycycline.
- Remember to prescribe treatment for the sexual partner(s).

American Board of Pediatrics Content Specifications:

- Know the differential diagnosis and etiology of urethritis in adolescent boys
- Know the appropriate treatment of urethritis in adolescent boys

Suggested Reading:

- Brill JR. Diagnosis and treatment of urethritis in men. Am Fam Physician. 2010;81:873-878
- Centers for Disease Control and Prevention. Diseases characterized by urethritis and cervicitis. MMWR Morb Mortal Wkly Rep. 2010;(RR-12):40-43
- Chandran L, Boykan R. Chlamydia) infections in children and adolescents. Pediatr Rev. 2009;30:243-250. doi:10.1542/pir.30-7-243
- Fortenberry JD, Neinstein LS. Overview of sexually transmitted diseases. In: Neinstein LS, Gordon CM, Katzman DK, Rosen DS, Woods ER, eds. Adolescent Health Care: A Practical Guide. 5th ed. Philadelphia, PA: Lippincott Williams & Wilkins, a Wolters Kluwer business; 2008:60:767-770
- Marcell AV, Wibbelsman C, Seigel WM, and the Committee on Adolescence Clinical Report. Male adolescent sexual and reproductive health care. Pediatrics. 2011;128:el658-el676

Item 21

A 14-year-old girl complains of nausea and right shoulder pain. She was well until 6 weeks ago, when she began to experience pain and swelling of the right knee while on vacation. She was evaluated at a local hospital, diagnosed with Lyme disease on the basis of her history and physical examination, and started on a course of ceftriaxone. Since that time, she has had some exercise intolerance and occasional knee pain. Recently, she has experienced postprandial nausea, without vomiting and for the past week has had discomfort in her right shoulder. She denies vomiting, diarrhea, jaundice, and abdominal pain. Physical findings demonstrate a well-developed, well-nourished adolescent. The remainder of the physical examination is unremarkable. You order laboratory tests that produce the following results:

- Hemoglobin, 12.8 g/dL (128 g/L)
- White blood cell count, 6,500/µL (6.5×10^9/L)
- Aspartate aminotransferase, 125 U/L; reference range, ≤40 U/L
- Alanine aminotransferase, 35 U/L; reference range, ≤30 U/L
- Total bilirubin, 2.0 mg/dL (34.2 pmol/L)

Results of the girl's urinalysis are normal.

Of the following, the MOST appropriate next step in evaluating this patient is

A. abdominal ultrasonography
B. magnetic resonance cholangiopancreatography
C. serum γ-glutamyltransferase
D. upper gastrointestinal endoscopy
E. Western blot testing for Lyme disease

Item 21 Preferred Response: A

The case presented in the vignette illustrates the importance of having an index of suspicion based on knowledge of adverse drug effects and a history of seemingly nonspecific symptoms. The girl was treated for Lyme disease with ceftriaxone, a third-generation cephalosporin and now complains of postprandial nausea and right shoulder pain. These vague symptoms are often associated with gallbladder and biliary tract disease, particularly cholelithiasis. Laboratory studies reveal a mild hyperbilirubinemia and transaminase elevation, further supporting this possibility. Ceftriaxone has been associated with the development of gallbladder sludge or stones after prolonged use. Although ceftriaxone-induced biliary precipitates are usually reversible after discontinuing use of the antibiotic, reports indicate that so-called ceftriaxone pseudolithiasis may simulate acute or chronic cholecystitis. In some patients, endoscopic management may be required to relieve common bile duct obstruction. On the basis of this information, the most appropriate next diagnostic study should be abdominal ultrasonography focusing on the biliary tract to evaluate the presence of echogenic foci indicating gallstones.

Cholelithiasis is an uncommon diagnosis in pediatrics. However, its frequency has been increasing because of the emerging obesity epidemic. The major risk factors for development of gallstones during childhood are presented in Item C21. Of these, cholelithiasis occurs with the highest prevalence in patients with sickle cell disease, with up to 40% of children with this diagnosis developing gallstones by 20 years of age. Although gallstones are rarely "pure;' comprising mixtures of calcium, bilirubin, cholesterol, and other substances secreted in bile, they are generally classified as follows:

- Cholesterol stones, which result from increased secretion of cholesterol into bile (common associated conditions include hyperlipidemia, pregnancy, obesity, and female sex) Black pigment stones, secondary to increased secretion of conjugated bilirubin into bile (hemolytic disease, ileal resection, Crohn disease, cystic fibrosis with pancreatic insufficiency, and total parenteral nutrition)
- Brown pigment stones, which form as a consequence of biliary stasis and increased mucin production (bacterial infection and parasitic infection)

A biliary tract ultrasonography should be the diagnostic study of first choice in the evaluation of possible cholelithiasis. If the results are inconclusive and a stone is still suspected, magnetic resonance cholangiopancreatography will provide enhanced views of the biliary tract. When an obstructing common duct stone is suspected, located at or near the ampulla of Vater, and associated with signs of cholestasis (direct hyperbilirubinemia, and elevated alkaline phosphatase and γ-glutamyltransferase levels) and often with obstructive pancreatitis (increased amylase and lipase levels), the newer technology of endoscopic ultrasonography may be required to achieve adequate stone visualization. Upper gastrointestinal tract endoscopy is of no value in the evaluation of presumed cholelithiasis, and confirming a Lyme disease diagnosis by Western blot testing will not assist in this evaluation. The gamma-glutamyltransferase level may be elevated if

cholestasis is present, but of the choices provided it would not be the best next step in evaluation.

PREP Pearls

- Ceftriaxone is commonly associated with pseudolithiasis and cholestasis.
- Biliary ultrasonography is the test of first choice when evaluating suspected cholestasis.

American Board of Pediatrics Content Specification(s):

- Know the risk factor associated with the development of cholelithiasis

Item C21. Major Risk Factors for Cholelithiasis in Children

- Obesity
- Female sex (postpubertal)
- Hemolysis
  - Sickle cell disease
  - Hereditary spherocytosis
  - Thalassemias
  - Wilson disease
- Ileal disease
  - Crohn disease
  - Ileal resection
- Cystic fibrosis with pancreatic insufficiency
- Total parenteral nutrition

  Drugs
  - Ceftriaxone

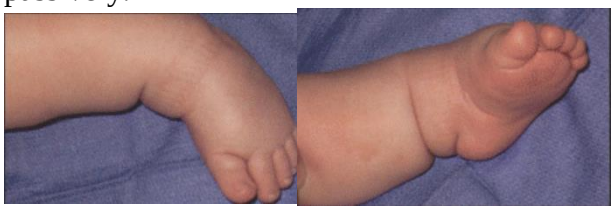
    Cyclosporine
  - Furosemide
  - Octreotide
  - Oral contraceptives
- Trisomy 21
- Hyperlipidemia
- Pregnancy
- Native American origin (Pima, Hopi, Araucan)

Suggested Reading:

- Broderick A. Gallbladder disease. In: Kleinman RE, Sanderson IR, Goulet 0, Sherman PM, Mieli-Vergani G, Shneider BL, eds. Walker's Pediatric Gastrointestinal Disease: Pathophysiology, Diagnosis and Treatment. 5th ed. Hamilton, Ontario, Canada: BC Decker, Inc; 2008:1173-1183
- Kaechele V, Wabitsch M, Thieve D, et al. Prevalence of gallbladder stone disease in obese children and adolescents: influence of the degree of obesity, sex, and pubertal development. J Pediatr Gastroenterol Nutr. 2006;42:66-70
- Kumar R, Nguyen K, Shun A. Gallstones and common bile duct calculi in infancy and childhood. Aust N Z J Surg. 2000;70:188-191
- Lebovics E, Halata MS, Rosario JA, Lantin J, Schwarz SM, Rosenthal WS. Endoscopic management of ceftriaxone pseudolithiasis involving the common bile duct and gallbladder. Gastrointest Endosc. 1994;40:246-248
- Wesdorp I, Bosman D, de GraaffA, Aronson D, van der Blij F, Taminiau J. Clinical presentations and predisposing factors of cholelithiasis and sludge in children. J Pediatr Gastroenterol Nutr. 2000;31:411-417

Item 22

Foot abnormalities are noted in a full-term newborn who is otherwise healthy. The ankle and forefoot are in cavus and adductus position, with the hindfoot varus and the ankle plantar flexed (equinus) (Item Q22). This condition cannot be corrected with positioning passively.



ITEM Q22: Findings as described for the infant in the vignette.

Of the following, the MOST appropriate management plan is

A. immediate referral to orthopedics for manipulation and serial casting
B. observation with follow-up in 1 month to determine if flexibility improves
C. physical therapy consult to teach the parents stretching exercises for the foot and ankle
D. placement in a bar and brace orthotic to hold feet abducted and dorsiflexed
E. referral to orthopedics at 1 month of age for anticipated tendon release surgery

Item 22 TE I-C Preferred Answer: A

The infant described in the vignette has the classic features of talipes equinovarus (clubfoot) and it is recommended that treatment begin immediately after birth. Clubfoot or talipes equinovarus is a common congenital deformity affecting approximately 1 per 1,000 live births. Boys are affected twice as often as girls, and in half the cases the deformity is bilateral. A family history of clubfoot increases the risk of subsequent affected infants, but the inheritance pattern is uncertain. Eighty percent of the deformities are isolated whereas 20% are associated with other conditions, including spina bifida, cerebral palsy, and arthrogryposis.

Although talipes equinovarus involves the entire foot, talar anomalies appear to be the major underlying deformities. On physical examination, the hindfoot is in varus (the heel turning in) and equinus (the foot points downward at the ankle). The forefoot and midfoot are adducted whereas the first metatarsal ray is plantar flexed; together this produces the cavus appearance characteristic of the distal foot.

The Ponseti method of weekly manipulation and casting is the most widely used treatment in North America. First developed in the 1940s, it involves a specific sequence of manipulations and stretching of immature collagen into a maximal correction that is then held in position with a cast. After 6 or more weeks of sequential manipulation and casting, the midfoot and forefoot are usually corrected. Persistent equinus deformity often (85%) requires Achilles tenotomy, which may be done in the orthopedist's office. This is followed by 3 weeks in a final cast. If correction is adequate, the patient is then placed in a "boots and bar" orthotic device that holds the feet externally rotated and dorsiflexed.

For the initial 3 months, the orthosis is used continuously. After 3 months, it is used only during sleep until age 3 to 4 years. Occasionally the deformity may recur, and manipulation and casting with or without tibialis anterior tendon transfer may be reinstituted. Although some reports indicate that delayed initiation of the Ponseti method may still be successful, immediate treatment is still the recommended approach.

Alternatives approaches include early surgical treatment and the French physiotherapy method. Studies directly comparing the Ponseti method to early surgery are limited, but outcomes with surgical management appear to be worse than with the Ponseti method. The French physiotherapy method is very resource intensive, requiring daily treatments by a therapist or physician. 'Outcomes are similar to those with the Ponseti method except for the most severely affected children in whom the Ponseti method produces better results. More extensive surgical procedures may also be required to achieve the best long-term result after initial treatment with the French method. In a child with a fixed deformity, passive stretching by the parents at home is unlikely to result in improvement. The brace and bar orthotic, as described above, is used after rather

PREP Pearls

- Talipes equinovarus is characterized by fixed in-turning of the hindfoot, plantar flexion at the ankle, and cavus appearance of the distal foot (adduction of the fore- and midfoot with first metatarsal ray plantar flexion).
- In cases of clubfoot, the Ponseti method of serial manipulation and casting should be initiated as soon after birth as possible.
- Outcomes for serial manipulation and casting appear to be better than those of a strictly surgical approach.

American Board of Pediatrics Content Specification(s):

- Recognize that the treatment for talipes equinovarus is casting or splinting of the affected foot
- Know that the most common component of clubfoot is equinovarus deformity
- Be aware that early treatment of clubfoot is critical

Suggested Reading:

- Bridgens J, Kiely N. Current management of clubfoot (congenital talipes equinovarus). BMJ. 2010;340:c355-359. doi:10.1136/bmj.c355
- Carroll NC. Clubfoot in the twentieth century: where we were and where we may be going in the twenty-first century. J Pediatr Orthop B. 2011;21:16. doi: 10.1097/BPB.0b013e32834a99f2
- Chotel F, Parot R, Seringe R, Berard J, Wicart P. Comparative study: Ponseti method versus French physiotherapy for initial treatment of idiopathic clubfood deformity. J Pediatr Orthop. 2011;31:320-325
- Van Bosse H. Ponseti treatment for clubfeet: an international perspective. Curr Opin Pediatr. 2011;23:41-45. doi:10.1097/MOP.ObO]3e328342112a

Item 23

A 15-year-old boy who is a patient in your practice collapsed during physical education class. He had no heart rate when he was first assessed, and cardiopulmonary resuscitation (CPR) was initiated promptly. An automatic external defibrillator was attached and detected an initial rhythm of ventricular fibrillation. A single shock was delivered with return of circulation and shallow breathing. The patient was transported to the local hospital and is now in the pediatric intensive care unit and receiving mechanical ventilation. After seeing patients in the office, you go to the hospital to check on the boy's condition and talk with his parents. The father, who is an internal medicine physician, asks you if their son should be "cooled" as a therapy to help preserve brain function.

Of the following, the MOST accurate statement regarding therapeutic hypothermia following cardiac arrest is that

A. the goal temperature should be below 30°C
B. it has been demonstrated to be beneficial for all age groups
C. it has been demonstrated to be effective only after traumatic brain injury
D. it should be continued for a minimum of 5 days
E. there are no completed randomized pediatric trials in this age group

Item 23 PBLI Preferred Response: E

Pediatric patients have poor outcomes of cardiac arrest, with approximately 35% of children surviving to hospital discharge after in-hospital cardiac arrest and fewer than 10% surviving after out-of-hospital cardiac arrest. In the United States, it is estimated that approximately 2,000 patients younger than 25 years die each year of sudden cardiac arrest. Hypoxic-ischemic injury is an important cause of post arrest morbidity and mortality and occurs after periods of impaired perfusion. Irreversible central nervous system injury may occur after as little as 3 to 5 minutes of interrupted blood flow or oxygen delivery. Both ischemia and hypoxia trigger numerous pathophysiologic processes that result in cellular injury and death and subsequent development of cerebral edema that compromises blood flow. Although many investigational therapies have been studied, the cornerstone of therapy remains supportive care, with assurance of adequate perfusion and oxygenation.

Therapeutic hypothermia (also known as targeted temperature management) is being used increasingly in neonatal and adult critical care settings after several studies showed improved outcomes after hypoxic-ischemic injury caused by birth asphyxia (neonates) or cardiac arrest secondary to ventricular fibrillation (adults). Although it would seem logical that children would benefit as well, to date no randomized trials have documented efficacy. A retrospective study of pediatric patients who experienced cardiac arrest demonstrated worse outcomes in those who were treated with therapeutic hypothermia but no differences in mortality were seen after adjusting for severity of injury. Currently a multicenter randomized trial to assess clinical effectiveness is being conducted but it will not be completed until 2015. In therapeutic hypothermia, the goal temperature is 32°C to 34°C (lower temperatures increase the risk of coagulation abnormalities, infectious complications, and arrhythmias) and treatment is continued for 72 hours. Early studies in treatment of pediatric traumatic brain injury have not demonstrated a benefit of therapeutic hypothermia.

Cerebral edema can result from ischemia and hypoxia and the resultant increased intracranial pressure is a medical emergency that must be recognized promptly to prevent potentially permanent neurologic injury or death. Physical examination findings in infants include a full or bulging fontanelle, widened sutures, and increasing head circumference, as well as findings that may be observed in older children, including sluggish or unequal pupils, impaired or absent upward gaze of the eyes ("sunsetting"), and papilledema. Changes in motor tone with eventual asymmetric movements and progression to decerebrate or decorticate posturing may be seen. The heart rate and blood pressure, which may improve cerebral blood flow, may be increased. The Cushing triad (hypertension, bradycardia, and abnormal respiration) is a late finding that usually indicates imminent brainstem herniation.

PREP Pearls

- Outcomes of pediatric cardiac arrest are poor, with fewer than 10% surviving after out-of-hospital cardiac arrest.
- Irreversible brain injury may occur after 3 to 5 minutes of interrupted blood flow or oxygen delivery.

- Cerebral edema must be recognized and treated promptly to prevent potentially permanent neurologic injury or death.
- Currently no guidelines exist for therapeutic hypothermia in the pediatric age group.

American Board of Pediatrics Content Specification(s):

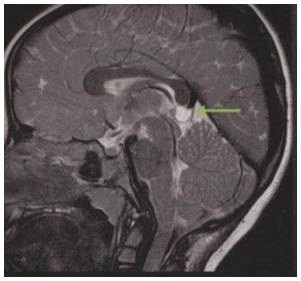
- Recognize cerebral edema in an asphyxiated patient

Suggested Reading:

- American Academy of Pediatrics Section on Cardiology and Cardiac Surgery. Policy statement: pediatric sudden cardiac arrest. Pediatrics. 2012;129:e1094-e1102. doi: 10.1542/peds.2012-0144
- Doherty DR, Parshuram CSo-Gaboury I, et al. Hypothermia after pediatric cardiac arrest. Circulation. 2009;119:1492-1500. doi: 10.1161/ circulationaha.108.791384
- Httchison JS, Ward RE, Lacroix J, et al. Hypothermia therapy after traumatic brain injury in children. N Engl I Med. 2008;358(23):2447-2456. doi:10.1056/NEJMoa0706930
- Kleinman ME, De Caen AR, Chameides L, et al; Pediatric Basic and Advanced Life Support Chapter Collaborators. 2010 International consensus on cardiopulmonary resuscitation and emergency cardiovascular care science with treatment recommendations: part 10—pediatric basic and advanced life support. Circulation. 2010;122:5466-5515. doi:10.1161/circulationaha.110.971093
- Nunnally ME, Jaeschke R, Bellingan J, et al. Targeted temperature management in critical care: a report and recommendation from five professional societies. Crit Care Med. 2011;39:1113 -1125

Item 24

A 5-year-old girl is noted to have breast budding. She has no pubic hair, axillary hair, deepening of her voice, or acne. Laboratory evaluation reveals an elevated baseline luteinizing hormone level of 4.6 mIU/mL (4.6 IU/L). Brain magnetic resonance imaging shows a large pineal gland cyst (Item Q24). Her mother is worried her daughter will have menses at a very young age.



ITEM Q24: Large pineal gland cyst for the girl described in the vignette.

Of the following, the BEST treatment for managing precocious puberty in this patient is

A. hydrocortisone
B. leuprolide acetate
C. medroxyprogesterone acetate
D. observation
E. tamoxifen

Item 24 I-C Preferred Response: B

Precocious puberty is the development of pubertal signs before the age of 8 years in girls and before the age of 9 years in boys and should warrant a referral to a pediatric endocrinologist. In 1999, the Pediatric Endocrine Society recommended new, lower age limits for girls: before 6 years for African Americans and before 7 years for whites. However, considerable controversy still exists concerning how best to define precocious puberty. A primary risk associated with precocious puberty is loss of final adult height due to early fusion of epiphyseal growth plates from sex steroid exposure. Many believe that there are additional psychosocial risks of precocious puberty. In some studies, precocious puberty has been associated with significant psychological stress due to appearing different from peers. Other studies note that long-term behavioral problems in school, early sexual activity, and difficulties with social adaptation can occur in girls with precocious puberty. Increased psychosomatic symptoms have been reported during menstruation in girls with precocious puberty; however, other studies dispute these findings, concluding that precocious puberty is not associated with any significant changes in psychological health.

Treatment is clearly indicated for boys with precious puberty and for girls (like the one described in this vignette) younger than 6 years old with evidence of progressive central precocious puberty. Treatment with a gonadotropin-releasing hormone (GnRH) agonist, such as leuprolide acetate, will suppress the pulsatile release of GnRH necessary to stimulate puberty and may increase final adult height by an average of 9 to 10 cm. Some studies have demonstrated that treatment may improve peer acceptance. There is no literature that examines psychological distress as it relates to age of menarche because this may be different according to family history and cultural perceptions. Therefore, it is important to discuss with families the specific concerns that led to evaluation and assess their capacity to help the child manage personal hygiene and cope with menstrual periods.

Treatment of girls who show signs of central precocious puberty between 6 and 8 years of age is far more controversial. In some studies, height gain has been modest, 4.5 cm to 7 cm beyond predicted adult height. Others studies indicate that larger gains in adult height are possible. Because most girls will have menarche between 2 to 3 years after thelarche, treatment should be tailored to individual family needs and ability of the child to cope effectively with menstrual periods within their social support structure.

Treatment after 8 years of age generally shows no significant benefit in adult height, and it is not clear that treatment would offer any psychological benefit.
Tamoxifen, hydrocortisone, or medroxyprogesterone acetate are not appropriate for the treatment of central precocious puberty. Observation would not be appropriate for a child younger than 6 years of age with clear signs of progressive central precocious puberty and a magnetic resonance imaging finding supporting the diagnosis, as noted in the girl in this vignette.

PREP Pearls

- Signs of precocious puberty should warrant referral to endocrinology.
- Treatment of precocious puberty is clearly indicated in all boys with progressive central precocious puberty and in girls younger than 6 years of age.
- Early treatment is best because treatment of precocious puberty in girls 6 to 8 years of age can be beneficial but may be less effective in preserving height when compared with the treatment of younger girls.

American Board of Pediatrics Content Specification(s):

- Recognize the tumors that may produce precocious puberty (eg, in liver, CNS, ovary, testes, adrenal glands)

Suggested Reading:

- Koplowitz P. Treatment of central precocious puberty. Curr Opin Endocrinol Diabetes Obes. 2009;16:31-36. doi:10.1097MED.ObO13e3283 20a650
- Kim EY, Lee MI. Psychosocial aspects in girls with idiopathic precocious puberty. Psychiatry Investig. 2012;9:25-28. doi:10.4306/pi.2012.9.1.25

Item 25

An 8-year-old boy is seen in the emergency department with a fever, depressed mental status, vomiting, and a stiff neck. Two days ago, he was involved in a motor vehicle accident and hit his head on the dashboard. His past medical history is unremarkable and immunizations are up to date. On physical examination, the boy is febrile (up to 39.3°C) and difficult to arouse. He has clear rhinorrhea and nuchal rigidity. Cerebrospinal fluid examination reveals the following:

- White blood cells, 2,100/pL (2.1 x 109/L), with 93% neutrophils
- Glucose, 10 mg/dL (0.6 mmol/L)
- Protein, 180 mg/dL (1.8 g/L)

Gram stain reveals pleomorphic gram-negative rods.

Of the following, the BEST antibiotic to treat this organism is

A. ampicillin
B. cefazolin
C. ceftriaxone
D. clindamycin
E. vancomycin

Item 25 TE Preferred Response: C

Nontypeable Haemophilus influenzae (NTHI) commonly are present in normal nasopharyngeal flora, with more than 50% of children colonized by 5 years of age. The organism has been associated with bacterial infections of the upper respiratory tract including sinusitis, otitis media, conjunctivitis, bronchitis, and community-acquired pneumonia. Disruption of the blood-brain barrier, as evidenced in the boy described in the vignette by clear rhinorrhea after head trauma sustained in a motor vehicle accident, and the description of pleomorphic gram-negative rods, indicate that NTHI is the most likely cause of the boy's meningitis. Haemophilus influenza type b would have the same appearance on Gram stain, but in a fully immunized normal host, this organism is extremely unlikely to cause colonization or infection.

The third-generation cephalosporin ceftriaxone provides the best coverage for NTHI and penetrates the blood-brain barrier at sufficient concentrations in meningitis doses. Cefotaxime or meropenem are acceptable alternatives.

A significant percentage of NTHI are ampicillin resistant, either by production of p-lactamase or an alternate mechanism (p-lactamase—negative ampicillin- resistant strains) such that ampicillin would not be an appropriate choice unless sensitivity test results are known. Hospital microbiology laboratories maintain antibiograms of local sensitivity patterns, which may be helpful while awaiting specific culture results.

Nontypeable Haemophilus influenzae are not sensitive to first-generation cephalosporins. In addition, cefazolin does not adequately penetrate the cerebrospinal fluid. Clindamycin similarly does not inhibit NTHI and does not reliably cross the blood-brain barrier. Vancomycin is not active against gram-negative organisms. It is often included in the empiric therapy of meningitis to provide coverage for penicillin-resistant Streptococcus pneumoniae.

PREP Pearls

- Nontypeable Haemophilus influenzae (NTHI) are normal nasopharyngeal flora associated with upper respiratory infections (eg, sinusitis, otitis media, conjunctivitis, pneumonia).
- Invasive infection with NTHI occurs rarely, but the risk is increased in the face of trauma with disruption of the blood-brain barrier.

American Board of Pediatrics Content Specification(s):

- Plan the treatment of a nontypeable Haemophilus influenzae infection

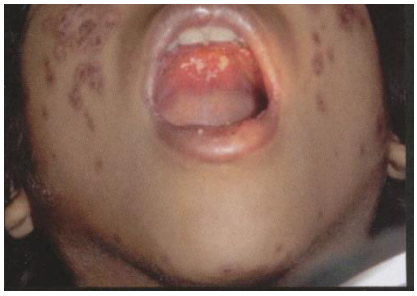
Suggested Reading:

- American Academy of Pediatrics. Haemophilus influenzae infections. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:345-352

- MacNeil JR, Cohn AC, Farley M, et al. Current epidemiology and trends in invasive Haemophilus influenzae disease—United States, 1989-2008. Clin Infect Dis. 2011;53(12):1230-1236. doi: 10.1093/cid/cir735
- Murphy TF, Faden H, Bakaletz LO, et al. Nontypeable Haemophilus influenzae as a pathogen in children. Pediatr Infect Dis J. 2009;28(1):43-48. doi:10.1097/INF.Ob013e318184dba2

Item 26

A 15-year-old, African-American boy is seen in your office with fever, rash, and joint pain. The patient has had 2 weeks of joint swelling and intermittent low-grade fevers. A facial rash has been present for 7 days. On physical examination, he has swollen, tender metacarpal phalangeal joints bilaterally, bilateral knee swelling, warmth, and pain with range of motion, oral ulcers, and a rash (Item Q26).



ITEM Q26: Rash as described for the patient in the vignette.

Of the following, the test MOST likely to establish the diagnosis is

A. anti-double-stranded DNA antibody
B. antinuclear antibody
C. erythrocyte sedimentation rate
D. monospot
E. rheumatoid factor

Item 26 Preferred Response: A

The patient described in the vignette has fever, joint pain, a typical discoid rash and mouth ulcers and most likely has systemic lupus erythematosus (SLE), an autoimmune disease. Anti-double-stranded DNA antibody is the test that is most specific for a diagnosis of lupus. While the antinuclear antibody is positive for most SLE patients, it is not specific to the diagnosis with elevations seen in some infections and cancers. Rheumatoid factor and erythrocyte sedimentation rate can be abnormal with lupus but are also not specific. The patient's rash and mouth ulcers are not consistent with a diagnosis of mononucleosis.

Laboratory findings associated with a diagnosis of SLE include the following:

- Positive antinuclear antibody (usually >! 1:160)
- Positive anti-double-stranded DNA antibody
- Positive anti-Smith antibodies
- Positive anti- ribonuclear protein antibodies
- Positive anti-Ro antibody
- Positive anti-La antibody
- Hypocomplementemia.
- Cytopenia.
- elevated erythrocyte sedimentation rate
- elevated liver transaminases
- elevated muscle enzymes
- proteinuria
- hematuria
- urine casts

Systemic lupus erythematosus can affect any organ system; therefore, the clinical symptoms of lupus are numerous and varied (Item C26A, page C-21).

Item (26A. Signs and Symptoms of Systemic Lupus Erythematosus

System	Clinical manifestations or clinical findings
Cardiac	Pericarditis, pericardial effusion, myocarditis, Libman—Sacks endocarditis, bacterial endocarditis
Constitutional	Fatigue, fever, weight loss, anorexia
Endocrine	Hypothyroidism, hyperthyroidism, irregular menses, delayed puberty
Gastrointestinal	Abdominal pain, serositis, vasculitis, pancreatitis, hepatitis, hepatomegaly, enteritis
Hematologic	Cytopenia, leukopenia usually secondary to lymphopenia, anemia (normocytic normochromic or Coombs-positive hemolytic), thrombocytopenia, antiphospholipid syndrome, splenomegaly
Mucocutaneous	Molar rash, discoid lupus, alopecia, bullous lupus, annular erythema, maculopapular rash, photosensitivity, oral ulcers
Musculoskeletal	Arthralgia, arthritis, avascular necrosis, bone-fragility fractures, myalgia, myositis, secondary pain amplification syndromes
Neuropsychiatric	Headache, cognitive dysfunction, psychosis, seizures, transverse myelitis, central nervous system vasculitis, stroke, aseptic meningitis, cerebrovascular disease, demyelinating syndrome, chorea, myelopathy, acute confusional state, anxiety disorder, mood disorder, acute inflammatory demyelinating polyradiculoneuropathy, autonomic disorder, mononeuropathy, myasthenia gravis, piexopathy, polyneuropathy
Pulmonary	Pleuritic, pleural effusion, pneumonitis, pulmonary hemorrhage, pulmonary hypertension

Renal	Lupus nephritis, hypertension, proteinuria, microscopic hematuria, elevated blood urea nitrogen, elevated creatinine, urinary casts
Vascular	Purpura, palmar erythema, petechiae, tender skin nodules, ulcerations, Raynaud phenomenon, nail fold capillary changes, livedo reticularis

Since SLE mimics many other disease processes, diagnoses such as infection and neoplasm must be ruled out; this is critical in that the immunosuppressive treatment used in SLE management can worsen or mask some conditions.
Most pediatric patients who have SLE will present with 4 or more of the criteria outlined by the American College of Rheumatology (Item C2613, page 22) and therefore meet criteria for the diagnosis for SLE. However, these criteria were not validated in the pediatric population; therefore children/adolescents can be diagnosed with SLE without meeting the criteria.

Item C2613. American College of Rheumatology Revised Criteria for the classification of Systemic Lupus Erythematosus*

System	Clinical manifestations or clinical findings
Malay rash	Fixed erythema, flat or raised, over the malar eminences, tending to spare the nasolabial folds
Discoid rash	Erythematous raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring
Photosensitivity	Skin rash as a result of unusual reaction to sunlight, by patient history or physician observation
Oral ulcers	Oral or nasopharyngeal ulceration, usually painless, observed by a physician
Arthritis	Nonerosive arthritis involving 2 or more peripheral joints, characterized by tenderness, swelling, or effusion
Serositis	A) Pleuritic: convincing history of pleuritic pain or rubbing heard by a physician, or evidence of pleural effusion OR B) Pericarditis: documented by ECG or rub or evidence of pericardial effusion
Renal disorder	A) Persistent proteinuria greater than **0.5** grams per day or greater than 3+ if quantitation not performed OR B) Cellular casts may be red cell, hemoglobin, granular, tubular, or mixed
Neurologic disorder	A) Seizures: in the absence of offending drugs or known metabolic derangements (eg, uremia, ketoacidosis, or electrolyte imbalance) OR B) Psychosis: in the absence of offending drugs or known metabolic derangements (eg, uremia, ketoacidosis, or electrolyte imbalance)
Hematologic disorder	A) Hemolytic anemia: with reticulocytosis OR B) Leukopenia: less than 4,000/mm' total on 2 or more occasions OR C) Lymphopenia: less than 1,500/mm' on 2 or more occasions OR D) Thrombo(ytopenia: less than 100,000/mm' in the absence of offending drugs
Immunologic disorder	A) Positive LE cell preparation OR B) Anti-DNA: antibody to native DNA in abnormal titer

	OR C) Anti-Sm: presence of antibody to Sm nuclear antigen OR D) False-positive serologic test for syphilis known to be positive for at least 6 months and confirmed by *Treponema pallidum* immobilization or fluorescent treponemal antibody absorption test
Antinuclear antibody	An abnormal titer of antinuclear antibody by immunofluorescence or an equivalent assay at any point in time and in the absence of drugs known to be associated with "drug-induced lupus" syndrome

Abbreviation: ECG, electrocardiography.
*Reprinted with permission from Hochberg MC. Updating the American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus. *Arthritis Rheum. 1997;40(9):1725.*

PREP Pearls

- Pediatric patients may be diagnosed with SLE without meeting diagnostic criteria.
- Because SLE mimics many other conditions, it is important to consider all possibilities, including infection and cancer.
- While antinuclear antibody test is often positive in SLE, it is not diagnostic and can be positive in many other disease processes. Anti—double-stranded DNA antibody and anti-Smith antibody are more specific for lupus.

American Board of Pediatrics Content Specification (s):

- Recognize the clinical manifestations of systemic lupus erythematosus

Suggested Reading:

- Levy DM, Kamphuis S. Systemic lupus erythematosus in children and adolescents. Pediatr Clin NAm. 2012;59(2):345-364. doi:10.1016/j. pcl.2012.03.007
- Weiss JE. Pediatric systemic lupus erythematosus: more than a positive antinuclear antibody. Pediatr Rev. 2012;33(2):62-73. doi:10.1542/pir.33-2-62

Item 27

A 9-year-old girl who has been living in her current foster home for the past 2 months is brought to your office by the foster parents. The couple is concerned because the girl has been "hearing and seeing things" around the house that aren't actually there. A few nights prior to the visit, she became very distressed and afraid, stating that she saw a man looking in her second-floor bedroom window. She hears her name being called and becomes distressed about hearing doors slamming when neither has occurred. The girl occasionally wakes up from nightmares. On several occasions, the foster parents have seen her appear to "space out" for several minutes during the day, after which she appears scared and states that she had been thinking about "bad things. ' She occasionally has dramatic and apparently unprovoked mood swings and often has a negative mood. At other times she can engage normally and can get along well with other children. Although they were provided no details, the foster parents were told that the girl witnessed domestic violence in her biological parents' home and that there is a family history of a mental health disorder. Results of her physical examination are within normal limits. Her interaction with you in the office is age appropriate.

Of the following, the MOST likely cause of this girl's symptoms is

A. bipolar disorder
B. depression
C. posttraumatic stress disorder
D. schizophrenia
E. seizure disorder

Item 27 Preferred Response: C

The young girl in the vignette is exhibiting symptoms commonly found in children who have experienced traumatic events, and she most likely has posttraumatic stress disorder (PTSD). Posttraumatic stress disorder is characterized by a history of exposure to a traumatic event of threatened death, injury, or sexual violence; and at least I event of each of the following 4 types of symptoms over the period of at least 1 month:
(1) Intrusive recollection, which involves re-experiencing the traumatic event in an intrusive fashion (for her, both nightmares and spacing out episodes or flashbacks);
(2) Hyperarousal, which may hypervigilance (exemplified by her listening for door slamming and monitoring what is outside the home's windows);
(3) avoidance, or numbing, symptoms presenting as persistent avoidance of stimuli associated with the trauma or numbing of general responsiveness (not present in the child described in the vignette), and
(4) negative alterations in trauma associated cognitions and mood (she does have negative moods). Because only 3 of the 4 required PTSD symptom types appear in the child described in the vignette, a diagnosis of PTSD cannot yet be confirmed, but it is the most likely diagnosis of the choices listed.

Young patients who have experienced trauma may have recurrent hallucinations that invoke the same emotional state as the child's past traumatic experience. For instance, ~ child who recurrently heard one parent striking the other may hallucinate hearing these same noises as a PTSD flashback while reexperiencing the associated emotional fear. [he sound of a door slamming shut, for instance, may be an abuse- associated noise that signaled to the child the likelihood that abuse was about to occur.

Other causes of hallucinations are less likely for this child. Bipolar disorder can cause hallucinations during the manic late, but it is quite uncommon at age 9 years; the child in the vignette has no symptoms of mania. Depression can cause hallucinations when it is severe and prolonged, but no symptoms of depression were noted. Traumatized children commonly experience mood swings and irritability that are not necessarily indicative of a concurrent mood or anxiety disorder. Schizophrenia causes hallucinations, but schizophrenia is very rare in prepubertal children, and the child n the vignette has no history of a schizophrenia prodrome ie, social withdrawal or flattened personality preceding psychosis) or a pervasive loss of awareness of reality that would be characteristic of schizophrenia. Partial seizures can rarely cause hallucinations, which would typically appear during the seizure (with an abrupt onset and offset) and are less complex in experience than in this vignette (eg, a single sensation such as a particular smell).

Many other possible causes of hallucinations exist, the most common of which would include substance abuse and delirium; neither of these causes would be likely for this patient.

PREP Pearls

- PTSD should be considered in pediatric patients who present with hallucinations in the absence of medical disease.
- Psychotic disorders like schizophrenia are not a common cause of hallucinations in young children.

AAP Mental Health Competency

- Know the likely differential diagnosis for a child presenting with complaint of hallucinations

Suggested Reading:

- American Psychiatric Association. Diagnostic and Statistical Manual f Mental Disorders, Fifth ed. Washington, DC: American Psychiatric association; 2013:271-280
- Arsenault L, Cannon M, Fisher HL, Polanczyk G, Moffitt T, Caspi A. Childhood trauma and children's emerging psychotic symptoms: a genetically sensitive longitudinal cohort study. Am J Psychiatry. 2011;168: 5-72. doi:10.1176/appi.ajp.2010.10040567
- !delson GA. Hallucinations in children and adolescents: considerations n the emergency setting. Am J Psychiatry. 2006;163:781-785. doi: 10.1176/ appi.ajp.163.5.781
- !lbridge D, Hillenbrand K, Serwint J. In brief: hallucinogens. Pediatr Rev. 006; 27:314-315. doi: 10.1542/pir.27-8-314

Item 28

A mother well known to your practice has just presented to the hospital with premature rupture of the membranes at 24 weeks' gestation. The medical student shadowing you in the office asks you if there is a clinical variable that is associated with improved survival and outcome at this very early gestational age.

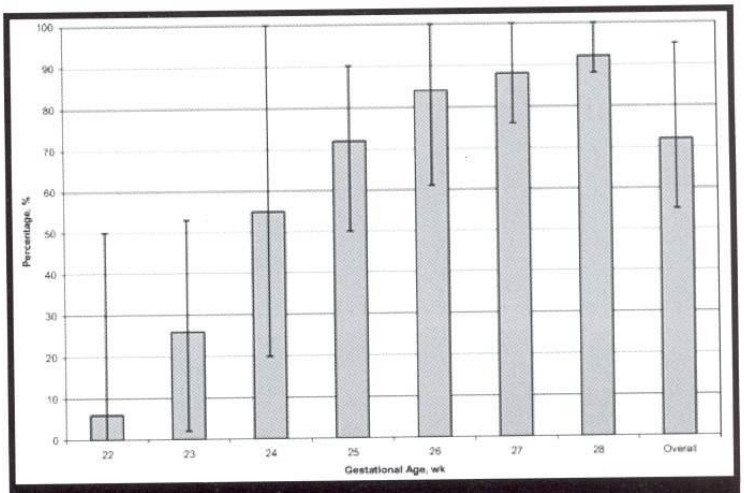
Of the following, the BEST response to the student's question is

A. antenatal corticosteroids
B. antenatal magnesium sulfate
C. male sex
D. multiple gestation
E. small for gestational age

Item 28 TE I-C SBP **Preferred Response: A**

Antenatal corticosteroids are associated with improved survival and outcome in extremely low-birthweight (ELBW) infants. Infants who are born at less than 1,000 g are considered ELBW These infants have higher mortality rates, ranging from 74% at 23 weeks' gestation to 16% at 26 weeks' gestational age (Item C28). ELBW infants who survive are at risk for neurodevelopmental morbidity, cerebral palsy, school difficulties, abnormal pulmonary function, visual impairment, hearing impairment, and growth issues.

Antenatal corticosteroids (betamethasone or dexamethasone) given to mothers accelerate fetal lung maturation and decrease respiratory distress syndrome in premature infants. In addition, their administration reduces rates of mortality, necrotizing enterocolitis, and intraventricular hemorrhage. The American Congress of Obstetricians and Gynecologists published a consensus statement in 1994 recommending the use of antenatal steroids in mothers between 24 weeks and 34 weeks of gestation who are at risk of delivering within 7 days. This statement was reaffirmed in 2000 and 2011. Recent data demonstrate the use of antenatal steroids in approximately 85% of ELBW deliveries.



Reprinted with permission from Stoll BJ, et al. *Pediatrics*. 2010; 126:443-456.

ITEM C28: Survival to discharge according to gestational age among 9,575 very-low-birthweight infants born in National Institute of Child Health and Human Development (NICHD) Neonatal Research Network (NRN) centers between January 1, 2003 and December 31, 2007.

Male sex, twin gestation, and small for gestational age are not associated with improved survival and outcome in the ELBW infant. ELBW infants who are small for gestational age have increased risk of developing chronic lung disease and secondary pulmonary hypertension. These infants also carry the additional risks of metabolic syndrome in later life including type 2 diabetes mellitus and hypertension. The antenatal administration of magnesium sulfate before preterm delivery has been suggested to decrease the risk of cerebral palsy in surviving infants but not improve overall survival. Prospective multicenter randomized trials are currently examining this potential benefit.

The National Institute for Child Health and Human Development has created a calculator for estimating outcome for infants born between 22 0/7 and 25 6/7 weeks of gestation (http://www.nichd.nih.gov/about/org/cdbpm/pp/prog-epbo/epbo–case.cfm).

PREP Pearls

- Antenatal corticosteroids are associated with improved survival and outcome in extremely low-birthweight infants.

American Board of Pediatrics Content Specification(s):

- Understand the prognostic factors for very-low-birth-weight infants

Suggested Reading:

- ACOG Committee on Obstetric Practice. ACOG committee opinion no. 475: antenatal corticosteroid therapy of fetal maturation. Obstet Gynecol. 2011;117:422-424. doi: 10.1097/A0G.06013e31820eee00.
- American College of Obstetricians and Gynecologists Committee on Obstetric Practice; Society for Maternal-Fetal Medicine. Committee opinion no.455: magnesium sulfate before anticipated preterm birth for neuroprotection. Obstet Gynecol. 2010;115:669-771. doi: 10.1097/ AOG.0b013e3181d4ffa5.
- Doyle LW, Saigal S. Long-term outcomes of very preterm or tiny infants. NeoReviews. 2009; 10: e130-e137. doi: 10.1542/neo.10-3-el30
- Stoll BJ, Hansen NI, Bell EF, et al; for the Eunice Kennedy Shriver National Institute of Child Health and Human Development Neonatal Research Network. Neonatal outcomes of extremely preterm infants from the NICHD Neonatal Research Network. Pediatrics. 2010;126:443-455. doi: 10.1542/peds.2009-2959
- Tyson JE, Parikh NA, Langer J, et al. Intensive care for extreme prematurity - moving beyond gestational age. N Engl J Med. 2008;358:1672-1681. doi: 10.1056/NEJMoaO73059

Item 29

A 10-year-old girl who has undergone a cardiac transplant 8 months ago is now hospitalized with fever and adenopathy. A biopsy of the lymph node shows post-transplant lymphoproliferative disease.

Of the following, the medication MOST likely to be associated with the development of this patient's symptoms is

A. enalapril
B. fluticasone
C. prednisone
D. tacrolimus
E. trimethoprim-sulfamethoxazole

Item 29 S Preferred Response: D

Of the medications listed, the one most likely associated with post-transplant lymphoproliferative disease (PTLD) is tacrolimus.

Patients who undergo organ transplantation receive immunosuppressive therapy, often lifelong, to prevent rejection. For pediatric cardiac transplant recipients, most centers combine multiple immunosuppressive agents, including calcineurin inhibitors (eg, cyclosporine and tacrolimus), azathioprine, mycophenolate mofetil, and steroids. Chronic immunosuppression has many consequences, such as infection, infertility, growth impairment, neurotoxicity, and PTLD. Post-transplant lymphoproliferative disease occurs as a result of immunosuppression in patients who have undergone solid organ or allogeneic stem cell transplantation (SCT). A total of 90% to 95% of cases of PTLD are associated with Epstein-Barr virus (EBV) proliferation in the setting of chronic immunosuppression and depressed T-cell function. In immunocompetent hosts, T cells eliminate most EBV-infected B cells; however, a small population of infected cells escapes immune surveillance by downregulating viral expression on the B-cell surface. These B cells remain latent but may proliferate in the setting of suppressed T-cell immunity. The origin of the EBV-infected cells can be from the donor or the recipient (host).

Post-transplant lymphoproliferative disease is one of the most common malignant conditions affecting solid organ transplant recipients (20% of all patients), but is much less common in allogeneic SCT recipients. This finding is in part due to the higher amount of and prolonged (lifelong) exposure to immunosuppression after solid organ transplants compared with after SCT (usually several months to prevent graft-vs-host disease). More than 80% of cases occur during the first year after transplantation. Risk factors for develop-ment of PTLD are degree of T-cell suppression and the EBV serostatus of the patient. Studies in pediatric patients have found an increased risk of developing PTLD with prolonged exposure to high doses of tacrolimus.
Patients with PTLD may present with fever, weight loss, fatigue, and an extranodal mass. They may have cytopenias; elevated lactate dehydrogenase, serum calcium, uric acid, and monoclonal protein levels in the serum or urine; and positive positron emission tomography results. Diagnosis is made by tissue biopsy. Treatment for PTLD includes reduction of immunosuppression, anti-CD20 monoclonal antibody (rituximab), chemotherapy, and radiation therapy.

The child described in the vignette has undergone a cardiac transplantation, which has a relatively high risk for PTLD given the amount of immunosuppression required. Of the medications she is taking, fluticasone, prednisone, and tacrolimus are the only ones with immunosuppressive properties. Of these, tacrolimus has the greatest degree of T-cell inhibition and has been specifically associated with a higher risk of developing PTLD. Enalapril and trimethoprim-sulfamethoxazole are not immunosuppressive agents.

PREP Pearls

- PTLD occurs as a result of immunosuppression in patients who have undergone solid organ or allogeneic SCT.
- PTLD is most likely to occur in the first year after transplantation.
- T-cell suppression (especially with tacrolimus) is associated with a higher risk of PTLD.

American Board of Pediatrics Content Specification(s):

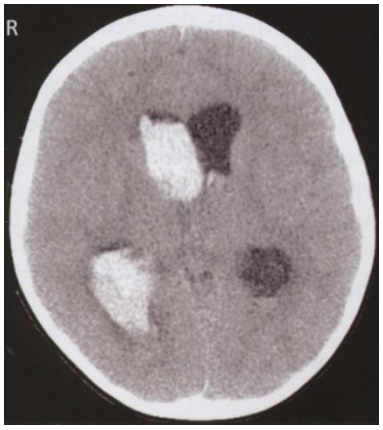
- Recognize the long-term risks of chronic immunosuppression

Suggested Reading:

- Friedberg JW, Aster JC. Epidemiology, clinical manifestations, and diagnosis of post-transplant lymphoproliferative disorders. UptoDate. Available online only for subscription
- Negrin RS, Brennan DC, Jessup M. Treatment and prevention of post-transplant lymphoproliferative disorders. UptoDate. Available online only for subscription
- Schnepp E, Kaminski D. Chronic immunosuppression. Pediatr Rev. 2012;33:481-482. doi:10.1542/pir.33-10-481
- Sudan D, Bacha EA, John E, Bartholomew A. Pediatr Rev. 2007;28:439-453. DOT: 10.1542/pir.28-12-439

Item 30

A 5-year-old boy complains of headache and neck pain and lies down for a nap. An hour later, his father tries to rouse him, but the boy can only mumble. He is brought to the emergency department, where on physical examination his blood pressure is 125/90 mm Hg, heart rate is 78 beats/ min, respiratory rate is 14 breaths/min, and temperature is 37.4°C. The boy briefly opens his eyes but otherwise does not respond during the examination. There is no sign of head injury, no nuchal rigidity, and no rashes. Neurologic examination shows that the pupils are round and equally reactive to light, the limbs are flaccid, the deep tendon reflexes are brisk, and the toes are upgoing on plantar stimulation. The parents report that all medications in the home are secured and deny any ingestions. Computed tomography of the head without contrast is obtained (Item Q30).



ITEM Q30: Computed tomography scan without contrast for the boy described in the vignette.

Of the following, the MOST likely cause of his symptoms is

A. arterial ischemic stroke
B. arteriovenous malformation
C. brain abscess
D. choroid plexus carcinoma
E. vein of Galen aneurysmal malformation

Item 30 TE **Preferred Response: B**

The boy in the vignette has a ruptured arteriovenous mal-formation (AVM). His slightly elevated blood pressure and coma are consistent with increased intracranial pressure. The computed tomography image shows intraventricular hemorrhage, and the ventricles are enlarged from early, non-obstructive hydrocephalus (Item C30A). As hydrocephalus progresses and intracranial pressure increases, the brain will begin to herniate. A neurosurgeon should be consulted immediately.

An AVM is an abnormally formed connection between arteries and veins. AVMs are congenital, not acquired, and are commonly found in the brain. Clinically, some AVMs can be detected by auscultation of an intracranial bruit through the orbit or an open fontanelle. Rarely, AVMs can present with high output cardiac failure because of abnormal shunting between the arterial and venous systems. On contrasted imaging, AVMs appear as a tangled clump of vessels (Item C30B). These can occur anywhere in the brain, including inside the ventricles as in this case.

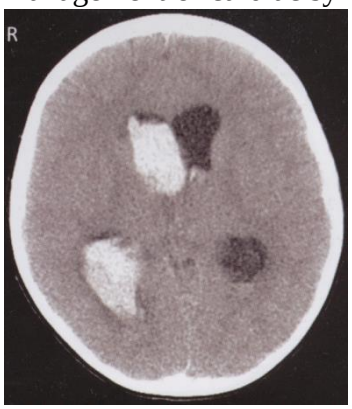
Unruptured AVMs can cause local brain ischemic. Because there is no capillary bed in the area of the AVM, the local brain tissue is ischemic. This can present with focal neurologic deficits, seizures, or headaches. Unruptured AVMs in children have a 2% rate of hemorrhage per year. When AVMs rupture, they produce a sudden, severe headache and loss of consciousness. The intracranial hemorrhage causes increasing intracranial pressure, which quickly leads to coma and death if no intervention occurs.

Arterial ischemic stroke typically presents with an acute focal neurologic deficit such as hemiparesis. In children, an acute ischemic stroke often presents with seizure. If the area of the stroke is large enough, consciousness can be impaired, leading to signs of increased intracranial pressure because of brain swelling. Computed tomography shows decreased attenuation in the brain parenchyma (Item C30C).

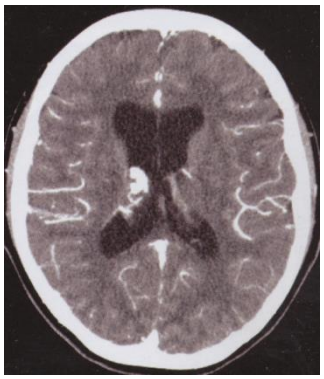
A brain abscess presents with signs of infection, such as fever, altered mental status, or nuchal rigidity. Seizures can also be a presenting symptom especially if the abscess is near the cerebral cortex. Bacteremia, cardiac defects with right to left shunt, recent neurosurgery, or dental procedures are risk factors for brain abscess. Computed tomography with contrast typically shows a round, enhancing mass (Item C30D, page C-26).choroid plexus carcinoma presents with persistent headache, vomiting, and lethargy arising from gradually progressive hydrocephalus. Altered mental status worsens over days, unlike the sudden loss of consciousness resulting from a ruptured AVM. On noncontrasted computed tomography, a mass is apparent in the region of the choroid plexus. Ventricles are often enlarged, reflecting nonobstructive hydrocephalus (ItemC30E, page C-26).

Vein of Galen aneurysmal malformation (VGAM) is a type of arteriovenous malformation that typically presents with high output cardiac failure in the neonatal period or infancy. Hydrocephalus, signs of increased intracranial pressure, seizures, and developmental delay can be present in infants and older children. Hemorrhage on presentation is rare. On imaging the dilated vein is visible (ItemC30F, page C-26).

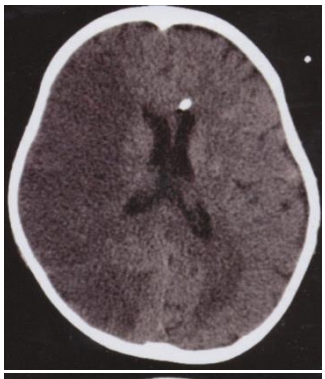
Treatment for VGAM is endovascular embolization of the malformation and management of cardiac symptoms.



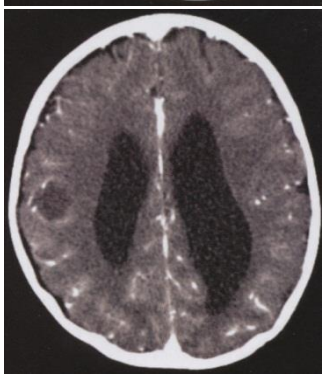
ITEM C30A: Computed tomography of the head without contrast, showing intraventricular hemorrhage, more on the right, and enlarged ventricles.



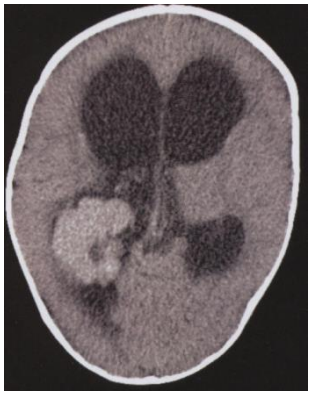
ITEM C30B: Computed tomography of the head with contrast, showing an arteriovenous malformation in the right lateral ventricle. Normally enhancing choroid plexus is visible in the left ventricle.



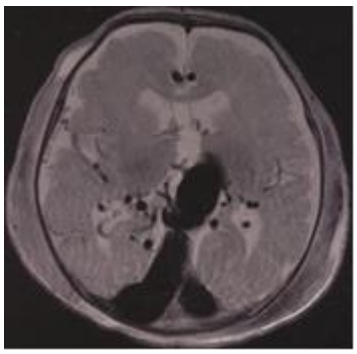
ITEM C30C: Computed tomography of the head without contrast, showing right-sided hypodensity in the brain parenchyma in the territory of the right middle cerebral artery territory, with effacement of the suld and blurring of the gray white junction in the right hemisphere. All of these findings together suggest an acute, arterial ischemic stroke. There is a ventricular catheter in the anterior left ventricle that is unrelated to the acute stroke. The crescent shaped area of hypodensity posterior to the left ventricle is normal white matter.



ITEM C30D: Computed tomography of the head with contrast, showing a round, ring-enhancing lesion in the right hemisphere. Consistent with an intraparenchymal brain abscess. The left ventricle is enlarged, which is unrelated to the abscess.



ITEM C30E: Computed tomography of the head without contrast, showing a lobulated mass in the choroid plexus of the right ventricle. The ventricles are enlarged due to hydrocephalus.



ITEM C30F: Magnetic resonance imaging of the head, T2 weighted, axial view showing a dilated vein of Galen in the midline.

PREP Pearls

- When central nervous system arteriovenous malformations (AVMs) rupture, they cause sudden, severe headache and loss of consciousness.
- Central nervous system AVM rupture is a neurosurgical emergency.

American Board of Pediatrics Content Specification(s):

- Identify the clinical features of CNS arteriovenous malformations of childhood

Suggested Reading:

- Fullerton HI, Achrol AS, Johnston SC, et al. Long-term hemorrhage risk in children versus adults with brain arteriovenous malformations. Stroke. 2005;36(10):2099-2104. doi: 10.1161/01.STR.0000181746.77149.2b
- Getzoff M, Goldstein B. Spontaneous subarachnoid hemorrhage in children. Pediatr Rev. 1999;20(4):141. doi 10.1542/pir.20-12-422
- Kochanek PM, Bell MJ. Neurologic emergencies and stabilization. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:296-304

Item 31

A 12-year-old female ice hockey player presents to your office for a preparticipation physical evaluation. This is her first visit with your practice. Her parents bring a copy of her medical records for your review. She would like to play in a hockey game later that day.

Of the following, the only ABSOLUTE contraindication to participation would be

A. absence of one ovary
B. blood pressure reading at the 90th percentile for her age
C. history of a seizure disorder
D. a soft systolic ejection murmur that disappears with standing
E. temperature of 38.8°C

Item 31 S Preferred Response: E

The main goal of the preparticipation physical evaluation (PPE) is to screen athletes for conditions that might interfere with safe sports participation. Ideally, the PPE should be performed at approximately 6 weeks before the athlete's sport season; this allows time for additional testing should the history and physical indicate the need for further evaluation. The PPE monograph, authored jointly by the American Academy of Pediatrics, American Academy of Family Physicians, and 4 sports medicine societies, provides expert opinion and evidence-based guidelines regarding the performance of the PPE and use of the results to guide participation in athletics. Of the response choices listed for this question, only fever is an absolute contraindication to participation. Children with fever have an increased risk of exertional heat illness and decreased exercise capacity. In addition, fever can be an indicator of illnesses, such as myocarditis, that increase the risk of sudden death. The risk of ovarian damage resulting from sports participation is very low; therefore, girls with a single ovary should not be restricted from any athletic activities.

Children with severely elevated blood pressure (above the 99th percentile for age) should be restricted from weightlifting and other activities with a high static component. A blood pressure above the 95th percentile for age merits additional evaluation in any case. A history of a well-controlled seizure disorder should not preclude participation in sports; however, coaches and athletic trainers should be aware that the athlete is at risk for having seizures. An athlete with a poorly controlled seizure disorder should not be permitted to participate in water sports, archery, riflery, or weightlifting; contact and collision sports may be permissible on a case-by-case basis. The cardiac murmur described in choice d is an innocent murmur and should not trigger additional evaluation or restriction from athletics.

PREP Pearls

- The presence of fever is an absolute contraindication to sports participation.
- Children and adolescents with moderately elevated blood pressure should not be restricted from sports participation.

American Board of Pediatrics Content Specification(s):

- Understand the factors that influence participation in contact sports by healthy children and adolescents

Suggested Reading:

- Bernhardt DT, Roberts WO, eds. Preparticipation Physical Evaluation. 4th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2010
- Peterson AR, Bernhardt DT. The preparticipation sports evaluation. Pediatr Rev. 2011;32(5):e53-e65. doi:10.1542/pir.32-5-e53
- Wingfield K, Matheson GO, Meeuwisse WH. Preparticipation evaluation: an evidence-based review. Clin J Sport Med. 2004;14(3):109-122

Item 32

In the regular nursery, you are examining a full-term new-born who was born yesterday evening. You note that he has proximal limb shortening. Because of your concerns about possible achondroplasia, you order a skeletal survey, which confirms this clinical diagnosis. You meet with the parents and discuss these findings and potential medical problems in infants and children with achondroplasia.

Of the following, before discharge from the hospital, you are MOST likely to recommend

A. DNA analysis for an FGFR3 mutation
B. head ultrasonography
C. magnetic resonance imaging of the thoracic and lumbar spine
D. polysomnography
E. renal ultrasonography

Item 32 Preferred Response: D

A newborn with achondroplasia is at increased risk for central apnea secondary to arterial compression at the level of the foramen magnum, so polysomnography is recommended for all those with this common skeletal dysplasia. In addition, a careful neurologic examination should be performed that looks for signs of significant hypotonia or sustained ankle clonus that may be suggestive of spinal cord compression. Since unexpected death secondary to central apnea occurs in 2% to 5% of infants with achondroplasia (including some who have had no neurologic symptoms), neuroimaging is also recommended. A computed tomography study of the head with bone windows and thin cuts may provide very good visualization of the foramen magnum, but it carries the risk of radiation exposure; magnetic resonance imaging allows for direct visualization of the brainstem and upper spinal cord but requires sedation. Therefore, clinicians need to decide which neuroimaging study is optimal for each patient.

Because of the potential for cervical cord problems as well as progressive thoracolumbar kyphosis in infants and young children with achondroplasia, the following recommendations have been made:

- Use an infant seat or carrier with a firm back and neck supports
- Use the car seat in the rear-facing position as long as possible
- Do not place the infant in mechanical swings or carrier slings
- During wakeful periods, make use of "tummy time" and also supine positioning, with activity bridges overhead to avoid prolonged upright or sitting positioning

Although molecular testing for an FGFR3 mutation can serve as confirmation of the diagnosis, it is not routinely required when the clinical diagnosis has been established. This molecular testing may be helpful for the individual with achondroplasia for family planning purposes. Testing is relatively straightforward because 99% of individuals with achondroplasia have an amino acid substitution at position 380 (Gly380Arg) in the FGFR3 gene. Molecular genetic testing may be in order if the infant or child has atypical features or the diagnosis of hypochondroplasia is under consideration. Head ultrasonography will not be helpful in visualizing the foramen magnum and is not recommended.

Magnetic resonance imaging of the thoracic and lumbar spine is not recommended even if there is clinical evidence of kyphosis unless there are accompanying neurologic symptoms. There are no known renal problems associated with achondroplasia.

PREP Pearls

- Polysomnography and head imaging studies (to visualize the spinal cord in the area of the foramen magnum) are recommended for infants with achondroplasia because of the risk of central apnea and sudden unexpected death.

American Board of Pediatrics Content Specification(s):

- Recognize the significance of apnea in a patient with achondroplasia

Suggested Reading:

- Pauli RM; National Institutes of Health, US Department of Health and Human Services. Achondroplasia. In: Pagon RA, Bird TD, Dolan CR, eds. GeneReviews. Seattle, Washington: University of Washington; 2013
- Trotter TL, Hall JG; Committee on Genetics. Health supervision for children with achondroplasia. Pediatrics. 2005;116(3):771-783. doi:10.1542/ peds.2005-1440

Item 33

You are taking weekend telephone calls for your practice when the father of a 12-year-old boy calls to let you know he is driving his son to the local hospital. The boy was working in the family garage when he drank an unknown substance out of a plastic bottle that he mistakenly thought contained water. Since swallowing some of the substance, the boy has complained of severe throat pain, burning pain in his chest, difficulty swallowing, and nausea. The father tells you that his son is drooling and that he can see a few white "sores" on his tongue and the roof of his mouth.

Of the following, the MOST likely substance to cause the boy's symptoms and physical findings is

A. antifreeze (ethylene glycol)
B. furniture polish (hydrocarbon)
C. insecticide (organophosphate)
D. toilet bowl cleaner (sodium hydroxide)
E. weed killer (glycophosphate)

Item 33 S Preferred Response: D

The adolescent boy described in the vignette is symptomatic after unintentional ingestion of an unknown household product that was improperly stored in an unlabeled container. Of the substances listed, sodium hydroxide, a corrosive product, is most likely responsible for his symptoms.

Most unintentional ingestions in pediatric patients occur in their homes, where toxic household products are often readily available. Many household cleaners, furniture polishes, garden chemicals, and automotive products carry the risk of significant toxic effects if ingested. Often, as in the vignette, a child's caregivers seek medical attention when symptoms indicate toxic effects from exposure to an unknown product.

Recognition of the typical signs and symptoms related to specific toxic exposures is critical to intervening appropriately and for providing appropriate anticipatory guidance. Corrosives are concentrated acid, alkaline, or oxidizing agents. Many are ingredients found within common household products, including toilet bowl cleaners, laundry detergents, stain and mildew removers, floor cleaners, oven cleaners, rust removers, phenol-based disinfectants, swimming pool products, and batteries. The Centers for Disease Control and Prevention estimate that more than a half million children are treated emergently each year for acute poisoning with corrosives. Ingestion of even small amounts of a corrosive alkaline substance, such as toilet bowl cleaner, drain cleaners, or rust remover, can result in serious penetrating injuries to mucosal and skin surfaces by liquefaction necrosis. Acidic substances tend to cause injury via coagulation necrosis, rather than liquefaction necrosis; thus, acids carry a lower risk of esophageal perforation but are still capable of producing serious corrosive injury.

Patients ingesting corrosive substances typically present with odynophagia, dysphagia, drooling, intraoral burns, or ulcerations, as experienced by the adolescent in the vignette. Additional findings may include vomiting with hematemesis, respiratory difficulty with stridor or wheezing, hoarseness, retrosternal chest pain, dyspnea, and burns on the face, hands, or chest. Significant burns to the eyes may occur with any ocular exposure. Because the primary mode of injury from contact with these toxins is direct tissue corrosion, systemic symptoms are rare, and hemodynamic instability generally does not occur. Apart from distress due to pain, mental status is usually normal in affected children. The most pressing clinical concern after a caustic ingestion is the potential for airway or esophageal injury. Early airway visualization and protection are indicated in any patient presenting with stridor or respiratory distress. Evaluation of the esophagus by upper endoscopy is indicated in patients who have intraoral burns, other symptoms related to the ingestion, or a history strongly suggesting ingestion of a corrosive product. In asymptomatic patients in whom caustic ingestion is uncertain, the need for endoscopy is controversial.

Ethylene glycol, the toxic ingredient in antifreeze, presents with a clinical syndrome typical of all alcohols. Depressed mental status is the initial manifestation, with nausea and vomiting as common associated symptoms. Because of accumulation of the toxic metabolites glycolaldehyde, glycolic acid, and oxalic acid, produced by the metabolism

of ethylene glycol, severe metabolic acidosis ensues after ingestion. Seizure and coma may manifest within a few hours after a significant ingestion. Affected patients may progress to frank coma and cardiopulmonary failure. Hypocalcemia is another common finding, resulting from the formation of calcium oxalate crystals by toxic metabolites; these calcium oxylate crystals may be deposited in all organs of the body. Crystalluria in patients with ethylene glycol poisoning is a late finding. Approximately 24 to 72 hours after ingestion, patients typically progress to renal failure, and hemodialysis may be warranted.

Glycophosphate-containing weed killers are irritants that may cause chemical conjunctivitis, cough with inhalation, and vomiting after ingestion. These products are not corrosive and generally do not cause mucosal injury.

Toxicity from insect repellants that contain organophosphate is marked by clinical signs and symptoms related to the overactivation of cholinergic receptors by excess acetylcholine. The classic features of cholinergic toxicity can be recalled using the mnemonic SLUDGE: salivation, lacrimation, urination, defecation or diarrhea, gastrointestinal upset, and emesis. Among the expected vital sign abnormalities are tachypnea and bradycardia. Pupillary constriction is a classic finding with organophosphate toxicity.
Ingestion of hydrocarbon -based products, such as furniture polish, primarily causes respiratory distress due to pulmonary aspiration. Patients present with acute coughing, gagging, and choking. Physical examination findings may include fever, tachypnea, cyanosis, and abnormal lung sounds, which may include crackles and wheezing. Direct central nervous system effects of the hydrocarbon may lead to lethargy, seizure, or even coma.

PREP Pearls

- Ingestion of even small amounts of a corrosive alkaline substance can result in serious penetrating mucosal and skin injuries.
- Patients ingesting corrosive substances typically present with odynophagia, dysphagia, drooling, intraoral burns, or ulcerations.
- The most pressing clinical concern after a caustic ingestion is the potential for airway or esophageal injury.
- All household chemical products should be stored in their original labeled containers and kept out of the reach of children.

American Board of Pediatrics Content Specification(s):

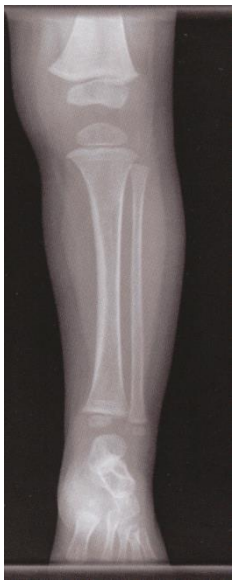
- Know the common household sources of acids and alkali

Suggested Reading:

- Brunnie C, Savage RR. Corrosive ingestions. Pediatr Rev. 2006;27:154-155. doi:10.1542/pir.27-4-154
- Ferry GD, Fishman DS. Caustic esophageal injury in children. UptoDate. Available online only for subscription
- 33-3 Osterhoudt, KC, Ewald MB, Shannon M, Henretig FM. Toxicologic emergencies. In: Fleisher GR, Ludwig S, eds. Textbook of Pediatric Emergency Medicine. 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2010:1171-1223

Item 34

A 16-month-old girl is seen for evaluation of a limp. For the past day, she has been crying and refusing to bear full weight on her left leg. She is afebrile and has no other symptoms. There is no known trauma, but the mother states that she is an active toddler. On physical examination, there is no erythema, bruising, warmth, or swelling of the extremity or joints. The patient cries when you attempt to have her bear weight. She has full range of motion at the hips, knees, and ankles but cries when you palpate the left lower leg. You obtain a radiograph of the left leg (Item Q34).



Of the following, the MOST likely diagnosis is

F. ankle sprain
G. neuroblastoma
H. osteomyelitis
I. toddler's fracture
J. transient synovitis

Item 34 Preferred Response: D

The radiologic findings typical of the girl described in the vignette reveal a nondisplaced spiral fracture of the distal tibia, commonly known as a toddler's fracture. This is one of the most common fractures in children younger than 4 years and is induced by a rotational injury. Often the injury is minor and occurs during any of the usual mishaps of toddler-hood; frequently the parents have no specific recollection of a traumatic event. In the absence of constitutional symptoms and with nonspecific findings localized to an extremity in a toddler, as in the girl in the vignette, the most helpful first diagnostic tool is a plain radiograph. Anterior–posterior and lateral plain radiographic views of the lower leg, including the knee and ankle, are the initial images of choice when a tibial or fibular fracture is suspected. An oblique view may be needed to detect the fracture and should be obtained if the initial images are negative and there is a high index of suspicion of toddler's fracture.

On physical examination, the subtle finding of localized tenderness with minimal swelling or bruising is common. Although an older child may be able to localize the area of pain, a toddler or infant may present only with irritability, pseudoparalysis, and refusal to walk or bear weight. The presence of a toddler's fracture does not necessarily indicate intentional trauma or inadequate supervision because this injury may occur in young children who twist when falling as they are learning to walk and run. However, the social situation should be explored.

Acute onset of a limp or refusal to bear weight is a common clinical problem that can be challenging to diagnose. The differential diagnosis includes infectious causes, such as osteomyelitis and septic arthritis; postinfectious entities, such as transient synovitis and rheumatic fever; malignant tumor; rheumatologic disorders; and injuries related to unintentional or intentional trauma. A careful history and physical examination help direct the evaluation. The history should include recent or recurrent febrile illness, rash, weight loss, or changes in urine or stools. Family history, social history, and travel should be elucidated. A thorough physical examination must be performed and should include observation for deformity, spontaneous movement of the limb, and position of the child at rest. Careful palpation of the entire limb and evaluation of range of motion of all joints of the affected limb are indicated. An assessment of the skin for bruising, erythema, or induration plus neurovascular competence of the injured areas should be performed.

Children's ligaments are stronger and withstand more mechanical force than adults, so sprains are not common in this age group. Neuroblastoma may present with bone pain, but the child in the vignette has no other suggestive physical findings, and the radiograph is not consistent with this diagnosis. Children with osteomyelitis would most likely be febrile and have localized erythema, warmth, or swelling noted on physical examination. Transient synovitis usually occurs after a viral illness. The girl in this vignette exhibited full range of motion at the hips and knees, which would not be typical in a case of transient synovitis.

PREP Pearls

- A nondisplaced spiral fracture of the distal tibia (toddler's fracture) is prevalent in children younger than 4 years.
- Often the rotational injury that causes this fracture is minor, so the history may not be revealing.
- The social situation should be explored carefully to evaluate for intentional trauma, although this fracture is often related to a minor fall.

American Board of Pediatrics Content Specification(s):

- Understand that occult fractures can cause gait disturbances in young children

Suggested Reading:

- Chapman 1, Cohan J. Tibial and fibular shaft fractures in children. UptoDate. 2012. Available online only for subscription
- Deeney VF, Moreland MS, Ward WT, Davis HW. Orthopedics: toddler's fracture. In: Zitelli BJ, Davis HW. Atlas of Pediatric Physical Diagnosis. 5th ed. Maryland Heights, MO: Mosby; 2007:806
- Dinolfo EA. Fractures. Pediatr Rev. 2004;25:218-219. doi: 10. 1 542/pir. 25-6-218
- Wells L, Sehgal K, Dormans JP. Fractures of lower extremity. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:2392

Item 35

A 2-year-old boy in your office is receiving an albuterol nebulizer treatment along with oxygen supplementation. He has been given albuterol every 1 to 2 hours at home to treat his cough and wheeze since his exacerbation began 6 hours ago. The boy's mother is concerned that he is excessively shaky and irritable. On his initial physical examination, he had an oxygen saturation of 85% on room air and a respiratory rate of 60 breaths/min with intercostal retractions and use of accessory muscles. On auscultation, he had loud wheezing in both lung fields. He has received an albuterol nebulizer treatment with oxygen and now has a heart rate of 120 beats/min, a respiratory rate of 40 breaths/min, and oxygen saturation of 97%. He has a mild tremor, minimal use of accessory muscles, and faint expiratory wheezing in both lung bases.

Of the following, the adverse effect this boy is MOST likely to experience is

A. arrhythmia
B. hypocalcemia
C. hypoglycemia
D. hypokalemia
E. seizure

Item 35 S Preferred Response: D

Of the options listed, the child described in the vignette is most likely to demonstrate hypokalemia as a side effect of β-agonist use. Beta-agonists are associated with several adverse effects. Tremor is the most frequent acute side effect reported and is more noticeable with oral therapy than with inhaled agents. Patient acceptance of tremor is often related to diminished awareness once steady-state levels are reached. Metabolic disturbances, such as hypokalemia and hyperglycemia (but not hypoglycemia), have been observed, although the clinical significance is unclear. Hypocalcemia has not been reported during β-agonist use. Increased heart rate and palpitations are dose dependent and are less common with the selective (β2-agonists (such as albuterol) than with nonselective agents (such as metaproterenol). While sympathomimetic amines such as (β-agonists should be used with caution in children with cardiac conditions (eg, coronary insufficiency, arrhythmias, or hypertension), rhythm disturbances are not a commonly reported concern in individuals without an underlying cardiac condition. Stress-induced cardiomyopathy has been associated with treatment of status asthmaticus in case reports. Seizures have not been reported as an adverse effect of excess selective β2-agonist use in normal individuals. Use of a spacer or chamber device reduces these adverse effects by reducing oral deposition and systemic absorption of medication.

PREP Pearls

- The most frequent clinical side effect of β-agonists is tremor.
- The most common metabolic disturbances, when using β-agonists include hypokalemia and hyperglycemia.
- Use of a spacer or holding chamber device can help minimize adverse effects from β-agonists.

American Board of Pediatrics Content Specification(s):

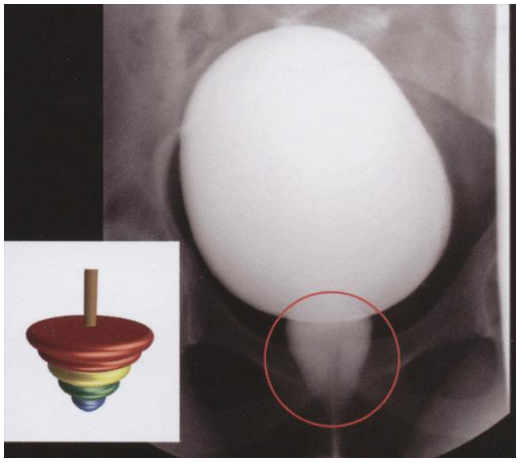
- Recognize the clinical manifestations of toxicity to adrenergic agonists (muscular tremor, tachycardia, hypokalemia)

Suggested Reading:

- National Asthma Education and Prevention Program. Expert Panel Report 3 (EPR-3): guidelines for the diagnosis and management of asthma—summary report 2007 [published correction appears in J Allergy Clin Immunol. 2008;121(6):1330]. J Allergy Clin Immunol. 2007;120(5 suppl):S94-S138. doi:10.1016/j.jaci.2007.09.029
- Osuorji I, Williams C, Hessney J, Patel D, Hsi D. Acute stress cardiomyopathy following treatment of status asthmaticus. South Med J. 2009;102(3):301-303. doi:10.1097/SMj.Ob013e31818f5bd8
- Sorkness CA. Beta-adrenergic agonists. In: Adkinson NF, Bochner BS, Busse WW, et al, eds. Middleton's Allergy: Principles and Practice. 7th ed. Philadelphia, PA: Mosby; 2009:1485-1503

Item 36

A 3-month-old female infant presents to your office for follow-up of pyelonephritis diagnosed 2 months ago. After treatment of her urinary tract infection, she had a contrast voiding cystourethrogram (VCUG) that showed narrowing of the distal urethra and a normal urinary stream upon voiding (Item Q36). Her physical examination is unremarkable. She is currently on oral amoxicillin for urinary tract infection prophylaxis.



ITEM Q36: Contrast voiding cystourethrography for the girl described in the vignette. Note the appearance of the abnormality mimics the appearance of a spinning toy top.

Of the following, the MOST appropriate next step in the management of this patient is

A. intravenous pyelography
B. referral to urology for surgical correction
C. repeat urine culture
D. repeat VCUG in 1 year
E. stop prophylactic antibiotics

Item 36 Preferred Response: E

The voiding cystourethrogram (VCUG) shown for the girl described in the vignette has the classic findings of spinning top urethral (STU) deformity. STU is caused by dilation of the proximal muscular urethra against a closed or narrow distal urethral sphincter. This is a rare variant seen in girls and young women.

Initially considered to be a normal variant, it has been recently associated with bladder dysfunction (detrusor sphincter dyssynergia: contraction of the detrusor against a closed urethral sphincter). In a 3-month-old infant, this appearance could be secondary to the age-appropriate immature voiding reflexes. The overactive bladder almost always resolves, but the time to resolution is highly variable. In this patient, the detrusor-sphincter dyssynergia will most likely resolve with maturation and development of normal voiding habits. Therefore there is no indication for urology referral or surgical correction at this time. Older patients usually present with symptoms of overactive bladder such as frequency, urgency, and urge incontinence. Constipation is a commonly associated symptom in such patients. It is currently unclear if urinary tract infections (UTIs) are a cause or effect of the overactive bladder. However, the appearance of the urethra on VCUG does not correlate with either urethral caliber or episodes of lower UTI. The VCUG in the infant in the vignette shows no evidence of reflux, therefore the next most appropriate step is to stop antibiotics. Currently a repeat urine culture in the presence of a normal VCUG is not recommended before stopping antibiotics. A repeat VCUG, after an initial normal VCUG also is not indicated in this patient.

Intravenous pyelography is a radiologic test for identifying the details of the urinary system including kidneys and ureters. The test has diagnostic significance for identifying upper urinary tract obstruction. The current availability of ultrasonography, computed tomography scan, and magnetic resonance urography has replaced intravenous pyelography in almost all diagnostic settings. In the patient described in the vignette, no further diagnostic imaging is indicated.

PREP Pearls

- Narrow urethra on voiding cystourethrogram (also termed spinning top urethral [STU] deformity) is caused by dilation of the proximal muscular urethra against a closed or narrow distal urethral sphincter.
- STU has been associated with bladder dysfunction arising from contraction of the detrusor muscle of the bladder against a closed urethral sphincter.
- Bladder dysfunction is common in infants; the development of normal voiding patterns is highly variable.

American Board of Pediatrics Content Specification (s):

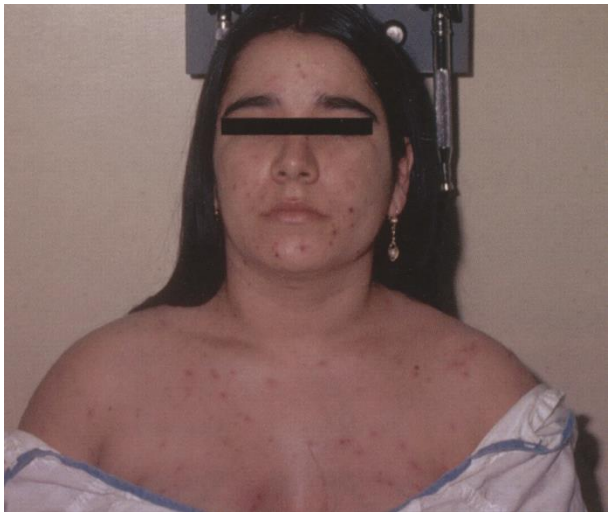
- Know that a girl with a narrow urethra needs no treatment

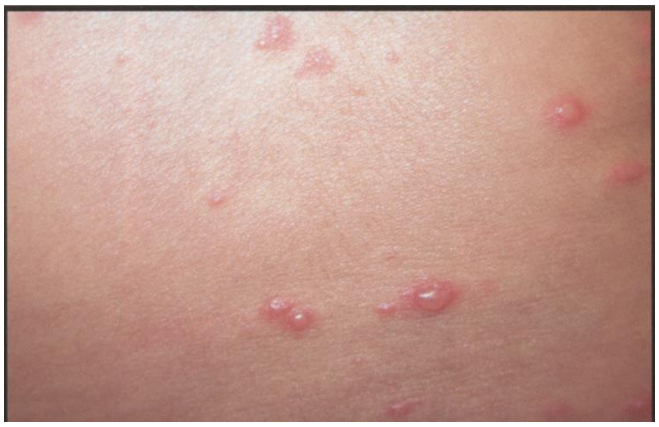
Suggested Reading:

- Elder JS. Voiding dysfunction. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011: 537:1847-1852
- Elder JS. Obstruction of the urinary tract. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:1838-1847
- Saxton HM, Borzyskowski M, Mundy AR, et al: Spinning top urethra: not a normal variant. Radiology. 1988;168:147-150

Item 37

A 2-day-old, term neonate discharged at 24 hours of age is brought into the emergency department because the mother has developed a rash (Item Q37A and Item C37B). The infant is afebrile and has normal vital signs for age. Findings on physical examination are unremarkable.





ITEM Q37B: Close-up view of the rash as described for the woman in the vignette.

Of the following, the BEST next step in the management of this patient is to administer

A. immune globulin intravenous
B. oral acyclovir
C. parenteral foscarnet
D. varicella vaccine
E. varicella-zoster immune globulin

Item 37 Preferred Response: E

The mother of the neonate described in the vignette has a rash due to the varicella-zoster virus (VZV; ie, chickenpox), and the infant is at risk for serious, potentially fatal (25%) infection. The best next step in the management of the neonate is to administer varicella-zoster immune globulin intramuscularly, ideally within 96 hours (but indicated up to 10 days) after exposure. Varicella-zoster immune globulin is a purified human immune globulin preparation that contains high levels of antibodies against varicella. It was approved by the US Food and Drug Administration in 2012 for varicella prophylaxis in people at high risk for severe varicella who are not eligible to get the vaccine. Varicella-zoster immune globulin is preferred over intravenous immunoglobulin (IGIV) because clinical data demonstrating effectiveness of IGIV for varicella postexposure prophylaxis are lacking. However, IGIV may be used for postexposure prophylaxis if varicella-zoster immune globulin is unavailable.

For infants born to mothers who develop VZV infection 5 days before to 2 days after delivery, there is insufficient time for the production and transplacental transfer of VZV-specific maternal IgG to the infant. In addition, the neonate's cellular immune system is immature, further increasing the risk of severe infection. Administration of varicella-zoster immune globulin within the 96-hour timeframe may be successful in preventing, ameliorating, or delaying varicella disease.

Varicella-zoster immune globulin for postexposure pro-phylaxis is recommended for the following high-risk individuals who have significant exposure to varicella and who cannot be immunized:

- Neonates born to mothers who develop chickenpox within 5 days before and 48 hours after delivery
- Neonates at greater than 28 weeks' gestation whose mothers lack evidence of immunity
- Neonates at less than 28 weeks' gestation or weighing less than 1000 g in birth weight regardless of maternal immunity
- Pregnant women without evidence of immunity
- Immunocompromised persons without evidence of immunity

Other people without evidence of varicella immunity in whom varicella-zoster immune globulin may be used for postexposure prophylaxis include household contacts, play-mates with significant contact, close hospital contacts, and those with intimate contact with a contagious person with varicella or zoster.

There are limited data on the use of acyclovir for post-exposure prophylaxis against varicella in healthy children. Some experts recommend the administration of prophylactic acyclovir if varicella-zoster immune globulin or IGIV is unavailable or not administered within 96 hours of exposure. Parenteral foscarnet is used almost exclusively in immunocompromised hosts with infections caused by acyclovir-resistant VZV. Varicella vaccine is recommended for postexposure prophylaxis for healthy people without evidence of immunity 12 months or older. The vaccine may prevent or ameliorate disease if administered within 72 (some data support up to 120) hours after exposure to varicella.

Varicella vaccine should not be administered to people with congenital or acquired T-cell immunodeficiency, including those with leukemia, lymphoma, other malignant tumors that affect the bone marrow or lymphatics, human immunodeficiency virus infection with a CD4+ T-lymphocyte percentage less than 15%, and long-term immune suppression, including high-dose (2 mg/kg of prednisone) systemic steroid use for more than 2 weeks. Varicella vaccine also should not be administered to pregnant women or those with a life-threatening reaction to any component of the vaccine.

PREP Pearls

- Intramuscular varicella-zoster immune globulin (within 96 hours after exposure) is recommended for high-risk individuals who have significant exposure to varicella and who cannot be immunized.
- Varicella vaccine (72-120 hours after exposure) is recommended for postexposure prophylaxis for healthy people without evidence of immunity 12 months or older.

American Board of Pediatrics Content Specification(s):

- Know the indications for the use of immune globulin and varicella vaccine in patients exposed to varicella

Suggested Reading:

- American Academy of Pediatrics. Varicella-zoster infections. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:774-789
- Centers for Disease Control and Prevention. A new product (VariZIG) for postexposure prophylaxis of varicella available under an investigational new drug application expanded access protocol. MMWR Morb Mortal Wkly Rep. 2006;55:209-210
- Centers for Disease Control and Prevention. FDA approval of an extended period of administering VariZIG for postexposure prophylaxis of varicella. Morbidity and Mortality Weekly Report. 2012;61(12):212
- Centers for Disease Control and Prevention. Prevention of varicella; recommendations of the Advisory Committee on Immunization Practices (ACIP). MMWR Recomm Rep. 2007;56(RR-4):1-40

Item 38

The mother of a 17-year-old girl made an appointment for her daughter to see you for a gynecologic examination. She learned that her daughter had 2 Chlamydia infections in the past year and requests that her daughter be "fully tested" and have a Papanicolaou (Pap) smear. The girl is otherwise healthy and asymptomatic today. She has received 3 doses of quadrivalent human papillomavirus (HPV) vaccine.

Of the following, the advice you are MOST likely to provide is that Pap smear testing should be

A. deferred until the girl reaches 18 years of age
B. deferred until the girl reaches 21 years of age
C. performed today because it has been requested
D. performed today because of the girl's history of Chlamydia infection
E. unnecessary because the girl has been immunized against HPV

Item 38 Preferred Response: B

Traditionally, screening with cervical cytologic testing for precancerous changes (ie, cervical dysplasia and cervical intraepithelial neoplasia) was routinely performed in all sexually active females using the Papanicolaou (Pap) smear. Recent recommendations from the US Preventive Services Task Force (USPSTF) have changed the timing of initiating screening to 21 years and older, and their recommendation is against screening for cervical cancer in women younger than 21 years because epidemiologic studies indicate that cervical cancer is rare in this age group. Exposure of the cervical cells to oncogenic types of human papilloma virus (HPV) during vaginal intercourse may eventually lead to cervical cancer. However, this process is not rapid; most often the virus is cleared and early lesions regress. These recommendations don't apply to those females who are immunocompromised Evidence indicates that there is more harm than benefit from Pap smear screening before 21 years of age. Complications from diagnostic procedures (eg, cervical biopsies), as documented in studies, include vaginal bleeding, pain, and infection. Abnormal results could result in short-term increase in anxiety from health concerns. Risks from the treatment procedure (eg, loop electrosurgical excision procedure [LEEP]) include the potential for adverse pregnancy outcomes, such as preterm delivery with an associated low birth-weight infant and an increased risk for perinatal death.

In their decision analyses, the USPSTF found little evidence of the influence of sexual history on the age at which to begin screening. Therefore, there is no need to screen patients earlier if they have had sexually transmitted infections. The fact that someone has been vaccinated against HPV does not negate the need for Pap smear screening starting at 21 years of age.

PREP Pearls

- Routine Pap smear testing should start at 21 years of age; a history of sexually transmitted infections does not change this recommendation.
- Diagnostic procedures on the cervix increase the risk for future negative pregnancy outcomes.

American Board of Pediatrics Content Specification(s):

- Know the indications for a Papanicolaou smear in adolescence

Suggested Reading:

- ACOG Committee on Practice Bulletins—Gynecology. ACOG Practice Bulletin no. 109: cervical cytology screening. Obstet Gynecol. 2009;114:1409-1420. doi:10.1097/A0G.0b0l3e3l81c6f8a4
- Greydanus DE, Omar H, Patel DR. What's new: cervical cancer screening in adolescents? Pediatr Rev. 2009;30:23-25. doi:10.1542/pir.30-1-23
- Moyer VA; on behalf of the U.S. Preventive Services Task Force. Screening for Cervical Cancer: U.S. Preventive Services Task Force Recommendation Statement. Ann Intern Med. 2012; 156:880-891, W312. doi:10.7326/0003- 4819-156-12-201206190-00424

- Saslow D, Solomon D, Lawson HW, et al; American Cancer Society; American Society for Colposcopy and Cervical Pathology; American Society for Clinical Pathology. American Cancer Society, American Society for Colposcopy and Cervical Pathology, and American Society for Clinical Pathology screening guidelines for the prevention and early detection of cervical cancer. Am J Clin Pathol. 2012;137:516-5142. doi:10.1309/AJCPTGD94EVRSJCG

Item 39

A 13-year-old boy is seen in the emergency department with a complaint of "something stuck in my chest. ' He was well until eating dinner, which is when the symptom occurred. The problem was relieved after he drank a large glass of apple juice. The past medical history demonstrates that he has been taking oral tetracycline for facial acne and is trying to lose weight. He does not complain of odynophagia, heartburn, or regurgitation. The boy's parents report that he seems to eat very slowly and drinks copious amounts of water with his meals. His body mass index is 27, but all physical examination findings are normal.

Of the following, the MOST appropriate next step you recommend is

A. abdominal ultrasonography
B. esophageal manometry
C. esophageal pH study
D. upper gastrointestinal endoscopy
E. videofluoroscopic swallowing study

Item 39 Preferred Response: D

Feeding and swallowing disorders during childhood often occur in conjunction with complex medical, neurologic, and developmental conditions. However, in a previously well child, a careful clinical history will often indicate the most likely cause of the symptom. In the adolescent described in the vignette, the use of tetracycline to treat facial acne and the swallowing problems during mealtimes strongly suggest dysphagia, occurring as the consequence of 1 of 2 diagnoses: food impaction or pill esophagitis. Considering these likely causes, the most appropriate diagnostic study, in both conditions, is an upper gastrointestinal tract endoscopy.

If a child has a suspected foreign-body ingestion (either intentional or unintentional), radiographic imaging generally should be conducted, unless the ingested object is known to be radiolucent. For the boy in the vignette, we are presented with a clear history of relatively long-standing solid food dysphagia, suggested by the consumption of copious amounts of water during meals. Food impaction, which prompted the emergency department visit, is most often associated with eosinophilic esophagitis, a disorder being recognized with increased frequency. The history is complicated by the use of tetracycline, the agent most commonly associated with pill esophagitis. In these conditions, endoscopy will serve a diagnostic and therapeutic function. Although foreign-body ingestions account for far greater than 20,000 emergency department visits yearly in the United States, estimates suggest that up to 75% of ingestions are either not witnessed or not reported. Although gastric foreign bodies are generally asymptomatic (and may never be identified because most pass uneventfully), esophageal foreign bodies should always be considered impacted, and they most often present with age-dependent symptoms. excessive drooling, vomiting and/or feeding refusal in infants and toddlers, and dysphagia in older children and adolescents. Because impacted esophageal foreign bodies are associated with significant morbidity, their urgent removal, usually within 12 hours, is indicated in all cases. An important exception is ingestion of button (disk) batteries. These objects must be removed from the esophagus within 2 hours of presentation because esophageal impaction for a longer period has been associated with significant complications, including esophageal ulceration, perforation, spondylodiscitis, and esophageal–vascular fistulization. Pointed objects should also be removed as soon as possible to reduce the risk of perforation. In the case of food impaction, meat is the most commonly reported impacted substance.

Eosinophilic esophagitis is a chronic, immune/antigen-mediated esophageal disease characterized by esophageal dysfunction and histopathologically by an eosinophil-predominant inflammatory response. A common mode of presentation is meat impaction. In all such cases, the impacted material should be endoscopically removed. "Pushing" the meat into the stomach with a nasogastric tube is contraindicated because of the risk of perforation of the inflamed esophagus. The use of meat tenderizers is similarly to be avoided because these compounds can also digest normal tissues.

Other diagnostic studies are unlikely to be of assistance in establishing a diagnosis for the boy in the vignette. In patients with known or suspected radiolucent foreign-body ingestion, abdominal ultrasonography may be considered, particularly if a gastric foreign

body is likely. However, available data do not support the use of ultrasonography in the evaluation of dysphagia. Additional, elective studies of esophageal function (manometry, pH monitoring, and videofluoroscopic swallowing study) should only be considered in cases in which endoscopy does not establish a diagnosis (eg, eosinophilic esophagitis and pill esophagitis) and an underlying motility disorder is suspected.

PREP Pearls

- In adolescent patients with eosinophilic esophagitis, dysphagia from food impaction is the most common presentation.
- Tetracycline should always be taken with copious amounts of water and never at bedtime because of the risk of pill esophagitis.
- Dysphagia commencing during adolescence should always signal an evaluation for eosinophilic esophagitis.

American Board of Pediatrics Content Specification (s):

- Know the treatment of an esophageal foreign body

Suggested Reading:

- American Society for Gastrointestinal Endoscopy. Guideline for the management of ingested foreign bodies. Gastrointest Endosc. 2002;55:802 806. doi: 10.1016/S0016-5107%2895%2970030-7
- Liacouras CA, Furuta GT, Hirano I, et al. Eosinophilic esophagitis: updated consensus recommendations for children and adults. I Allergy Clin Immunol. 2011;128:3-20. doi:10.1016/j.jaci.2011.02.040
- Litovitz T. Emerging battery-ingestion hazard: clinical implications. Pediatrics. 2010;125:1168-1177. doi:10.1542/peds.2009-3037
- Mas E, Olives I. Toxic and traumatic injury of the esophagus. In: Kleinm, RE, Sanderson IR, Goulet 0, Sherman PM, Mieli-Vergani G, Shneider BL, eds. Walker's Pediatric Gastrointestinal Disease: Pathophysiology, Diagnosis and Treatment. 5th ed. Hamilton, Ontario, Canada: BC Decker Inc; 2008:105-116
- Orenstein SR. Oral, pharyngeal, and esophageal motor disorders in infantand children. GI Motility Online. May 16, 2006. doi:10.1038/gimo38

Item 40

A 6-week-old infant is being discharged after a hospitalization for an apparent life-threatening event (ALTE). He was born at term and had no medical problems before this event. During the hospitalization, no underlying cause for the ALTE was found, and he remained well, had a normal physical examination, and had no further events. His parents ask about home apnea monitoring.

Of the following, the MOST accurate information to give these parents is

A. the benefits of home monitoring to the infant outweigh the negative psychosocial effects on parents
B. the psychosocial changes the parents will experience are likely to remain constant during the entire time the infant is monitored
C. the parents are likely to experience early increases in depression and hostility once home monitoring is instituted
D. the parents are likely to experience increases in depression starting about 6 months after home monitoring is instituted
E. the parents are likely to report feeling that home monitoring of their infant is not helpful

Item 40 I-C S Preferred Response: C

Evidence does not indicate that home apnea monitors have saved lives or had any effect on the incidence of sudden infant death syndrome. Despite the lack of consensus on the indications, timing, and duration of monitoring, home cardiorespiratory monitoring is still an intervention that many pediatricians and neonatologists consider for individual patients. The psychological effects on families by the presence of a home apnea monitor, both good and bad, should be one factor that the practitioner considers when contemplating prescribing this intervention.

Reports show family psychosocial responses to be some-times conflicted when a home apnea monitor is prescribed for an infant. Any family with a newborn infant experiences stress, sleep deprivation, and fatigue, but one expects that these effects would be magnified if the infant has a medical problem that requires home monitoring. Studies have demonstrated increased parental anxiety, increased mood disturbances in mothers, and social isolation especially if the family does not have access to respite care. However, in other studies, parents report that the presence of a monitor is a source of comfort, especially if they had previously lost a child or if the current infant had a cyanotic episode before the monitor was instituted. Parents have frequently described the monitor as helpful to them.

A 1999 study compared 2 groups of parents of infants discharged from a neonatal intensive care unit: 1 group discharged with monitors and the other without monitors. The most striking finding was that depression and hostility increased in the first 2 weeks after hospital discharge for the monitor group in contrast to the no-monitor group in whom depression decreased and hostility stayed constant during the same period. In the monitor group, depression and hostility steadily decreased between 2 weeks and 6 months after discharge. Interestingly, the no-monitor group had increased feelings of hostility by 3 to 6 months after hospital discharge. The authors hypothesized that this may be because mothers returned to work outside the home by that time and were having difficulty juggling multiple roles. This, however, was speculation and was not studied. There was no change in family functioning scores in either group.

PREP Pearls

- There is no evidence that home apnea monitors are effective in decreasing mortality or preventing sudden infant death syndrome.
- Use of apnea monitors in the home may increase parental anxiety, mood disturbance, and social isolation, especially immediately after hospital discharge.
- Parents often describe home apnea monitors as helpful and a source of comfort.

American Board of Pediatrics Content Specification(s):

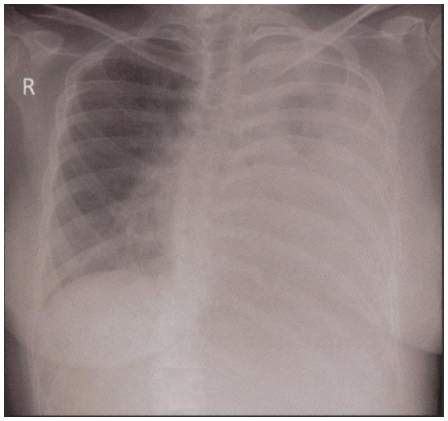
- Recognize the psychosocial issues surrounding the use of home monitors

Suggested Reading:

- Abendroth D, Moser DK, Dracup, K, Doering LV. Do apnea monitors decrease emotional distress in parents of infants at high risk for cardiopulmonary arrest? JPediatr Health Care. 1999;13:50-57. doi:10.1016/ S0891-5245(99)90053-6
- American Academy of Pediatrics, Committee on Fetus and Newborn. Apnea, sudden infant death syndrome and home monitoring. Pediatrics. 2003;111(4 pt 0:914-916
- Silvestri JM. Indications for home apnea monitoring (or not). Clin Perinatol. 2009;36:87-99

Item 41

A 15-year-old girl was admitted to the hospital 3 days ago for a large left-sided pneumonia with associated pleural effusion (Item Q41). At the time of admission, she was started on ceftriaxone and placed on oxygen via nasal cannula. Over the last 3 days, she has had daily fevers along with a persistent cough. Her vital signs this morning are temperature 39.0°C, heart rate 100 beats/min, and respiratory rate 35 breaths/ min. Her oxygen saturation is 94% on 3 liters/min of oxygen via nasal cannula. Daily chest radiographs, which are unchanged since admission, demonstrate the persistence of the pneumonia and pleural effusion. On physical examination, she has mild respiratory distress, prolonged expiration, and unchanged decreased breath sounds over the left side of her chest. She has a grade 2/6 systolic ejection murmur.



ITEM Q41: Radiographic findings as described for the girl in the vignette.

Of the following, the MOST appropriate next step is

A. bronchoscopy with bronchoalveolar lavage
B. echocardiogram
C. open lung biopsy
D. sputum culture
E. video-assisted thoracoscopic surgery (VATS)

Item 41 TE Preferred Response: E

Most patients who have bacterial pneumonia recover uneventfully with appropriate treatment, but 10% of patients may require hospital admission and an even smaller percent-age develop pleural and parenchymal complications, such as empyema, lung abscess, necrotizing pneumonia, pneumothorax, and bronchopleural fistulas. The patient in the vignette was admitted to the hospital for acute pneumonia with a pleural effusion and started on appropriate antibiotic therapy. She failed to improve over 3 days as evidenced by persistence of clinical signs and symptoms and no change in her pneumonia and effusion on radiographic examination.

Children who have pneumonia who are not responding to initial therapy after 48 to 72 hours should have a clinical, laboratory, and radiographic reassessment to help determine the cause of the lack of response and plan further investigation and treatment. In children with moderate to large pleural effusions, especially those that impair respiratory function, these effusions should be drained. For the girl in the vignette, drainage may have been warranted on admission but is clearly indicated now. The choice of video-assisted thoracoscopic surgery or thoracostomy (fibrinolytic agents will need to be added if there is evidence of loculation) is institution dependent. Sputum culture may be of diagnostic benefit in older children who can cough and produce sputum but drainage of the effusion would be a higher priority. Bronchoscopy with alveolar lavage is indicated for obtaining specimens for Gram stain and culture in a child receiving mechanical ventilation who has an unknown infection. However, performing bronchoscopy in the patient in the vignette may prompt respiratory failure and therefore drainage of the effusions would be preferred. More invasive procedures such as open lung biopsy and percutaneous lung aspiration could be considered in critically ill children with an unknown diagnosis who are failing to improve. In the absence of suspected cardiac disease, echocardiography would not be indicated.

PREP Pearls

- Approximately 10% of pediatric patients with bacterial pneumonia may require hospital admission.
- Children with pneumonia who do not respond to initial therapy after 48-72 hours should have a clinical, laboratory, and radiographic reassessment.
- Moderate to large pleural effusions, especially those that impair respiratory function, should be drained.

American Board of Pediatrics Content Specification(s):

- Know that invasive studies (eg, bronchoscopy, lung aspiration, open lung biopsy) may be indicated in patients with acute pneumonia

Suggested Reading:

- Bradley JS, Byington CL, Shah SS, et al. The management of community-acquired pneumonia in infants and children older than 3 months of age: clinical practice guidelines by the Pediatric Infectious Diseases Society and the Infectious Diseases Society of America. Clin Infect Dis. 2011;7:e25-e76. doi:10.1093/cid/cir531

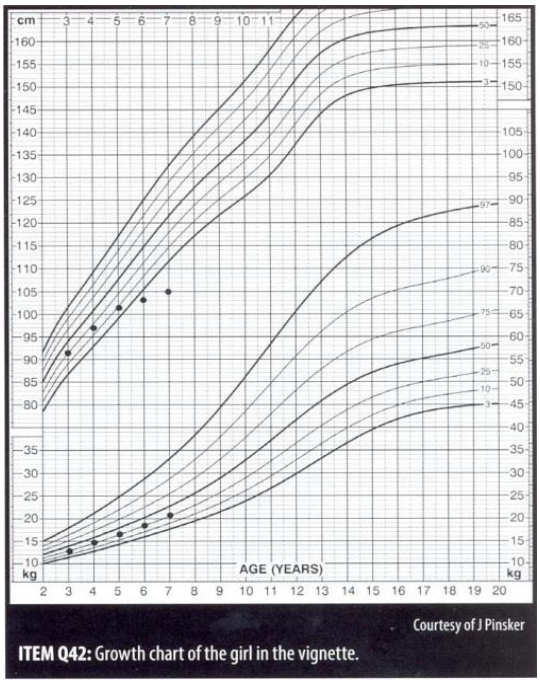
- Durbin WJ, Stille C. Pneumonia. Pediatr Rev. 2008;29:147-160. doi:10.1542/pir.29-5-147. http://pedsinreview.aappublications.org/content/29/5/147. full
- Efrati 0, Barak A. Pleural effusions in the pediatric population. Pediatr Rev. 2002:23:417-426. doi:10.1542/pir.23-12-417
- Li ST, Tanredi DJ. Empyema hospitalizations increased in US children despite pneumococcal conjugate vaccine. Pediatrics. 2010;125:26-33. doi:10.1542/peds.2009-0184
- Schultz KD, Fan LL, Pinksy J, et al. The changing face of pleural empyemas in children: epidemiology and management. Pediatrics. 2004;113:1735-1740
- Winnie GB, Lossef SV. Pleurisy, pleural effusions, and empyema. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. 2011:1505-1509

Item 42

During a health supervision visit, a 7-year-old girl is noted to be growing poorly. She has had a minimal increase in height during the last 2 years, but her weight continues to track at the 25th percentile (Item Q42). The mother reports that her daughter is a picky eater but is otherwise well. The girl has no constipation, diarrhea, or other signs of malabsorption. Her parents are not concerned because her mother is 5 feet, 1 inch (155 cm) tall and her father is 5 feet, 6 inches (168 cm) tall, and both had delayed puberty and had growth spurts in their late teenage years.

Of the following, the MOST likely diagnosis is

A. constitutional delay of growth
B. Crohn disease
C. familial short stature
D. hypothyroidism
E. inadequate caloric intake



ITEM Q42: Growth chart of the girl in the vignette.

Item 42 Preferred Response: D

When linear growth arrests, particularly as weight is maintained or increases, an underlying endocrine cause is likely. Hypothyroidism is the only endocrine disorder listed that fits the growth pattern seen in the girl described in this vignette. Other conditions that can follow this pattern include growth hormone deficiency and genetic disorders, such as Turner syndrome.

Gastrointestinal disorders, such as inflammatory bowel disease (especially Crohn disease) or other chronic disease—mediated causes of malnutrition often present initially with poor weight gain and impaired linear growth, especially if the period of inadequate nutriture is prolonged.

Despite a family history of short stature and pubertal delay, this growth chart is not consistent with familial short stature or constitutional delay of growth. In these conditions, linear growth often resets to the bottom of the growth chart in the first 18 months to 2 years of life and then tracks at a normal growth velocity thereafter. Children with familial short stature will continue this growth pattern until their final adult height is reached. They would not reset their growth pattern at 5 years of age, as noted in the girl in the vignette. Children with constitutional delay of growth will continue growing at a prepubertal growth rate beyond the expected age and then have catchup growth later, consistent with their family history.

PREP Pearls

- An endocrine cause of poor growth is likely when linear growth arrests but weight gain is either normal or increasing.
- In most cases where poor growth is related to undernutrition, falloff in weight gain will precede falloff in linear growth.

American Board of Pediatrics Content Specification(s):

- Distinguish among constitutional short stature, genetic (familial) short stature, and growth hormone or thyroid deficiencies by growth chart evaluation

Suggested Reading:

- Rosenfeld RG, Cohen P. Disorders of growth hormone/insulin-like growth factor secretion and action. In: Sperling M, ed. Pediatric Endocrinology. 3rd ed. Philadelphia, PA: Saunders; 2008:281-305
- Weintraub B. Growth. Pediatr Rev. 2011;32(9):404-406. doi:10. I 542/pir. 32-9-404

Item 43

A 17-year-old girl in your practice returned 3 weeks ago from a semester as an exchange student in Ghana. She complains of having mild abdominal discomfort since she returned. The discomfort has progressed to severe abdominal pain in association with the development of grossly bloody stools. She has had intermittent, low-grade fever (up to 38.2°C) and reports a 4- to 5-pound weight loss. On physical examination, she appears uncomfortable but nontoxic. Her abdominal examination reveals increased bowel sounds and diffuse tenderness without rebound. There is no enlargement of the liver or spleen.

Of the following, the MOST likely cause of this patient's symptoms is infection with

A. Entamoeba histolytica
B. enterotoxigenic Escherichia coli
C. Escherichia coli 0157:H7
D. Giardia intestinalis
E. rotavirus

Item 43 S Preferred Response: A

The gradual development of gastrointestinal symptoms over 3 weeks, progressing to development of dysenteric, bloody stools, in someone returning from a developing country, is most consistent with amebiasis caused by the protozoan Entamoeba histolytica. The gastrointestinal manifestations of amebiasis run the gamut from asymptomatic infection or nonspecific intestinal tract complaints to amebic dysentery. Weight loss is frequently reported because of the gradual onset. Fever is generally absent or low-grade. Rarely, progressive involvement of the colon may lead to fulminant colitis, toxic megacolon, or ulceration of the colon. A palpable intraabdominal mass (most commonly in the cecum) or ameboma, may also occur as an isolated manifestation of this infection.

Treatment of amebiasis involves eliminating both the active, invading trophozoites and intestinal cysts of the organism. Invasive colitis, as well as extraintestinal infections, is generally treated with metronidazole for 7 to 10 days. Tinidazole and nitazoxanide are potential alternatives to metronidazole. This treatment is then followed by an intraluminal agent (paromomycin or iodoquinol) to eliminate the remaining intestinal cysts and prevent reinfection and transmission.

A small proportion of E. histolytica infections result in extraintestinal involvement often in the absence of intestinal symptoms. Liver abscesses are the most common extraintestinal amebic infection and may present with high fever, hepatic tenderness, and hepatomegaly in acute conditions or with vague abdominal symptoms, weight loss, and irritability in chronic conditions. Once in the liver, dissemination to lung, pleura, or pericardium may occur. Hematogenous spread to the brain has also been rarely reported. Prevention of amebic infection in persons traveling to developing countries involves avoidance of untreated water and uncooked foods such as fruits and vegetables. ("Boil it, peel it, or forget it:') Sexual practices, such as anal inter- course, which may contribute to fecal-oral transmission, also Enterotoxigenic Escherichia coli is associated with travelers' diarrhea, which is typically a self-limited 1- to 5-day illness with fever, watery stools, and abdominal cramps. E coli 0157:H7 is a shiga toxin-producing strain associated with diarrhea, hemorrhagic colitis, and hemolytic-uremic syndrome. The course of illness with this organism is much more acute than described for the girl in the vignette, with bloody stools developing over 3 to 4 days after the onset of symptoms. Giardia intestinalis (previously called G lamblia) infection may be subacute or chronic in presentation but typical symptoms are abdominal bloating, watery diarrhea, and malabsorption, not bloody stools or severe abdominal pain as in this case. Symptomatic rotavirus infection primarily affects infants and young children, and presents as an acute illness characterized by fever, vomiting, and diarrhea.

PREP Pearls

- Intestinal manifestations of amebiasis present gradually, over weeks, and may run the gamut from asymptomatic to severe dysentery.
- Extraintestinal manifestations of amebiasis are less common, with liver abscess being the most common such presentation.

- Treatment of amebic infection involves eradicating the active trophozoites (with metronidazole) and the intraintestinal cysts (with paromomycin or iodoquinol).

American Board of Pediatrics Content Specification(s):

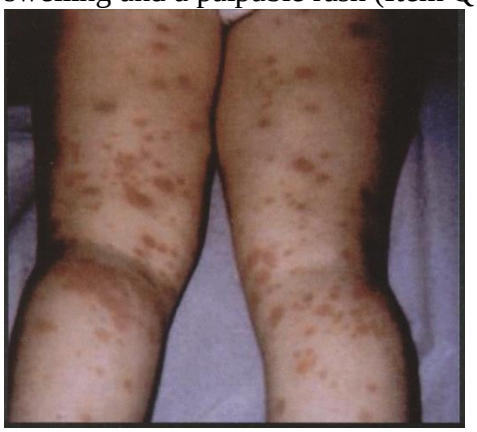
- Recognize the clinical manifestations of amebiasis

Suggested Reading:

- American Academy of Pediatrics. Amebiasis. In: Pickering LK, Baker Cj, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:222-225
- American Academy of Pediatrics. Escherichia coli diarrhea (including hemolytic-uremic syndrome). In: Pickering LK, Baker Cj, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:324-328
- Leder K, Weller PF. Intestinal Entamoeba histolytica amebiasis. UptoDate. 2013. Available online only for subscription

Item 44

A previously healthy, 5-year-old boy presents to your office with a 3-day history of bilateral knee pain and swelling. This morning, he developed a rash over his buttocks and lower extremities. There is no history of trauma or recent illness. On physical examination, he is well-appearing. His vital signs are unremarkable. He has bilateral knee swelling and a palpable rash (Item Q44).

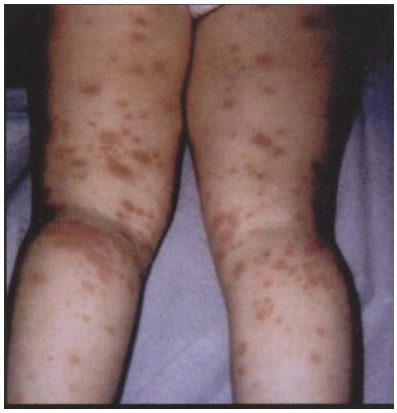


Of the following, the MOST likely diagnosis is

A. infectious mononucleosis
B. Henoch-Schonlein purpura
C. erythema infectiosum
D. exanthem subitum
E. meningococcemia

Item 44 Preferred Response: B

The rash and constellation of symptoms in the boy described in the vignette are consistent with a diagnosis of Henoch-Sch6nlein purpura (HSP). HSP is a leukocytoclastic vasculitis that primarily affects small blood vessels. The European League Against Rheumatism (EULAR)-Paediatric Rheumatology Europe Society (PRES) criteria for HSP include purpura or petechiae with lower limb predominance (Item C44, page C-35) and at least 1 of the following: arthritis or arthralgia, abdominal pain, histopathology demonstrating immunoglobulin A deposition, or renal involvement demonstrated by hematuria or proteinuria. Occasionally, the purpura are preceded by an urticarial or maculopapular rash that fades. The vasculitis rash follows, usually within 24 hours, with a purpuric, necrotic, or deep bruising appearance. The purpuric rash is most often on dependent areas but can be seen on arms, face, and ears.



ITEM C44: *Well-defined and slight petechial lesions in a boy with Henoch-Schonlein purpura.*

Arthritis affects about 75% of children with HSP. Most often, the knees and ankles are affected. The arthritis of HSP is usually oligoarticular, nondeforming, and self-limited. Approximately 15% of patients have arthritis as the presenting feature of HSP.
Abdominal pain occurs in over half of patients who have HSP and can precede the pathognomonic rash by as much as 2 weeks. Other gastrointestinal manifestations include intestinal bleeding and intussusception. While intussusception is rare and only occurs in 1% to 5% of children, it should be considered in this clinical setting. Intestinal bleeding occurs in 33% of HSP patients.

The most common renal manifestation of HSP is microscopic hematuria. While renal involvement is relatively common, occurring in 20% to 60% of patients, chronic renal impairment occurs in 2% to 15% and end-stage renal disease occurs in only 1% of HSP patients. Most renal involvement presents within the first 6 weeks of disease, and 97% occurs within 6 months.

Rare manifestations seen in HSP include scrotal edema, periorbital edema, hand swelling, pulmonary hemorrhage, seizure, stroke, and mental status changes. The clinical symptoms of HSP can last up to a month and can recur in one-third of patients. Management of HSP is primarily supportive and includes analgesics and nonsteroidal anti-inflammatory drugs (NSAIDs). Early use of corticosteroids in the hospital setting should be considered in the case of gastrointestinal comorbidities. Arthritis should be managed with NSAIDs; however, corticosteroids may be used in severe or nonresponsive cases. The use of corticosteroids in mild cases of HSP remains controversial. The rare, life-threatening, or severe manifestations of HSP, including acute renal failure, intussusceptions, and severe arthritis, often require management by a subspecialist.

The purpuric rash seen in the boy in this vignette is not consistent with infectious mononucleosis, erythema infectiosum, or erythema subitum. He is well appearing and afebrile, which would not be consistent with meningococcemia.

PREP Pearls

- Gastrointestinal symptoms may be a presenting feature of HSP.
- Arthritis may be the presenting feature of HSP and can precede the rash by up to 2 weeks.
- Patients with HSP should be monitored for the occurrence of renal disease for 6 months after diagnosis.

American Board of Pediatrics Content Specification(s):

- Recognize that Henoch-Schonlein purpura may present with initial abdominal pain or joint complaints, and manage appropriately

Suggested Reading:

- Eleftheriou D, Brogan PA. Vasculitis in children. Pediatric Endocrinology. 2009;23(3):309-323. doi:10.1016/j.berh.2009.02.001
- Gedalia A, Cuchacovich R. Systemic vasculitis in childhood. Curr Rheumatol Rep. 2009;11(6):402-409. doi:10.1007/s11926-009-0059-4
- Weiss P. Pediatric vasculitis. Pediatr Clin N Am. 2012;59(2):407-423

Item 45

An 8-year-old boy is seen in your office for problem behavior. He will not stay in his seat at school, does not pay attention well in class, and does not seem to be learning as expected. Last year, he was treated with methylphenidate followed by dextroamphetamine with no significant effect. He is becoming increasingly disruptive in class and refusing to do work. Notes are now being sent home repeatedly reporting aggressive behavior with others. Upon questioning, you learn that there appears to be no new social stressors, he has not been bullied, and there is no particular family disruption. His school and home behavior have worsened each year, but he is far less disruptive at home. According to his mother, he achieved his early motor milestones at the expected times. His physical examination is unremarkable, with no dysmorphic features, and normal vision and hearing.

Of the following, the MOST appropriate next step is to

- perform a blood lead level test
- recommend an electroencephalogram
- recommend genetic testing
- recommend a psychoeducational evaluation
- refer the boy for evaluation of possible child abuse

Item 45 SBP Preferred Response: D

The ineffectiveness of both methylphenidate and dextroamphetamine to treat what was initially presumed to be attention-deficit/hyperactivity disorder (ADHD) suggests that something other than ADHD is causing problems for the boy described in the vignette. Behavior problems at school that are far greater than at home, overall worsening with each year, and very poor school performance are suggestive of a learning disability. Children who cannot learn as easily as others in the class often develop increasing frustration and feelings of inadequacy, which can be expressed as disruptive behavior and even aggression. Schools are required by special education law to respond to a parent's learning concerns for their child (particularly if that concern is expressed in writing rather than verbally) but are notably not required to respond to a physician's concerns about a learning disability. Therefore, encouraging the child's parent to reach out to the school to obtain a psychoeducational evaluation is the most effective next step. Other worthwhile steps might include performing screening assessments for both vision and hearing to make sure the child has no sensory problems affecting his ability to learn.

Checking a lead level may be warranted in a young child who has ADHD symptoms or learning difficulties and who is living in a home, or spending time in an environment that might contain lead paint (ie, contains paint from before 1977) or other sources of lead exposure. However, the steady worsening of symptoms in an 8-year-old child is unlikely to be caused by lead toxicity unless that child is continuing to increase his exposure to toxic lead. Pica, as the primary path-way to lead toxicity, is fairly common in children younger than age 5 years but uncommon in school age children unless they have a major developmental impairment or suffer from severe neglect. Although children with seizure disorders can have comorbid learning disabilities, the absence of a seizure history makes electroencephalography unlikely to be helpful. Although genetic syndromes are often associated with learning disabilities, the absence of dysmorphic features or hallmark traits suggestive of a genetic disorder makes genetic testing unlikely to be helpful. Although inattention and behavior problems at school could result from child abuse, no information in this child's history suggests abuse. Referral for an abuse evaluation in a case where there is no reason to suspect abuse is likely to cause unnecessary distress for the family and potential mistrust of clinicians.

PREP Pearls

- Learning disabilities are common in children with ADHD.
- For children who appear to have ADHD but fail to respond to stimulants, evaluation for a learning disability should be considered.

AAP Mental Health Competency:

- Recognize the risk factors behind chronic aggression, such as the high rate of learning disorders and academic underachievement in conduct disordered children

Suggested Reading:

- American Academy of Child and Adolescent Psychiatry. Practice parameters for the assessment and treatment of children and adolescents with language and learning disorders. J Am Acad Child Adolesc Psychiatry. 1998;37(10 suppl):465-62S.
- American Academy of Pediatrics. Learning disabilities, dyslexia, and vision: a subject review. Pediatrics. 2009;124(2):837-844. doi: 10.1542/ peds.2009-1445

Item 46

During morning rounds, the nurse informs you that a 3.8-kg neonate has just been delivered by caesarean section due to breech presentation. The pregnancy was notable for well-controlled maternal type I diabetes. The maternal glycated hemoglobin A,, values ranged between 5% to 6% before conception and during pregnancy. Level II screening ultrasonography done at 18 weeks of gestation was normal. Your assessment at 40 minutes after birth reveals a pink, well-perfused neonate with normal tone, strong suck, and good color. Cardiac examination reveals a grade 1/6 systolic murmur at the left lower sternal border with a preductal oxygen saturation of 97% on room air. The mother is anxious to breastfeed.

Of the following, the MOST appropriate next step in management is to immediately

A. determine blood glucose concentration
B. initiate breastfeeding
C. obtain a chest radiograph
D. perform an echocardiogram
E. supplement with formula

Item 46 TE Preferred Response: B

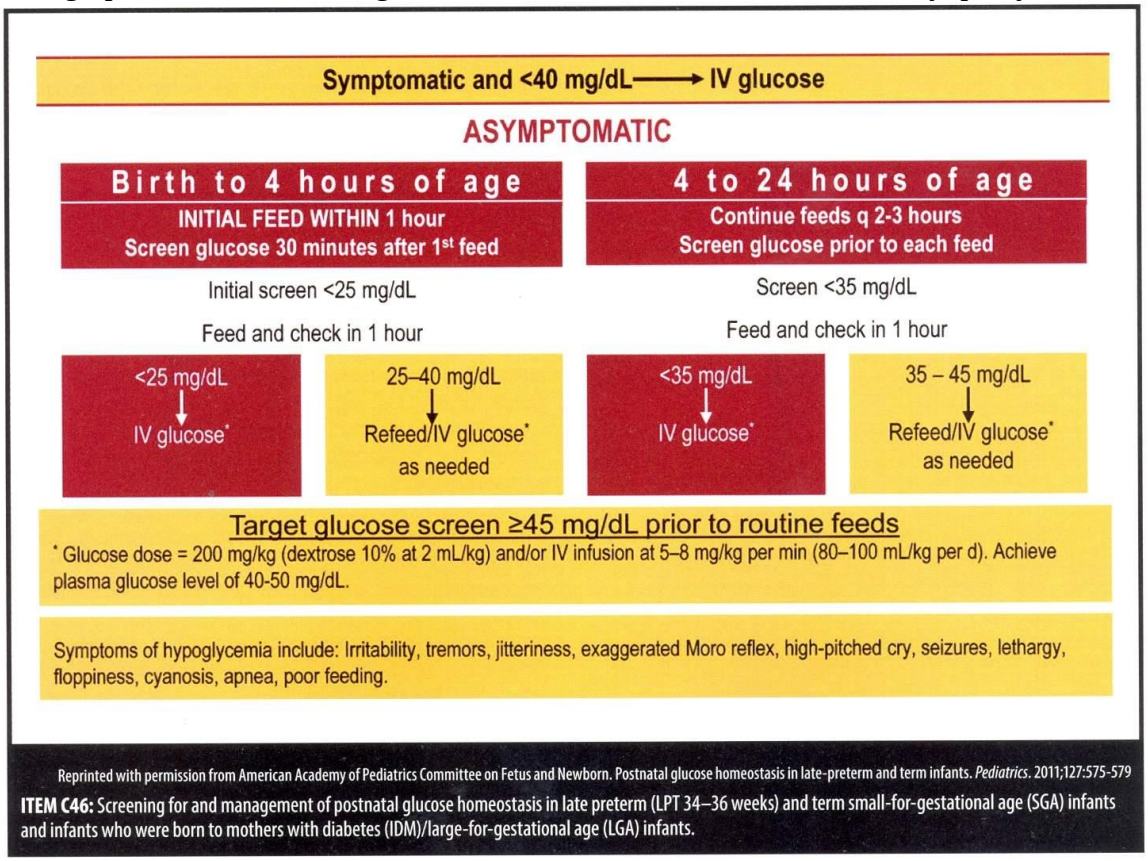
An infant of a diabetic mother (IDM) who has no symptoms of hypoglycemia should be breastfed within the first hour after birth. The Committee on Fetus and Newborn of the American Academy of Pediatrics published a clinical report in 2011 outlining screening and management strategies for hypoglycemia in the late-preterm and full-term infant (Item C46). Early feeding, within the first hour after birth, with a screening glucose value obtained 30 minutes after the first feeding, is the first step in the management of the asymptomatic infant described in this vignette. Glucose concentrations normally decline in the first 1 to 2 hours after birth following the discontinuation of maternal glucose influx with umbilical cord clamping. Plasma glucose values as low as 30 mg/dL (1.7 mmol/L) may be seen transiently during this period. The plasma glucose concentrations then routinely increase above 45 mg/dL (2.5 mmol/L) by 12 hours after birth. Infants at risk for neonatal hypoglycemia, including late-preterm, small for gestational age, large for gestational age, and IDM, should be monitored closely during this period for clinical symptoms of hypoglycemia. These signs include jitteriness, tremors, high-pitched cry, irritability, lethargy, floppiness, poor suck, apnea, cyanosis, exaggerated Moro reflex, and seizure activity. Screening glucose values should be obtained in infants with symptoms and intravenous glucose initiated if the value is less than 40 mg/dL (2.2 mmol/L).

Maternal hyperglycemia is hypothesized to produce hyperglycemia in the developing fetus, leading to fetal pancreatic stimulation, islet cell hyperplasia, and hyperinsulinemia in the IDM. While contributing to the hypoglycemia seen in the IDM at birth, the elevated insulin levels also cause increased growth in insulin-sensitive tissues such as the heart, liver, and muscle leading to fetal macrosomia. The cardiomyopathies associated with IDMs, which include thickening of the intraventricular septum and ventricular walls, are believed to be related to the direct effect of fetal insulin on cardiac muscle growth. Fetal hyperinsulinemia is also linked to decreased surfactant production, with respiratory distress syndrome (RDS) more common in IDMs. Well-controlled maternal diabetes in pregnancy is associated with less severe hypoglycemia and a decreased risk of RDS in the IDM, but no effect has been reported on macrosomia. Polycythemia and hypocalcemia are associated with IDMs, but the mechanisms underlying these findings are not fully known.

Preconception counseling and optimization of metabolic control is essential for women with type 1 diabetes who are considering pregnancy. Well-managed maternal diabetes before conception has been demonstrated to decrease the frequency of congenital anomalies associated with the IDM including congenital heart disease (transposition of the great vessels, ventricular septal defect), caudal regression syndrome, and neural tube defects (anencephaly, spina bifida). In spite of preconceptual management, up to 5% of IDMs may have small left colon syndrome.

The IDM in the vignette is at risk for hypoglycemia. Because the infant is asymptomatic, he should be allowed to breastfeed within 1 hour of birth with a screening glucose value obtained 30 minutes after feeding. The infant does not require formula supplementation at this time, with further glucose management tailored to meet the needs of the infant while supporting the mother-infant dyad and breastfeeding. A low rapid bedside

screening glucose value should be confirmed in the laboratory by determining the serum glucose concentration, but treatment should not be delayed while awaiting the results. Although IDMs are at an increased risk for respiratory problems at birth, the infant in the vignette is not demonstrating any respiratory symptoms and does not require chest radiography immediately. If the murmur persists or respiratory distress develops, a chest radiograph and/or echocardiogram can be obtained to screen for cardiomyopathy.



Reprinted with permission from American Academy of Pediatrics Committee on Fetus and Newborn. Postnatal glucose homeostasis in late-preterm and term infants. *Pediatrics*. 2011;127:575-579

ITEM C46: Screening for and management of postnatal glucose homeostasis in late preterm (LPT 34–36 weeks) and term small-for-gestational age (SGA) infants and infants who were born to mothers with diabetes (IDM)/large-for-gestational age (LGA) infants.

PREP Pearls

- An infant of a diabetic mother who has no symptoms of hypoglycemia should be fed, preferably by breast, within the first hour after birth and undergo a screening glucose measurement 30 minutes after the feeding.

American Board of Pediatrics Content Specification (s):

- Understand the management of a newborn whose mother has type 1 diabetes

Suggested Reading:

- American Academy of Pediatrics Committee on Fetus and Newborn. Postnatal glucose homeostasis in late-preterm and term infants. Pediatrics. 2011;127:575-579. doi: 10.1542/peds.2010-3851
- Carlo W. Infants of diabetic mothers. In: Kleigman RM, Stanton BF St Genre JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:627-629

- Ogata ES. Problems of the infant of the diabetic mother. NeoReviews. 2010;11:e627-e631. doi: 10.1542/neo.11-11-e627
- Srinivason G, Pildes RS, Cattamanchi G, Voora S, Lilien LD. Plasma glucose values in normal neonates: a new look. I Pediatr. 1986;109:114-117

Item 47

A medical student is working with you in clinic. He is preparing to see a 2-month-old infant whose older sister has sickle cell disease. The infant's medical record has the results of the state newborn screen, which shows the hemoglobin fractionation to be an F, A, S pattern.

Of the following, the MOST appropriate counseling for the family of this patient is that the infant

A. has a less severe phenotype of sickle cell disease
B. has the most common form of sickle cell disease
C. has sickle cell trait
D. must be referred to a hematologist immediately
E. must be retested at 4 to 6 months of age for a definitive diagnosis

Item 47 S SBP I-C Preferred Response: C

The newborn described in this vignette has a hemoglobin fractionation pattern consistent with sickle cell trait. The result of the hemoglobin fractionation on newborn screen reflects the relative expression of hemoglobin types present from greatest to least. Therefore, an FAS pattern indicates that the hemoglobin with the highest expression is F hemoglobin, followed by A hemoglobin, and then S hemoglobin. All newborns, even those with hemoglobinopathies, will have a predominance of F hemoglobin at birth. A newborn with sickle cell disease would have S hemoglobin as the next most abundant hemoglobin type and therefore would have a pattern such as FS, FSA, or FSC. An infant with the most common type of sickle cell anemia (ie, SS disease) would have an FS pattern on a newborn screen, indicating that there is no production of hemoglobin A. The types of sickle cell disease associated with a milder phenotype are SC disease (newborn screen pattern FSC) and S β^+-thalassemia (newborn screen pattern FSA), in which patients may have mild or no anemia. The families, and eventually the patient, should be counseled on the autosomal recessive inheritance pattern of sickle cell disease and the possibility of double heterozygous states that can also lead to disease (eg, SC disease, S β-thalassemia). The family should be offered genetic counseling if considering future pregnancies.

All US states, territories, and the District of Columbia test for sickle cell disease in their newborn screening programs. This requirement is largely due to the high prevalence of sickle cell disease (ie, 1 in 375 African American newborns) and the benefit of early prophylactic penicillin for infants with sickle cell disease in decreasing mortality. The US Preventive Services Task Force recommends that infants found to have sickle cell anemia receive prophylactic penicillin (125 mg by mouth twice daily) by 2 months of age and receive pneumococcal vaccines at recommended intervals. Most states use either thin-layer isoelectric focusing or high-performance liquid chromatography as the initial screening test, both of which have extremely high sensitivity and specificity for sickle cell anemia. Repeat testing at 6 months of age is not necessary in a patient whose newborn screen pattern is consistent with sickle cell trait. Patients with sickle cell trait are generally asymptomatic, with normal hematologic values and a normal life span; therefore, immediate referral to a hematologist is not indicated. There is emerging literature that patients with sickle cell trait may be more susceptible than those without sickle cell trait to rhabdomyolysis after extreme physical exertion, hyphema and glaucoma after eye trauma, renal dysfunction, and thrombosis; however, these associations are not yet definitive. This controversy has led to discrimination and prevention of participation among athletes and military personnel who have sickle cell trait. For these reasons, referral to a hematologist may be warranted at the family's request but is not mandatory.

PREP Pearls

- Infants diagnosed as having sickle cell disease should be given prophylactic penicillin by 2 months of age to decrease mortality.
- Infants diagnosed as having sickle cell disease (FS, FSA, FSC) on newborn screening should be referred to a pediatric hematologist.
- Sickle cell trait may be associated with eye, renal, and thrombotic complications, although causality has not been determined.
- Sickle cell trait can be diagnosed on newborn screen with high sensitivity and specificity.

American Board of Pediatrics Content Specification(s):

- Understand that sickle cell disease can be diagnosed at birth

Suggested Reading:

- Screening for Sickle Cell Disease in Newborns: U.S. Preventive Services Task Force Recommendation Statement, 2007
- Kavanaugh PL, Sprint PG, Vinci SR, Bauchner H, Wang Q. Management of children with sickle cell disease: a comprehensive review of the literature. Pediatrics. 2011;128:e1552-el574. doi:10.1542/peds.2010-3686
- Key NS, Derebail VK. Sickle-cell trait: novel clinical significance. Hematology Am Soc Hematol Educ Program. 2010;2010:418-422. doi:10.1 182/asheducation-2010.1.418
- Section on Hematology/Oncology and Committee on Genetics. Health supervision for children with sickle cell disease. Pediatrics. 2002;109:526535. doi:10.1542/peds.2010-3686

Item 48

You are evaluating a 4-year-old boy who complains of "bouncing eyes:' His mother reports seeing his eyes jiggle for a few seconds at a time. There are no abnormal head, neck, trunk, or limb movements. He is awake during these episodes and complains that his eyes are "bouncing. " On physical examination, the pupils are round and equally reactive to light, and eye movements are conjugate and intact in all directions. On downward gaze, there is downward nystagmus. The remainder of his physical examination findings, including hair and skin, are unremarkable.

Of the following, the MOST likely diagnosis is

A. Chiari I malformation
B. congenital nystagmus
C. neurofibromatosis type 1
D. phenytoin ingestion
E. spasmus nutans

Item 48 TE Preferred Response: A

The boy described in the vignette has downbeat nystagmus, a subtype of vertical nystagmus. Vertical nystagmus is never normal and is usually the result of a brainstem abnormality. (Horizontal nystagmus can sometimes be normal, but a clinical evaluation is necessary, if it is a new finding.)

In this case the most likely cause is a Chiari I malformation. Chiari I malformation is defined as the descent of the cerebellar tonsils at least 5 mm below the foramen magnum. Although Chiari I malformation is often an incidental finding, it can cause neurologic problems. Symptomatic Chiari I malformation can cause brainstem compression, which presents with dysphagia, dysarthria, upbeat or downbeat nystagmus, or limb weakness with hyperreflexia. Headache with Valsalva. maneuver (coughing, straining, or laughing, for example) can also be a symptom. Chiari I malformation can also be associated with spinal cord syrinx. In a clinically unstable patient, computed tomography of the head is the best test to evaluate for structural brain abnormality; in stable patients for whom magnetic resonance imaging is safe, this modality will yield greater information. The treatment for symptomatic Chiari I malformation is surgical decompression.

Congenital nystagmus is present at birth and persists throughout life. The nystagmus is most often in the horizontal direction but can be vertical. No oscillopsia—the subjective sensation of objects moving in the visual field—is reported. Congenital nystagmus is often an isolated, benign finding, but can be associated with rare neurogenetic disorders such as Pelizaeus-Merzbacher disease or brain malformations.
Neurofibromatosis type 1 is associated with optic nerve gliomas and asymptomatic brain lesions, termed focal areas of signal abnormality, but neither of these cause downbeat nystagmus.

Phenytoin can cause horizontal nystagmus especially when given in high or loading doses. It does not cause vertical nystagmus.
Spasmus nutans, an idiopathic condition of infancy, is characterized by horizontal nystagmus, head tilting, and head nodding. It does not cause downbeat nystagmus. If the clinical diagnosis is uncertain, head and neck imaging should be performed to evaluate for structural abnormalities of the eye, brain, or neck.

PREP Pearls

- Vertical nystagmus can be caused by Chiari I malformation and is never normal; evaluation for brainstem abnormality is required.
- In a clinically unstable patient, computed tomography of the head is the best test to evaluate for structural brain abnormality; in stable patients for whom magnetic resonance imaging is safe, this modality will yield greater information.

American Board of Pediatrics Content Specification(s):

- Recognize that nystagmus may signify important eye or central nervous system pathology

Suggested Reading:

- Lin JH, Arora B, Sethuraman U, Roy-Bornstein C. Index of suspicion. Pediatr Rev 2010;31(6):257-261. doi 10.1542/pir.31-6-257
- Olitskey SE, Hug d, Plummer LS, Stass-Iern M. Disorders of eye movement and alignment. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:2057-2162

Item 49

A 14-year-old girl presents to your office for a preparticipation physical evaluation for the school soccer team. During a recent ophthalmologic examination, she was noted to have a corrected visual acuity of 20/20 in the right eye and 20/50 in the left eye.

Of the following, the MOST appropriate recommendation for this patient would be that she

A. does not need any special eye protection when playing soccer
B. should not participate in soccer due to her poor visual acuity on the left side
C. should wear hard contact lenses to protect her eyes during soccer
D. should wear her regular glasses secured to the head with a strap while playing soccer
E. should wear sports goggles with polycarbonate lenses when playing soccer

Item 49 S Preferred Response: E

Eye injuries that occur as a result of participation in sports and recreational activities represent 25% of all ocular trauma seen in United States emergency departments. In a joint policy statement, the American Academy of Pediatrics (AAP) and the American Academy of Ophthalmology (AAO) "strongly recommend" appropriate protective eye-wear for athletes who participate in sports with a risk of eye injury. For participants in high-risk and some moderate-risk sports, the AAP and AAO recommend use of an appropriate face mask or sports safety goggles with polycarbonate lenses that meet the standards of the American Society of Testing and Materials. Polycarbonate is more shatter-resistant than CR-39, the plastic polymer typically used for eyeglasses. High-risk sports include those that involve small, fast projectiles, such as paintball, and sports with hard projectiles or sticks, such as hockey and baseball. Boxing and full-contact martial arts are also considered high risk. The AAP and AAO make specific recommendations about sports eyewear for the following sports in the moderate-risk category: racquet sports, soccer, water polo, and football. For children requiring vision correction when playing low-risk sports, eyeglasses with CR-39 lenses, used with a strap to fasten them to the head, afford adequate eye protection. Soccer is considered a moderate-risk sport for eye injuries. However, the girl in the vignette is a functionally 1 -eyed athlete because she has a corrected vision of less than 20/40 in 1 eye. Functionally 1-eyed athletes should use polycarbonate protective eyewear for all sports activities and should not participate in any sports with a risk of eye injury that do not allow the use of protective eyewear, such as full-contact martial arts. Contact lenses do not protect the eyes from trauma. Athletes who wear contact lenses and participate in sports with a risk of eye injury should wear appropriate eye protection in addition to their contact lenses.

PREP Pearls

- Athletes who participate in moderate- or high-risk sports should use appropriate AAP–AAO recommended protective eyewear to reduce their risk for eye injuries.
- Functionally 1 -eyed athletes require eye protection for all sports and should not participate in full-contact sports that do not allow the use of protective eyewear.

American Board of Pediatrics Content Specification (s):

- Know the indications for the use of goggles for eye protection in sports activities

Suggested Reading:

- American Academy of Pediatrics Committee on Sports Medicine and Fitness. Protective eyewear for young athletes. Pediatrics. 2004;113(3 pt 1):619-622
- Bernhardt DT, Roberts WO, eds. Preparticipation Physical Evaluation. 4th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2010
- Pollard KA, Xiang H, Smith GA. Pediatric eye injuries treated in US emergency departments, 1990-2009. Clin Pediatr (Phila). 2012;51(4):374381. doi:10.1177/0009922811427583

Item 50

A full-term newborn was delivered after an uneventful pregnancy to a gravida 2, para 2 woman by normal spontaneous vaginal delivery. His birth weight is 3,150 g. Findings on physical examination are unremarkable except for bilateral ear pits and a small branchial sinus on the left neck with no drainage noted.

Of the following, prior to discharge, you are MOST likely to order a(n)

A. complete blood cell count
B. complete metabolic profile
C. echocardiogram
D. head ultrasonography
E. renal ultrasonography

Item 50 Preferred Response: E

The infant described in this vignette has clinical features suggestive of branchio-oto-renal (BOR) syndrome. This autosomal dominant disorder caused by mutations in EYA1 is associated with preauricular pits (70%-80%), branchial cysts or fistulas (30%-60%), and structural renal anomalies (12%-20%). Therefore, renal ultrasonography is recommended in light of the other 2 clinical findings. In addition to these features, individuals with BOR syndrome may have other external ear malformations, and at least 75% have some degree of hearing loss.

Other syndromes typically associated with external ear malformations include CHARGE syndrome (Coloboma of the eye, Heart defects, Atresia of the choanae, Retardation of growth and development, Genital and Renal anomalies, Ear abnormalities), Townes-Brocks syndrome, oculo-auriculo-vertebral spectrum (Goldenhar syndrome), Treacher-Collins syndrome, Nager syndrome, Miller syndrome, and diabetic embryopathy (Item C50, page C-40). A large survey of patients with external ear anomalies, including preauricular pits, demonstrated that almost 30% had concomitant renal anomalies and 92% represented a multiple congenital anomaly syndrome. For individuals with apparent isolated ear anomalies, approximately 11% will be found to have a hidden renal malformation. Therefore, renal ultrasonography must be performed in infants with ear anomalies (including tags, pits, lop ear, cupped ear, microtia or anotia) in addition to any of the following findings: dysmorphic facies, facial asymmetry, ocular or eyelid colobomas, choanal atresia, micrognathia (small jaw), branchial cysts or sinuses, cardiac murmur, limb anomalies, or an imperforate or anteriorly placed anus. Renal ultrasonography should also be performed on an infant with external ear malformations if there is a family history of hearing loss and ear or renal malformations, or if there is a history of maternal diabetes during gestation.

A complete blood cell count or head ultrasonography would not be indicated under these circumstances. A complete metabolic profile might uncover renal dysfunction, but most renal malformations identified in patients with BOR syndrome would not present with renal failure. An echocardiogram would only be indicated if the infant had a cardiac murmur, cyanosis, low oxygen saturation, or other signs of a cardiac defect.

Item C50. Syndromes Associated with External Ear Malformations

Condition or Syndrome	Ear Findings	Other Clinical FIndings	Mode of Inheritance
Branchio-oto-renal	Abnormal pinna, preauricular pits, auditory canal stenosis, mixed hearing loss	Branchial fistulas or cysts, lacrimal duct stenosis or aplasia, renal malformations	Autosomal dominant *EYA1* (50%) Variable expression
CHARGE	*E*, abnormal pinna (often lop ear), mixed variable hearing loss	*C*, colobomas; *H*, heart defects; *A*, atresia choanae; *R*, retardation of growth and development; *G*, genital and renal anomalies	Autosomal dominant *CDH7* de novo mutations
Townes-Brocks	Abnormal pinna, pre-auricular tags, variable sensorineural hearing loss	Abnormal thumbs, polydactyly, renal anomalies, imperforate anus, bony fusions	Autosomal dominant *SALL1* Variable expression
Oculo-auriculo-vertebral spectrum	Microtia, preauricular tags and pits, variable hearing loss	Hemifacial microsomia; epibulbar dermoids; vertebral anomalies; heart, genitourinary, cleft defects	Sporadic Likely vascular or field defect in utero
Treacher-Collins	Microtia/malformed auricles and external ear canals, conductive hearing loss	Malar hypoplasia, lower eyelid colobomas, micrognathia, cleft palate	Autosomal dominant *TCOF1*
Nager	Preauricular tags, atresia of the external ear canals, conductive hearing loss	Micrognathia, cleft palate, absent thumbs, radioulnar synostosis, downslanting palpebrae	Mostly sporadic Some autosomal dominant and recessive families reported
Miller	Hypoplastic or cup- shaped ears, occasional conductive hearing loss	Malar hypoplasia, lower eyelid colobomas, absent fifth digits, ulnar and radial hypoplasia	Autosomal recessive
Diabetic embryopathy	Abnormal pinna including microtia in most severe cases	Variable anomalies of the heart, brain, neural tube, kidneys, limbs along with macrosomia	Teratogenic

PREP Pearls

- Infants with external ear malformations have an 11 % risk for also having a structural renal malformation and should be evaluated for other birth defects.
- Renal ultrasonography should be performed if an infant has external ear malformations plus at least 1 other significant clinical finding.

American Board of Pediatrics Content Specification (s):

- Recognize that malformed external and middle ears maybe associated with renal anomalies, craniofacial malformations, and inner ear malformations

Suggested Reading:

- Khoury MJ, Becerra JE, Cordero IF, Erickson JD. Clinical-epidemiological assessment of patterns of birth defects associated with human teratogens: application to diabetic embryopathy. Pediatrics. 1989;84(4):658-665
- Roth DAE, Hildesheimer M, Bardenstein S, et al. Preauricular skin tags and ear pits are associated with permanent hearing impairment in newborns. Pediatrics. 2008;122(4):e884-e890. doi:10.1542/peds.2008-0606
- Wang RY, Earl DL, Ruder RO, Graham JM. Syndromic ear anomalies and renal ultrasounds. Pediatrics. 2001;108(2):e32. doi:10.1542/peds.108.2.e32

Item 51

A 16-year-old boy is brought to the emergency department following an all-terrain vehicle (ATV) collision. He was driving the vehicle at a high rate of speed when he lost control and crashed into a tree. He was not wearing a helmet and sustained significant trauma to his forehead and face. The boy experienced an initial loss of consciousness at the time of the accident, but he is now awake and has a Glasgow Coma Score of 14. His airway, breathing, and circulation are fully intact. Computed tomography of his head and facial bones reveals fractures involving both the anterior and posterior tables of his frontal sinus.

Of the following, the BEST next step in management is

A. antibiotic therapy
B. decongestants
C. irrigation of the nasal passages with sterile saline
D. observation in the hospital
E. surgical consultation

Item 51 Preferred Response: E

The teenage patient described in the vignette has fractures involving the anterior and posterior tables of the frontal sinus after sustaining a direct, forceful impact to his head and face in an all-terrain vehicle crash. Prompt neurosurgical consultation is indicated to reduce the risk of complications. Although minor injuries to the face and head in pediatric patients are common, the incidence of facial fractures in children is much lower than in the adult population. Children account for approximately 5% to 15% of maxillofacial trauma overall. The incidence of pediatric facial fractures is lowest in children younger than 5 years and peaks during adolescence with increased participation in unsupervised sports and activities, including the operation of motorized vehicles. Nasal bone fractures are the most common type of facial fractures sustained by children, followed by mandibular fractures. Frontal sinus fractures in particular are quite rare in children. Trauma forceful enough to result in frontal sinus fractures generally involves a high-velocity collision between the individual's head and face with a moving object, such as a ball or baseball bat, or impact of the face against the windshield or dashboard of an automobile during a motor vehicle collision. Frontal sinus fractures may involve both he anterior and posterior tables of the sinus. These fractures re typically described as displaced or nondisplaced. In cases of significant facial trauma, physical examination generally reveals ecchymosis, swelling, and pain at the site of injuries. Palpation of the facial bones may reveal step-offs and crepitus when displacement of fracture segments has occurred. Cranial nerve integrity, especially sensation and movement of the face, must be evaluated carefully in facial trauma patients because branches of the facial nerve may be injured in midface and frontal fractures. Associated intracranial and ophthalmologic injuries are also common. In one case series of 120 children with frontal sinus fractures, more than 60% of the patients included were found to have an associated intracranial injury. In the same case series, all included children had concomitant orbital fractures. Although plain radiographs may reveal facial bone fractures, computed tomography of the facial bones is the recommended imaging modality for evaluation of children with suspected facial fractures. Computed tomography is not only more sensitive in detecting fractures but also useful in revealing the degree of fracture segment displacement and associated soft tissue injuries. Computed tomography of the head is also indicated in most patients with significant facial trauma to evaluate for concomitant intracranial injury.

Proper evaluation, management, and follow-up of frontal sinus injuries in children are essential to prevent the occurrence of significant short-term and long-term complications. In addition to the high incidence of associated intracranial injury, children with frontal sinus fractures are at risk for cerebrospinal fluid (CSF) leaks, cosmetic deformities, and infectious sequelae, including the development of sinusitis, meningitis, brain abscesses, and orbital abscesses. Although linear, nondisplaced fractures that involve only the anterior wall of the frontal sinus may be treated with observation rather than immediate surgical management, injuries that involve the posterior wall of the frontal sinus, as seen in the patient in the vignette, require prompt neurosurgical consultation. Consultation with a maxillofacial specialist is also prudent given the high incidence of associated facial injuries.

Although antibiotics may play a role in preventing infectious complications that arise from frontal sinus fractures, they do not address the underlying injury that may lead to these complications if repair is delayed. There is no evidence to support the role of decongestants or saline irrigation of the sinuses in patients with frontal sinus fractures. Observation alone in patients with a displaced anterior table frontal sinus fracture, as well as in patients with a fracture of the posterior table, increases the risk of cosmetic deformity, functional complications (CSF leak), and development of central nervous system infections.

PREP Pearls

- Intracranial injury should be strongly considered in children with frontal sinus fractures.
- Computed tomography of the facial bones is the recommended imaging modality for children with suspected facial fractures.
- Injuries that involve the posterior wall of the frontal sinus require prompt neurosurgical consultation.
- In addition to intracranial injury, children with frontal sinus fractures are at risk for CSF leaks, cosmetic deformities, sinusitis, meningitis, brain abscesses, and orbital abscesses.

American Board of Pediatrics Content Specification (s):

- Know that trauma involving fracture of the frontal sinus requires surgical repair and that delay may lead to later CNS infection

Suggested Reading:

- York J, Colucciello SA. Maxillofacial trauma. In: Wiebe RA, Ahrens WR, Strange GR, Schafermeyer RW, eds. Pediatric Emergency Medicine. 3rd ed. New York, NY: McGraw-Hill; 2009:289-296
- Ilgen JS. Facial injuries. In: Cline DM, Ma 0J, Cydulka RK, Meckler GD, Handel DA, Thomas SH, eds. Tintinalli's Emergency Medicine Manual. 7th ed. New York, NY: McGraw-Hill; 2012
- Ginsberg CM. Frontal sinus fractures. Pediatr Rev. 1997;18:120-121. doi:I 0. 1 542/pir.18-4-120

Item 52

A 6-year-old boy is having problems in school. As part of the evaluation for special education services, the school performed a full individual evaluation. On a standardized achievement test, his IQ score is 60. His birth history, past medical history, and physical examination are unremarkable. Hearing and vision screen are normal. Parents report the patient has some early language developmental delay, but they deny any regression of milestones. There is a family history of some adults with learning difficulties.

Of the following tests, the BEST next step is to perform

A. electroencephalogram
B. genetic testing
C. magnetic resonance imaging of the brain
D. serum amino acids
E. urine for cytomegalovirus

Item 52 Preferred Response: B

Many standardized tests, such as the IQ, are based on a mean population score of 100, and 1 standard deviation (SD) equals 15 points. A score of less than 70 points (>2 SDs below the mean) represents intellectual disability (ID). ID is synonymous with and preferred over the older term mental retardation. As defined by the American Association of Intellectual and Developmental Disabilities and the Diagnostic and Statistical Manual of Mental Disorders, 4th edition, text revision, ID is a general term for a lifelong condition characterized by significant impairment of cognitive and adaptive development with onset before 18 years of age. ID is defined as mild when the IQ decreases 2 to 3 SDs below the mean (range, 55-69), moderate when the IQ is 40 to 54, severe when the IQ is 25 to 39, and profound when the IQ is less than 25.

The prevalence of ID in the general population is estimated to be 1% to 3%. ID is found more commonly in males than females (1.4:1.0). The age at identification varies, depending on the degree of severity. It is common that children with mild ID do not get identified until school age, as in the boy in the vignette. Early language skills, especially receptive language, are a good predictor of intelligence. Some children will have associated medical conditions or characteristic features of a genetic syndrome, but many have normal histories and physical examinations.

Attempts should be made to identify the cause of the ID for several reasons: (1) the condition may be treatable, (2) screening for associated health risks may lessen morbidity and mortality, (3) there may be genetic implications for the family, and (4) planning for the future and accessing social supports may be enhanced. The diagnostic evaluation of each case must be individualized and should follow a staged approach. A thorough history, including prenatal, birth, and family history, and a comprehensive physical examination remain the most important first steps in identifying a cause for ID. When an abnormality is found, further investigations should be directed toward this potential cause. When the history and physical examination do not suggest the cause, further evaluation should be offered to the family in a thoughtful stepwise manner. Normal physical examination findings do not rule out an underlying genetic cause. One may choose to begin genetic testing in this situation by obtaining a karyotype and molecular analysis for fragile X syndrome. If these test results are normal, additional genetic evaluation, such as chromosomal microarray or whole exome sequencing, may be warranted. If a certain specific syndrome, such as Williams syndrome, is suspected, fluorescence in situ hybridization assessment may be the most cost-effective approach. Sometimes testing of parents or an affected family member is needed to better define the disorder. The approach to genetic testing is evolving, so it is important to be familiar with the capabilities of the laboratory used. In addition, additional genetic testing should be pursued as newer technologies emerge.

Routine electroencephalography is not recommended but would be indicated in cases in which there is a concern for seizures. There is disagreement about the value of routine neuroimaging in children with ID and normal history and physical examination findings. Rarely are the findings informative or causative, so magnetic resonance imaging should be reserved for those patients with specific findings, such as neurocutaneous lesions,

abnormal hair patterns, midline facial defects, microcephaly or macrocephaly, abnormal neurologic examination findings, seizures, or significant birth history. Although ID is a feature of some inborn errors of metabolism, most affected children have other clinical manifestations and developmental regression. In addition, newborn screening programs in the United States today would identify most conditions. Urinalysis for cytomegalovirus would not be helpful in identifying causality of ID in a 6-year-old.

PREP Pearls

- In patients with intellectual disability, genetic testing may be used when there is not an identified cause.
- Many standardized tests, such as the IQ, are based on a mean population score of 100, and 1 SD equals 15 points.
- Intellectual disability is synonymous with and preferred over the older term mental retardation.
- Intellectual disability is defined as mild when the IQ decreases 2 to 3 SDs below the mean (range, 55-69), moderate when the IQ is 40 to 54, severe when the IQ is 25 to 39, and profound when the IQ is less than 25.

American Board of Pediatrics Content Specification (s):

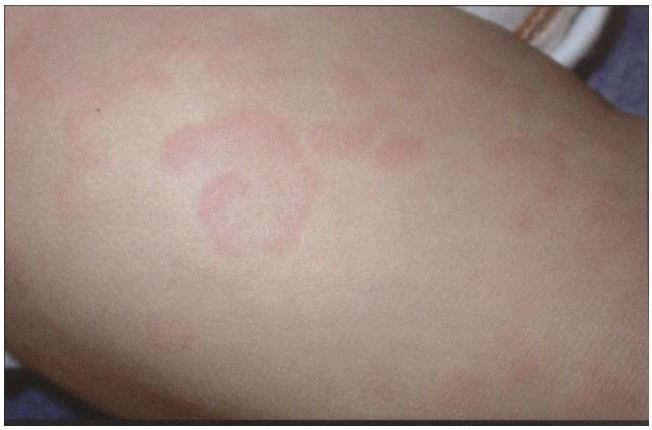
- Understand how to interpret the scores on standardized achievement tests

Suggested Reading:

- American Association of Intellectual and Developmental Disabilities. www.aaidd.org.
- Braaten EB, Norman D. Intelligence (IQ) testing. Pediatr Rev. 2006;27:403408. doi:10.1542/pir.27-11-403
- Pivalizza P. Intellectual disability (mental retardation) in children: definition; causes; and diagnosis. UptoDate. 2012. Available online only for subscription
- Pivalizza P. Intellectual disability (mental retardation) in children: evaluation. UptoDate. 2012. Available online only for subscription
- Shea S. Intellectual disability (mental retardation). Pediatr Rev. 2012;33:110-121doi:10. 1542/pir.33-3-1 10

Item 53

A 9-year-old girl presents with symptoms of an itchy, raised rash. The rash started 2 weeks ago, but she has had similar episodes intermittently for the previous 4 months. The rash consists of multiple erythematous, slightly elevated lesions ranging in size from a dime to a golf ball (Item Q53).



The lesions resolve in a few hours without bruising or discoloration. Diphenhydramine as needed and daily cetirizine help minimize the frequency and severity of her symptoms, but she still gets breakthrough episodes. Her parents have not found any association of the eruption with foods, cosmetics, or medications. At times, the rash occurs after the girl becomes hot and sweaty. The parents have heard that food allergies might be a trigger for their daughter's episodes and wonder if she should undergo testing.

Of the following, the MOST appropriate response is to

A. order allergen- specific IgE tests to milk, egg, soy, wheat, fish, shellfish, peanuts, nuts, and food additives
B. order allergen- specific IgG4 tests to milk, egg, soy, wheat, fish, shellfish, peanuts, nuts, and food additives
C. reassure the parents that allergy testing is not needed; an undiscovered allergy to foods or food additives is unlikely to be the cause
D. recommend an elimination diet of home-cooked rice or oats, chicken or turkey, and vegetables for a trial period of 2 weeks
E. refer them to an allergist for skin testing to milk, egg, soy, wheat, fish, shellfish, peanuts, nuts, and food additives

Item 53 Preferred Response: C

The child described in the vignette is unlikely to have a new-onset allergy to food/food additives as the cause of the chronic urticaria (CU). Chronic urticaria is defined by the presence of urticaria (hives) on most days of the week for a period of 6 weeks or longer. While acute urticaria and angioedema may be manifestations of IgE-mediated allergic reactions, no external cause can be identified in 80% to 90% of people affected by CU. While milk, egg, soy, wheat, fish, shellfish, peanuts, and nuts are the most common triggers of food-allergic reactions, IgE-mediated reactions to foods and food additives are not a cause of CU and allergy testing is not warranted. Nonstandardized tests such as allergen-specific IgG4 are not recommended for the routine evaluation of IgE mediated food allergic disorders and do not have a role in the evaluation of chronic urticaria.

Patients with CU may perceive food-associated reactions; but their perceptions have not been validated in studies using placebo-controlled challenges. They may also often report that rich meals, fermented foods, and alcohol worsen the condition transiently. This may be related to the histamine content or innate histamine-releasing properties of these foods, as well as the vasodilatory effects of alcohol and certain spices. Food additives are rarely, if ever, confirmed to cause or contribute to flares of CU in carefully performed studies. While temporary avoidance of the offending foods discussed above may be considered, elimination diets have not been shown to help and are not recommended.

Nevertheless, every effort should be made to determine the etiology of these symptoms. A detailed history should be taken and repeated periodically if the CU persists. The history should be geared toward (1) differentiating between vasculitic (duration of lesions >72 hours, pain, discoloration, or residual scarring) and nonvasculitic urticaria (duration <72 hours and absence of above signs), (2) identifying possible underlying cause, and (3) ensuring that the patient does not have evidence of a more serious systemic disease (fever, weight loss, arthralgias, and other constitutional symptoms). It is helpful to approach patients on the basis of broad categories of mechanisms such as IgE-dependent mechanisms (eg, drug, food, insect venom, and latex exposure) and complement-mediated mechanisms (eg, hereditary angioedema and serum sickness). Other factors to consider include (1) physical urticarias; (2) underlying infection; (3) autoimmune etiology; (4) possible hormonal effects, especially when hives in women occur on a cyclic basis; (5) association with malignant tumor; (6) pertinent occupational exposure; (7) multiple/repetitive or late onset reactions to insect stings/bites; (8) direct contact of skin or oropharynx with foods, chemicals, animal saliva, and other substances; (9) familial pattern/ hereditary syndromes; and (10) psychologic stresses.

Initial screening tests for disorders most commonly associated with urticaria include: a complete blood count with differential, markers of inflammation (C-reactive protein or erythrocyte sedimentation rate), liver function tests, urine analysis, and a thyroid-stimulating hormone level. Further testing should be based upon the results of these tests or as dictated by the history, physical examination, and review of systems. Biopsy of a fresh lesion may be indicated if urticaria) vasculitis is suspected. Skin or serum-specific IgE allergy testing is typically not indicated unless a specific trigger is suspected on the basis of history. At this stage of the evaluation it is reasonable to define chronic urticaria

angioedema as idiopathic because this is a diagnosis by exclusion of underlying etiologies. If treatment is ineffective at this point, referral to an allergist/ immunologist or dermatologist might be considered.

PREP Pearls

- Chronic urticaria is defined by the presence of hives on most days of the week for a period of 6 weeks or longer.
- Typically, unless a specific trigger is suspected, chronic urticaria does not warrant allergy testing.

American Board of Pediatrics Content Specification (s):

- Recognize that chronic urticaria does not warrant allergy testing

Suggested Reading:

- Joint Task Force on Practice Parameters. The diagnosis and management of urticaria: a practice parameter, part II: chronic urticaria/angioedema. Ann Allergy Asthma Immunol. 2000; 85(6 pt 2):S521-S544
- Khan DA. Chronic urticaria: diagnosis and management. Allergy Asthma Proc. 2008;29(5):439-446. doi:10.2500/aap.2008.29.3151
- NIAID-Sponsored Expert Panel; Boyce JA, Assa'ad A, Burks AW, et
- al. Guidelines for the diagnosis and management of food allergy in the United States: report of the NIAID-Sponsored Expert Panel. I Allergy Clin Immunol. 2010;126(6 suppt):SI-S58. doi:10.1016/j.jaci.2010.10.007

Item 54

A 7-year-old boy has significant daytime and nocturnal enuresis since birth. Physical examination reveals a temperature of 37.8°C, heart rate of 76 beats/min, respiratory rate of 16 breaths /min, blood pressure of 98/50 mm Hg, and normal growth parameters. He has a tuft of hair above the gluteal cleft. His urinalysis demonstrates a specific gravity of 1.005, pH of 6.0, and no blood, leukocyte esterase, protein, or nitrites.

Of the following, the MOST likely cause of enuresis in this patient is

A. bladder dysfunction
B. central diabetes insipidus
C. chronic renal failure
D. nephrogenic diabetes insipidus
E. urinary tract infection

Item 54 Preferred Response: A

Enuresis is diagnosed in children aged 5 years or older who void in bed or on clothes twice or more per week for 3 consecutive months. Primary enuresis occurs in children with no period of sustained dryness. Secondary enuresis is identified in children with a period of sustained dryness for 6 months (for nocturnal enuresis) or 3 months (for diurnal enuresis). The 7-year-old boy in the vignette has an abnormal voiding pattern (primary daytime and nocturnal enuresis), which needs further evaluation.

A detailed neurologic examination including examination of the spine is vital in any patient presenting with abnormal voiding patterns. Skin abnormalities of the spine such as tuft of hair, vascular lesions (hemangioma), or discoloration of the skin overlying the spine are suggestive of an underlying vertebral or spinal lesion. In the boy in the vignette, the tuft of hair on the spine in the presence of enuresis is highly suspicious of an underlying neural (spinal) cause for the bladder dysfunction. Spinal cord lesions (even very low sacral lesions associated with normal lower extremity function) are associated with bladder dysfunction because bladder control is below the level for lower extremity function in the spinal cord. Therefore all patients with spina bifida need a detailed and regular evaluation of the bladder capacity and pressures. Bladder function is evaluated with renal bladder ultrasonography, voiding cystourethrogram, and urodynamic studies.

Bladder management in patients with spinal disorders is aimed at lowering bladder pressures and episodes of upper urinary tract infection (UTI) to decrease renal injury and thereby prevent or slow down the progression of chronic renal injury. Therefore, it is important to identify bladder dysfunction (presenting with enuresis) at an earlier age; if unrecognized it may progress to chronic renal failure. Also, it is important to note that enuresis in a patient with spinal injury indicates bladder dysfunction, which if untreated, can lead to chronic renal failure. Magnetic resonance imaging examination of the spine is indicated in this case for evaluation of the spinal lesion.

For the child described in the vignette, UTI is unlikely in the absence of fever or urinary symptoms such as dysuria, flank pain, or burning micturition. Also, the absence of pyuria, nitrites, and bacteria on urinalysis rules out UTI as the underlying cause of primary enuresis in the patient. Normal growth parameters and absence of proteinuria in the boy in this vignette make chronic renal failure less likely.

Patients with diabetes insipidus may have enuresis, but they usually present with polyuria and polydipsia which are absent in the boy in the vignette. Patients with diabetes insipidus may also present more commonly with recurrent episodes of hypernatremic dehydration and failure to thrive.

PREP Pearls

- Enuresis is diagnosed in children aged 5 years or older who void in bed or on clothes twice or more per week for 3 successive months.
- Primary enuresis occurs in children with no period of sustained dryness. Secondary enuresis is identified in children with a period of sustained dryness for 6 months (for nocturnal enuresis) or 3 months (for diurnal enuresis).
- Skin abnormalities of the spine such as tuft of hair, vascular lesions (hemangioma), or discoloration of the skin overlying the spine suggests an underlying vertebral or spinal lesion. Spinal lesions can be associated with bladder dysfunction because bladder control is below the level for lower extremity function in the spinal cord.
- It is important to identify bladder dysfunction (presenting with enuresis) at an earlier age; if unrecognized it may progress to chronic renal failure.

American Board of Pediatrics Content Specification(s):

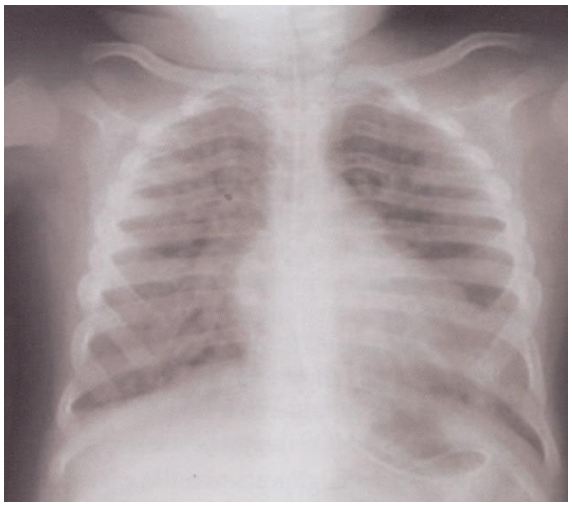
- Know the importance of skin abnormalities in the sacral area when evaluating patients with primary enuresis

Suggested Reading:

- de Jong TP, Chrzan R, Klijn Al, Dik P. Treatment of the neurogenic bladder in spina bifida. Pediatr Nephrol. 2008;23:889-896. doi: 10.1007/ s00467-008-0780-7
- Fritz G, Rockney R, Bernet W, et al; Work Group on Quality Issues, AACAP. Practice parameter for the assessment and treatment of children and adolescents with enuresis. J Am Acad Child Adolesc Psychiatry. 2004;43(12):1540-1550. doi:10.1097/01.chi.0000142196.41215.cc
- Nev6us T, von Gontard A, Hoebeke P, et al. The standardization of terminology of lower urinary tract function in children and adolescents: report from the Standardisation Committee of the International Children's Continence Society. J Urol. 2006;176(1):314-324. doi:10.10161 S0022-5347(06)00305-3

Item 55

A 3-year-old boy diagnosed with pneumonia 3 days ago returns to your office with ongoing fever and cough. Because he received amoxicillin for otitis media 1 month ago, you prescribed an oral third-generation cephalosporin. His mother reports that he has been taking the medication but that he will not eat. He has been drinking some fluids and seems to have normal urine output. He has not been vomiting, and his cough is nonproductive.



On physical examination, his temperature is 39°C, heart rate is 130 beats/min, respiratory rate is 40 breaths/min, and blood pressure is 90/45 mm Hg. Oxygen saturation is 98% in room air. His mucous membranes are mildly dry, but capillary refill is less than 2 seconds. He is mildly tachypneic without retractions or nasal flaring. Auscultation of his lungs reveals good air entry throughout with diffuse crackles. He is tachycardic but has no murmur. The remainder of his physical examination findings are normal. A white blood cell count is 10.0 x 101/µL (10.0 x 10'/L), with 30% polymorphonuclear leukocytes, 55% lymphocytes, and 15% monocytes. You obtain a chest radiograph (Item Q55).

Of the following, the MOST appropriate next step in the management of this patient is to

A. add azithromycin to his regimen
B. admit him for intravenous ceftriaxone
C. discontinue antibiotics
D. obtain a chest computed tomography scan
E. send a blood culture

Item 55 Preferred Response: C

The patient described in the vignette has pneumonia characterized by fever, nonproductive cough, and diffuse interstitial infiltrates without parenchymal involvement on chest radiography. The most likely cause of the patient's pneumonia is a virus, and discontinuing antibiotics and providing supportive care are recommended for management. Azithromycin would be appropriate for treating Mycoplasma or Chlamydia pneumoniae (ie, atypical) pneumonia, but these pathogens are unlikely causes of the patient's illness. Children with atypical pneumonia generally are school-aged or older, are not highly febrile, and have a dry cough that develops later in the illness, as symptoms of headache, myalgias, and malaise improve. Intravenous antibiotics are not helpful for treating viral pneumonia but may be warranted in patients with bacterial (eg, Streptococcus pneumoniae, group A Streptococcus, and Staphylococcus aureus) pneumonia requiring hospitalization because of young age (<3 months), hypoxemia, vomiting and dehydration, underlying serious medical condition, complications (eg, empyema and abscess), or toxic appearance. In patients with presumed bacterial pneumonia requiring hospitalization, a blood culture may be helpful in determining the cause, but blood cultures are not recommended for patients with viral pneumonia or those well enough to be treated in the outpatient setting. Chest computed tomography is generally reserved for patients with suspected suppurative complications of pneumonia in whom surgical intervention is being considered.

Viruses are the most common cause of pneumonia in children 3 months to 4 years of age. Likely pathogens include respiratory syncytial virus, influenza virus, parainfluenza virus, adenovirus, human metapneumovirus, and rhino-virus. The patient described in the vignette was ultimately diagnosed as having pneumonia due to adenovirus infection by a positive result on a rapid antigen assay performed on respiratory tract secretions. There are no distinguishing clinical characteristics in the patient's presentation to suggest adenovirus over other viral origins. Common manifestations of adenovirus infection include the common cold, pharyngitis, tonsillitis, conjunctivitis (sometimes hemorrhagic), and otitis media. Croup, bronchiolitis, pneumonia, a pertussis-like syndrome, hemorrhagic cystitis, and gastroenteritis also occur. Adenovirus sepsis, severe pneumonia, meningitis, and encephalitis are uncommon and more likely to occur in infants and immunocompromised patients.

PREP Pearls

- Viruses are the most common cause of pneumonia in children 3 months to 4 years of age and antibiotics are not warranted.
- Viral pneumonia in children are likely caused by respiratory syncytial virus, influenza virus, parainfluenza virus, adenovirus, human metapneumovirus, or rhinovirus.

American Board of Pediatrics Content Specification(s):

- Recognize the clinical manifestations of adenovirus infection

Suggested Reading:

- American Academy of Pediatrics. Adenovirus infections. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:204-206
- Cherry JD, Chen TK. Adenoviruses. In: Feigin RD, Cherry JD, DemmlerHarrison GJ, Kaplan SL, eds. Feigin and Cherry's Textbook of Pediatric Infectious Diseases. 6th ed. Philadelphia, PA: Elsevier Saunders; 2009:1949-1972
- Durbin WJ, Stille C. Pneumonia. Pediatr Rev. 2008;29:147-158. doi:10.1542/pir.29-5-147

Item 56

You are seeing a 15-year-old boy for a health supervision visit. His parents complain that he has been irritable lately and spends too much time playing his guitar and hanging out with his friends. He seems less interested in family activities and his grades have dropped from A+'s to B's in most subjects. They ask that you screen him for drug use. The boy is upset with this conversation and says his parents don't give him any breathing space.

Of the following, the BEST next step is to

A. advise the parents to restrict the boy's privileges until his grades improve
B. interview the boy alone to discuss his school, peer, and family functioning
C. obtain a psychiatric consultation to evaluate the boy for depression
D. obtain a urine drug screen per the parents' request
E. reassure the parents that this is normal adolescent behavior

Item 56 Preferred Response: B

To gauge whether behavior exhibited by an adolescent is normal, it is useful to understand the 3 stages of development (early, middle, and late adolescence) and the behaviors they exhibit in each of these stages in relation to their families, peer group, and the school or work environment. The developmental tasks of adolescents include separation from one's parents, achieving economic independence, and the development of a stable self and sexual identity. The boy in this vignette is in middle adolescence, and his behavior is fairly typical. However, to gauge his mental health status and make sure there is no other cause for concern, it is important that the physician interview him alone after discussing the limits of confidentiality. This is important to gain the adolescent's trust and help with any needed behavior change. Information need only be shared with parents if there is a history of abuse or homicidal or suicidal ideation. A decision to ground him or obtain a psychiatric consultation or drug test should be deferred until after the interview. Behavior change, if necessary, will require the youth's cooperation, and thus proceeding without his knowledge or consent will not improve the situation.

PREP Pearls

- Family, school, peers, and a work environment are the major spheres of influence in an adolescent's life, and a functional assessment for each of these areas is important.
- Providing a confidential setting is essential to getting a complete history from an adolescent.
- Behavior exhibited needs to be compared with norms for the stage of psychosocial development.

American Board of Pediatrics Content Specification(s):

- Understand that assessment of the functional status of an adolescent peer relationships, school, work, family relationships) is a specific task of the adolescent visit
- Understand the importance of obtaining a confidential sexual and substance use history during a health care visit for an adolescent

Suggested Reading:

- Goldenring JM, Rosen DS. Getting into adolescent heads: an essential update. Contemp Pediatr. January 1, 2004. http://www2.aap.org/pubserv/PSVpreview/pages/Files/HEADSS.pdf
- Hazen E, Schlozman S, Beresin E. Adolescent psychological development: review. Pediatr Rev. 2008;29:161-168. doi:10.1542/pir.29-5-161
- Ozer EM, Adams SH, et al. Increasing the screening and counseling of adolescents for risky health behaviors: a primary care intervention. Pediatrics. 2005;115: 960-968. doi:10.1542/peds.2004-0520
- Williams S13, O'Connor EA, Eder M, Whitlock EP. Screening for child and adolescent depression in primary care settings: a systematic evidence review for the US Preventive Services Task Force. Pediatrics. 2009;123:e7l6-e735. doi:10.1542/peds.2008-2415.
- Zahrt DM, Melzer-Lange MD. Aggressive behavior in children and adolescents. Pediatr Rev. 2011;32:325-332. doi:10.1542/pir.32-8-325

Item 57

An 8-year-old boy is brought to the emergency department because of hematemesis. He has recently been experiencing upper respiratory tract symptoms, including cough and nasal congestion, which have been treated by his parents using an over-the-counter cold preparation. Upon awakening this morning, he ate normally. Several hours later he complained of abdominal pain and vomited several times. The vomitus contained both bright red blood and clots. He was brought to the emergency department, where his temperature is 38°C, heart rate is 130 beats/min, respiratory rate is 18 breaths/min, and blood pressure is 80/40 mm Hg. He appears pale, his liver edge is palpated at the right costal margin, and the spleen tip is palpated 2.5 cm below the left costal margin.

Of the following, the MOST likely cause of this child's bleeding is

A. esophageal varices
B. Helicobacter pylori- associated gastritis
C. Mallory-Weiss tear
D. nasopharyngeal bleeding
E. nonsteroidal anti-inflammatory drug-induced ulcer

Item 57 Preferred Response: A

The child who has hematemesis presents a frightening situation for both parents and primary care physicians. Although in all such cases discovering the cause of bleeding is of utmost importance, to institute an appropriate therapeutic plan, the initial goal of management must be to ensure or restore hemodynamic integrity and to determine the extent and severity of bleeding. The boy in the vignette experienced abdominal pain followed by vomiting fresh blood and clots. The initial physical examination is remarkable for signs of cardiovascular instability (tachycardia and hypotension) and splenomegaly. When a patient presents with these findings, portal hypertension with bleeding esophageal varices must always be suspected. Nevertheless, in this case and in every case of suspected upper gastrointestinal (GI) tract bleeding, the initial approach should focus on patient stabilization, including the following:

1. Establishing venous access for fluid resuscitation
2. Performing nasogastric lavage with room temperature normal saline, using a large-bore nasogastric tube

These procedures will stabilize hemodynamic status and allow for assessment of bleeding severity. Lavage fluid that rapidly clears or returns only "coffee-ground" material indicates the cessation of active bleeding. Under these conditions, additional GI evaluation and further studies (ie, endoscopy) may be conducted on an urgent or a semielective basis. Continued aspiration of red blood confirms active, ongoing blood loss, and emergency GI consultation is required for diagnostic evaluation and possible therapeutic intervention.

Many reviews of GI bleeding discuss the differential diagnosis based on patient age at presentation. However, because these diagnoses may occur in more than one age group, it is helpful to consider causes of bleeding that are common or uncommon (Item C57). Hematemesis in infants, children, and adolescents may result from swallowed blood (maternal blood in newborns or nasopharyngeal bleeding), upper GI tract mucosal lesions, variceal bleeding, or rarely, hemorrhage into the biliary tract (hemobilia). As is the case for the boy in the vignette, upper GI tract bleeding is often the initial presentation of esophageal varices secondary to portal hypertension. Therefore, variceal bleeding should be entertained in a child who has splenomegaly (with or without hepatomegaly), ascites, or jaundice. In the absence of chronic liver disease, a history of hepatitis, blood transfusion, right heart failure, or portal vein thrombosis (associated with abdominal surgery, sepsis, shock, exchange transfusion, omphalitis, or umbilical vein catheterization) will provide further diagnostic clues. An octreotide infusion should be started in any patient who is experiencing bleeding from varices.

Other diagnoses should be considered for the boy in the vignette but are less likely in view of the presence of splenomegaly. A careful physical examination should identify causes of swallowed blood from a nasopharyngeal source. A Mallory–Weiss tear is an acute mucosal laceration, usually at the gastroesophageal junction. Hematemesis follows forceful retching, vomiting, or coughing; however, abdominal pain is uncommon unless there is associated musculoskeletal injury from forceful vomiting. Over-the-counter cold medicines that contain nonsteroidal anti-inflammatory agents, even at low dose, have

been associated with acute gastric ulceration and bleeding. Hemodynamically significant bleeding, however, is unusual. Finally, Helicobacter pylori–associated gastritis is an important cause of gastritis and peptic ulceration in both children and adults; however, bleeding is an infrequent complication.

Item C57. Causes of Hematemesis in Childhood

Common Problems	Uncommon Problems
• Swallowed blood - Maternal blood (newborn) - Tonsillectomy - Dental work - Epistaxis • Vitamin K deficiency (newborn) • Esophagitis - Erosive (peptic) esophagitis - Eosinophilic esophagitis - Caustic ingestion - Pill esophagitis • Tetracyclines • Nonsteroidal anti-inflammatory drugs • Bisphosphonates - Viral (in an immunocompromised host) • Foreign-body ulcer • Mallory-Weiss tear • Portal hypertension - Esophageal varices - Gastric varices - Portal gastropathy • Prolapse gastropathy • Hemorrhagic gastropathy - Sepsis - Coagulopathy • Peptic ulcer - Stress - Postsurgical - Central nervous system, systemic disease - Crohn disease - Nonsteroidal anti-inflammatory drugs - *Helicobacter pylori*	• Gastric duplication • Tumors - Leiomyosarcoma - Adenocarcinoma • Hemobilia • Henoch–Schönlein purpura • Multiple hemangiomas • Vasculitis • Vascular malformation - Angiodysplasia - Hemangioma - Dieulafoy lesion - Hereditary hemorrhagic telangiectasia

PREP Pearls

- Suspected upper GI bleeding should always be initially assessed by nasogastric intubation.
- Coffee-ground nasogastric lavage findings indicate the resolution of active bleeding.
- In patients with upper GI bleeding, splenomegaly indicates the presence of portal hypertension and variceal bleeding.

American Board of Pediatrics Content Specification (s):

- Know the differential diagnosis of vomiting bright red blood

Suggested Reading:

- Boyle JT. Gastrointestinal bleeding in infants and children. Pediatr Rev. 2008;29:39-52. doi:10.1542/pir.29-2-39
- Chawla S, Seth D, Mahajan P, Kamat D. Upper gastrointestinal bleeding in children. Clin Pediatr (Phila). 2007;46:16-21.
- doi:10.1 177/1084713806297151
- Eroglu Y, Emerick KM, Whitington PF, Alonso EM. Octreotide therapy for control of acute gastrointestinal bleeding in children. J Pediatr Gastroenterol Nutr. 2004;38:41-47
- Fox VL. Gastrointestinal bleeding in infancy and childhood. Gastroenterol Clin NAm. 2000;29:37-66. doi:10.1016/S0889-8553%2805%2970107-2 Squires RH. Gastrointestinal bleeding. Pediatr Rev. 1999;20:95-101. doi:10.1542/pir.20-3-95

Item 58

A 10-month-old has been hospitalized for the third time for persistent vomiting and weight loss. His length and head circumference are at the 50th percentile, while his weight is at less than the 3rd percentile. His history and physical examination do not suggest a clear cause for his symptoms, and past evaluations, including infectious disease, neurology, immunology, and gastroenterology consults, have not yielded a diagnosis. His mother is friendly with the staff and doctors and has readily agreed to all suggested diagnostic studies.

Of the following, the MOST appropriate next step in this infant's management is to

A. do a thorough review of all medical records with a multidisciplinary team, including a child abuse specialist
B. exclude all other potential organic causes before considering a factitious cause
C. initiate total parenteral nutrition to improve caloric intake and support growth
D. obtain a pediatric surgery consult for fundoplication and gastric tube insertion
E. obtain a psychiatric evaluation to determine if the parent has an intention to injure the child

Item 58 TE S I-C Preferred Response: A

Munchausen syndrome by proxy is a rare form of child abuse with a prevalence of 0.4 in 100,000 children younger than 16 years old and 2 in 100,000 children younger than 1 year old. The mean age at diagnosis is 40 months. It can involve physical abuse, medical neglect, and/or psychological maltreatment. The 4 characteristics necessary to make the diagnosis are that: (1) a caregiver fabricates the illness; (2) the child is seen for medical evaluation recurrently and frequently, often for diagnostic and therapeutic procedures; (3) the perpetrator denies knowledge of the cause of the child's condition; and (4) the signs and symptoms resolve when the child is removed from the care of the perpetrator. Between 94% and 99% of perpetrators are mothers, but other caregivers including fathers, babysitters, and health care workers occasionally are involved. The perpetrator is often described as friendly, concerned, very involved in the child's care, and agreeable to medical procedures. The caregiver has a health care background in 14% to 30% of cases, and about 14% of the caregivers have a psychiatric diagnosis, but the medical team providing care for the child is often unaware of this.

Although still commonly called Munchausen syndrome by proxy, newer designations have been proposed for describing this phenomenon. Because in no other form of child abuse is the perpetrator's motivation considered in the diagnosis, the American Professional Society on the Abuse of Children stresses that it is important to differentiate the abuse done to the victim from the perpetrator's intention. Consequently, what happens to the child may be called pediatric disease falsification, and the offending caregiver has been labeled as having factitious disorder by proxy. The Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition, has just recommended another slight alteration of this nomenclature to "factitious disorder imposed on another." Factitious disorder imposed on another involves fabricated complaints projected onto a dependent person (usually a child but could involve a dependent adult) that are convincing enough to lead to medical intervention. Although evaluation of the offending caregiver is an important part of the overall care of the family, assessment of the perpetrator's intention is not necessary for the diagnosis and care of the child victim.

This condition does not have a single characteristic presentation, and a spectrum of concerns may be noted. Some children may be brought in for concerns that are exaggerations of normal symptoms and behaviors. Other caregivers will fabricate a severe symptom (eg, apnea) that cannot be easily verified except by direct observation. In other cases, a caregiver induces a symptom, for example by suffocating the child, injecting poisons, or administering laxatives to induce diarrhea or ipecac to produce vomiting. Alternatively, the perpetrator may falsify laboratory results such as placing pebbles in urine to simulate renal stones or contaminating a urine sample with table sugar or blood. The unifying factors are the caregiver's assertion that there is a significant medical problem, that there is no medical evidence to support that assertion, and that the child can be harmed by the assertion or resulting medical intervention.
The most important response when the diagnosis of pediatric disease falsification is considered, as seen in the child in this vignette, is to convene a multidisciplinary team including all physicians providing care to the child, and, whenever possible, a pediatrician expert in child abuse. In addition, nursing, social service, and

physical/occupational therapy professionals should participate, and a hospital legal representative may be helpful. Child protective services representatives and law enforcement must be involved as the diagnosis proceeds. Because these families may seek care from multiple or sequential physicians, it is important to obtain records from all medical providers who have seen the child. Further surgical or invasive diagnostic procedures should not be done unless necessary to exclude logical alternative diagnoses. However, the diagnosis of pediatric disease falsification is not a diagnosis of exclusion, and pursuing all diagnoses even if they are unlikely, is not a reasonable approach.
The prognosis for victims of pediatric disease falsification may be poor, but placing them in a stable and nurturing long-term home and providing individual and family counseling may improve the outcome. Family reunification can rarely be achieved. Case reports describe situations in which the victim and perpetrator share the psychiatric disorder, and s/he may corroborate the perpetrator's story. Some victims develop outright factitious disorder as adults. In addition, the child victim may be left with physical impairments because of either the original "symptom" or the diagnostic and therapeutic interventions performed to address the factitious disease.

PREP Pearls

- The diagnosis of Munchausen by proxy (also known as pediatric disease falsification or factitious disorder imposed on another) is made when the following 4 characteristics are present:
    - The caregiver fabricates the illness.
    - The child is seen frequently and recurrently for medical evaluation.
    - The caregiver denies knowledge of the cause of the child's symptoms.
    - Signs and symptoms resolve when the child is removed from the perpetrator's care.
- The caregiver's motivation or psychiatric diagnosis is not a criterion in the diagnosis of pediatric disease falsification. Assessment of the perpetrator's intentions is not necessary for making the diagnosis.
- The diagnosis of Munchausen by proxy is best made with the involvement of a multidisciplinary team that includes all of the child's physicians and, whenever possible, a pediatrician expert in child abuse.

American Board of Pediatrics Content Specification(s):

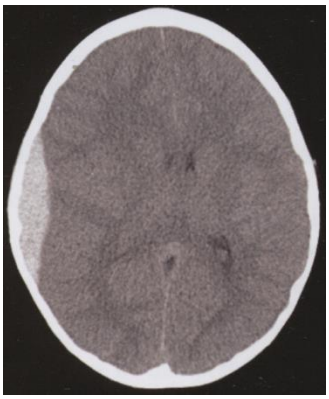
- Recognize the signs of factitious disorder (Munchausen syndrome) by proxy: recurrent sepsis from injecting fluids, chronic diarrhea from laxatives, false renal stones from pebbles, fever from heating thermometer, rashes from trauma, sugar or blood in the urine, etc
- Recognize the features of the parent of a child with factitious disorder (Munchausen syndrome) by proxy
- Recognize that children with factitious disorder (Munchausen syndrome) by proxy may exhibit significant ongoing psychologic problems

Suggested Reading:

- Brown P, Tierney C. Munchausen syndrome by proxy. Pediatr Rev. 2009;30:414-415. doi: 10.1542/pir.30-10-414
- Shapiro M, Nguyen M. Psychological sequelae of Munchausen syndrome by proxy. Child Abuse Negl. 2011;35:87-88. doi:10.1016/j.chiabu.2010.10.005
- Shaw RJ, Dayal S, Hartman JK, DeMaso DR. Factitious disease by proxy: pediatric condition falsification. Hary Rev Psychiatry. 2008;16:215-224. doi: 10.1080/10673220802277870
- Stirling J. Beyond Munchausen syndrome by proxy: identification and treatment of child abuse in a medical setting. Pediatrics. 2007;119:10261030. doi: 10.1542/peds.2007-0563

Item 59

You are evaluating a 15-year-old boy in the emergency department who collided with a parked car while sledding. The accident was witnessed by his friends who report that the patient was not wearing a helmet and was unresponsive after impact. Emergency medical service providers intubated the boy at the scene and report that he moved all extremities in response to stimulation. The boy has a temperature of 37.6°C and a heart rate of 120 beats/min and is being mechanically ventilated at a rate of 20 breaths/min. He has received 1 small dose of fentanyl for agitation and presumed pain. His pupils are 2 mm in size and equal. A large area of swelling over his right temporal area is noted, and computed tomography of his head is shown (Item Q59).



During ongoing observation, you note that his right pupil becomes markedly dilated and unresponsive to light. He now only moves his right arm and leg in response to stimulation.

Of the following, the MOST likely cause of the pupillary finding is

A. compression of the fourth cranial nerve
B. compression of the third cranial nerve
C. disruption of sympathetic nerve activity
D. inflammation of the second cranial nerve
E. narcotic stimulation of opiate receptors

Item 59 Preferred Response: B

Increased intracranial pressure can result in herniation of the cerebral contents. Transtentorial herniation results from downward or upward displacement of the brain through the tentorium at the level of the incisura. Temporal lobe (uncal) herniations are a subcategory of transtentorial herniations that are characterized by dilation of a unilateral pupil because of compression of cranial nerve III (oculomotor) as described for the boy in the vignette. In addition, uncal herniation is accompanied by contralateral paresis. Uncal herniations usually occur with rapid expansion of the contents of the temporal lobe fossa such as seen with epidural hematomas, focal injury, or infection.

Inflammation of cranial nerve II (optic) can arise from various diseases and leads to a decrease in visual acuity. Cranial nerve IV (trochlear) innervates the superior oblique muscle, and injury to the nerve produces difficulty in ocular movement and resultant diplopia. Pupillary response to sympathetic nerve stimulation results in dilation (mydriasis), and thus unopposed parasympathetic stimulation would result in constriction (miosis). Narcotic administration also results in miosis. None of these conditions would be characterized by unilateral pupillary dilation.

PREP Pearls

- Temporal lobe (uncal) herniations are characterized by dilation of a unilateral pupil due to compression of cranial nerve III.
- Uncal herniations usually occurs secondary to trauma such as epidural hematomas.
- Uncal herniation is accompanied by contralateral paresis.

American Board of Pediatrics Content Specification(s):

- Recognize a unilateral dilated pupil as a sign of uncal herniation

Suggested Reading:

- Avner JR. Altered states of consciousness. Pediatr Rev. 2006:27:331-338. doi: 10.1542/pir.27-9-331. http://pedsinreview.aappublications.org/cgi/content/full/27/9/331
- Carlo WA. Intracranial-intraventricular hemorrhage and periventricularleukomalacia. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:566-568
- Kochanek PM, Bell MJ. Neurological emergencies and stabilization. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:296-304

Item 60

You are called by your state newborn screening program for a low total thyroxine (T4) level found on newborn screening. The level was drawn 24 hours after a term, healthy female neonate was born. Pregnancy was uncomplicated, and there was no maternal history of thyroid disease. Repeat testing in your office at 7 days after birth shows a thyroid-stimulating hormone level of 200 mIU/L and a low free thyroxine (FT4) level of 0.2 ng/dL (2.57 pmol/L) in this otherwise healthy girl. Her weight is 3 kg. Findings on physical examination are unremarkable.

Of the following, the BEST next step in the management of this patient is to

A. begin treatment with levothyroxine (LT4)
B. begin treatment with liothyronine (LT3)
C. measure serum thyroglobulin level
D. order a thyroid radionuclide uptake and scan
E. order thyroid ultrasonography

Item 60 S Preferred Response: A

The infant described in this vignette has had 2 abnormal thyroid function test results indicating profound hypothyroidism. Therefore, treatment should be initiated with levothyroxine (LT4) immediately. In clear cases of congenital hypothyroidism (CH), treatment should never be delayed to obtain another study, such as a thyroid uptake scan or ultrasonography. These studies can be performed after treatment is initiated.

The best developmental outcomes occur with levothyroxine therapy started by 2 weeks of age at 10 µg/kg or more per day, compared with lower doses or later start of therapy. There are only minor differences in intelligence, school achievement, and neuropsychological test results in adults with CH that was treated early with levothyroxine compared with control groups of classmates and siblings. However, even with early treatment impaired visuospatial processing, selective memory and sensorimotor defects can occur.

In contrast, the prognosis for normal mental and neurologic performance is less certain for infants with CH that is not detected early by newborn screening or treated later on. If treatment is delayed even a few months, 77% of infants show some signs of developmental delay and may have impairment of arithmetic ability, speech, or fine motor coordination in later life.

Knowledge of the normal range for thyroid function tests in the first year of life is important in deciding whom to treat and how soon treatment is needed to prevent adverse developmental outcomes. Shortly after birth, there is a cold-stimulated surge of thyrotropin (TSH) in the infant that peaks by 30 minutes of life. TSH levels then gradually decrease, so that by 24 hours the TSH level is typically less than 20 to 25 mIU/L. If the TSH level is greater than 25 mIU/L at 24 hours of life, a confirmatory test should be performed. Confirmatory serum testing should be performed in infants before 2 weeks of age, when the upper TSH range has decreased to approximately 10 mIU/L. For persistent elevation of TSH (as seen in the infant in this vignette), treatment should be started with levothyroxine immediately at 10 to 15 pg/kg per day. There is no current recommendation for using liothyronine (LT3) in the routine care of infants with congenital hypothyroidism.

The treatment of infants with TSH elevations between 5 and 10 mIU/L that persist after the first month of life is controversial. A TSH range of 1.7 to 9.1 mIU/L has been reported for children 2 to 20 weeks of age. Other studies report that repeated episodes of TSH levels greater than 5 mIU/L after the age of 6 months were the most important variables associated with developmental delay. Thus, if treatment is begun for a mild elevation of TSH between 5 and 10 mIU/L in the first year of life, consideration should be given to an underlying transient cause, and a trial off therapy at 3 years of age should be considered.

Measurement of serum thyroglobulin (TBG) can be performed if confirmatory testing reveals a normal free thyroxine (FT4) level when the initial total thyroxine (T4) level on screening was low. TBG deficiency can lead to a low total T4 measurement due to a

decreased amount of thyroid hormone bound to TBG, but this is not clinically relevant because the FT4 will be normal.

PREP Pearls

- Suspected congenital hypothyroidism should always be treated with levothyroxine as early as possible and can be weaned later if a transient cause of hypothyroidism is suspected.

American Board of Pediatrics Content Specification(s):

- Know the prognosis for a patient with congenital or acquired hypothyroidism

Suggested Reading:

- American Academy of Pediatrics, Rose SR; Section on Endocrinology and Committee on Genetics, American Thyroid Association, Brown RS; Public Health Committee, Lawson Wilkins Pediatric Endocrine Society, Foley T, Kaplowitz PB, Kaye CI, Sundararajan S, Varma SK. Update of newborn screening and therapy for congenital hypothyroidism. Pediatrics. 2006;117:2290-2303. doi:10.1542/peds.2006-0915
- LaFranchi SH. Approach to the diagnosis and treatment of neonatal hypothyroidism. I Clin Endocrinol Meta b. 2011;96:2959-2967. doi:10.1210/jc.2011-1175

Item 61

A 17-year-old boy in your practice has an 18-mm induration response to a tuberculin skin test that was placed as part of a precollege physical examination. He is clinically well, with no cough, night sweats, fever, or weight loss. He is well-appearing and findings on his physical examination are unremarkable. He lives with his parents and 14-year-old sister, all of whom are reported as healthy.
He immigrated to the United States from India at 3 years of age. He received a Bacillus Calmette-Guerin vaccine as an infant. He has not travelled internationally since immigrating into the United States. His grandparents visited from India 6 months ago but are reportedly healthy.

Of the following, the BEST next step in management of this patient is to

A. begin isoniazid preventive therapy
B. obtain a chest radiograph
C. order an interferon-y release assay
D. repeat the tuberculin skin test in 6 months
E. repeat the tuberculin skin test in 2 weeks

Item 61 S Preferred Response: B

Bacillus Calmette-Guerin (BCG) is an attenuated vaccine derived from a strain of Mycobacterium bovis that is administered at birth in many countries around the world to prevent tuberculosis disease. Most people who receive this vaccine develop some degree of short-term reaction to a tuberculin skin test (TST). However, more than 10 years after the receipt of BCG vaccine, an 18-mm induration as described for the boy in the vignette should not be attributed to BCG vaccine; the finding would be considered a positive TST reaction especially in a person from a region with high rates of tuberculosis.
In general, a routine TST in low-risk populations is not indicated. A history of epidemiologic risk for exposure to tuberculosis including contact with an active case of tuberculosis, incarceration, institutionalization, or living in a high-risk area of the world are indications for performing a TST.

In the face of a positive TST result in a well-appearing individual, the best next step in the evaluation would be a chest radiograph. If the chest radiograph is normal or reveals only calcification consistent with a walled-off past infection, a diagnosis of latent tuberculosis infection is made and then preventive therapy with isoniazid would be indicated.

Interferon-γ release assays (IGRAs) detect reaction to Mycobacterium tuberculosis but not BCG. However, the extent of induration to the TST and the nation of origin described in this case are sufficient to define tuberculosis and additional testing is unnecessary. Although IGRAs may be useful for testing in adolescents, data to recommend their use in children younger than 5 years are insufficient. Positive IGRA and TST results do not differentiate tuberculosis infection from tuberculosis disease.
In view of the magnitude of the reaction to the TST, further repeat testing is not indicated. In addition, conducting a repeat TST 2 weeks after a prior test could boost a borderline test result to an apparent positive result, thus making interpretation difficult.

PREP Pearls

- Although prior receipt of BCG vaccine may lead to a reaction to a TST, this wanes with time and is expected to be <10 mm by 10 years after the BCG.
- Interferon-γ release assay results are not affected by previous BCG vaccination.

American Board of Pediatrics Content Specification(s):

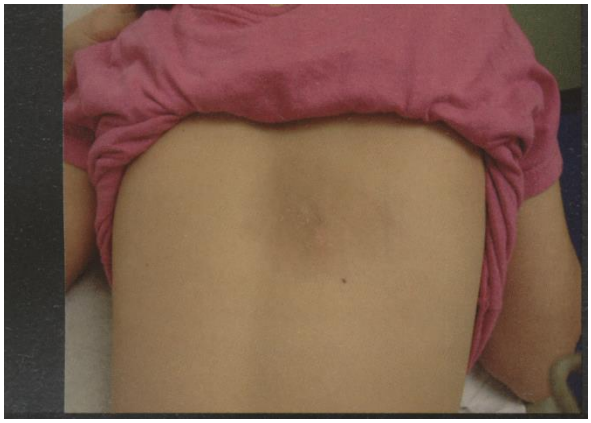
- Understand the effect of the BCG vaccine on the tuberculin skin test

Suggested Reading:

- American Academy of Pediatrics. Tuberculosis. In: Pickering LK, Baker CI, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:736-759
- Machingaidze S, Wiysonge CS, Gonzalez-Angelo Y, et al. The utility of an interferon gamma release assay for diagnosis of latent tuberculosis infection and disease in children: a systematic review and meta-analysis. Pediatr Infect Dis J. 2011;30(8):694-700. doi:10.1097/INF.0b013e318214b915
- Mazurek GH, Jereb J, Vernon A, et al. Updated guidelines for using interferon gamma release assays to detect Mycobacterium tuberculosis infection—United States, 2010. MMWR Recomm Rep. 2010;59 (RR-5):1- 26

Item 62

A 10-year-old girl presents to your office with a lesion diagnosed 4 years ago as a nevus simplex (salmon patch) on her back. Recently, the lesion has enlarged, darkened, and developed an area that is hard and white (Item Q62).



Of the following, the BEST diagnosis based on the clinical findings is

A. lichen scleroses
B. localized scleroderma
C. neurofibromatosis
D. tinea versicolor
E. vitiligo

Item 62 Preferred Response: B

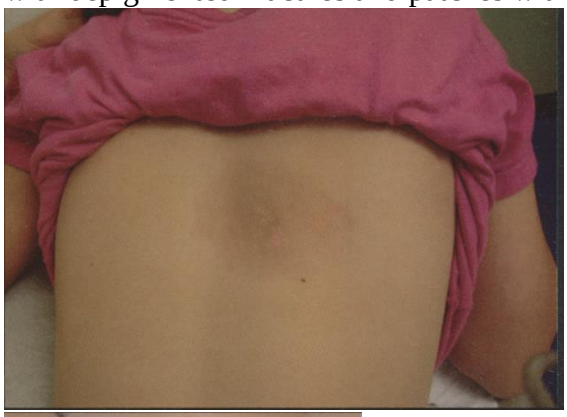
The child described in the vignette has localized scleroderma (LS). Localized scleroderma has several subtypes including plaque morphea, generalized morphea, bullous morphea, linear morphea, and deep morphea—that are characterized by the pattern and depth of skin involvement. Early lesions of LS are characterized by a lilac ring or violaceous inflammatory border. The skin is indurated throughout the lesion. The lesions may enlarge and new lesions may occur. Because of uncontrolled inflammation, tissue damage accumulates as the diseases progresses, causing skin thickening and, commonly, an ivory-colored center of sclerosis (Item C62A). Dermal and subcutaneous atrophy can develop later in the disease course, resulting in visible veins, a flat or concave "cliff-drop" appearance to the skin (depression of the underlying subcutaneous tissues), and a lack of hair growth. Postinflammatory hyperpigmentation or hypopigmentation often occurs. The changes are often described as a "bruise that does not heal."

Linear scleroderma is a subtype of scleroderma that presents as a single linear band that affects the head, trunk, or extremities. Linear scleroderma is usually unilateral. If the linear band of scleroderma crosses joints, it can affect the underlying structures, including muscle, tendon, joint capsule, and growth plate. These lesions can result in atrophy or shortening of the limb and joint contractures. Both linear scleroderma affecting the scalp and forehead (also called en coup de sabre) and Parry-Romberg syndrome (a form that causes hemifacial atrophy) can involve the central nervous system (CNS). Affected individuals are at risk for seizures, chronic headache, and optic neuritis and should undergo cranial magnetic resonance imaging (MRI) if CNS symptoms are present. Findings on MRI can include calcifications, white matter changes, vascular malformations, and signs of CNS vasculitis. Several other subtypes of LS exist, varying from single lesions (morphea) to more generalized involvement of the limbs (pansclerotic morphea). Untreated LS can result in growth abnormalities, disfiguring lesions, and joint contractures.

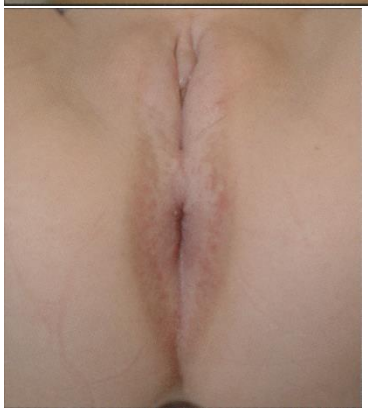
Diagnosis of LS can be made by clinical examination or skin biopsy. Treatment for small isolated lesions can be topical with phototherapy, topical steroids, or other topical immunomodulating agents. Large or multiple lesions, especially those involving joints or the face, are often treated systemically to prevent permanent damage and disfigurement. No standardized treatments exist; however, most rheumatologists use a combination of steroids and methotrexate to control the disease. Expectations for improvement in the lesions are dependent on the stage of disease at the time of treatment. Once sclerosis and scarring have occurred, very little improvement can be expected. Referral to dermatology or rheumatology in order to improve outcomes is warranted for these patients.

Lichen sclerosus lesions are scaly and typically hypopigmented, with a cigarette paper-like wrinkled appearance and varying degrees of sclerosis (Item C62B). Lichen sclerosis can be present concurrently with morphea. Neurofibromatosis can present with ash leaf spots but without evidence of induration. Tinea versicolor presents as erythematous, hypopigmented, or hyperpigmented macules that are most commonly over the trunk and

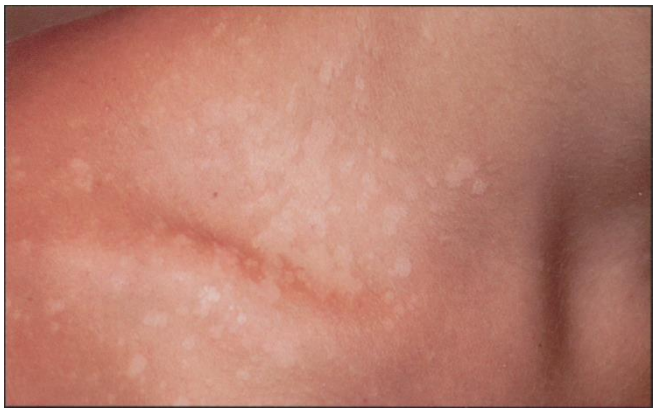
upper extremities but without skin thickening (Item C62C, page C-50). Vitiligo presents with depigmented macules and patches without skin thickening (Item C62D, page C-50).



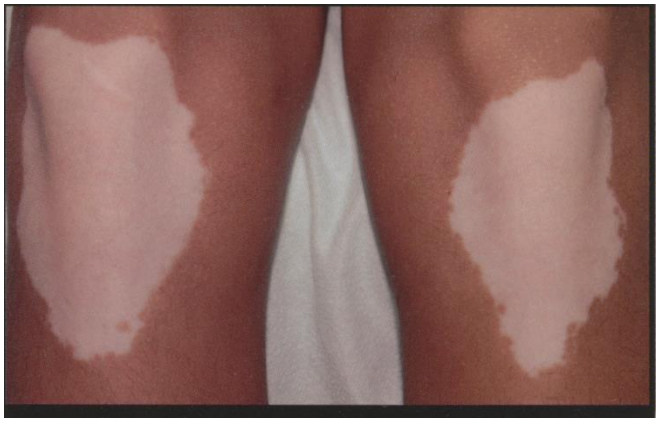
ITEM C62A: Localized scleroderma, note the ivory-colored central area.



ITEM C62B: Lichen sclerosus lesions are scaly and typically hypopigmented.



ITEM C62C: Tines versicolor presents as erythematous, hypopigmented, or hyperpigmented macules that are most commonly over the trunk and upper extremities but without skin thickening.



ITEM C6D): Vitiligo is characterized by a complete loss of pigmentation.

PREP Pearls

- Early diagnosis of localized scleroderma and treatment by a rheumatologist or dermatologist are key in preventing long-term disfigurement.
- Localized scleroderma overlying limbs and joints can cause functional deficits.
- Early localized scleroderma lesions are characterized by induration of the skin and erythema or a violaceous border.

American Board of Pediatrics Content Specification (s):

- Recognize the clinical manifestations of localized scleroderma (morphea and linear scleroderma)

Suggested Reading:

- Torok KS. Pediatric scleroderma: systemic or localized forms. Pediatr Clin NAm. 2012;58:381-405. doi:10.1016/j.pcl.2012.03.011

Item 63

A 16-year-old boy complains of a depressed mood of 3 months' duration associated with loss of interest in previously enjoyed activities, low energy, poor appetite, difficulty concentrating, and thoughts of hopelessness. He has started to have some suicidal thoughts but has no history of suicidal plans or attempts. His school performance has been significantly worsening over the past 3 months. The boy goes to bed around 11:00 PM in a quiet, dark room but does not fall asleep until 4:00 AM. He wakes at 6:00 or 7:00 AM. Shortly before he developed his current depressed mood, he had a 1-week period of elevated mood with a high energy level and a decreased need for sleep. He was disruptive in class during that time, which resulted in disciplinary action by his teacher. For the past 6 weeks, the boy has been seeing a psychologist who has referred him to you for consideration of a medication trial because of his worsening condition. While awaiting an available appointment with a child psychiatrist, you decide to initiate a medication trial.

Of the following, the MOST appropriate treatment strategy is to initiate:

A. amitriptyline
B. fluoxetine
C. lamotrigine
D. lamotrigine
E. valproic acid

Item 63 S Preferred Response: D

The patient described in the vignette is currently suffering from both depression and insomnia with only 2 to 3 hours of sleep per night. Three months ago he had a 1 -week period before the onset of his depression that likely represented a manic episode (high energy, little need for sleep, and negative impact on daily functioning); however, based on the information in the vignette, a definitive diagnosis of bipolar disorder cannot be made. The possibility that he might have bipolar disorder significantly complicates the recommended medication treatment plan.

Insomnia to this degree is a problem that perpetuates his current depression, and if he has bipolar disorder, the insomnia would place him at high risk for switching back into a manic state. Therefore, his insomnia should be the first treatment target to improve both the depression and reduce the risk of a manic switch. Among the medication choices listed, lorazepam given at bedtime to help initiate sleep would be the best immediate option because of the high likelihood of improving sleep with the first administration. It would be important to follow up with this patient within a few days of starting lorazepam to adjust dosage as needed for a reasonable sleep duration (for instance >5 hours per night). Benzodiazepines are not recommended for long-term use to treat insomnia, so a weaning plan should quickly follow the short-term effective restoration of sleep. If he is confirmed to have a bipolar disorder, the more preferred treatment for insomnia would be a sedating atypical antipsychotic such as risperidone, which can both restore sleep and treat a manic mood disturbance.

Because the patient in this vignette is not definitively diagnosed as having bipolar disorder, it would not be advisable to initiate treatment with valproic acid because of the high incidence of side effects and the less predictable or immediate effect on sleep patterns. Also, valproic acid is a less preferred initial bipolar medication than other options in young people. Amitriptyline can cause sedation, but its effect is less reliable. Also, amitriptyline is specifically not recommended for the treatment of depression in children and adolescents. Tricyclic antidepressants have not been shown to be effective in treating depression in this age group and have a high risk of fatality in case of an overdose. Lamotrigine can be helpful for treating bipolar depression in adults but it is not clearly helpful for insomnia. Also because of the risk of developing Stevens-Johnson rash with rapid titration, it takes about 2 months to safely increase lamotrigine dosage to full therapeutic levels—far too long to be helpful for this patient's current problem. Although fluoxetine is a preferred first-line treatment for most adolescents with unipolar depression, it would be inappropriate in this case. Fluoxetine does not treat insomnia quickly, and all selective serotonin reuptake inhibitors should be avoided in patients with suspected bipolar disorder because they can increase the frequency of manic episodes. Because of their complex treatment issues, it is recommended that adolescents with bipolar depression be under the principal care of a mental health specialist.

PREP Pearls

- The use of medications to restore sleep in a bipolar patient with insomnia is important to prevent switching into mania.
- Selective serotonin reuptake inhibitors should be avoided in bipolar depression.

AAP Mental Health Competency

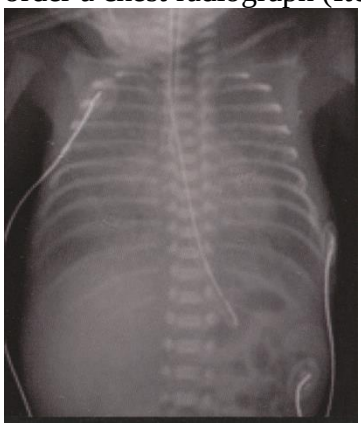
- Identify how to treat depression in a patient suspected of having true bipolar disorder

Suggested Reading:

- American Academy of Child and Adolescent Psychiatry. Practice Parameter for the Assessment and Treatment of Children and Adolescents With Bipolar Disorder. www.aacap.org
- Kowatch RA, DelBello MP. Pediatric bipolar disorder: emerging diagnostic and treatment approaches. Child Adolesc Psychiatr Clin N Am. 2006;15(issue?):73-108. doi:10.1016/j.chc.2005.08.013

Item 64

A neonate is being assessed for increasing work of breathing. The infant was born weighing 2.8 kg at 36 weeks of gestation to a 32-year-old primigravida by urgent caesarean section due to severe preeclampsia. Artificial rupture of membranes occurred at delivery and revealed clear amniotic fluid. He emerged vigorous, with the development of grunting, retracting, and flaring at 30 minutes after birth. The neonate required initiation of a 30% oxygen hood two hours after birth to maintain oxygen saturation values greater than 94%. His clinical status was unchanged until 12 hours after birth, when his oxygen requirement escalated to 40% and was accompanied by increased grunting and retracting. Physical examination reveals a neonate with moderate grunting and retractions who has a temperature of 37°C, heart rate of 140 beats/min, respiratory rate of 76 breaths/min, blood pressure of 60/36 mm Hg, and a pulse oximetry reading of 90% in a 40% oxygen hood. The examination is notable for diminished breath sounds bilaterally. The white blood cell count is 12,000/pL (12.0 x 109/L), with 50% polymorphonuclear leukocytes, 5% bands, 30% lymphocytes, and 15% monocytes. You order a chest radiograph (Item Q64).

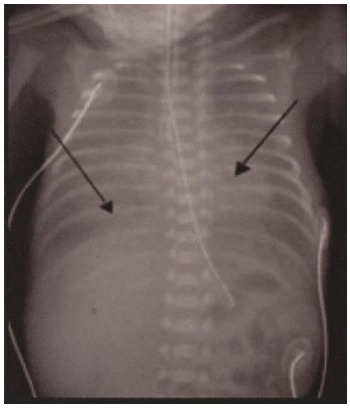


Of the following, the MOST likely cause of this neonate's signs and symptoms is

A. cystic adenomatoid malformation
B. group B streptococcal pneumonia
C. respiratory distress syndrome
D. retained fetal lung liquid syndrome
E. total anomalous pulmonary venous return

Item 64 Preferred Response: C

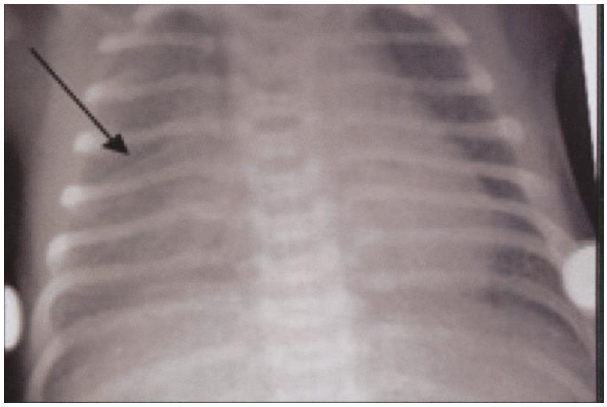
The infant described in the vignette has the clinical and radiographic findings seen in respiratory distress syndrome (RDS). Respiratory distress syndrome is caused by decreased production of surfactant by type II pneumocytes in the lung. Surfactant provides surface tension to the alveoli, improving compliance and minimizing alveolar collapse. Surfactant deficiency leads to atelectasis and progressive respiratory compromise. Clinical signs of respiratory distress seen in affected infants include tachypnea, grunting, nasal flaring, retractions, and hypoxia. These findings escalate over time as the degree of atelectasis increases. The classic chest radiograph of an infant with RDS demonstrates a "ground glass" appearance related to microatelectasis with superimposed air bronchograms (Item C64A).



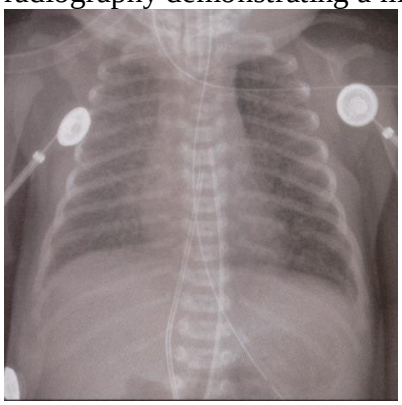
ITEM C64A: Respiratory distress syndrome. Note the "ground glass" appearance, which represents diffuse microatelectasis, and air bronchograms (arrows).

Although RDS is generally associated with the premature infant born before 30 weeks of gestation, it has been recognized that the late preterm infant (34 0/7 to 36 6/7 weeks' gestation) is at increased risk of developing RDS compared with a full-term infant. Late preterm infants account for up to 9% of all deliveries in the United States and are often treated in newborn nurseries. The incidence of RDS in this population ranges from 11% at 34 weeks to 3% at 36 weeks. RDS must be considered in the differential diagnosis of late preterm infants with respiratory distress. Affected infants may require intubation and delivery of exogenous surfactant through the endotracheal tube to improve their respiratory status.

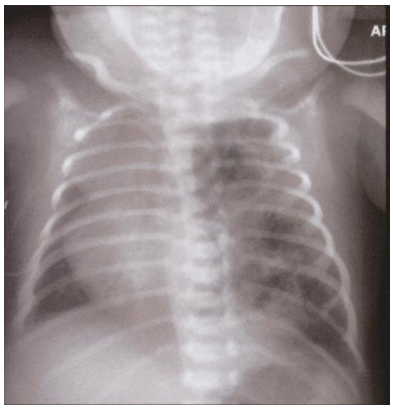
Retained fetal lung liquid syndrome (RFLLS) (also referred to as transient tachypnea of the newborn) also presents with the clinical findings of respiratory distress. Because of incomplete resorption of fluid from the lungs, affected infants typically present shortly after birth and have tachypnea for up to 72 hours without escalation of their clinical findings. Unlike the infant in the vignette, the classic chest radiograph of an infant with RFLLS demonstrates prominent perihilar vascular markings and fluid in the fissures (Item C64B). Group B



ITEM C64111: Retained fetal lung liquid syndrome. Note the increased interstitial markings and fluid in the interlobar fissure on the right (arrow).streptococcal (GBS) pneumonia should be considered in any infant with respiratory distress. The appearance of the chest radiograph in GBS is highly variable and may appear similar to RDS or RFLLS. Although the prenatal history and benign complete blood count make this diagnosis less likely in the infant in the vignette, administration of antibiotics would be appropriate. Infants with total anomalous pulmonary venous return often present with severe hypoxia and respiratory failure. Obstruction of the pulmonary veins may cause pulmonary congestion, with chest radiography demonstrating pulmonary edema (Item C64C, page C-52). Cystic adenomatoid malformation of the lung is a rare condition caused by abnormal embryogenesis leading to cystic overgrowth of the terminal bronchioles. Affected infants may present with respiratory distress, with chest radiography demonstrating a mass with air-filled cysts (Item C64D, page C-52).



ITEM (64C: In obstructed TAVPR total anomalous pulmonary venous return bilateral interstitial opacities and a normal heart size are noted.



ITEM C64D: Mixed lucencies with mass effect, left to right shift.

PREP Pearls

- The classic chest radiograph of an infant with respiratory distress syndrome demonstrates a "ground glass" appearance resulting from microatelectasis with superimposed air bronchograms.
- Respiratory distress syndrome should be included in the differential diagnosis of a late preterm infant with respiratory distress.

American Board of Pediatrics Content Specification(s):

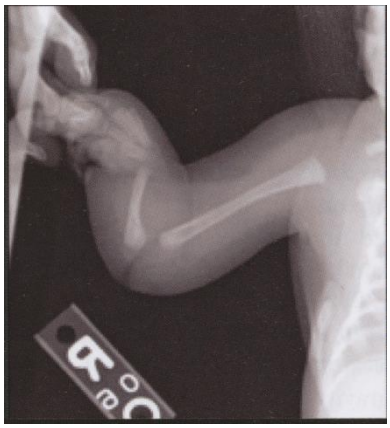
- Recognize the characteristic clinical and radiographic appearance of respiratory distress syndrome

Suggested Reading:

- Consortium on Safe Labor; Hibbard JU, Wilkins I, Sun L, et al. Respiratory morbidity in late preterm births. /AMA. 2010;304:419-425. doi: 10.1001/jama.2010.1015
- Guglani L, Lakshminrusimha S, Ryan RM. Transient tachypnea of the newborn. Pediatr Rev. 2008;29:e59-e65. doi: 10.1542/pir.29-11-e59
- Warren JB, Anderson JD. Newborn respiratory disorders. Pediatr Rev. 2010; 487-496. doi: 10.1542/pir.31-12-487

Item 65

A 1-day-old neonate, with Apgar scores of 8 at one minute and 9 at five minutes, was born to a healthy, 17-year-old with no known medical problems. The mother did not receive any prenatal care. Birth weight was 3,050 grams, and estimated gestational age was 40 weeks. On physical examination, the baby is in no distress and has a few scattered petechiae on the face, neck, and shoulders. The upper extremities are shortened in the forearm. A radiograph is shown (Item Q65). The remainder of the physical examination is normal. The patient is afebrile and vital signs are normal. Head ultrasonography is unremarkable. The results of the baby's complete blood cell count are as follows:



- White blood cell count, 12,500/µL (12.5 x 10^9/L), with 45% polymorphonuclear leukocytes, 45% lymphocytes, 7% monocytes, and 3% eosinophils
- Hemoglobin, 16.3g/dL (163 g/L)
- Platelet count, 30 x 10^3/µL (30 x 10^9/L)
- Mean platelet volume, 9 fL; normal range, 6 to 10 fL

An evaluation to rule out sepsis and empiric parenteral antibiotics have been started.

Of the following, the MOST appropriate next treatment for this patient is

A. anti-D antibody
B. corticosteroids
C. γ-globulin intravenous
D. plasmapheresis
E. platelet transfusion

Item 65 Preferred Response:

The neonate in the vignette has severe thrombocytopenia (<50 x 10^3/µL [<50 x 10^9/L]) and needs an immediate platelet transfusion because of the high incidence of life-threatening hemorrhage (eg, intracranial bleed) associated with neonatal platelet counts in this range. The normal platelet count ranges from 150 to 450 x 10^3/µl, (150-450 x 10^9/µL). Thrombocytopenia can be categorized into disorders of increased destruction or decreased production. Increased platelet destruction usually leads to a predominance of enlarged platelets on the peripheral blood smear, as indicated by an increased mean platelet volume (MPV). Megakaryocytes are large platelet precursors in the bone marrow and are increased as a compensatory mechanism for increased platelet destruction in the peripheral circulation. Young platelets are generally large when they are released from the bone marrow; therefore, when there is increased turnover of platelets due to destruction, the MPV tends to be high. Examples of mechanisms of platelet destruction include immune-mediated destruction (idiopathic thrombocytopenic purpura or from the transplacental effect of maternal antibodies against the infant's platelets), mechanical platelet destruction (disseminated intravascular coagulation or hemolytic uremic syndrome), and platelet sequestration (hypersplenism). In contrast, patients with problems associated with decreased platelet production have normal MPV and decreased numbers of megakaryocytes in the bone marrow. Decreased platelet production can be due to infiltration (leukemia), bone marrow failure (aplastic anemia or congenital marrow failure), infection, nutritional deficiencies, genetic disorders, and cyanotic heart disease.

The absent radii seen on the radiograph with the presence of thumbs on physical examination is consistent with the diagnosis of thrombocytopenia with absent radii (TAI syndrome. TAR syndrome is a congenital disorder of variable inheritance. The genetic cause is unclear, and the inheritance pattern is uncertain, with reports of both autosomal recessive and autosomal dominant patterns. The diagnosis is made based on clinical features of bilateral radial dysplasia (with thumbs present) and thrombocytopenia, which can be severe in the first week of life. Most infants will become symptomatic within the first 4 months; however, the thrombocytopenia tends to improve and can resolve after the first year of life. The MPV is normal, and the bone marrow examination reveals decreased size and number of megakaryocytes. Other congenital anomalies associated with TAR syndrome may affect the gastrointestinal, skeletal, genitourinary, and cardiac systems.

Other congenital disorders with associated cytopenias, such as Fanconi anemia, should be ruled out. Fanconi anemia is a rare autosomal recessive bone marrow failure syndrome with congenital anomalies, such as abnormal thumbs, abnormal skin pigmentation, microcephaly, and urogenital abnormalities. The pancytopenia seen in Fanconi anemia usually appears later in childhood at a mean age of 6 to 9 years but does not improve.

Anti-D antibody is used for infants who have maternally derived antibodies to the Rh antigen on the red blood cells. Steroids, intravenous immunoglobulin, or plasmapheresis may be recommended in cases of immune-mediated thrombocytopenia, however, plasmapheresis is not feasible in a neonate because of the patient's small blood volume and the minimum blood requirement needed to prime the pheresis apparatus.

In neonates without a clear immune origin, the mainstay treatment for thrombocytopenia is usually immediate platelet transfusion.

PREP Pearls

- In neonates, the mainstay of treatment for thrombocytopenia is immediate platelet transfusion to prevent life-threatening hemorrhage.
- Neonates with thrombocytopenia should be examined thoroughly for dysmorphic features, which could be part of a syndrome.
- Sepsis should always be ruled out in any newborn while other causes of thrombocytopenia are being investigated.

American Board of Pediatrics Content Specification(s):

- Manage the thrombocytopenia associated with TAR syndrome

Suggested Reading:

- Chakravorty S, Roberts I. How I manage neonatal thrombocytopenia. Br J ematol. 2011;156:155-162. doi:10.1111/j.1365-2141.2011.08892.x
- Consolini DM. Thrombocytopenia in infants and children. Pediatr Rev. 111;32:135-152. doi:10.1542/pir.32-4-135
- De Ybarrondo L, Barratt MS. Thrombocytopenia absent radius syndrome. ,diatr Rev. 2011;32:399-400. doi:10.1542/pir.32-9-399
- Stanworth SJ, Clarke P, Watts T, Ballard S, et al. Prospective, observational ady of outcomes in neonates with severe thrombocytopenia. Pediatrics. 109;124:e826-e834. doi:10.1542/peds.2009-0332

Item 66

A 3-year-old girl suddenly refuses to walk. There is no reported history of injury or ingestions. She has been well, although several children in her preschool class have been absent because of illness. Physical examination shows a temperature of 37.8°C, blood pressure of 88/62 mm Hg, heart rate of 96 beats/min, and respiratory rate of 20 breaths/min. She is crying loudly but calms down when her mother holds her.

The girl's neck is supple and there are no skin lesions. Her neurologic examination shows conjugate eye movements in all directions. She has strong, symmetric facial movements when crying and strong, symmetric limb movements when she is resisting examination. After being calmed again, her deep tendon reflexes are found to be absent. She can sit independently, but, when placed standing, she wobbles, immediately adopts a wide-based stance, refuses to take steps, and collapses to the floor while crying. Results of magnetic resonance imaging of the brain with and without contrast are normal.

Of the following, the MOST likely diagnosis is

A. acute cerebellar ataxia
B. ataxia telangiectasia
C. Friedreich ataxia
D. Guillain-Barre syndrome
E. opsoclonus-myoclonus-ataxia syndrome

Item 66 Preferred Response: A

The girl described in the vignette has acute onset of ataxia. At her age, and with no other historic, neurologic, or imaging findings, the most likely cause is acute cerebellar ataxia. In addition to an abrupt onset of ataxia, horizontal nystagmus and hyporeflexia may be seen. Mental status is normal, but the child may be uncooperative or fussy because of anxiety. Normal mental status differentiates acute cerebellar ataxia from toxic ingestion, which. is the most common cause of acute ataxia in childhood. Other causes of acute ataxia such as stroke or brain tumor were not seen on imaging. No signs of concurrent infection such as meningitis or encephalitis were noted. The history of abrupt onset of symptoms is incompatible with diseases that cause progressive ataxia. Acute cerebellar ataxia is thought to be a postviral syndrome; often patients have a recent history of viral illness, especially varicella infection; since the advent of varicella vaccination the incidence of acute cerebellar ataxia has declined. Symptoms typically improve over weeks to months and rarely recur.

Ataxia-telangiectasia is an autosomal recessive disorder that presents in young toddlers with slowly progressive gait ataxia. Eye movement abnormalities develop over time, including oculomotor apraxia, a diminished ability to voluntarily move the eyes, while nonvoluntary eye movement is preserved. This can be seen by rapidly turning a child's head and observing eye movements; however, when asked, the child is only able to move his or her eyes with effort. Telangiectasias are seen in the face and neck as the child reaches age 3 to 5 years. Many children have recurrent respiratory infections because of the associated immune deficiency. Neurologic deterioration occurs over years, with the development of dysarthria, dysphagia, and movement disorders. There is no specific treatment for ataxia-telangiectasia.

Friedreich ataxia is another autosomal recessive disorder that presents with progressive ataxia. Onset ranges from toddlers to adults. In addition to cerebellar ataxia, patients with Friedreich ataxia have loss of vibration and proprioception; this presents with worsened ataxia in dark rooms or when the patient's eyes are closed (abnormal Romberg test). Optic neuropathy, eye movement disorders, cardiomyopathy, and diabetes mellitus commonly develop. Medication trials for Friedreich ataxia have not shown clear benefit. Death is usually the result of cardiomyopathy or complications of dysphagia.
The Miller Fisher variant of Guillain Barre syndrome is characterized by the abrupt onset of ataxia, ophthalmoplegia, and areflexia. In this case, the lack of eye movement abnormalities makes this diagnosis less likely. The laboratory and imaging findings in the Miller Fisher variant are the same as in Guillain Barre syndrome: brain and spine imaging findings are normal, and the cerebrospinal fluid can be normal or show cytoalbuminologic dissociation (elevated protein with normal white blood cell count). Treatment is typically with immunoglobulin intravenous, and recovery occurs over weeks to months.

Opsoclonus-myoclonus-ataxia (OMA) syndrome is a paraneoplastic syndrome typically presenting in young children I to 2 years of age. Opsoclonus is an abnormal, rapid, conjugate, jerky eye movement that happens spontaneously and also when the child looks quickly in any direction. Myoclonus is a brief, shocklike jerk of the body or limbs, and

ataxia presents with truncal titubation or instability. In childhood, OMA syndrome is associated with neuroblastoma. In addition to investigation and treatment for neuroblastoma, many children are treated with steroids, immunoglobulin intravenous, or other immunosuppressive therapies for the neurologic symptoms.

PREP Pearls

- Acute cerebellar ataxia is a common cause of acute ataxia in toddlers generally considered postviral.
- Diagnostic evaluation for acute ataxia should include brain imaging to evaluate for acute stroke or tumor. Computed tomography is the best modality in an unstable patient but magnetic resonance imaging will give more information.

American Board of Pediatrics Content Specification(s):

- Know the common causes of acute ataxia

Suggested Reading:

- Augustine EF, Mink JW. Movement disorders. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:2053-2061
- Ryan MM, Engle EC. Acute ataxia in childhood. J Child Neurol. 2003;18(5):309-316. doi: 10.1177/08830738030180050901

Item 67

A 16-year-old boy is seen for a sports preparticipation physical evaluation. He is an 11th grader preparing for the wrestling season, and he would like to wrestle at a weight class that is lower than his current weight. His parents ask you to counsel him about safe weight control practices during the wrestling season. Upon review of his records, you note that this teenager has consistently been around the 70th percentile for weight and the 50th percentile for height.

Of the following, the statement MOST appropriate to include in your discussion with this boy is that

A. athletes at his age and grade level should not implement a weight loss plan for sports
B. an athlete's build does not significantly affect his or her performance, except for athletes in the extreme ranges for body composition
C. cycling between higher and lower weights will lead to an increased metabolic rate and make weight control easier
D. he should plan his training and weight loss to achieve between 5% and 10% body fat
E. mild hypohydration will allow him to safely reduce his weight before events and will not interfere with performance

Item 67 S Preferred Response: B

Many young athletes attempt to lose weight in the belief that weight reduction will enhance their athletic performance. Children and adolescents who participate in activities that emphasize leanness or a thin physique, such as dance, gymnastics, wrestling, and distance running, are more likely to attempt to cut weight. For most athletes, build has not been shown to predict success in sports and other physical activities. However, being significantly overweight or underweight may be detrimental to an athlete's health and physical performance. In their policy statement "Promotion of Healthy Weight-Control Practices in Young Athletes:' the American Academy of Pediatrics (AAP) advises that children should not implement a weight loss plan for sports before 9th grade. Adolescent athletes who would like to lose weight should work with their physicians to develop a gradual weight loss program that incorporates a healthy, diverse diet with adequate caloric intake. The nutritional program should be paired with an appropriate physical training regimen that includes strength training and aerobic exercise. Cyclic weight loss and gain can be an indicator of inappropriate weight loss methods, such as binging and purging or voluntary dehydration. Dehydration is especially dangerous for young athletes; children and teens have a higher body surface area to body mass ratio and decreased sweating capacity and are therefore more susceptible to heat illness. Additionally, even mild hypohydration has been shown to impair physical performance in both adults and children.. The use of body mass index to assess weight in relation to height in children, particularly those with a muscular build, may result in the inappropriate classification of some young athletes as being overweight. Body composition measurements combined with height and weight measurements more accurately reflect body type in young people. Adolescent male athletes should have at least 7% to 10% body fat. There are no recommendations for minimum body fat in female adolescents; however, it is generally accepted that female athletes should maintain a higher percentage of body fat than should male athletes. A body fat content of 14% to 17% is considered "very low" for female athletes. Physicians should counsel young athletes who want a weight loss program about minimum acceptable weights and healthy weight control practices.

PREP Pearls

- The AAP position statement "Promotion of Healthy Weight-Control Practices in Young Athletes" provides guidelines for appropriate weight control measures for child and adolescent athletes.
- Unhealthy weight control practices can be especially detrimental to children and teens.

American Board of Pediatrics Content Specification(s):

- Understand the role of fluids in weight control for athletes
- Know the appropriate amount of weight loss per week for athletes who participate in sports with weight categories
- Know that many athletes who participate in sports with weight categories practice weight control that may be pathogenic and pathologic

Suggested Reading:

- American Academy of Pediatrics Committee on Sports Medicine and Fitness. Promotion of healthy weight-control practices in young athletes [published correction appears in Pediatrics. 2006;117(4):1467]. Pediatrics. 2005;116(6):1557-1564. doi:10.1542/peds.2005-2314
- Kiningham RB, Gorenflo DW. Weight loss methods of high school wrestlers. Med Sci Sports Exerc. 2001;33(5):810-813
- Turocy PS, DePalma BF, Florswill CA, et al. National Athletic Trainers' Association position statement: safe weight loss and maintenance practices in sport and exercise. J Athl Train. 2011;46(3):322-336

Item 68

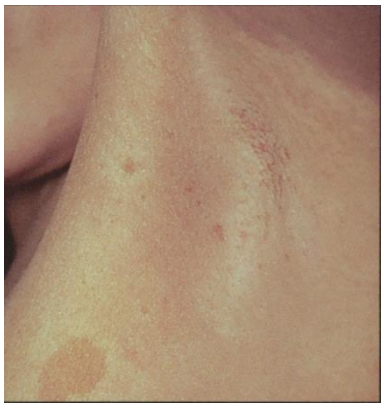
While examining a 2-year-old for a routine health supervision visit at least 18 hyperpigmented macules greater than 0.5 cm in diameter are noted on her trunk and extremities. No axillary or inguinal freckles are noted and no subcutaneous or cutaneous lesions suggestive of neurofibromas are observed. Findings on examination by a pediatric ophthalmologist are normal, with good visual acuity in both eyes and no Lisch nodules noted. The history is negative for anyone else in the family with cafe au lait spots or other signs of neurofibromatosis.

Of the following, you are MOST likely to tell her parents that

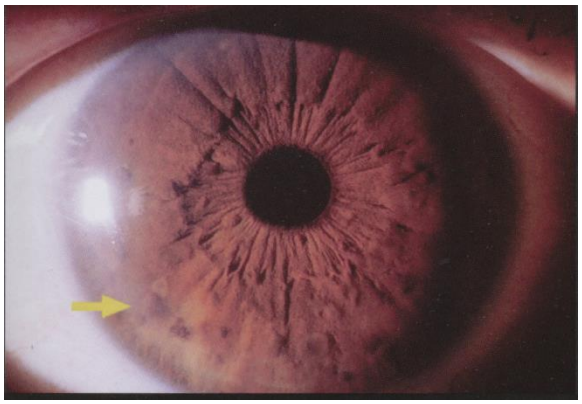
A. she has neurofibromatosis because she has more than 6 cafe au lait spots
B. she possibly has neurofibromatosis because she is only 2 years old and other findings are likely to appear over time
C. she probably does not have neurofibromatosis because the family history is negative for neurofibromatosis
D. she probably does not have neurofibromatosis because she has no cutaneous findings other than the spots
E. she probably has neurofibromatosis because she has 18 spots, which is significantly greater than the minimum number of 6

Item 68 TE Response: B

The young girl described in the vignette has 18 or more cafe au lait spots (CLSs) greater than 0.5 cm in diameter. While this is a significant finding suggestive of neurofibromatosis type 1 (NFI), this diagnosis cannot be confirmed without a second clinical finding. Because she is only 2 years old, if she has NF1, there is a good chance that additional clinical features will appear over time. For example, axillary and inguinal freckles may appear for the first time in older children and neurofibromas may not appear until preadolescence through adulthood. However, there are individuals with many CLSs who never develop a second sign of NF1 and are eventually cleared after appropriate investigations. While 6 is the minimum number to count as a clue for the diagnosis of NF1, greater than 6 CLSs does not increase the likelihood of an eventual diagnosis. A second sign, including: axillary or inguinal freckles (Item C68A, page C-55), 1 plexiform or 2 simple neurofibromas, an optic nerve glioma, Lisch nodules (Item C68B, page C-55), sphenoid bone dysplasia, a bony pseudarthrosis, or a first-degree relative with NF 1; must be present for a diagnosis of NF I to be made in any patient who has 6 or more CLSs. A family history that is positive for NF1 is quite helpful, especially in confirming a diagnosis in an infant or young child (eg, the girl in the vignette) who has only CLSs. However, approximately 50% of individuals with NF1 are the first person in their family with this condition. In such cases, the gene mutation occurs as a result of a de novo event at the time of conception. Lisch nodules (ie, benign iris freckles) are almost never present in young children but are seen in most patients who have NF1 after 10 years of age.



ITEM (68A: Axillary freckling in a patient with neurofibromatosis type 1.



ITEM C68B: Lisch nodules (arrow) are hamartomas of the iris.

Children, such as this girl, must be monitored closely for other signs of NF1, including axillary or inguinal freckles, optic nerve gliomas, and bony malformations such as thinning or curving of a long bone (eg, the tibia). Because clinically significant optic nerve gliomas most often present before age 5 years, a careful eye examination by an experienced pediatric ophthalmologist may be critical to assess the optic nerves and visual acuity. Regular eye examinations may permit earlier identification of optic nerve gliomas in young children who cannot adequately describe visual difficulties, thereby allowing for prompt treatment and improved visual outcomes. Macrocephaly is also quite common in infants, children, and adults who have NF1 but may not necessitate head imaging studies unless measurements are sequentially crossing percentiles or are associated with neurologic symptoms. Younger children should also be assessed for the need for services, such as speech, occupational, or physical therapies. Children and teenagers who have NF1 must be monitored closely for developmental problems because of an increased risk for learning disabilities and attention difficulties. Children and teenagers must also be monitored closely for scoliosis, since the incidence in this population is quite high and scoliosis may be rapidly progressive in patients who have NF1.

More recently, molecular diagnostic testing has proven helpful in confirming a diagnosis of NF1 in patients with only a single finding and for family planning purposes. Neurofibromin gene sequencing, however, is not generally available through large commercial laboratories and must be performed at one of several genetic specialty laboratories. As opposed to older methodologies, gene sequencing has a mutation detection rate of about 95%. However, a negative molecular genetic test does not exclude a diagnosis of NFI. Patients should still be monitored for additional features of NF1. If by the age of 10 years an individual has no eye findings (including Lisch nodules), axillary or inguinal freckles, neurofibromas, or other characteristic features, the diagnosis of NF 1 is unlikely.

PREP Pearls

- In order to confirm a diagnosis of NF1, at least 2 of the following clinical features must be present: axillary or inguinal freckles, 6 or more cafe au lait spots, neurofibromas, an optic nerve glioma, Lisch nodules, specific bony lesions, or a first-degree relative with NF1.
- Molecular testing can be helpful in confirming a diagnosis of NF1 in some cases and may be useful for family planning purposes.

American Board of Pediatrics Content Specification(s):

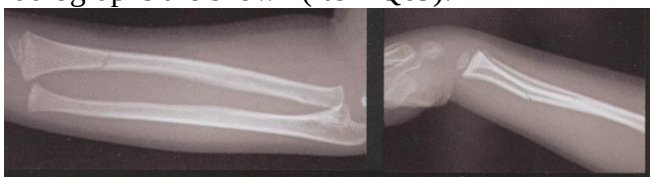
- Know the clinical features of neurofibromatosis

<u>Suggested Reading:</u>

- Hersh JH. Health supervision for children with neurofibromatosis. Pediatrics. 2008;121(3):633-642. doi:10.1542/peds.2007-3364

Item 69

An 8-year-old girl presents for evaluation shortly after falling down her front porch steps and landing on her outstretched left arm. A complete physical examination is remarkable only for mild swelling and tenderness over the midportion of her left forearm. Arm radiographs are shown (Item Q69).



Of the following, the MOST likely diagnosis is a

A. buckle fracture
B. greenstick fracture
C. metaphyseal chip fracture
D. Salter–Harris type I fracture
E. spiral fracture

Item 69 Preferred Response: B

Anatomical and physiologic differences in the musculoskeletal systems of children differ considerably from those seen in adults. An understanding of these differences and of the various fracture patterns commonly seen in children is crucial for pediatric practitioners to accurately diagnose and treat orthopedic injuries.

The girl described in the vignette has sustained an incomplete fracture occurring at the diaphyseal–metaphyseal junction of her left radius, with the cortex remaining intact on one side. This type of fracture is commonly called a greenstick fracture. Greenstick fractures occur because of the pliable nature of children's bones, which results in bowing rather than a complete disruption of the bony cortex. Because only one side of the bony cortex is disrupted, the appearance of the injured bone on plain radiographs often resembles an immature or "green" tree branch that breaks in a similar manner when bent. The typical mechanism of a greenstick fracture is a fall onto an outstretched hand. Greenstick-type injuries are the most common fracture pattern in children, accounting for up to half of fractures in children before 12 years of age. Angulation and rotation are common, but these fractures are not displaced because the bone ends are not separated. Greenstick fractures with angulation of greater than 15° may require closed reduction, with immobilization in a forearm sugar-tong splint and orthopedic follow-up. Radiographs should be repeated within 7 to 10 days to ensure that the reduction has been maintained. The long-term prognosis for patients with greenstick fractures is generally good; complications are rare in children because of the remarkable capacity of their bones to heal and remodel rapidly.

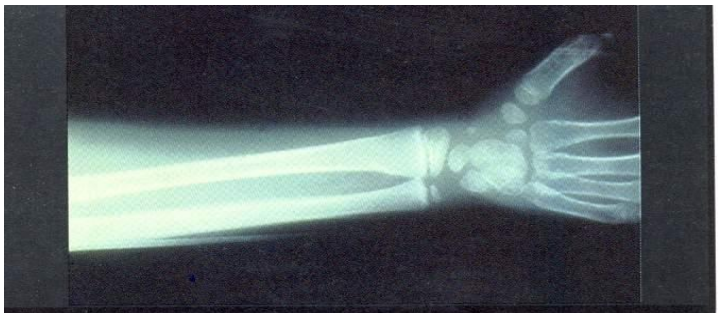
Buckle fractures, also known as torus fractures, are also common fractures sustained by young children. These fractures occur in the metaphyseal region of the bone due to a compressive load, typically from a fall onto an outstretched hand. The cortex of the bone buckles in a small area but is not disrupted, resulting in a stable fracture pattern (Item C69A). Torus fractures are usually nondisplaced and may be difficult to identify on radiographs because the findings may be subtle. These fractures may be immobilized in a short-arm volar splint, and patients should be referred for outpatient orthopedic follow-up within 5 days.

The Salter-Harris classification system describes various fractures that involve the epiphysis or growth plate. Growth plate injuries constitute nearly 20% of all pediatric skeletal injuries. The Salter-Harris system for classifying physeal injuries is based on the extent of involvement of the physis, epiphysis, and joint (Item C69B). Physeal damage due to trauma may disrupt bone growth and carries the potential for limb length discrepancies at the injured site from 1% to 10% of the time. A nondisplaced Salter-Harris type I fracture indicates bony injury across the physis only, with no fracture through the adjacent metaphyseal or epiphyseal regions. These types of fractures generally heal well with few complications. When suspected, Salter-Harris type 1 fractures should be immobilized by the pediatric practitioner, and affected children should be referred for orthopedic follow-up.

Spiral fractures run in a diagonal direction down the long axis of a bone. True spiral fractures arise from a severe rotational force that causes the bone to splinter, disrupting it

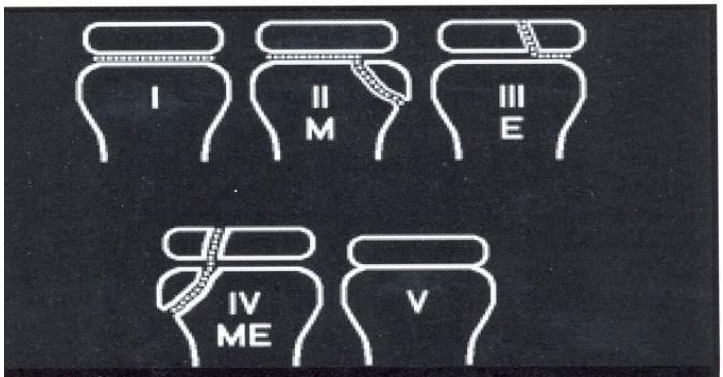
in a characteristic pattern that involves a fracture line that travels in 2 different oblique directions (Item C69C, page C-57). Although spiral fractures may occur as a result of unintentional injury, they may also arise from abusive trauma. Spiral fractures in nonambulating children or those occurring in patients where the injury identified is inconsistent with the reported history or developmental stage of the child, should raise the clinician's suspicion for child abuse.

Metaphyseal chip fractures, also known as corner or bucket-handle fractures, occur when a child's extremity is pulled or twisted forcibly or when the child is shaken. These fractures are frequently bilateral and result from periosteal avulsion of bone and cartilage secondary to violent twisting forces or downward pull on an extremity. A chip of bone or larger bucket-handle fragment may be present on plain radiographs (Item C691), page C-57). Children with metaphyseal corner fractures are often asymptomatic. These fractures are virtually diagnostic of child abuse. When metaphyseal chip fractures are diagnosed, affected children should receive a comprehensive evaluation for abuse.



Courtesy of M. Rimsza

ITEM C69A: Buckle fracture.



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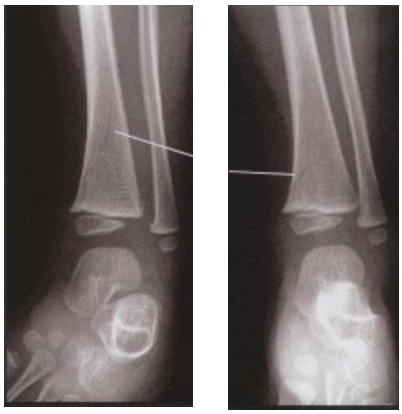
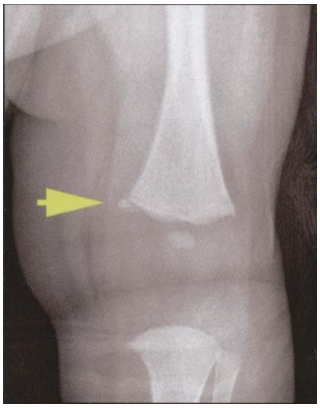
ITEM C69B-1: Salter-Harris classification system for fractures of the growth plate. See Item C69B-2 for description of fractures, treatment, and prognosis. M=metaphysis, E=epiphysis

Salter-Harris Type	Description	Treatment	Prognosis
I	Fracture through the physis	Splint or cast immobilization, rest, and ice	Excellent
II	Fracture through a portion of the physis extending into the metaphysis	Splint or cast immobilization, rest, and ice	Excellent
III	Fracture through the physis extending into the epiphysis	Reduction to achieve anatomic alignment, operative intervention may be necessary, cast immobilization, non-weightbearing	Moderate risk for long -term deformity due to physeal growth disturbance
IV	Facture crossing the metaphysis, physis, and epiphysis	Reduction to achieve anatomic alignment, operative intervention may be necessary, cast immobilization, non-weightbearing	Significant risk for deformity due to physeal growth arrest
V	Physeal crush injury	Cast immobilization, nonweightbearing, close orthopedic follow-up	Significant risk for deformity due to physeal growth arrest

ITEM C69B-2: Salter-Harris Fractures: Description, Management, and Prognosis.



ITEM C69C: Oblique (left) and anteroposterior (right) views of the distal tibia reveal a nondisplaced spiral fracture.



ITEM C69D: Metaphyseal corner (chip) fracture. May be seen in victims of nonaccidental trauma.

PREP Pearls

- Greenstick-type fractures account for up to half of the fractures in children before 12 years of age.
- Greenstick fractures with angulation of greater than 15° may require closed reduction, with immobilization in a forearm sugar-tong splint and orthopedic follow-up.
- Complications of greenstick fractures are rare in children because of the capacity of their bones to rapidly heal and remodel.

American Board of Pediatrics Content Specification (s):

- Identify a greenstick fracture

Suggested Reading:

- Bachman C, Santora S. Musculoskeletal trauma. In: Fleisher GR, Ludwig S, eds. Textbook of Pediatric Emergency Medicine. 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2010:1335-1375
- Chasm RM, Swencki SA. Pediatric orthopedic emergencies. Emerg Med Clin NAm 2010;28:907-926. doi:10.1016/j.emc.2010.06.003
- Dinolfo EA. Fractures. Pediatr Rev. 2004;25:218. doi:10.1542/pir.25-6-218
- Scherl SA, Endom EE. Orthopedic aspects of child abuse. UptoDate. Available online only for subscription

Item 70

A 20-month-old girl is being evaluated for low-grade fever and upper respiratory symptoms that have been present for 3 days. In addition, her parents noted drainage from the right ear this morning. This is her first episode of ear drainage since surgery. The child had tympanostomy tubes placed 3 months ago for persistent middle ear effusion with conductive hearing loss following acute otitis media. She had 3 episodes of otitis media prior to the surgery and has had no serious systemic infections. She attends child care and is fully immunized. On physical examination, you note purulent drainage from the right tympanostomy tube.

Of the following, the BEST next step in management is

A. culture and sensitivity of the drainage
B. evaluation for immunodeficiency
C. oral antimicrobial therapy
D. topical antifungal therapy
E. topical steroid therapy

Item 70 Preferred Response: C

For patients who have undergone tympanostomy tube placement, an occasional bout of otorrhea is not uncommon. Frequently, these episodes are temporally related to an upper respiratory tract infection, as in the girl described in the vignette. Acute tympanostomy tube otorrhea in young children is usually caused by the pathogens that cause acute otitis media (Streptococcus pneumoniae, Moraxella catarrhalis, and Haemophilus influenzae). Chronic suppurative otitis media is defined as discharge through a perforated tympanic membrane that persists for more than 6 weeks despite appropriate treatment. The most common pathogens isolated in these cases are Pseudomonas aeruginosa, Staphylococcus aureus, enteric gram-negative bacilli or anaerobes.

Initiating antimicrobial therapy is the next best step in management. Oral antibiotics may be preferred in a young child (<2 years of age), patients with fever and certainly in patients with known immunodeficiencies. Oral antibiotics may be chosen to improve delivery of therapy when there are copious amounts of purulent drainage; clearance of the external canal before instillation of topical antibiotics may prove challenging. Topical antibiotics allow for delivery of a high concentration of drug at the site of the infection without systemic adverse effects, so they may be preferred over oral antibiotics, depending on the age of the patient and severity of the illness. Topical quinolone antibiotics are the treatment of choice since they are the most effective and least ototoxic. Culture and sensitivity of the drainage is not recommended at this time; however, it is warranted if antibiotic therapy fails to resolve the otorrhea. Any specimen collected must be obtained from active drainage, from the patent tube or tympanic membrane perforation, and not from debris present in the external ear canal.

Children with refractory tympanostomy tube otorrhea or chronic suppurative otitis media with drainage should be referred to their otolaryngologist. Evaluation for immunodeficiency may be considered in select cases. Next steps in therapy should be based on results of culture and sensitivities of the purulent drainage to specifically address potential drug resistance or fungal overgrowth. The otolaryngologist will also be able to examine carefully for the presence of granulation tissue, which would be an indication for the use of topical steroids, or a cholesteatoma, which requires surgical intervention.

PREP Pearls

- Acute tympanostomy tube otorrhea in young children is usually caused by the same pathogens that cause acute otitis media.
- Antimicrobial therapy, oral or topical, is the first step in the management of acute otorrhea.
- Children with refractory otorrhea should be referred to their otolaryngologist for further evaluation.

American Board of Pediatrics Content Specification(s):

- Know the causes of ear drainage in a child with a perforated tympanic membrane or a tympanostomy tube

Suggested Reading:

- Baum ED. Tonsillectomy and adenoidectomy and myringotomy with tube insertion. Pediatr Rev. 2010;31:417-426. doi:10.1542/pir.31-10-417
- Gould JM, Matz PS. Otitis media. Pediatr Rev. 2010;31:102-116. doi:10.1542/pir.31-3-102
- Isaacson GC. Prevention and management of tympanostomy tube otorrhea in children. UptoDate. Available online only for subscription
- Kerschner JE. Otitis media: tube otorrhea. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:2208-2209
- Lieberthal AS, Carroll AE, Chonmaitree T, et al. American Academy of Pediatrics Clinical Practice Guideline. The Diagnosis and Management of Acute Otitis Media. Pediatrics. 2013; 131 (3):e964-e999doi: 10.1542/ peds.2012-3488

Item 71

A 9-year-old girl is seen in the emergency department for an asthma exacerbation. She received 3 breathing treatments at home, 20 minutes apart, before coming to the emergency department. On physical examination, she has a respiratory rate of 40 breaths/min, an oxygen saturation of 92% on room air, intercostal and suprasternal retractions, and bilateral expiratory wheezes. Her peak expiratory flow is 40% of the predicted value. You administer a nebulized albuterol treatment.

Of the following, the best NEXT step in the management of this patient is to

A. add ipratropium bromide to the nebulized albuterol
B. observe her to assess response to albuterol
C. order heliox administration
D. order a loading dose of an oral corticosteroid
E. order subcutaneous terbutaline

Item 71 Preferred Response: D

The child described in the vignette has an acute asthma exacerbation with significant airway obstruction that has not responded adequately to repeated short-acting β2-agonist (SABA) treatments. The best next step is to administer oral glucocorticoids. The goals of therapy for acute severe asthma include rapid reversal of airflow obstruction, correction of hypoxemia or severe hypercapnia, and reduction of likelihood of recurrence. The 2007 National Heart, Lung, and Blood Institute guidelines recommend that systemic glucocorticoids be given to patients who have moderate to severe exacerbations and do not respond completely to initial SABA therapy. corticosteroids are potent anti-inflammatory agents that can reduce bronchial hyperreactivity, hasten the resolution of airflow obstruction and edema, and improve lung function. They also prevent the late asthmatic response, attenuate the rate of relapse, and may reduce hospitalizations.

If this child does not respond to the albuterol treatment given in the emergency department, a trial of ipratropium bromide may be considered. Because this child has already received 3 albuterol treatments at home, in this situation, oral steroids should be administered before the trial of ipratropium bromide. The National Heart, Lung, and Blood Institute guidelines recommend the addition of ipratropium bromide be considered with each of the first 3 albuterol treatments, or with the second and third treatments, in children with moderate to severe asthma exacerbations. Terbutaline is an injectable β-agonist and may be used if inhaled albuterol is not available, such as by emergency medical services personnel. The use of terbutaline would not provide additional benefit because the mechanism of action is similar to albuterol and the child is able to use the nebulizer device appropriately. Adjunct treatments (magnesium sulfate or heliox) are indicated to decrease the likelihood of intubation only if the child fails to respond to the first-line treatments (SABA, oxygen, systemic steroids, and trial of ipratropium bromide).

PREP Pearls

- Systemic glucocorticoids, typically via the oral route, should be given to patients who have moderate to severe exacerbations and do not respond completely to initial short-acting β-agonist therapy
- The addition of ipratropium bromide may be considered with each of the first 3 albuterol treatments, or with the second and third treatments, in children with moderate to severe asthma exacerbations.

American Board of Pediatrics Content Specification(s):

- Plan the most appropriate treatment for a patient with an acute exacerbation of asthma that is severe and unresponsive to adrenergic agonist therapy (ie, systemic corticosteroids)

Suggested Reading:

- National Asthma E&cation and Prevention Program. Expert Panel Report 3 (EPR-3): guidelines for the diagnosis and management of asthma—summary report 2007 [published correction appears in JAllergy Clin Immunol. 2008;121(6):1330]. JAllergy Clin Immunol. 2007;120(5 suppl):S94-S138. doi:10.1016/j.jaci.2007.09.029
- Rowe BH, Spooner C, Ducharme FM, BretzlafFJA, Bota GW. Early emergency department treatment of acute asthma with systemic corticosteroids. Cochrane Database Syst Rev 2001;(I):CD002178. doi:10.1002/14651858.CDO02178

Item 72

A 1-year-old with genitourinary malformations recently underwent corrective urological surgery and was discharged home in stable condition with an indwelling urinary catheter. The patient presents 10 days after discharge with fever and vomiting. Physical examination is significant only for a febrile infant (40.1°C) with mild dehydration. The urine is cloudy, and a spot urine test strip analysis shows a pH of 6.0, specific gravity of 1.040, 4+ leukocyte esterase, and no nitrites, blood, or protein. The patient is admitted to the hospital and parenteral antibiotics are started.

Of the following, the MOST appropriate empiric antibiotic choice for this patient is intravenous

A. ampicillin
B. ampicillin and ceftriaxone
C. ceftriaxone
D. cefuroxime and gentamicin
E. gentamicin

Item 72 TE Preferred Response: B

Early treatment of patients with suspected pyelonephritis is important to decrease the risk of renal damage. Children presenting with fever (especially >39°C [102.2°F] or for >48 hours), appearing ill at presentation, having costovertebral angle tenderness, immune deficiency, or a known urologic abnormality are at increased risk for renal scarring if the urinary tract infection (UTI) is not promptly treated.
The choice of appropriate empiric antibiotic therapy before urine culture results become available is based on the individual risk factors for UTI and the knowledge of the most common organisms associated with UTI and their prevalent sensitivities.

Escherichia coli is the most common cause of community-acquired UTI. However, the patient in the vignette has an indwelling catheter, which puts him at an increased risk for UTI with Enterococcus. (Enterococci are most commonly isolated from the urinary tract and there is increased risk for enterococcal UTI associated with catheterization, instrumentation, and obstruction.) Also, the urine dipstick test was negative for nitrites, thus increasing the possibility of enterococcal infection. The appropriate antibiotic therapy before urine culture and sensitivity results are available should include coverage for both E coli and Enterococcus in this patient. Third- generation cephalosporins (cefotaxime, ceftriaxone) and aminoglycosides (gentamicin) are appropriate first-line agents for empiric treatment of UTI in children. However, these drugs do not treat enterococcal infections effectively and amoxicillin or ampicillin should be added to the initial treatment.

Management in this case would include consideration of catheter removal after consulting with the operating surgeon. Removal of the urinary catheter has been shown to improve recovery and decrease colonization in enterococcal infections.

PREP Pearls

- Prompt treatment of patient suspected of having urinary tract infection (UTI) with appropriate empiric antibiotics is important.
- Enterococcal UTI should be suspected in children with indwelling catheters and urine dipstick analysis showing negative nitrites.
- Empiric antibiotics in patients suspected of having enterococcal UTI should include a combination of ampicillin and third-generation cephalosporin or aminoglycoside.

American Board of Pediatrics Content Specification(s):

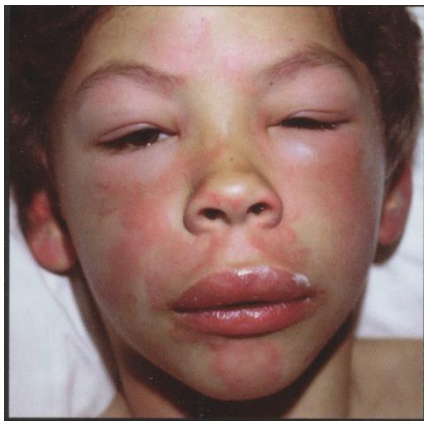
- Know the appropriate initial antimicrobial drugs for acute pyelonephritis before urine culture results area available

Suggested Reading:

- American Academic of Pediatrics, Subcommittee on Urinary Tract Infection, Steering Committee on Quality Improvement and Management. Urinary tract infection: clinical practice guideline for the diagnosis and management of the initial UTI in febrile infants and children 2 to 24 months. Pediatrics. 2011;128(3);595-610. doi: 10.1542/peds.2011-1330
- Feld LG, Mattoo TK. Urinary tract infections and vesicoureteral reflux in infants and children. Pediatr Rev. 2010;31(11):451-463. doi: 10.1542/pir. 31-11-451
- Haslam DB. Enterococcus. In: Kliegman RM, Stanton BF, St Geme JW III, Scher NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; ; 2011:928.e2-928.e5
- Ismaili K, Wising KM, Lolin K, et al. Characteristics of first urinary tract infection with fever in children: a prospective clinical and imaging study. Pediatr Infect Dis J. 2011;30(5):371-374. doi:10.1097/INF.Ob013e318204dcf3

Item 73

A 10-year-old boy has a fever, rash, and facial swelling. Just before the onset of these symptoms, he had a 4-day period of low-grade fever and cough, for which he took an over-the-counter cough medication. On physical examination, his temperature is 38.5°C, heart rate is 85 beats/min, and respiratory rate is 18 breaths/min. He has nonpruritic, erythematous macules and patches on his face, chest, arms, and back, as well as facial swelling (Item Q73). He has mild nasal congestion, and auscultation of his lungs reveals coarse scattered rales bilaterally. The remainder of his physical examination findings is normal.



Of the following, the MOST likely cause of the boy's illness is

A. adverse drug effect
B. group A Streptococcus infection
C. herpes simplex virus infection
D. influenza virus infection
E. Mycoplasma pneumoniae infection

Item 73 Preferred Response: E

The boy described in the vignette has facial swelling and a rash consistent with erythema multiforme that developed shortly after the onset of an illness characterized by nonproductive cough and congestion with low-grade fever. The most likely cause of these findings is infection due to Mycoplasma pneumoniae. Mycoplasma is a common cause of upper respiratory tract infection and bronchitis in school-aged children. The fever associated with Mycoplasma infection is usually low grade, and the cough is nonproductive. Otitis media, pharyngitis, malaise, and headache also can occur. Approximately 10% of school-age children with Mycoplasma infection will develop pneumonia; 10% of these children can develop a maculopapular rash. However, polymorphous mucocutaneous eruptions are uncommon and can be mild (eg, erythema multiforme minor as described in the vignette) to severe (eg, Stevens–Johnson syndrome). Other unusual extrapulmonary manifestations of Mycoplasma infection include hemolytic anemia, arthritis, carditis, or nervous system disease (eg, encephalitis, aseptic meningitis, cerebellar ataxia, transverse myelitis, and peripheral neuropathy).

Erythema multiforme (EM) is an immune-mediated skin condition characterized by erythematous and edematous papules or targetlike lesions. EM minor consists of skin eruptions with minimal or no mucosal involvement, whereas EM major involves the mucous membranes. More extensive blistering or mucous membrane erosions are seen in Stevens–Johnson syndrome. In children, infections are the most common cause of EM. Herpes simplex virus (HSV) is the most common cause of EM in young adults. EM minor can be triggered by subclinical HSV infection, or oral HSV lesions can be present. The presence of cough, congestion, and low-grade fever and the absence of oral lesions in the child described in the vignette better support Mycoplasma infection as the cause of EM. Influenza virus infection is a less likely cause of EM because of the mild nature of the child's respiratory tract illness and is generally a much less common cause of EM than is Mycoplasma. Infection due to group A Streptococcus has been associated with the development of EM, but the presence of cough and congestion, as well as the absence of oropharyngeal disease, in the patient described in the vignette argues against this cause. Finally, certain medications (eg, nonsteroidal anti-inflammatory drugs, sulfonamides, antiepileptics, and antibiotics) have been associated with EM, but over-the-counter cough suppressants are uncommon triggers.

PREP Pearls

- Mycoplasma infection is a cause of erythema multiforme.
- Extrapulmonary manifestations (eg, arthritis, polymorphous skin eruptions, hemolytic anemia, carditis, and nervous system disease) of Mycoplasma infection are uncommon.

American Board of Pediatrics Content Specification(s):

- Identify the extrapulmonary manifestations (eg, pharyngitis, rash, Stevens-Johnson syndrome, hemolytic anemia, arthritis, CNS disease) of a Mycoplasma infection

Suggested Reading:

- American Academy of Pediatrics. Mycoplasma pneumoniae and other Mycoplasma species infections. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:518-521
- Zaleznik DF, Vallejo JG. Mycoplasma pneumoniae infection in children. UptoDate. Available online only for subscription

Item 74

An adolescent girl is being treated for a Chlamydia infection that was discovered on routine screening. She is asymptomatic and believes she acquired the infection from a previous partner. After addressing antibiotic treatment for the girl and her current partner, you discuss prevention of future infections.

Of the following, the MOST effective prevention message for her at this time is to

A. begin and consistently use hormonal contraception
B. douche after sexual intercourse
C. maintain a monogamous relationship
D. undergo frequent testing for sexually transmitted infections
E. use condoms consistently

Item 74 I-C Preferred Response: E

Consistent and correct use of male condoms remains the most effective contraceptive method to prevent sexually transmitted infections (STIs). All barrier methods are important in prevention of STIs, but those other than male condoms (eg, female condoms and diaphragms) do not have good compliance rates. Male and female condoms should not be used together because slippage or tears are likely to occur as a result of friction. Nonoxynol-9 (N-9), the most commonly used spermicide, is present in the lubricant film in approximately 45% of condoms. It is not effective as a microbicide and offers no protection against human immunodeficiency virus (HIV), gonorrhea, or chlamydia. Concerns have been raised that N-9, the active ingredient in most contraceptive creams, jellies, foams, gel, film and suppositories and often used alone as a contraceptive by women, may actually increase the risk of HIV and human papillomavirus infection as a result of the vaginal irritation it causes when used very frequently.

Exploring the option of abstinence is important for every patient, especially if the desire for sexual activity rests mainly with the partner. However, adolescents who are voluntarily sexually active in a noncoercive relationship are very unlikely to stop all activity. Therefore, abstinence would not be a realistic choice for most sexually active adolescents. Regular screening for asymptomatic infection is important but does not supersede primary prevention. It would be important to remind the girl that although she may strive to maintain a monogamous relationship it does not guarantee that her partner will do so. As a result, the safest course of action would be to use condoms consistently. Although hormonal contraceptives are more effective at pregnancy prevention than barrier methods, they do not offer effective protection against STIs. Although there has been some speculation that the thickening of the cervical mucous plug that occurs with hormonal contraception may prevent ascending spread of cervical infections, this has not been confirmed. In fact, the delay in maturation of the cervical transition zone that may result from hormonal contraceptive use can increase the risk of acquiring STIs, especially Chlamydia trachomatis. Sex steroids have been reported to have a direct effect on gene expression on CD4+ lymphocytes, but an increased risk of HIV acquisition has only been found in commercial sex workers using hormonal contraception. Use of the intrauterine device and long-acting progesterone methods of contraception has not been found to be associated with an increased risk of acquiring HIV or other STIs.

PREP Pearls

- Counseling a sexually active teenager requires a discussion of both contraception and prevention of STIs.
- Use of male condoms is the most effective contraceptive method used to prevent sexually transmitted infections.
- Frequent use of spermicides alone may increase the risk of HIV acquisition.

American Board of Pediatrics Content Specification (s):

- Know the relationship between contraceptive choice and the prevention of sexually transmitted diseases, including HIV/AIDS

Suggested Reading:

- Committee on Adolescence. Contraception and adolescents. Pediatrics. 2007;120:1135-1148. doi:10.1542/peds.2007-2535
- Heikinheimo 0, Liihteemndki P. Contraception and HIV infection in women. Hum Reprod Update. 2009;15:165-176
- Herold BC, Mesquita PM, Madan RP, Keller MJ. Female genital tract secretions and semen impact the development of microbicides for the prevention of HIV and other sexually transmitted infections. Am J Reprod Immunol. 2011;65:325-333. doi:10.1111/j.1600-0897.2010.00932.x
- O'Leary A. Are dual-method messages undermining STI/HIV prevention? Infect Dis Obstet Gynecol. 2011;2011:691210

Item 75

An 11-year-old girl presents to your office because of left knee pain. She was in her usual state of health until 1 month ago, when she began experiencing pain in her right knee that was associated with limitation in motion. This problem resolved, but she is now complaining of similar symptoms in her left knee. Her parents also are concerned because she occasionally complains of periumbilical abdominal pain and they have noticed a decrease in her appetite. The child has 4 to 6 bowel movements per week, which are occasionally loose but without blood or mucus. The family has recently started to purchase all of their groceries at a local organic food market, and they have increased their consumption of whole grains. On physical examination, you note that her height and weight percentiles have steadily decreased over the past 3 years from the 25th percentile to the 10th percentile. Physical examination demonstrates a well-appearing girl, and there is mild swelling of her left knee without warmth, redness, or limitation of range of motion. Abdominal examination shows mild, direct tenderness to palpation in the right lower quadrant and periumbilical regions. You obtain the following laboratory test results:

- Hemoglobin, 10.3 g/dL (103 g/L)
- White blood cell count, 7,500/µL (7.5×10^9/L)
- Erythrocyte sedimentation rate, 36 mm/h
- Aspartate aminotransferase, 24 U/L; reference range,<40 U/L
- Alanine aminotransferase 14 U/L; reference range,<30 U/L
- Albumin, 3.2 g/dL (32 g/L)

Of the following, the MOST appropriate next diagnostic test is

A. abdominal ultrasonography
B. antineutrophil cytoplasmic antibody (ANCA)
C. colonoscopy
D. small-bowel radiographic series
E. tissue transglutaminase antibody

Item 75 Preferred Response: D

The girl described in the vignette presents with symptoms that suggest a possible rheumatologic illness. However, a thorough history and physical examination demonstrate anorexia, a gradual falloff in her growth percentiles, and abdominal pain and tenderness. This constellation of signs and symptoms should immediately suggest a disorder that affects multiple organ systems. Laboratory studies that indicate anemia, hypoalbuminemia, and an ongoing inflammatory process (elevated erythrocyte sedimentation rate) strongly suggest a diagnosis of Crohn disease with significant small-bowel involvement. Accordingly, the next step in this evaluation should be to obtain an upper gastrointestinal tract radiographic series, with small-bowel follow-through.
Crohn disease (CD) is 1 of the 2 idiopathic illnesses (with ulcerative colitis) termed inflammatory bowel disease (IBD). In the past 20 years, IBD has emerged as a relatively common childhood disorder. Today, more than 1 million individuals in the United States have IBD and approximately 1 in 4 patients is younger than 20 years of age. With a peak incidence in childhood during the second decade of life, the clinical manifestations of IBD often occur during a critical period of growth and development. Symptoms directly related to intestinal disease activity include abdominal pain, vomiting (with upper gastrointestinal tract involvement), diarrhea, bleeding, weight loss or failure to gain weight (secondary both to anorexia and to increased energy expenditure), and malabsorption (in approximately 20% of patients with CD). However IBD, and especially CD, may cause a wide array of extraintestinal complications, affecting multiple tissue and organ systems, and can masquerade as other disorders. Thus, pediatric gastroenterologists frequently see patients with IBD who are initially considered to have an endocrinopathy (resulting in failure to thrive), a rheumatologic illness (bone or joint involvement and unexplained fevers), or an infectious disorder (fever of unknown origin). Children with CD may present with diverse clinical disorders in association with active bowel disease (Item C75, page C-61). Rarely, these problems may present or persist when bowel disease is quiescent (eg, sclerosing cholangitis after colectomy for ulcerative colitis). Knowledge of the gastrointestinal and extraintestinal manifestations of IBD is essential for the primary care physician who is likely to encounter affected patients relatively early in the illness. Prompt and accurate diagnosis requires a high index of suspicion and will facilitate the earliest possible treatment necessary to minimize both short- and long-term complications.

As part of the IBD evaluation, other investigations should be considered. The antineutrophil cytoplasmic antibody (pANCA) and the anti-Saccharomyces cerevisiae antibody (ASCA) test are useful in differentiating between CD (ASCA often positive, pANCA negative) and ulcerative colitis (pANCA often positive, ASCA negative). In conjunction with other autoantibody titers, these tests may also be used to predict the likelihood of IBD-related complications. However, the IBD serologies should not be used as a means of establishing an IBD diagnosis because many patients with CD, particularly children, will demonstrate a negative test results. A colonoscopy should be performed in this girl as part of her IBD evaluation. Assessment of disease involvement (ie, continuous vs skip areas) will help differentiate CD from ulcerative colitis. In addition, the identification of granulomata on biopsy specimens will confirm a Crohn diagnosis, although, this finding is present in only approximately 25% of patients undergoing

colonoscopy. Because this child's symptoms (hypoalbuminemia, growth disturbance, joint involvement, and absence of gross bleeding) suggest small bowel involvement, the radiographic series remains the first-choice diagnostic study. Much like CD, celiac disease may also masquerade as other illnesses, and an evaluation for this disorder (including a tissue transglutaminase antibody) will be indicated if the IBD evaluation is negative. Abdominal ultrasonography, however, is not part of the standard testing for either of these illnesses.

Item C75. Extraintestinal Manifestations of Inflammatory Bowel Disease

Site	Manifestations
Skin	Erythema nodosum, pyoderma gangrenosum, metastatic Crohn disease
Liver	Steatosis, aminotransferase elevations, sclerosing cholangitis, cholelithiasis, acalculous cholecystitis, Budd–Chiari syndrome
Bones	Osteopenia, aseptic necrosis
Joints	Arthralgias, arthritis, ankylosing spondylitis, sacroiliitis
Eyes	Uveitis, episcleritis, keratitis
Kidneys	Nephrolithiasis, obstructive hydronephrosis, enterovesicular fistula, nephritis, amyloidosis
Blood	Nutritional anemia (iron, folate, vitamin 1312), autoimmune hemolytic anemia, thrombocytosis, thrombocytopenia
Vascular	Hypercoagulability (thrombosis, thrombo-phlebitis, portal vein thrombosis)
Pancreas	Pancreatitis
Other inflammatory'	Anorexia, growth delay, pubertal delay, increased risk of colonic malignant tumor

Adapted from Hyams JS. Inflammatory bowel disease. *Pediatr Rev.* 2005;26:314-320. doi: 10. 1 542/pi r26-9-314.

PREP Pearls

- The peak incidence for inflammatory bowel disease is during the second decade of life,
- Poor growth and weight gain in previously healthy children, with or without GI symptoms, should alert you regarding the possibility of IBD, particularly

Crohn disease.3. IBD serologies are often negative early in the course of disease, and should not be used to establish or rule out an IBD diagnosis.

American Board of Pediatrics Content Specification(s):

- Know the clinical manifestations of Crohn disease

Suggested Reading:

Hendrickson BA, Gokhale R, Cho JH. Clinical aspects and pathophysiology of inflammatory bowel disease. Clin Microbiol Rev. 2002;15:80-91. doi:10.1128/CMR.15.1.79-94.2002

Hyams JS. Inflammatory bowel disease. Pediatr Rev. 2005;26:314-320. doi:10.1542/pir.26-9-314

Kim S, Ferry G. Inflammatory bowel diseases in children. Curr Probl Pediatr. 2002;32:103-132

Item 76

While examining a term neonate in the newborn nursery, it is noted that the red reflex is markedly less bright on one side than it is on the other. The remainder of the physical examination, including growth parameters, is normal. You have requested an ophthalmologic evaluation, but you are considering whether additional evaluation is needed.

Of the following, the information MOST likely to be helpful in determining the cause of this abnormality is that the

A. infant's father has a history of a cataract at birth
B. infant's mother is rubella nonimmune
C. infant's mother is being treated for hypothyroidism
D. maternal serum α-fetoprotein levels were elevated during the pregnancy
E. mother smoked an average of 4 cigarettes per day during the pregnancy

Item 76 TE S Preferred Response: A

The patient described in the vignette has findings characteristic of a cataract. Because this is frequently an autosomal dominant condition, a positive family history aids in the diagnosis. A cataract is a lens opacity, but some would restrict the definition to visually significant opacities. Cataracts may involve a number of locations within the lens, with those involving central structures having the greatest effect on visual development. The lens is made up of an outer capsule enclosing the cortex that in turn surrounds the nucleus of the lens. Y sutures are lens fibers that mark the boundary of the cortex and the nucleus. A slit lamp examination can help differentiate cataracts as subcapsular (anterior and posterior), cortical (punctate and lamellar), sutural, and membranous types. These locations can help identify the cause of cataracts.

Because early detection of congenital cataracts is necessary to preserve normal visual development, every newborn should have an examination of his or her red reflexes before being discharged from the nursery and at each routine health supervision visit thereafter. The clinician should examine each red reflex individually and both together (Bruckner test) using the ophthalmoscope at approximately 18 inches from the subject. Abnormal findings suggesting cataract include absent, dulled, or asymmetric red reflexes; a white or opaque reflex; and dark spots. In addition, structural abnormalities of the eyes (eg, coloboma) should raise the possibility of lens anomalies. Infants with these abnormal findings need immediate referral to a pediatric ophthalmologist so that early treatment can be initiated before the critical period of visual development has passed.
The causes of cataracts in children are myriad, however, approximately 60% of congenital cataracts are idiopathic and 10% to 25% are genetic. Cataracts may be associated with infections, metabolic disease, genetic conditions, structural eye anomalies, trauma, and drugs/toxins. Many metabolic disorders can lead to cataracts; galactosemia is one of the more common causes, and in this situation, the cataracts may be reversible with appropriate dietary control. The classic infectious cause was congenital rubella, but as that disease has decreased in frequency with immunization, other TORCH (toxoplasmosis, other infections, rubella, cytomegalovirus, herpes simplex virus) congenital infections as well as varicella zoster, influenza, and polio have been identified as causes. Genetic causes include chromosomal, single gene, and multisystem conditions. Common chromosomal causes include trisomy 21 and trisomy 13; a low α-fetoprotein level during pregnancy is a marker for trisomy 21 but an elevated α-fetoprotein level is not linked to cataracts.

As with the patient in the vignette, family history of congenital cataracts is a risk factor for cataracts in offspring because the most common Mendelian mode of transmission is autosomal dominant. Therefore, the offspring or siblings of a patient who has familial congenital cataracts should be evaluated by an ophthalmologist. WAGR syndrome (Wilms tumor, aniridia, genital abnormalities, and retardation) is an example of a multisystem condition associated with cataracts, and in addition to early ophthalmologic intervention, it requires frequent renal ultrasounds to detect Wilms tumor. Endocrine associations with cataracts include prenatal exposure to maternal hypoparathyroidism and history of chronic hypoglycemia or diabetes mellitus in the child. Neither maternal hypothyroidism nor tobacco use during pregnancy is a risk factor for congenital cataracts.

Prematurity is a risk factor, with low birthweight infants having a threefold to fourfold increase in the rate of congenital cataracts.
Most acquired cataracts in children are associated with trauma. Toxin exposure, particularly corticosteroid use, is another risk factor for developing cataracts at a later age.

PREP Pearls

- Every newborn should be examined for evidence of lens opacity before discharge from the nursery and at each health supervision visit thereafter.
- Physical examination findings suggestive of cataracts include absent, dulled, or asymmetric red reflexes; a white or opaque reflex; and dark spots.
- Cataracts can be genetic, infectious, metabolic, structural, traumatic, or drug/toxin-related. About 60% of congenital cataracts are idiopathic.
- The most common genetic transmission pattern for familial cataracts is autosomal dominant, and therefore, the offspring or siblings of a patient who has familial congenital cataracts should be evaluated by an ophthalmologist.

American Board of Pediatrics Content Specification(s):

- Understand the risk factors for the development of cataracts

Suggested Reading:

- American Academy of Pediatrics, Section on Ophthalmology. Red reflex examination in neonates, infants and children. Pediatrics. 2008;122:14011404. doi: 10.1542/peds.2008-2624
- Davenport DM, Patel AA. Cataracts. Pediatr Rev. 2011;32:82-83. doi: 10.1542/pir.32-2-82
- Olitsky SE, Hug D, Plummer LS, Stass-Isern M. Abnormalities of the lens. In: Kliegman RM, Stanton BMD, St Gema 1, Schor N, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Elsevier Saunders; 2011: 2169-2171
- Trumler AA. Evaluation of pediatric cataracts and systemic disorders. Curr Opin Ophthalmol. 2011; 22:365-379. doi:10.1097/ ICU.Ob013e32834994dc

Item 77

A 3-year-old boy is brought to the emergency department (ED) 30 minutes after ingesting one of his grandmother's medications. The boy's mother reports that the grandmother takes medications for depression and hypertension. During the first hour of being monitored in the ED, the boy becomes increasingly lethargic and develops mydriasis and a dry mouth. You notice on the cardiopulmonary monitor that his heart rate is 130 beats/min and he now appears to have a prolonged QRS duration. An electrocardiogram demonstrates a QRS duration of 110 milliseconds, right axis deviation, and prolonged PR and QT intervals.

Of the following, the MOST likely medication ingested by the patient is

A. amitriptyline
B. clonidine
C. fluoxetine
D. labetalol
E. nicardipine

Item 77 S Preferred Response: A

Tricyclic antidepressants (TCAs), such as amitriptyline and imipramine are widely used for treating depression, chronic pain, and attention-deficit disorders. Pediatric patients can be exposed to potential accidental or intentional ingestions of their own, their siblings, or an adult relative's medications. Tricyclic antidepressants have strong anticholinergic activity and can produce symptoms such as dry mouth, blurred vision, tachycardia, urinary retention, constipation, dizziness, and vomiting. Overdoses of TCAs can produce other more serious side effects, including central nervous system abnormalities (irritability, lethargy, coma, and seizures), respiratory depression, hypotension, and cardiac dysrhythmias. Symptoms may occur as early as 30 minutes and are usually seen within 6 hours of ingestion.

Electrocardiographic changes (secondary to sodium channel blockade) are usually seen within 6 hours of ingestion and include widened QRS interval, right axis deviation, prolonged PR interval (first-degree heart block), abnormal T waves and ST segments, or atrioventricular block. These changes may persist for days. Sinus tachycardia is the most common rhythm but more serious cardiac complications such as bradydysrhythmias, supraventricular tachycardia, ventricular tachycardia, and ventricular fibrillation may occur within 24 hours of ingestion. A QRS duration greater than 100 milliseconds and right axis deviation appear to be the strongest predictors of cardiac toxicity.

Clonidine overdoses are also of significant pediatric toxicology concern. Central nervous system depression, pinpoint pupils, hypotension, bradydysrhythmias, and respiratory depression are characteristic findings. The electrocardiographic changes seen in the patient in the vignette are not consistent with a clonidine overdose. Ingestion of selective serotonin reuptake inhibitors (SSRIs), such as fluoxetine, is associated with less toxicity compared to TCAs. Agitation, delirium, tachycardia, hyperthermia, and muscle rigidity may be seen, but again the electrocardiographic changes exhibited by the patient in the vignette are not consistent with an SSRI overdose. β-blocker overdoses, such as that caused by labetalol, can produce both central nervous system changes (lethargy, coma, and seizures) as well as electrocardiographic changes, including a widening of the QRS interval and prolongation of the PR interval. Dysrhythmias seen can include ventricular tachycardia and asystole. However, the characteristic anticholinergic symptoms and signs of TCA overdose would not be present. Overdoses of calcium channel blockers, such as nicardipine, cause myocardial depression and cardiac conduction changes, including PR interval prolongation and bradydysrhythmias. The anticholinergic and central nervous symptoms that are seen with TCA overdose are not seen in calcium channel poisoning.

PREP Pearls

- Pediatric patients can be exposed to accidental ingestions of their own, their siblings', or an adult relative's medications.
- Overdoses of tricyclic antidepressants (TCAs) can cause central nervous system abnormalities, respiratory depression, hypotension, and cardiac dysrhythmias.
- An electrocardiogram demonstrating a QRS duration greater than 100 milliseconds and right axis deviation appears to be the strongest predictor of cardiac toxicity in TCA overdoses.

American Board of Pediatrics Content Specification(s):

- Understand that a danger of tricyclic antidepressant treatment is accidental ingestion by siblings
- Understand that cardiac dysrhythmias may occur late after ingestion of tricyclic antidepressants

Suggested Reading:

- O'Donnell KA, Ewald MB. Poisonings. In: Kliegman RM, Stanton BMD, St Geme J, Schor N, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Elsevier Saunders; 2011: 264
- Traub SJ. Tricyclic antidepressant poisoning. UptoDate. Available online only for subscription

Item 78

A 17-year-old boy develops altered mental status and is rapidly brought to the emergency department. Four days ago, he complained of polyuria, polydipsia, and fatigue. He has lost 4.5 kg of body weight in the last week. Vital signs taken in the emergency department show a blood pressure of 146/53 mm Hg, pulse rate of 133 beats/min, and respiratory rate of 16 breaths/min. He is afebrile. His body mass index is 30. His current medications include olanzapine, 20 mg once daily, started a few weeks ago for schizophrenia, and sertraline and valproic acid, both of which he has taken for a long time. Initial laboratory evaluation shows a glucose level of 660 mg/dL (36.6 mmol/L), serum sodium of 161 mEq/L (161 mmol/L), potassium of 4.8 mEq/L (4.8 mmol/L), and bicarbonate of 23 mEq/L (23 mmol/L).

Of the following, the MOST likely diagnosis is

A. diabetic ketoacidosis
B. hyperglycemic hyperosmolar syndrome
C. maturity onset diabetes of the young
D. medication overdose
E. toxic ingestion

Item 78 S Preferred Response: B

The adolescent described in this vignette has symptoms of diabetes mellitus (polyuria, polydipsia, weight loss, and fatigue) and laboratory evidence of hyperglycemia. However, he also has hypernatremia and is not acidotic. These findings are most consistent with the diagnosis of hyperglycemic hyperosmolar syndrome (HHS). Although acidosis may occur in the setting of HHS, this complication is more characteristic of diabetic ketoacidosis. The patient's clinical presentation is not consistent with maturity-onset diabetes of the young, a milder form of inherited diabetes, or a specific toxic ingestion or medication overdose.

Hyperosmolar syndrome is the most severe presentation of type 2 diabetes and, although uncommon, has been reported in children. In almost all reported cases of HHS in children, there was no preexisting diabetes, and a precipitating factor, such as steroid or other medication use, is implicated as a possible trigger for the presentation. Labeling for atypical antipsychotic medications, such as olanzapine, includes a recommendation to perform fasting blood glucose testing at the initiation of, and periodically during, treatment because their use has been linked to an increased risk of type 2 diabetes, ketoacidosis, or hyperosmolar coma.

In addition to screening patients who are taking any medication that predisposes them to diabetes mellitus, current American Diabetes Association (ADA) guidelines recommend screening for type 2 diabetes mellitus starting at 10 years of age or at onset of puberty and every 3 years thereafter in children who are overweight (body mass index >85th percentile for age and sex, weight for height >85th percentile, or weight >120% of ideal for height) and have at least 2 risk factors for diabetes (family history, higher risk race or ethnicity, signs of insulin resistance, or maternal history of diabetes). Children who already have evidence of impaired glucose tolerance (fasting glucose, 100-125 mg/dL (5.6-6.9 mmol/L); 2-hour glucose on oral glucose tolerance test [OGTT], 140-199 mg/dL [7.8-11.0 mmol/L]) also need more frequent monitoring.

A fasting glucose level of 126 mg/dL (7.0 mmol/L) or an 2-hour glucose reading during OGTT of 200 mg/dL (11.1 mmol/L) or higher is diagnostic for diabetes mellitus. In some cases the fasting plasma glucose level will be normal, but the postprandial glucose level may be elevated, in which case an OGTT should be performed if there is a high index of suspicion for type 2 diabetes mellitus. In the absence of unequivocal symptoms, testing should be repeated to confirm the diagnosis because there are numerous causes of transient hyperglycemia. However, with unequivocal symptoms of diabetes mellitus, a random plasma glucose level of 200 mg/dL or higher confirms the diagnosis.

Recently, guidelines from the ADA have added hemoglobin A,, as a supplemental screening test for diabetes, with a result of 6.5% or greater considered diagnostic of diabetes mellitus in adults. Although very specific, this screening method is not very sensitive in children, and results should be interpreted with caution.

PREP Pearls

- Overweight children with at least 2 risk factors for type 2 diabetes mellitus should be screened starting at puberty and then every 3 years.
- Children with overt symptoms of diabetes mellitus or taking medications that put them at high risk of developing diabetes should have regular follow-up of glucose levels.

American Board of Pediatrics Content Specification(s):

- Plan appropriate screening tests for type 2 diabetes

Suggested Reading:

- American Diabetes Association. Antipsychotic medications and the risk of diabetes and cardiovascular disease. American Diabetes Association www.diabetes.org
- American Diabetes Association. Standards of medical care in diabetes-2012. Diabetes Care. 2012;3(suppl 1):Sll-S63. doi:10.2337/ dc12-s011
- Rosenbloom AL, Silverstein JH, Amemiya S, Zeitler P, Klingensmith GJ; International Society for Pediatric and Adolescent Diabetes. ISPAD Clinical Practice Consensus Guidelines 2006-2007. Type 2 diabetes mellitus in the child and adolescent. Pediatr Diabetes. 2008;9:512-526
- Rosenbloom AL. Hyperglycemic hyperosmolar state: an emerging pediatric problem. JPediatr. 2010;156:180-184. doi:10.1016/j. jpeds.2009.11.057
- Vehik K, Cuthbertson D, Boulware D, Beam CA, Rodriguez H, Legault L, Hyytinen M, Rewers MJ, Schatz DA, Krischer JP; the TEDDY, TRIGR, Diabetes Prevention Trial-Type 1, and Type 1 Diabetes TrialNet Natural History Study Groups. Performance of HbAlc as an early diagnostic indicator of type 1 diabetes in children and youth. Diabetes Care. 2012;35 :1821-1825. doi:10.2337/dcl2-0111

Item 79

A 16-year-old boy is brought to the emergency department with a 1-day history of illness characterized by a diffuse, erythematous rash, nausea and vomiting, 3 loose stools, marked abdominal pain, and increasing confusion. A week ago, he cut his hand climbing a fence but did not seek medical attention.

Physical examination reveals a temperature of 39.2°C; heart rate of 130 beats/min; blood pressure of 80/50 mm Hg; respiratory rate of 24 breaths/min; diffuse erythematous, blanching rash; supple neck without significant adenopathy; and a laceration on the right hand with surrounding erythema and edema. Head, eyes, ears, nose, and throat examination reveal injected sclerae and inflamed lips and tongue. Lungs are clear to auscultation. Cardiac auscultation reveals tachycardia without murmur, rub, or gallop. Upon neurologic examination, the boy is sleepy and difficult to arouse. The laboratory test results are as follows:

- White blood cell count, 14,400/µL (14.4 x 10^9/L), with 80% neutrophils, 8% band neutrophils, and 12% lymphocytes
- Hemoglobin, 11.0 g/dL (110 g/L)
- Hematocrit, 34.6% (0.34)
- Platelets, 86 x 10^3/µL (86 x10^9/L)
- Sodium, 132 mEq/L (132 mmol/L)
- Potassium, 4.6 mEq/L (4.6 mmol/L)
- Chloride, 106 mEq/L (106 mmol/L)
- Bicarbonate, 16 mEq/L (16 mmol/L)
- Blood urea nitrogen, 52 mg/dL (18.6 mmol/L)
- Creatinine, 2.3 mg/dL (203.3 4mol/L)
- Alanine aminotransferase, 128 U/L
- Aspartate aminotransferase, 211 U/L
- Total bilirubin, 2.2 mg/dL (37.6 µmol/L)
- Direct bilirubin, 0.6 mg/dL (10.3 µmol/L)

He is infused with 20 mL/kg of intravenous normal Saline

Of the following, the BEST initial therapy for this boy's condition is

A. cefazolin
B. dobutamine
C. immune globulin intravenous
D. pentoxifylline
E. vancomycin

Item 79 SBP Preferred Response: E

The findings described for the young man in the vignette are consistent with a diagnosis of toxic shock syndrome (TSS) as evidenced by his fever, hypotension, diffuse erythroderma, elevated liver enzymes, and renal dysfunction (Item C79). Fluid resuscitation to maintain venous return and cardiac filling is the most important initial step in the treatment of the patient with TSS.

Antimicrobial therapy to cover Staphylococcus aureus, including community-acquired methicillin-resistant strains (CA-MRSA) and group A Streptococcus would be an important part of the initial management. Of the choices listed, vancomycin provides the best coverage for these organisms. In serious infections such as this, addition of clindamycin or nafcillin or oxacillin might be warranted. Clindamycin inhibits protein synthesis, which might offer a theoretical advantage as part of the initial treatment regimen to decrease toxin production by the bacteria.

First-generation cephalosporins such as cefazolin do not provide coverage for CA-MRSA and would be inadequate, pending culture and sensitivity results. Blood pressure support with dobutamine might be needed, if fluid resuscitation does not adequately improve the patient's condition, but would be addressed after a trial of fluid replacement and institution of antibiotic therapy.

Immune globulin intravenous has been tried in the treatment of TSS but the data supporting its use are not definitive. However, if used it is only considered for cases refractory to other therapy after several hours, with an undrainable focus of infection or in the presence of persistent oliguria with pulmonary edema.

Pentoxifylline is a xanthine derivative that has been used to improve blood flow in certain conditions such as intermittent claudication. This drug inhibits the production of superantigens and certain cytokines including tumor necrosis factor a, interleukin (IL) 6, and ILI 1, which might be helpful in controlling aspects of the systemic inflammatory response triggered in TSS. It has not been studied in people for this condition and would not be a routine part of management in TSS.

PREP Pearls

- Toxic shock syndrome (TSS) presents as a multisystem disease with fever, hypotension, diffuse rash, and multiple organ involvement (eg, nausea, vomiting, renal involvement, hepatitis, central nervous system dysfunction, severe myalgias).
- Management of TSS involves monitoring blood pressure, extensive volume support, and supportive care.
- Antibiotic therapy in TSS is primarily aimed at eliminating the offending organism to prevent recurrences.
- Bacteremia (especially with group A Streptococcus) can be associated with TSS and should be treated appropriately.

American Board of Pediatrics Content Specification(s):

- Understand the management of a patient with staphylococcal toxic shock syndrome

Item C79. Clinical Definition of Toxic Shock Syndrome*

Fever (temperature >38.9°C)

Hypotension

Diffuse macular erythroderma followed by desquamation 1 to 2 weeks after onset, particularly on palms, soles, fingers, toes

Multisystem organ involvement (≥3) of the following:

1. Gastrointestinal: Vomiting or diarrhea at onset of illness
2. Muscular: Severe myalgia or creatinine phosphokinase >2 times normal
3. Mucous membranes: Vaginal, oropharyngeal, or conjunctival hyperemia
4. Renal: Serum urea nitrogen or creatinine concentration >2 times the upper limit of normal or urinalysis with >5 WBC/hpf in the absence of urinary tract infection
5. Hepatic: Total bilirubin, aspartate transaminase, or alanine transaminase >2 times upper limit of normal
6. Hematologic: Platelet count $\leq 100 \times 10^3/\mu L$ ($100 \times 10^9/L$)
7. Central nervous system: Disorientation or alterations in consciousness without focal neurologic signs in the absence of fever and hypotension

Abbreviations: HPF, high-power field; WBC, white blood cell.

*Adapted from Staphylococcal infections, in Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. *Red Book: 2012 Report of the Committee on Infectious Diseases*. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics.

Suggested Reading:

- American Academy of Pediatrics. Staphylococcal infections. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:653-658
- Jamart S, Denis 0, Deplano A, et al. Methicillin-resistant Staphyloccus aureus toxic shock syndrome. Emerg Infect Dis. 2005;11:636-637
- Lappin E, Ferguson AT. Gram-positive toxic shock syndromes. Lancet. 2009;9:281-290. doi:10.1016/Sl473-3099(09)70066-0

Item 80

A 3-year-old girl presents for a routine health supervision visit. She was recently diagnosed with juvenile idiopathic arthritis. Her mother asks why the rheumatologist has referred her to an ophthalmologist.

Of the following, the BEST indication for ophthalmology referral is

A. assessment for corneal abrasions
B. assessment of depth perception
C. evaluation for cataracts
D. screening for eye infections before starting a new treatment
E. screening for uveitis

Item 80 Preferred Response: E

The child described in this vignette should be screened for uveitis. Uveitis is a serious complication of juvenile idiopathic arthritis (JIA), previously known as juvenile rheumatoid arthritis. Uveitis results from chronic nongranulomatous inflammation of the anterior eye chamber and can affect the iris and ciliary body of the eye. The uveitis associated with JIA is usually clinically silent with an insidious onset. Risk factors for developing uveitis in JIA patients include JIA subtype, age at onset of disease, and antinuclear antibody (ANA) status. The highest risk group is female patients who have pauciarticular JIA (4 or fewer joints in the first 6 months of diagnosis) and are ANA positive and diagnosed at younger than 4 years of age. Screening guidelines based on risk have been developed for uveitis (Item C80, page C-65). The severity of uveitis does not correlate well with arthritis activity; therefore, the status of joint disease should not affect the frequency of screening. Complications of uveitis include band keratopathy, corneal clouding, cataracts, glaucoma, and loss of visual acuity (including blindness). Uveitis should be managed by an ophthalmologist in conjunction with a rheumatologist. Topical corticosteroids are often used to help manage disease; however, long-term use of these medications can have significant adverse effects. Disease-modifying agents, such as low-dose methotrexate, or biologic agents are often required to treat uveitis associated with JIA.

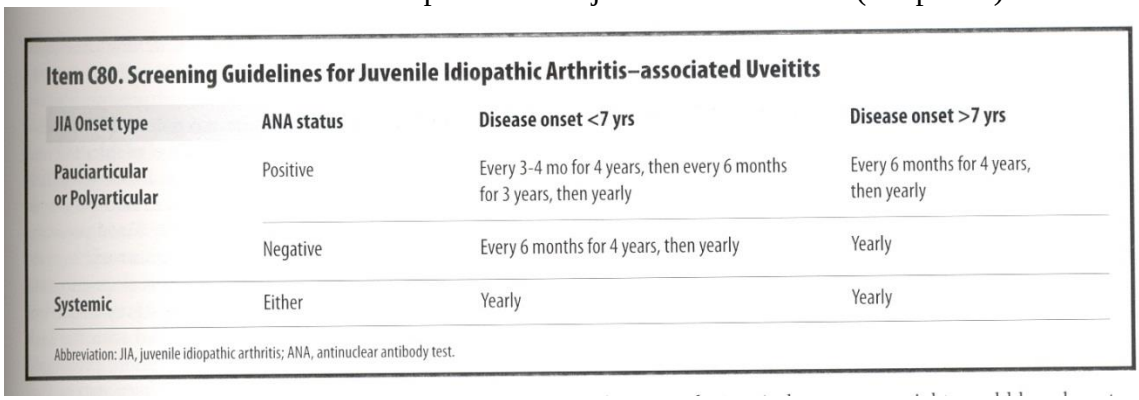
Juvenile idiopathic arthritis is not associated with problems of depth perception, eye infection, or corneal abrasion. Cataracts are a late complication of chronic uveitis or chronic ophthalmic steroid use. A young patient recently diagnosed would be unlikely to have this complication.

PREP Pearls

- Uveitis is associated with juvenile idiopathic arthritis (JIA) and is clinically silent.
- Frequency of screening for uveitis should be based on a patient's risk factors.
- The severity of uveitis does not correlate with the level of active joint inflammation in JIA.

American Board of Pediatrics Content Specification(s):

- Know the ocular complications of juvenile rheumatoid (idiopathic) arthritis



Item C80. Screening Guidelines for Juvenile Idiopathic Arthritis–associated Uveitits

JIA Onset type	ANA status	Disease onset <7 yrs	Disease onset >7 yrs
Pauciarticular or Polyarticular	Positive	Every 3-4 mo for 4 years, then every 6 months for 3 years, then yearly	Every 6 months for 4 years, then yearly
	Negative	Every 6 months for 4 years, then yearly	Yearly
Systemic	Either	Yearly	Yearly

Abbreviation: JIA, juvenile idiopathic arthritis; ANA, antinuclear antibody test.

Suggested Reading:

- Espinosa M, Gottlieb B. Juvenile idiopathic arthritis. Pediatr Rev. 2012;33:303-313. doi:10.1542/pir.33-7-303
- Goldmuntz EA, White PH. Juvenile idiopathic arthritis: a review for the pediatrician. Pediatr Rev! 2006;27(4):e24-e32. doi:10.1542/pir.27-4-e24
- Gowdie PJ, Tse SML. Juvenile idiopathic arthritis. Pediatr Clin N Am. 2012;59(2):301-327. doi:10.1016/j.pcl.2012.03.014

Item 81

A 15-year-old girl has a 5-month history of sad mood, poor appetite, social withdrawal, poor concentration, and a sense of hopelessness. She admits to occasionally wishing she would "just die" but emphatically asserts she would never consider suicide because of her religious beliefs. She describes having little energy, difficulty getting out of bed each morning, and difficulty falling asleep before 3:00 AM each night. Because she has a hard time falling asleep, she watches a lot of television, uses social media frequently, and browses the internet at night. For the past 2 months, she has been seeing a counselor who is providing cognitive behavioral therapy, and although she reports liking this counselor, she expresses frustration that she is not getting better. The patient and her mother are very much against the idea of using prescription antidepressant medication.

Of the following, the BEST next step in care is to recommend that the girl

- begin St John's wort supplements
- remove refined sugars and high fructose corn syrup from her diet
- stop using electronic devices at night
- take naps during the day
- transfer her care to a different psychological counselor

Item 81 Preferred Response: C

The girl described in the vignette is suffering from depression. She has stalled in her progress with cognitive behavioral therapy (CBT), perhaps in large part because she has chronic insomnia that is negatively affecting her mood. Insomnia could be a direct symptom of her depression, but it is likely being exacerbated by her excessive use of social media over-night. Sleep specialists recommend treating insomnia first by exploring the patient's sleep habits, and having them discontinue any sleep-interfering behaviors. Stopping all use of electronic devices in her room at night would be a key step toward improving this child's sleep hygiene.

There is no clear evidence that St John's Wort herbal supplements are helpful for treating depression, and no high-quality studies have reported its use in treating adolescent depression. Dietary changes have not been found to affect depression or insomnia. Daytime naps have a negative impact on overall sleep patterns, and clearly decrease overnight sleep time.

Transferring psychotherapy to a different counselor may be warranted when a patient lacks a "therapeutic alliance" with the current provider, but in this vignette the girl appears to like her current provider. This is a relevant issue because research demonstrates that a positive therapeutic alliance is one of the most consistent predictors of psychotherapy treatment success. A stalling of the progress of psychotherapy is common and indicates that there may be a new issue that needs to be addressed (such as this patient's insomnia).

PREP Pearls

- Depression treatment is assisted by improving sleep hygiene.
- Restricting access to electronic devices overnight is a simple way to improve sleep hygiene.
- Daytime naps do not improve overnight insomnia.

AAP Mental Health Competency

- Identify depression treatments besides referring to a therapist that a pediatrician can recommend

Suggested Reading:

- Birmaher B, Brent D; AACAP Work Group on Quality Issues, et al. Practice parameter for the assessment and treatment of children and adolescents with depressive disorders. JAm Acad Child Adolesc Psychiatry. 2007;46:1503-1526. doi:10.1097/chi.Ob013e318145aelc
- Jorm AF, Allen NB, O'Donnell CP, Parslow RA, Purcell R, Morgan Al. Effectiveness of complementary and self-help treatments for depression in children and adolescents. Med I Aust. 2006;185(7):368-372
- Mindell J, Owens J. A Clinical Guide To Pediatric Sleep: Diagnosis and Management of Sleep Problems. 2nd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2009
- Sherill IT, Kovacs M. Nonsomatic treatment of depression. Child Adolesc Psychiatr Clin N Am. 2002;11:579-593

Item 82

While performing a newborn examination in the mother's room 4 hours after birth, you notice the neonate appears dusky in dim lighting. He was born at term by spontaneous vaginal delivery and weighs 4.1 kg. The prenatal course was unremarkable, with a negative result for group B Streptococcus screening. Artificial rupture of the membranes occurred 4 hours before delivery. The neonate nursed well shortly after birth and has been sleeping since that time. Assessment in the well-lit normal nursery reveals a ruddy, cyanotic neonate with no respiratory distress. Vitals signs include a temperature of 36.8°C, heart rate of 140 beats/min, and respiratory rate of 60 breaths/min. Pulse oximetry performed on room air is 78% in the right hand, with an increase to 82% when the neonate is placed in a 100% oxygen hood. The physical examination reveals clear lungs and a normal cardiac examination. A blood gas obtained from the right radial artery while the infant is in the 100% hood demonstrates pH 7.32, PCO_2 40, PO_2 33, bicarbonate 19 and base deficit -4.

Of the following, the MOST likely cause of the neonate's findings is

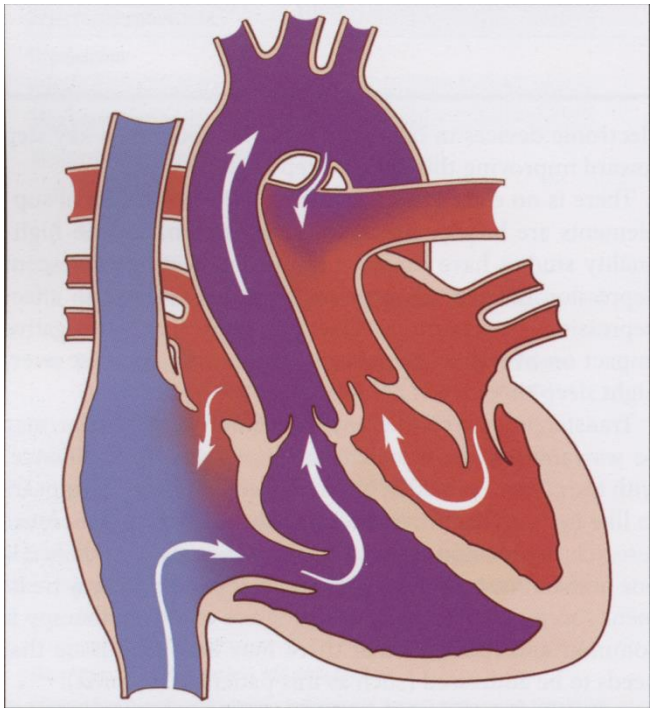
A. group B streptococcal pneumonia
B. methemoglobinemia
C. persistent pulmonary hypertension of the newborn
D. polycythemia
E. transposition of the great arteries

Item 82 Preferred Response: E

Infants without respiratory distress who present with severe cyanosis within hours of birth and fail to improve their arterial oxygen content with 100% oxygen therapy are likely to have transposition of the great arteries (TGA). In TGA, the great arteries carrying blood away from the heart are switched, with the aorta arising from the right ventricle and the pulmonary artery arising from the left ventricle (Item C82). This leads to severe cyanosis as the systemic deoxygenated blood returns to the right side of the heart and is unable to flow to the lungs while the oxygenated blood returns to the left side of the heart and passes directly back into the lungs. This had been described as 2 circuits in parallel. The degree of cyanosis is influenced by the degree of mixing that occurs between the 2 circuits at the level of the patent ductus arteriosus (PDA), patent foramen ovale (PFO), and ventricular septal defect (VSD). Infusion of prostaglandin E1 to maintain patency of the PDA is essential until a Rashkind balloon atrial septostomy is performed to enhance mixing.

Other forms of congenital heart disease that can present in the immediate newborn period with severe cyanosis are pulmonary atresia and Ebstein malformation. Both have severely restricted pulmonary blood flow and require initiation of prostaglandin El to maintain ductal patency. Infants with TGA, pulmonary atresia, and Ebstein malformation fail to improve their arterial oxygen saturation with the delivery of 100% oxygen because of the fixed restriction of pulmonary blood flow. Examination often reveals a "happy blue baby" with no respiratory distress or significant murmur. Clinical suspicions of congenital heart disease should be confirmed with echocardiography.

Persistent pulmonary hypertension of the newborn (PPHN) may also present with cyanosis and no respiratory distress. Increased pulmonary blood pressure due to failed neonatal transition or abnormal pulmonary vasculature causes right to left shunting at the level of the PFO and PDA, with resultant cyanosis. Unlike the infant described in the vignette, delivery of 100% oxygen to an infant with PPHN often leads to decreased pulmonary blood pressure, increased pulmonary blood flow, and improvement in arterial oxygen content. Group B streptococcal pneumonia must always be considered in the infant with cyanosis, but the absence of respiratory distress combined with the lack of maternal group B streptococcal colonization, prolonged rupture of the membranes, or maternal fever makes this diagnosis less likely in this infant. Methemoglobinemia caused by either congenital methemoglobin reductase deficiency or exposure to oxidants occurs in a blue-grey infant in whom cyanosis fails to respond to 100% oxygen delivery. Pulse oximetry readings are variable in methemoglobinemia, but the partial pressure of oxygen (PaO,) obtained in an arterial blood gas measurement will be normal in room air and increase when the infant is exposed to 100% oxygen. Infants with polycythemia appear to have cyanosis because of their elevated hemoglobin concentration, but pulse oximetry and the PaO, values are normal. The infant in the vignette had a low PaO, value that is not consistent with either methemoglobinemia or polycythemia.



ITEM C82: *In transposition of the great arteries, the pulmonary artery originates from the left ventricle and the aorta from the right ventricle.*

PREP Pearls

- Infants without respiratory distress who present with severe cyanosis within hours of birth and fail to improve their arterial oxygen content with 100% oxygen therapy may have transposition of the great arteries, pulmonary atresia, or Ebstein malformation.

American Board of Pediatrics Content Specification(s):

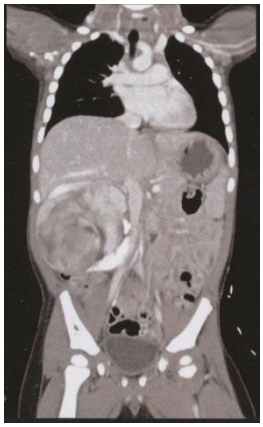
- Recognize that the absence of improvement in arterial oxygen content with 100% oxygen in comparison with room air is compatible with the diagnosis of cyanotic congenital heart disease

Suggested Reading:

- Eichenwald EC. Overview of cyanosis in the newborn. UptoDate. Available online only for subscription
- Silberbach M, Hannan D. Presentation of congenital heart disease in the neonate and young infant. Pediatr Rev. 2007;28:123-131. doi: 10.1542/ pir.28-4-123
- Steinhorn RH. Evaluation and management of the cyanotic neonate. Clin Pediatr Emerg Med. 2008;9:169-175. doi: 10.1016/j.cpem.2008.06.006

Item 83

A 3-year-old boy is brought to your office by his mother, who reports that she felt a lump in his belly while giving him a bath. There has been no fever, vomiting, diarrhea, abdominal pain, or dysuria. His axillary temperature is 38.2°C, pulse rate is 90 beats/min, respiratory rate is 20 breaths/min, and blood pressure is 120/80 mm Hg. On examination, the child is in no apparent distress. You palpate a firm, smooth, nontender mass on the right side of the abdomen. There is no costovertebral angle tenderness. The remainder of the physical examination is normal. A computed tomography scan of the abdomen with contrast is shown (Item Q83). The following are the results of the child's laboratory tests:



Complete blood cell count:

- White blood cell count, 11,500/pL (11.5 x 10^9/L), with 42% polymorphonuclear leukocytes, 49% lymphocytes, 6% monocytes, and 3% eosinophils
- Hemoglobin, 10.0 g/dL (100 g/L)
- Mean corpuscular volume, 75/pm3 (75 fL)
- Platelet count, 390 x 10^3/µL (390 x 10^9/L)

Urinalysis:

- 3+ heme
- Red blood cells, too numerous to count

Of the following, the MOST likely diagnosis in this child is

A. hepatoblastoma
B. neuroblastoma
C. renal abscess
D. renal cell carcinoma
E. Wilms tumor

Item 83 TE Preferred Response: E

The child described in the vignette with a right-sided renal mass most likely has Wilms tumor. Wilms tumor accounts for approximately 95% of renal tumors in young children, with a peak incidence at 2 to 4 years of age. In adolescents older than 15 years, renal cell carcinoma is the most common renal malignant disease. Wilms tumor usually presents with an abdominal mass (frequently painless), gross or microscopic hematuria, fever, and/or hypertension. Ninety-five percent of cases are unilateral. Approximately 10% to 12% of patients with Wilms tumor have congenital abnormalities, such as cryptorchidism, hypospadias, hemihypertrophy, or aniridia, which may be associated with syndromes such as WAGR (Wilms tumor, aniridia, genitourinary anomalies, and retardation), Beckwith-Wiedemann syndrome, Denys Drash syndrome, and hemihypertrophy. Patients diagnosed as having these syndromes may require ultrasonography screening for Wilms tumors every 4 months until 8 years of age. The initial evaluation for suspected Wilms tumor includes computed tomography with contrast or magnetic resonance imaging of the chest, abdomen, and pelvis to look for bilateral involvement, vascular extension, and metastases (most commonly to lymph nodes, lung, and liver). If the tumor appears to extend into the inferior vena cava, ultrasonography or echocardiography is recommended before surgery. Tissue evaluation at the time of nephrectomy is useful in determining whether the histologic findings are associated with a favorable or unfavorable prognosis. Wilms tumor staging is based on surgical staging and radiographic evaluation for disease spread (Item C83).

Other predictors of poorer prognosis include age older than 2 years, tumor weight greater than 550 g, and loss of heterozygosity on chromosomes 1p and 16q. Treatment for Wilms tumor involves a combination of surgery, chemotherapy, and radiation therapy, depending on the risk category. Overall, the survival rate for Wilms tumor is approximately 90%; therefore, current protocols are aimed at decreasing long-term sequelae of treatment without compromising the excellent survival rate.

Item C83. Wilms Tumor Staging

Stage I	Unilateral tumor, limited to kidney without capsular or lymph node involvement, completely resected without tumor spill
Stage II	Unilateral tumor, with capsule extension or invasion of adjacent structures, no lymph node involvement, completely resected without tumor spill
Stage III	Unilateral tumor, with lymph node involvement, tumor rupture or spill, incomplete resection
Stage IV	Metastasis to lungs, liver, bone, brain, distant lymph nodes
Stage V	Bilateral renal tumors

This child has a painless palpable abdominal mass, hematuria, and hypertension. In this age group, the most common renal tumor would be Wilms tumor. In children older than 15 years, renal cell carcinoma is the more prevalent renal malignant disease. Neuroblastoma most often occurs in the adrenal gland and presents at a median age of 19 months; 89% of cases are diagnosed by 5 years of age. Neuroblastoma in infants portends a better prognosis, whereas advanced disease is often found in children whose conditions are diagnosed when they are older than 30 months. Neuroblastoma can present as an incidentally discovered palpable abdominal mass (eg, adrenal mass with hypertension due to compression of the renal vessels), but hematuria is not common. Because of its

tendency to metastasize to bone and bone marrow, neuroblastoma frequently also presents with bone pain and cytopenia. Hepatoblastoma is the most common pediatric malignant tumor of the liver; however, it still comprises only 2% of all pediatric malignant tumors. It is associated with prematurity and very low birth weight (<1000 g), hemihypertrophy, and cancer predisposition syndromes, such as Beckwith-Wiedemann, Gardner, and familial adenomatous polyposis syndromes. Although hepatoblastoma can present with a painless abdominal mass, it is not typically associated with hematuria and hypertension. A renal abscess may appear as a mass on imaging; however, there is usually enhancement around the abscess on computed tomography with contrast. Furthermore, a patient with a renal abscess would likely have pyuria, leukocytosis, and high fever as well.

PREP Pearls

- The most common renal malignant tumor in young children is Wilms tumor.
- Wilms tumor, hepatoblastoma, and neuroblastoma can present as painless abdominal masses.
- Wilms tumor can present with abdominal mass, hypertension, and hematuria.
- In the presence of hemihypertrophy, screening for Wilms tumor with ultrasonography every 4 months is indicated through 8 years of age

American Academy of Pediatrics Content Specification:

- Understand that Wilms' tumor usually presents as an abdominal mass and may cause hypertension and/or hematuria

Suggested Reading:

- Buckley KS. Pediatric genitourinary tumors. Curr Opin Oncol. 2012;24:291-296. doi:10.1097/CC0.0b0l3e32835265c9
- Davenport KP, Blanco FC, Sandler AD. Pediatric malignancies: neuroblastoma, Wilm's tumor, hepatoblastoma, rhabdomyosarcoma, and sacroccygeal teratoma. SurgClin NAm. 2012;746-767. doi:10.1016/j. suc.2012.03.004
- Fernandez C, Geller JI, Ehrlich PF, et al. Renal tumors. In: Principles and Practice of Pediatric Oncology. 6th Ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2011:861-885

Item 84

An 8-year-old girl has a seizure at school. She was sitting at her desk when suddenly her whole body stiffened and she fell to the floor and had jerking movements of her limbs. The event lasted about 2 minutes and then stopped. The girl was sleepy for about 30 minutes and then returned to normal. This has never happened before, she has been healthy all her life, and there is no one in the family who has seizures. You order an electroencephalogram and receive a report that describes "right and left centrotemporal spikes, consistent with benign rolandic epilepsy." You refer her to a pediatric neurologist and her appointment is next week.

Of the following, the MOST appropriate advice to provide the girl's parents at this time is

A. brain damage is common even after a single seizure
B. children who have epilepsy should not participate in contact sports
C. children who have epilepsy should not take baths alone
D. risk of sudden death is high in children who have epilepsy
E. their daughter will likely need lifelong seizure medications

Item 84 I-C S Preferred Response: C

Seizure precautions should be discussed with the parents and the child, if appropriate, after a first seizure and typically at every office visit after that. Anyone with a seizure should not be in or around water by themselves. Children with seizures can shower alone because the risk of drowning in the shower is low, but they should not take a bath unattended and the bathroom door should remain unlocked. Typical school sports such as football or soccer are not restricted; however, participation in high-velocity sports, such as motocross or gymnastics should be discussed with parents and the child.
Most parents fear their child will have a brain injury or die because of a seizure. There is no evidence that a single, short (<30 min) seizure causes measurable brain damage to a child. The risk of sudden, unexplained death in epilepsy is very low in adults and even lower in children. Preventing accidental drowning is the most important intervention parents can do to protect their child.

In general, there is a 45% recurrence risk after a first, unprovoked seizure. The true risk of seizure recurrence, and thus the usefulness of starting seizure medications, depends on the cause of the seizures. The history should elucidate causes of provoked seizures, such as head injury, electrolyte abnormality, hypoglycemia, infection, etc.
Electroencephalography (EEG) can help identify an underlying epilepsy syndrome. If the seizure has focal onset, magnetic resonance imaging of the brain should also be performed to evaluate for a brain lesion. Unless the child has a history pointing to metabolic imbalances (vomiting, diarrhea, for example) it is not helpful to perform routine laboratory tests. Depending on the clinical situation, a seizure medication can be started to prevent further seizures while the workup is undertaken.

For the girl described in the vignette, the clinical history and EEG confirm a diagnosis of benign rolandic epilepsy (BRE). In this syndrome, seizures typically occur during sleep only and the child outgrows the seizure tendency within 2 years of diagnosis. In many cases of BRE, seizure medications are not used; seizure precautions are followed closely until the child has outgrown the seizure tendency.

PREP Pearls

- Seizure precautions including water safety should be reviewed with families of children with seizure.
- Most patients with a new seizure can be treated by their pediatrician. Referral to a neurologist is helpful if an abnormality is identified on imaging or electroencephalography.

American Board of Pediatrics Content Specification(s):

- Know how to manage a child following a first seizure

Suggested Reading:

- Camfield R Camfield C. Special considerations for a first seizure in childhood and adolescence. Epilepsia. 2008;49(suppl 1):40-44. doi: 10.1111/j.1528-1167.2008.01449.x

Item 85

A 9 year-old girl has an acute right ankle injury that occurred 3 days ago. She reports that she "rolled her ankle" during a basketball practice and had immediate sharp pain over the lateral aspect of the ankle. She was able to bear weight after the injury but was unable to continue playing because of pain. On physical examination, you observe mild swelling and bruising over the lateral aspect of the ankle and point tenderness over the lateral malleolus. The remainder of the examination is normal. Radiographs of the right ankle are normal.

Of the following, the MOST likely diagnosis is

A. avulsion fracture involving the distal fibula
B. grade 1 ankle sprain involving the posterior talofibular ligament
C. peroneal tendinitis
D. Salter–Harris type I fracture of the fibula
E. talar dome fracture

Item 85 Preferred Response: D

In skeletally immature children and teens, the cartilage growth plates of the skeleton are especially vulnerable to injury. Adults tend to sustain injuries to the tendons and ligaments, whereas children are prone to injuries involving primary and secondary growth centers, the physes and apophyses. The child in the vignette sustained an inversion ankle injury and has tenderness over the lateral malleolus. At her age, with this mechanism and the noted physical findings, a bony injury involving the fibula is very likely.

In a child with negative radiographs and bony tenderness over the physis, physicians should presume that the injury is a Salter–Harris type I injury involving the physis. The Salter–Harris type I classification denotes a separation at the physis without involvement of the adjacent metaphysic or epiphysis. This type of injury generally heals with 4 to 6 weeks of ankle immobilization (eg, with a walker boot). If symptoms have not resolved after 6 weeks, further evaluation would be warranted.

With a talar dome injury, the child would be expected to have tenderness over the anterior aspect of the ankle. Peroneal tendinitis (tendinopathy) does not generally develop acutely and would lead to tenderness behind and inferior to the lateral malleolus. This child's mechanism of injury could lead to an avulsion injury; however, the avulsed fragment is generally visible on radiographs. Skeletally immature children can have ligament sprains and these can be difficult to differentiate from mild growth plate injuries. Recent studies have suggested that lateral ankle ligament injuries in children are more common than previously thought. The anterior talofibular ligament is the most commonly sprained ankle ligament. This child could have an anterior talofibular ligament sprain; however, her pain is predominantly over the malleolus. She does not have tenderness over the ligament itself. Even in adults, the posterior talofibular ligament rarely is injured in isolation, so a posterior talofibular ligament sprain would be unlikely in this child.

PREP Pearls

- In a skeletally immature child or adolescent, the presence of a physeal injury should be considered, even when radiographs are normal.
- The anterior talofibular ligament is the most commonly sprained ankle ligament.

American Board of Pediatrics Content Specification(s):

- Recognize that an ankle injury in a prepubertal adolescent maybe a growth plate fracture rather than an ankle sprain

Suggested Reading:

- Boutis K, Narayanan UG, Dong FF, et al. Magnetic resonance imaging of clinically suspected Salter-Harris I fracture of the distal fibula. Injury. 2010;41(8):852-856. doi:10.1016/j.injury.2010.04.015
- Dayan PS, Vitale M, Langsam DJ, et al. Derivation of clinical prediction rules to identify children with fractures after twisting injuries of the ankle. Acad Emerg Med. 2004;11(7):736-743. doi:10.1197/j.aem.2004.02.517
- Sankar WN, Chen J, Kay RM, Skaggs DL. Incidence of occult fracture in children with acute ankle injuries. J Pediatr Orthop. 2008;28(5):500-501. doi: 10.1097/l3P0.0bO13e31817b9336

Item 86

A child in your practice was identified with phenylketonuria (PKU) on routine newborn screening, and the diagnosis was confirmed on follow-up testing. She was started on a low-phenylalanine diet and has been followed regularly by the metabolic specialist. She is doing quite well and shown excellent weight gain and normal developmental milestones for age. Her parents have gone online and read about potential problems with growth and cognitive development and risks to a developing fetus for adult women with PKU who have been maintained on the PKU diet. They are uncertain about the sources of this information and wish to know which, if any, of these issues are still a concern with optimal dietary management.

Of the following, you are MOST likely to tell the parents that with proper treatment their child will have

A. normal bone mineral densities in adulthood if maintained on the PKU diet
B. normal cognitive development if the diet is well managed until 10 years of age
C. normal height and head circumference measurements if maintained on the PKU diet
D. no teratogenic effects in her offspring if phenylalanine levels are well-controlled
E. some neurocognitive or psychosocial deficits in childhood despite good dietary management

Item 86 Preferred Response: E

The child described in this vignette was identified shortly after birth to have phenylketonuria (PKU), and dietary management was promptly initiated. However, despite early introduction of dietary therapy, long-term studies suggest that growth, neurocognitive outcomes, and bone health are still suboptimal in treated patients.

Untreated PKU is associated with severe neurocognitive deficits; however, the combination of newborn screening and maintenance of a therapeutic, phenylalanine-restricted diet has ameliorated but not eliminated the most serious clinical consequences. While some children on phenylalanine restricted diets were in the past permitted to go off diet after 10 years of age, studies have clearly demonstrated significant neurocognitive consequences of relaxing dietary intake, even for older children and adults. Therefore, lifelong dietary adherence is now recommended for all persons with PKU. In addition to a phenylalanine-restricted diet, use of oral sapropterin dihydrochloride as an adjuvant therapy has further helped to control blood phenylalanine levels in some patients.

Many children and young adults who had early dietary restriction and continued management still exhibit higher rates of executive functioning deficits and attention problems as well as reduced processing speed when compared to age-matched controls. Treated adults also may have decreased verbal memory, expressive naming, and verbal fluency despite optimal management. These individuals are also at greater risk for social and emotional difficulties, including low self-esteem, depression, generalized anxiety, phobias, and social isolation. Brain imaging of patients who have diet-treated PKU demonstrates a higher incidence of white matter abnormalities, decreased cerebral protein synthesis, altered L-DOPA uptake, volumetric changes in grey matter and altered cerebral metabolism.

Studies of diet-treated children and adolescents who have PKU have demonstrated reduced height and head circumference measurements, presumably due to the low dietary content of natural proteins. These patients are also subject to micronutrient deficiencies of zinc, copper, and selenium as well as deficient intake of calcium, cholesterol, and preformed long-chain polyunsaturated fatty acids (PLC-PUFAs). PLCPUFAs are critical to normal brain and retinal development; deficiencies of these fatty acids can lead to cognitive and visual dysfunction.

Bone metabolism is also disrupted in children and teens on the PKU diet, with evidence of slower bone formation and resorption in these individuals. This likely accounts for the higher incidence of osteopenia and osteoporosis in adults with PKU who have endured years of dietary deficiencies of protein, calcium, vitamin D, and trace elements.

Maternal PKU effects continue to be a significant concern for adult women with PKU who are pregnant or contemplating a pregnancy. Optimally, women with PKU should begin to lower their blood phenylalanine levels with dietary changes and medical input 3 months before conception. The goal is to reduce the maternal blood phenylalanine level to between 2.0 and 6.0 mg/dL (120 and 360 µmol/L) before the 8th to 10th week of

gestation and to maintain this level throughout pregnancy. Failure to achieve this metabolic tar-get by this point in gestation significantly increases the risks for congenital cardiac defects, microcephaly, cognitive deficits, and behavioral problems in offspring. However, because of daily blood phenylalanine fluctuations in even the most adherent mother-to-be, one cannot guarantee that optimal dietary management will completely negate potential teratogenic effects.

PREP Pearls

- Children and adults who have PKU are still at increased risk for cognitive, growth, and behavioral problems despite optimal dietary management.
- Adults who have PKU are at increased risk for osteopenia and osteoporosis.
- Women who have PKU and are of childbearing age should be cautioned about the need for additional preconceptional and prenatal dietary restrictions to minimize potential teratogenic influences of high levels of circulating maternal blood phenylalanine.

American Board of Pediatrics Content Specification(s):

- Know the natural history of treated and untreated phenylketonuria

Suggested Reading:

- Enns G, Koch R, Brumm V, Blakely E, Suter R, Jurecki E. Suboptimal outcomes in patients with PKU treated early with diet alone: revisiting the evidence. Mol Genet Metab. 2010;101(2-3):99-109. doi:10.1016/j. ymgme.2010.05.017
- Kaye CI, Committee on Genetics. Newborn screening fact sheets. Pediatrics. 2006;118(3):e934-e963 doi: 10.1542/peds.2006-1783
- Levy HL. Historical background for the maternal PKU syndrome. Pediatrics. 2003;112:1516-1518
- Widaman KF, Azen C. Relation of prenatal phenylalanine exposure to infant and childhood cognitive outcomes: results from the international maternal PKU collaborative study. Pediatrics. 2003;112:1537-1543

Item 87

The parents of a 4-year-old girl bring her to the emergency department for care of a laceration involving her upper lip. An hour ago, the girl tripped and fell into the corner of a wooden bookshelf, sustaining a 1-cm vertical laceration to her upper lip that extends approximately 4 mm beyond the vermillion border. The girl did not lose consciousness when she fell, and her parents state that she has been behaving normally since the injury occurred. On physical examination, you find no injuries other than the lip laceration. The laceration is not through and through, and it is not actively bleeding. A medical student you are working with asks about the approach you will use to treat this child's laceration.

Of the following, the MOST appropriate response is that

A. absorbable suture material should be used for the best possible outcome
B. consultation with a plastic surgeon may be required to achieve an acceptable outcome
C. lidocaine should be infiltrated liberally around the wound edges to provide adequate analgesia
D. suture placement is not needed since the laceration primarily involves a mucosal surface
E. use of a cyanoacrylate tissue adhesive would be a good option for laceration repair given the patient's young age

Item 87 Preferred Response: B

The girl in the vignette presents with a laceration to her upper lip with extension through the vermilion border. Lip lacerations require special attention for closure because they can result in significant cosmetic defects if not repaired properly. The vermilion border, the junction of the dry oral mucosa of the lip and the facial skin, serves as an important landmark for proper repair of a lip laceration when involved. Misalignment of the vermilion border by as little as 0.5 mm is easily noticeable. Although pediatric practitioners with prior training and experience in management of lip lacerations may possess the skill needed to properly repair lip injuries involving the vermilion border, consultation with an orofacial or plastic surgeon is indicated for most practitioners. When involved, the vermilion border should be the first area approximated in repair of a lip laceration. It is essential to identify and mark the vermilion border before initiating infiltration anesthesia or wound debridement. Infiltration of local anesthetic around the wound edges may lead to soft tis-sue swelling and tissue distortion, which can interfere with proper tissue apposition. This can be avoided through the use of regional nerve blocks to anesthetize the wound.

When parted, the vermilion border should be reapproximated precisely using a 6-0 nonabsorbable nylon suture. In general, lip lacerations should be closed in layers, depending on the depth of the wound. Full-thickness lip lacerations require a 3-layer repair.

Although small lip lacerations that involve only the inner mucosal surfaces of the lip may not require suture placement, the vermilion border must be precisely approximated whenever involved to avoid an unacceptable cosmetic outcome. Cyanoacrylate tissue adhesives are an acceptable option for repair of some uncomplicated facial lacerations, but they are not recommended for repair of injuries that involve the oral mucosa and would not facilitate exact alignment of the vermilion border.

PREP Pearls

- Lip lacerations that involve the vermilion border require special attention for closure. Misalignment of the vermilion border by as little as 0.5 mm is easily noticeable and can result in permanent cosmetic defects.
- Consultation with an orofacial or plastic surgeon may be indicated for lip lacerations that involve the vermilion border.
- In managing a child with a lip laceration, it is essential to identify and mark the vermilion border before initiating infiltration anesthesia or wound debridement.

American Board of Pediatrics Content Specification(s):

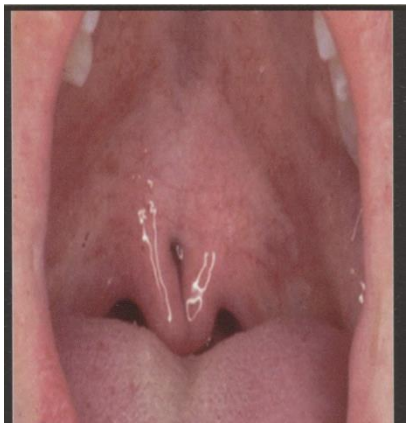
- Recognize the problem with a laceration through the vermilion border of the lip

Suggested Reading

- Attia MW, Loiselle J. Management of soft-tissue injuries of the mouth. In: King C, Henretig FM, eds. Textbook of Pediatric Emergency Procedures. 2nd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2008:680-687
- Harter DA, Miller S. Management of specific soft tissue injuries. In: Reichman EF, Simon RR, eds. Emergency Medicine Procedures. New York, NY: McGraw-Hill; 2004
- Sagerman PL Wounds. Pediatr Rev. 2005;26:43-49. doi:10.1542/pir.26-2-43
- Selbst SM, Attia MW. Minor trauma—lacerations. In: Fleisher GR, Ludwig S, eds. Textbook of Pediatric Emergency Medicine. 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2010:1256-1270

Item 88

During routine examination of a well 4-year-old boy in your office, you notice a bifid uvula (Item Q88). On palpation of the soft palate, you suspect a submucous cleft palate.



ITEM Q88: Bifid uvula and a submucous cleft palate for the child described In the vignette.

Of the following, this child is at GREATEST risk for

A. conductive hearing loss
B. dental anomalies
C. dysphagia
D. problems with articulation
E. recurrent sinusitis

Item 88 SBP Preferred Response: A

Patients with a bifid uvula, such as the boy described in the vignette, are at increased risk of having a submucosal cleft palate (SMCP) (Item C88). Careful examination of the posterior palate should be performed and may reveal a visible dimpling or notching or a palpable defect. The classic SMCP is the triad of bifid uvula, diastases of the muscles in the midline of the soft palate with intact mucosa, and notching of the posterior border of the hard palate. However, not all these features must be present for the condition to be diagnosed. Occult SMCP has been reported in the absence of a cleft uvula; conversely, bifid uvula without other physical signs of SMCP or velopharyngeal incompetence is reported to occur in approximately 0.1% to 10% of the general population.

Only a small percentage of cases of SMCP are symptomatic, but one of the most common consequences of SMCP is recurrent acute otitis media or chronic serous otitis media, which may be associated with conductive hearing loss. Early recognition is important so that appropriate management can be instituted, including speech therapy, careful middle ear examinations, and regular audiometric screening. The dentition is not affected by this mild palatal abnormality. Although SMCP may cause some slowness of feeding in infants and nasal regurgitation of liquids, it is not usually associated with dysphagia. Hypernasal speech secondary to velopharyngeal incompetence may occur, but articulation problems are not common. Adenoidectomy is contraindicated in these patients because of the risk of acquired velo-pharyngeal insufficiency after the procedure and worsening of existing hypernasal speech. The anatomical severity of the SMCP will not, by itself, predict which children will develop poor speech. Therefore, it is prudent to defer surgery to repair the cleft until the child is 4 to 6 years old and is mature enough to undergo adequate speech evaluation and assessment of velopharyngeal competence. Bifid uvula and SMCP are not commonly associated with an increased prevalence of sinusitis.

PREP Pearls

- Patients with a bifid uvula are at increased risk of having a submucosal cleft palate.
- Submucosal cleft palate may be detected by a visible dimpling or notching of the posterior palate or by palpating a defect on physical examination.
- Conductive hearing loss, which may be associated with recurrent acute otitis media or chronic serous otitis media, is one of the most common consequences of submucosal cleft palate.

American Board of Pediatrics Content Specification(s):

- Know that a bifid uvula is associated with submucous cleft palate and middle ear effusion

Suggested Reading:

- Berera G. Index of suspicion: case 1: submucous cleft palate. Pediatr Rev. 1993;14::191-193
- Drutz JE. The pediatric physical examination: HEENT. UPtoDate. 2012. Available online only for subscription
- Tinanoff N. Disorders of the oral cavity associated with other conditions: cleft lip and palate. In: Kliegman RM, Stanton BMD, St Geme J, Schor N, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Elsevier Saunders, 2011:1251-1253

Item 89

While discussing the care of a child who has asthma, the resident you are supervising asks if formoterol can be used as rescue therapy. You explain the differences in the kinetics of short-acting β-agonists (SABAs), such as albuterol, and long-acting β-adrenergic agonists (LABAs), such as formoterol and salmeterol.

Of the following, the MOST accurate statement regarding these differences is

A. LABAs have bronchodilating effects lasting up to 6 hours
B. levoalbuterol is a SABA modified to have the same duration of action as a LABA
C. regular use of LABAs can lead to dimunition of bronchoprotective effect
D. regular use of SABAs does not lead to dimunition of bronchoprotective effect
E. SABAs have bronchodilating effects lasting up to 2 hours

Item 89 Preferred Response: C

Diminution of the bronchoprotective effect occurs with the frequent use of both long-acting β-adrenergic agonists (LABAs) (eg, salmeterol and formoterol) and short-acting β-agonists (SABAs) (eg, albuterol). The bronchoprotective effect is described as the ability to protect against broncho-constriction in response to stimuli such as methacholine, exercise, or allergen exposure. Although the clinical significance of this attenuation is debated, it is associated with in vivo changes such as increased sputum eosinophilia and mediator release and worsening of the late phase asthmatic response. Typically, the allergic cascade involves a stage of sensitization to an allergen, followed by an early-phase response upon reexposure to an allergen. This may be followed by the late phase response usually 2 to 24 hours after the original reaction triggered by TH2 (allergic) cytokines provoking eosinophil and other cellular mediator release.
The decreased bronchoprotection associated with the chronic use of β-agonists is thought to be similar to "desensitization" (ie, decreased cellular responsiveness due to constant stimulation). The current recommendation of 2007 National Heart, Lung, and Blood Institute guidelines is to prescribe SABAs as needed for symptom control rather than on a regular schedule.

Short-acting (β-agonists are bronchodilators whose effects last 4 to 6 hours. Long-acting β-adrenergic agonists (salmeterol and formoterol) are bronchodilators whose effects last 10 to 12 hours. Levalbuterol is the R-isomer of albuterol. Albuterol is a racemic mixture with a 1:1 ratio of the isomers R-albuterol (levalbuterol) and S-albuterol; the R-isomer is responsible for the drug's bronchodilating activity. Levalbuterol (SABA) therefore has the same duration of broncho-dilation as albuterol (4-6 hours).

Variations in the molecular structure of β-agonists affect the onset and duration of bronchodilation. As an example, prolongation of the bronchodilator effect with LABAs is achieved by modifications that decrease degradation by catechol-O-methyl transferase and monoamine oxidase. In addition, the long, lipophilic side chains of formoterol and salmeterol attach to the plasma membrane and increase the duration of binding of the drugs to the adrenergic receptor.

All LABA medications currently carry a black box warning and the Food and Drug Administration (FDA) specifically recommends against use of LABAs alone for rescue therapy. Formoterol or salmeterol therefore should not be used as rescue therapy or in situations of acute worsening of symptoms. Recent review of LABAs carried out by the FDA demonstrated that "concomitant" use of inhaled corticosteroids and LABA (in separate inhalers) had an increased risk of exacerbations requiring emergency department visits and hospitalizations, particularly in the 4- to 11-year-old age group. The risk was not higher than other age groups when used "as assigned" (inhaled corticosteroids and LABA in the same inhaler device). National Heart, Lung, and Blood Institute guidelines do not recommend regularly scheduled, daily, chronic use of SABAs.

PREP Pearls

- SABAs have quick onset of action (within 15 minutes) and duration of action of 4-6 hours.
- LABAs may have a similar onset of action to albuterol (formoterol) or take up to 30 minutes (salmeterol). The duration of action is approximately 12 hours.
- Regular use of LABAs and SABAs can lead to dimunition of the bronchoprotective effect.

American Board of Pediatrics Content Specification(s):

- Know the kinetics of short- and long-acting inhaled beta-adrenergic agonists

Suggested Reading:

- McMahon AW, Levenson MS, McEvory BW, Mosholder AD, Murphy D. Age and risks of FDA-approved long-acting P2-adrenergic receptor agonists. Pediatrics. 2011;128(5):e1147-e1154
- National Asthma Education and Prevention Program. Expert Panel Report 3 (EPR-3): guidelines for the diagnosis and management of asthma—summary report 2007 [published correction appears in]Allergy Clin Immunol. 2008;121(6):1330).1 Allergy Clin Immunol. 2007;120(5 suppl):S94-5138
- Sorkness CA. Beta-adrenergic agonists. In: Adkinson NF, Bochner BS, Busse WW, et al, eds. Middleton's Allergy: Principles and Practice. 7th ed. Philadelphia, PA: Mosby; 2009:1485-1503
- US Food and Drug Administration. FDA drug safety communication: drug labels now contain updated recommendations on the appropriate use of long-acting inhaled asthma medications called long-acting betaagonists (LABAs)

Item 90

An 8-year-old boy is seen for a routine health supervision visit. Physical examination reveals a temperature of 37.8°C, heart rate of 60 beats/min, respiratory rate of 16 breaths/min, blood pressure of 150/90 mm Hg, and normal growth parameters. He has multiple cafe au laic spots and peripheral neurofibromas as well as axillary freckling. He is not taking any medications currently. His urinalysis demonstrates a specific gravity of 1.035, pH of 6.0, 1+ blood, and no leukocyte esterase, protein, or nitrites. His urine microscopy shows 2 to 5 red blood cells per high-power field, less than 5 white blood cells per high-power field, and no crystals or bacteria. Serum electrolytes are within normal range.

Of the following, the MOST likely cause of elevated blood pressure in this boy is

A. acute nephritis
B. chronic renal failure
C. pseudoaldosteronism (Liddle syndrome)
D. pseudohypoaldosteronism (Gordon syndrome)
E. renal artery stenosis

Item 90 TE Preferred Response: E

Secondary hypertension in children can be renal, cardiac, or endocrine in origin, with renal disease and renovascular anomalies being the most common reported cause in about 80% to 90% of children.

Very young children, children with stage 2 hypertension, or children and adolescents with clinical signs that suggest systemic conditions with hypertension are at higher risk for secondary hypertension and require detailed investigation. In the 8-year-old child in the vignette, who has significantly elevated blood pressures along with the clinical phenotype consistent with neurofibromatosis type 1 (NF1), an underlying cause for hypertension should be suspected. Patients with NF1 are at increased risk for renal artery stenosis leading to renovascular hypertension compared with the general population. The gold standard for diagnosing renal artery stenosis in children is intraarterial angiography. Also available for evaluating such patients are magnetic resonance angiography, computed tomographic angiography, and duplex Doppler ultrasonography. These patients should be evaluated and treated by clinicians with experience in pediatric hypertension and the imaging performed at a radiology center with pediatric experience in these screening techniques. If the workup, for renal artery stenosis in patients with NF1 who have hypertension has negative results, then the possibility of pheochromocytoma leading to hypertension should be considered.

Normal urinalysis findings without significant hematuria or pyuria make the diagnosis of acute nephritis less likely. In view of normal growth parameters, specific gravity of 1.035 indicating intact concentrating ability of the kidney (patients with advanced chronic renal failure usually have a fixed specific gravity of 1.010 or less), and normal electrolytes, underlying chronic renal failure is unlikely.

Gordon syndrome (pseudohypoaldosteronism) and Liddle syndrome (pseudoaldosteronism) are rare genetic disorders associated with abnormalities in the renal tubules leading to hypertension. Gordon syndrome is characterized by hypertension, hyperkalemia, metabolic acidosis, and normal renal function. Genetic renal tubular anomalies lead to increased sodium chloride reabsorption in the distal tubule, causing volume expansion and hypertension in association with diminished renin secretion. This also leads to reduced potassium and hydrogen excretion, which accounts for the hyperkalemia and acidosis seen in these patients. Liddle syndrome is a rare autosomal dominant condition in which a primary increase is noted in collecting tubule sodium reabsorption and potassium secretion. Affected patients have electrolyte abnormalities consistent with mineralocorticoid excess and typically present with hypertension, hypokalemia, and metabolic alkalosis.

PREP Pearls

- Renal disease and renovascular anomalies are the most common causes of secondary hypertension in children.
- Patients with NF1 are at increased risk for renal artery stenosis.

American Board of Pediatrics Content Specification (s):

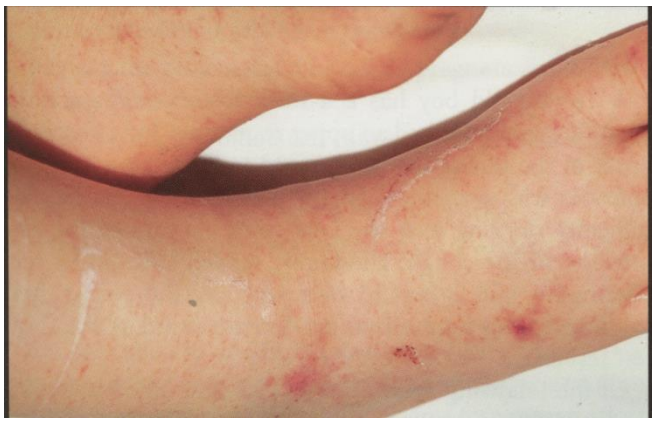
- Know the causes of renal hypertension

Suggested Reading:

- Feld LG, Corey H. Hypertension in childhood. Pediatr Rev. 2007;28(8):283298. doi:10.1542/pir.28-8-283
- Fossali E, Signorina E, Intermits RC et al. Renovascular disease and hypertension in children with neurofibromatosis. Pediatr Nephrol. 2000;14(8-9):806
- Londe MB. Systemic hypertension.. In: Kliegman RM, Stanton BF, St Genre JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier, 2011:1639-1647
- National High Blood Pressure Education Program Working Group on High Blood Pressure in Children and Adolescents. The fourth report on the diagnosis, evaluation, and treatment of high blood pressure in children and adolescents. Pediatrics. 2004;114:555-576

Item 91

A 7-year-old girl is seen in the emergency department with 3 days of fever, headache, myalgias, and vomiting. She also complains of mild abdominal pain and loose stools and now has a rash on her ankles and feet. The family just returned from a camping trip to North Carolina. On physical examination, her temperature is 39.5°C, heart rate is 135 beats/ min, respiratory rate is 25 breaths/min, and blood pressure is 90/55 mm Hg. She is ill-appearing but answers questions appropriately. Kernig and Brudzinski signs are absent. Her pupils are equal and reactive to light, but her mucous membranes are dry. The oropharynx, nares, and tympanic membranes are normal. Auscultation of the lungs reveals coarse breath sounds bilaterally with normal effort. The girl is tachycardic, but there is no murmur. The abdomen is mildly tender to palpation throughout without rebound or guarding. There is no organomegaly. Results of her neurologic examination are unremarkable. Examination of the skin reveals a rash on her ankles and feet, including the soles (Item Q91).



Laboratory findings include the following:

- White blood cell count, 5,600/µL (5.6×10^9/L), with 20% neutrophils, 68% lymphocytes, and 12% monocytes
- Hemoglobin, 10 g/dL (100 g/L)
- Platelet count, 75×10^3/µL (75×10^9/L)
- Aspartate aminotransferase, 55 U/L
- Alanine aminotransferase, 38 U/L
- Sodium, 128 mEq/L (128 mmol/L)
- Potassium, 3.0 mEq/L (3.0 mmol/L)
- Chloride, 102 mEq/L (102 mmol/L)
- Bicarbonate, 20 mEq/L (20 mmol/L)
- Glucose, 90 mg/dL (5.0 mmol/L)
- Blood urea nitrogen, 20 mg/dL (7.14 mmol/L)
- Creatinine, 0.5 mg/dL (38.1 µmol/L)

A chest radiograph is unremarkable.
Of the following, the MOST appropriate antimicrobial therapy for this patient is

A. chloramphenicol
B. ciprofloxacin
C. doxycycline
D. penicillin
E. trimethoprim-sulfamethoxazole

Item 91 Preferred Response: C

The girl described in the vignette has an illness characterized by fever, headache, and a petechial rash on her ankles and feet (Item C91) (including the soles) after traveling to North Carolina. Her clinical presentation and laboratory study results (thrombocytopenia, anemia, and hyponatremia) are most consistent with infection due to Rickettsia rickettsii, the cause of Rocky Mountain spotted fever (RMSF). Doxycycline is the drug of choice for treating RMSF, even in children younger than 8 years. Doxycycline also is effective against ehrlichiosis, which can be clinically similar to RMSF. Chloramphenicol may be less effective for the treat ment of RMSF and does not treat ehrlichiosis. In addition, chloramphenicol is associated with serious adverse effects and is not available orally in the United States. Data on the use of fluoroquinolones (eg, ciprofloxacin and levofloxacin) for treating RMSF are limited. Neither penicillin nor trimethoprim–sulfamethoxazole has activity against R rickettsii.

Rocky Mountain spotted fever is a systemic, small vessel vasculitis characterized by fever, severe headache, myalgias, vomiting, and rash that typically begins before the sixth day of illness. Classically, the rash begins on the wrists and ankles and spreads to the trunk. Involvement of the palms and soles is typical. However, up to 20% of patients may not have a rash. Thrombocytopenia and hyponatremia are common, and leukopenia and anemia also can occur. Untreated, the case-fatality rate is approximately 25%. In the United States, R rickettsii is transmitted to humans through the bite of the American dog tick (Dermacentor variabilis), Rocky Mountain wood tick (Dermacentor andersoni), and brown dog tick (Rhipicephalus sanguineus). Rocky Mountain spotted fever has been reported throughout most of the United States; however, Arkansas, Missouri, North Carolina, Oklahoma, and Tennessee account for most cases. Rocky Mountain spotted fever also occurs in Mexico, South America, Central America, and Canada. Rocky Mountain spotted fever is most common in the summer months but can occur year-round in endemic areas.

PREP Pearls

- Doxycycline is the treatment of choice for Rocky Mountain spotted fever even in young children.
- In 80% of cases, Rocky Mountain spotted fever is accompanied by a petechial rash starting on the ankles and wrists and involving the palms and soles.
- Most cases of Rocky Mountain spotted fever occur in Arkansas, Missouri, North Carolina, Oklahoma, and Tennessee.

American Board of Pediatrics Content Specification(s):

- Understand the geographical distribution of Rocky Mountain spotted fever

Suggested Reading:

- American Academy of Pediatrics. Rickettsia) diseases. In: Pickering LK, Baker CI, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: 2012:620-622
- American Academy of Pediatrics. Rickettsialpox. In: Pickering LK, Baker Cj, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: 2012:622-623
- American Academy of Pediatrics. Rocky Mountain spotted fever. In: Pickering LK, Baker CI, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: 2012:623-625
- Centers for Disease Control and Prevention. Rocky Mountain Spotted Fever, www.cdc.gov

Item 92

A 15-year-old girl is seen in your office as a follow-up from a recent visit to the emergency department (ED) for vulvar pain. You check the results of testing done in the ED and find a positive test result for herpes simplex virus type 2. The girl was given a pain medication but no antiviral medication, pending the results of her tests. Physical examination reveals scattered shallow ulcers on the vulva. You discuss sexual transmission and the natural history of the infection.

Of the following, the BEST course of action would be to prescribe

A. oral acyclovir for an initial infection
B. oral acyclovir suppressive therapy
C. oral acyclovir to begin with the next recurrence
D. topical acyclovir for the current infection
E. topical acyclovir to begin with the next recurrence

Item 92 Preferred Response: A

The adolescent described in the vignette is experiencing her first clinical episode of genital herpes. Despite having a mild infection, the girl should receive antiviral therapy as recommended by the Centers for Disease Control and Prevention (CDC) because she may develop severe or prolonged symptoms during this first clinical episode. If genital herpes is a consideration, therapy should be started before test results are available to better control the symptoms and signs and decrease the number of days of viral shedding. There is no available treatment to eradicate the virus. The 3 currently available medications include acyclovir, valacyclovir, and famciclovir. Recommended regimens are presented below (Items C92A, 92B, 92C) and are available on the CDC Web site at www.cdc.gov.

To be effective, episodic treatment should be started within a day of the onset of lesions and preferably during the prodrome preceding the outbreak. Therefore, it is best for an infected person to have medication available rather than delay treatment while seeking a prescription.

Antiviral agents may also be used to suppress recurrences and to prevent asymptomatic shedding of herpes simplex virus 2, which might lead to infection of a partner. The decision to use suppressive therapy may be guided by the patient's preference (ie, tolerance of recurrences) rather than by the specific number or frequency of recurrences. Because recurrences tend to decrease over time, the need for suppressive therapy should be revisited on a yearly basis and consideration given to stopping use of the medication. Topical therapies have limited efficacy, and their use is discouraged. It is important to discuss with the patient the natural history of the infection and its transmissibility, in addition to pain relief when addressing treatment.

Item (92A• Recommended Regimens for a First Clinical Episode of Genital Herpes

Medication	Dose	Duration
Acyclovir	400 mg orally 3 times a day	7-10 days
Acyclovir	200 mg orally 5 times a day	7-10 days
Famciclovir	250 mg orally 3 times a day	7-10 days
Valacyclovir	1 g orally twice a day	7-10 days

Reprinted with permission from Workowski KA, Berman S; Centers for Disease Control and Prevention. Sexually transmitted diseases treatment guidelines, 2010. *MMWR Recomm Rep.* 2010;59(RR-12):20-25. www.cdc.gov/std/treatment/2010/default.htm

Item C9213• Episodic Therapy for Recurrent Genital Herpes

Medication	Dose	Duration
Acyclovir	400 mg orally 3 times a day	5 days
Acyclovir	800 mg orally twice a day	5 days
Acyclovir	800 mg orally 3 times a day	2 days
Famciclovir	125 mg orally twice a day	5 days
Famciclovir	1000 mg orally twice a day	1 day
Valacyclovir	500 mg orally twice a day	3 days
Valacyclovir	1 g orally once a day	5 days

Reprinted with permission from Workowski KA, Berman S; Centers for Disease Control and Prevention. Sexually transmitted diseases treatment guidelines, 2010. MMWR *Recomm Rep.* 2010;59(No. RR-12):20-25. www.cdc.gov/std/treatment/2010/default.htm

Item (92• Recommended Regimens for Suppressive Therapy for Recurrent Genital Herpes

Medication	Dose
Acyclovir	400 mg orally twice a day
Famciclovir	250 mg orally twice a day
Valacyclovir	500 mg orally once a day
Valacyclovir	1 g orally once a day

Reprinted with permission from Workowski KA, Berman S; Centers for Disease Control and Prevention. Sexually transmitted diseases treatment guidelines, 2010. MMWRRecomm *Rep.* 2010;59(No. RR-12):20-25. www.cdc.gov/std/treatment/2010/default.htm.

PREP Pearls

- There is no currently available treatment to eradicate the herpes simplex virus.
- The dose and duration of antiviral therapy vary with different stages of infection.
- An infected person is best served by having medications available to start taking immediately with a recurrence.

American Board of Pediatrics Content Specification(s):

- Know the indications for and limitations of oral acyclovir treatment for genital herpes

Suggested Reading:

- American Social Health Association; Herpes Resource Center at www.ashastd.org
- Burstein GR, Workowski KA. Herpes genitalis. In: Neinstein LS, Gordon CM, Katzman DK, Rosen DS, Woods ER, eds. Adolescent Health Care: A Practical Guide. 5th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2008:65:834-841
- Chayavichitsilp P, Buckwalter JV, Krakowski AC, Friedlander SF. Herpes simplex. Pediatr Rev. 2009;30:119-130. DOI: 10.1542/pir.30-4-119
- Workowski KA, Berman S; Centers for Disease Control and Prevention. Sexually transmitted diseases treatment guidelines, 2010. MMWR Recomm Rep. 2010;59(No. RR-12):20-25

Item 93

A 16-year-old girl who has hemoglobin SS disease and has undergone cholecystectomy presents with the acute onset of abdominal pain, fever, and vomiting. Physical examination demonstrates an icteric and uncomfortable adolescent who has moderate right upper quadrant and epigastric tenderness to palpation. Laboratory testing demonstrates the following:

- Hemoglobin, 9.2 g/dL (92 g/L)
- White blood cell count, 16,500/uL (16.5 x 10^9/L)
- Total bilirubin, 18.5 mg/dL (316.4 µmol/L)
- Direct bilirubin, 10.8 mg/dL (184.7 umol/L)
- Amylase, 650 U/L; reference range, < 100 U/L
- Lipase, 900 U/L; reference range, 5100 U/L

Abdominal ultrasonography shows a dilated common bile duct without intrahepatic ductal dilatation. The pancreas appears edematous and without cysts or calcifications.

Of the following, the MOST appropriate next step in evaluating this patient is

A. abdominal computed tomography
B. endoscopic retrograde cholangiopancreatography
C. hepatobiliary scintigraphy
D. magnetic resonance cholangiopancreatography
E. percutaneous transhepatic cholangiography

Item 93 Preferred Response: D

The girl described in the vignette has SS disease and presents with signs and symptoms of extrahepatic biliary tract obstruction, marked by direct hyperbilirubinemia and secondary acute pancreatitis. Confirmation of this diagnosis is provided by abdominal ultrasonography, which demonstrates common bile duct dilation and an edematous pancreas. Considering the patient's underlying hemolytic disorder, the most likely cause of her presentation is choledocholithiasis. On the basis of the biochemical and ultrasonography findings, a stone obstructing the common bile duct near the ampulla of Vater is suspected. This finding should always be strongly considered in postcholecystectomy patients with a history of cholelithiasis, who present with signs suggestive of extrahepatic obstruction. Although several diagnostic modalities may be used to identify common duct stones, the endoscopic ultrasonography (EUS) is emerging as the most sensitive and specific test, particularly when transabdominal ultrasonography fans to identify an obstructing stone.

Cholelithiasis remains an uncommon diagnosis during childhood and adolescence; however, patients with hemolytic disorders are at particular risk. Accordingly, although cholesterol stones are the most common type in adults, black pigment stones represent the most common type in pediatric patients. The frequency of cholelithiasis in children with SS disease is almost double that of the general population; pigmented gallstones occur in approximately 50% of these children by 22 years of age. Black pigment stones are formed when bile becomes supersaturated with calcium bilirubinate, the calcium salt of unconjugated bilirubin.

In adults, up to 80% of gallstones are believed to be asymptomatic; however, pediatric data suggest that symptoms occur in more than 50% of children with cholelithiasis. Therefore, gallstone disease should be considered in the evaluation of nonspecific abdominal pain in children with risk factors, including chronic hemolysis, obesity, ileal disease, a positive family history, pregnancy, and those who have undergone a period of total parenteral nutrition. Although abdominal pain may be poorly localized in young children, older patients may experience symptoms in the right upper quadrant. Unexplained right scapular or shoulder pain may also be associated with gallstones. Clearly, cholelithiasis must be ruled out in a symptomatic child with hemolytic disease and should be considered in children with jaundice and transaminase elevations. Choledocholithiasis develops in up to 20% of patients with gallbladder stones; however, the diagnosis may be challenging even when signs and symptoms strongly suggest a stone obstructing the common bile duct at or near the ampulla of Vater (as was the case for the girl in the vignette). Although abdominal ultrasonography is highly sensitive for identification of gallbladder stones, its sensitivity decreases to less than 40% for common duct stones. Until recently, endoscopic retrograde cholangiopancreatography (ERCP) was considered the diagnostic study of choice in patients with suspected choledocholithiasis because it has the advantage of therapeutic intervention if a stone is discovered. However, ERCP may fail to identify small stones, the technique is highly invasive, and it may cause or exacerbate pancreatitis. The morbidity of this procedure may outweigh its benefits if the patient has already passed the obstructing stone. Therefore, every effort should be made to identify stones before recommending ERCP. In adult studies, EUS

detected more than 90% of common bile duct stones. In patients with acute pancreatitis, the sensitivity and specificity of this procedure were approximately 98%.

Other biliary tract imaging studies would be of little benefit to the girl in the vignette. Hepatobiliary scintigraphy may indicate biliary obstruction, but its sensitivity for stone identification is low. Similarly, computed tomography will fail to identify small stones. In patients who demonstrate intrahepatic bile duct dilation, percutaneous transhepatic cholangiography may be used in the evaluation of biliary obstructive disease. However, its usefulness may be supplanted by the emerging technology of magnetic resonance cholangiopancreatography (MRCP), and the technique is not of value in the evaluation of cholelithiasis.

PREP Pearls

- In patients with hemoglobin SS disease, cholelithiasis should always be suspected in the presence of signs or symptoms of cholestasis.
- Endoscopic retrograde cholangiopancreatography should not be attempted in the presence of pancreatitis.
- In patients with signs of cholestasis and pancreatitis, evaluation for gall stones at the ampulla of Vater should include magnetic resonance cholangiopancreatography or endoscopic ultrasonography.

American Board of Pediatrics Content Specification(s):

- Recognize the signs and symptoms of cholelithiasis and choledocholithiasis

Suggested Reading:

- Alonso MH. Gall bladder abnormalities in children with sickle cell disease: management with laparoscopic cholecystectomy. J Pediatr. 2004;145:580581. DOI: 10.1016/j.jpeds.2004.08.041
- Canto MI, Chak A, Stellato T, Sivak MV Jr. Endoscopic ultrasonography versus cholangiography for the diagnosis of choledocholithiasis. Gastrointest Endosc. 1998;47:439-448. DOI: 10.1016/S00165107%2898%2970242-1
- Garrow D, Miller S, Sinha D, Conway J, Hoffman BJ, Hawes RH, Romagnuolo J1. Endoscopic ultrasound: a meta-analysis of test performance in suspected biliary obstruction. ain Gastroenterol Hepatol. 2007;5:616-623. DOI: 10.1016/j.cgh.2007.02.027
- Tse F, Liu L, Barkun AN, Armstrong D, Moayyedi P. EUS: a meta-analysis of test performance in suspected choledocholithiasis. Gastrointest Endosc. 2008;67:235-44. doi: 10.1016/j.gie.2007.09.047. Abstract accessed February 2013 at: http://www.ncbi.nlm.nih.gov/pubmed/18226685
- Wesdorp I, Bosman D, de Graaff A, Aronson D, van der Blij F, Taminiau J. Clinical presentations and predisposing factors of cholelithiasis and sludge in children. J Pediatr Gastroenterol Nutr. 2000;31:411-417

Item 94

You are giving a presentation to a community group about environmental effects on pediatric health. One of the participants asks about the effects of air pollution in developing countries.

Of the following, the statement you are MOST likely to make is

A. adult men are most susceptible to air pollution effects in the developing world because of occupational exposures
B. air pollution causes proven respiratory effects in industrialized nations but has not been shown to be a health hazard in the developing world
C. exposure to household air pollution caused by burning plant or animal materials for cooking fuel increases a child's risk of developing pneumonia
D. the health effects of air pollution in the developing world are primarily restricted to congested urban areas
E. in the developing world, outdoor air pollution accounts for a greater impact on health and disability than does indoor air pollution

Item 94 S SBP Preferred Response: C

According to the World Health Organization (WHO), 3 billion people, mostly in developing countries, depend on biomass or solid fuels for cooking, lighting, and heating. Biomass fuels include wood, crop residue, charcoal, and animal dung, whereas solid fuel is primarily coal. People dependent on these fuel sources live at the lower end of the energy ladder, producing their energy needs locally and inefficiently in their own home, whereas affluent populations depend on more efficient natural gas and centrally produced electricity. In developing countries, these unprocessed, locally used energy sources may be found in both urban and rural areas. Biomass and solid fuels present both health and environmental hazards attributable largely to incomplete combustion and inadequate ventilation. Because these fuels are inefficient, large amounts must be used, contributing to deforestation, whereas incomplete combustion leads to carbon dioxide release into the general environment. Perhaps the most important effects, however, are the health consequences on women and children from exposure to open burning of these materials. Men are often physically removed from household fuel burning during the day, but women, who do most of the cooking, and the children they care for, can be exposed for many hours per day to levels of byproducts that far exceed the safe exposure levels designated by the US Environmental Protection Agency (US EPA). The byproducts of incomplete combustion include particulate matter, carbon monoxide, sulfur dioxide, nitrogen oxides, polyaromatic hydrocarbons, chlorinated dioxins, arsenic, lead, fluorine, vanadium, and more than 200 additional chemical compounds.

The WHO designates indoor or household air pollution as a leading environmental cause of death, accounting for 2.7% of the annual worldwide global burden of disease. Approximately 5% of deaths in countries using unprocessed fuel are attributable to household air pollution, or nearly 2 million deaths annually. As many as half of these deaths occur among children younger than 5 years of age, for whom the best documented health consequence is an increase in acute lower respiratory infections (ARI), especially pneumonia. Several studies show a two- to threefold increased risk for ARI among children exposed to solid fuel burning compared with unexposed children after adjusting for confounding variables, including socioeconomic status. In addition, asthma rates are suggested to be higher among exposed children, but data are still emerging with some contradictory studies published. However, particulates, which are elevated with exposure to open solid fuel burning, have been associated with asthma symptoms, so it is very possible that exposure to indoor air pollution does have an effect on asthma symptoms.

Among adults, the leading health effects of open solid fuel exposure are non-smokers' chronic obstructive pulmonary disease and lung cancer. Women exposed to household air pollution have double the risk of developing lung cancer as nonexposed women; household air pollution is the primary risk for lung cancer in women in developing countries whereas cigarette smoking and occupational exposure were the leading associations for men.

Additional health associations have been noted for household air pollution, though the data are still emerging for several of these diseases. Other conditions that have been associated include otitis media, cataracts, and tuberculosis. Several studies have linked

household air pollution to lower birthweight; a proposed mechanism is that elevated maternal carbon monoxide levels lead to hypoxia and limit fetal growth.

Although the primary effects of household air pollution are seen in the developing world, more affluent populations are not entirely immune to the consequences of indoor exposures. Many families in the developed world continue to burn solid fuels (eg, fireplaces and wood stoves) for heating. Intriguing new information suggests that byproducts of gas cooking in the developed world may have an effect on infant neurodevelopment. Two studies from Spain found lower scores on Bayley Scales of Infant Development among children who lived in households with gas cookers.

To address the health and environmental effects of household air pollution, the United Nations Foundation established the Global Alliance for Clean Cookstoves (http:// cleancookstoves.org). The goal of this effort is to engage developing populations in designing and distributing efficient cookstoves and fuels with plans to have 100 million homes adopting their use by 2020.

PREP Pearls

- Indoor air pollution, largely the result of incomplete combustion of biomass and solid cooking fuels, poses a major health risk to children in developing countries.
- Among children, the primary health consequence of exposure to household air pollution is an increase in acute lower respiratory infections.
- Indoor air pollution is a leading cause of non-tobacco-related lung cancer, particularly among women.

American Board of Pediatrics Content Specification(s):

- Recognize that household fumes (eg, from cooking) may be harmful to children

Suggested Reading:

- Kaplan C. Indoor air pollution from unprocessed solid fuels in developing countries. Review Environ Health. 2010;25(3):221-242
- Kodgule R, Salvi S. Exposure to biomass smoke as a cause for airway disease in women and children. Curr Opin Allergy Clin Immunol. 2012;12(1):82-90. doi:10.1097/ACI.0b013e32834ecb65
- Martin WJ, Glass RI, Balbus JM, Collins FS. A major environmental cause of death. Science. 2011;334(6053):180-181. doi:10.1126/science.1213088.
- Vrijheid M, Martinex D, Aguilera I, et al. Indoor air pollution from gas cooking and infant neurodevelopment. Epidemiology. 2012;23:23-32. doi:10.1097/EDE.0b013e31823a4023

Item 95

A 4-month-old boy has an atrioventricular septal defect and underwent cardiac catheterization earlier today in preparation for surgical repair in 2 weeks. His current vital signs are a temperature of 37.4°C, a heart rate of 112 beats/min, blood pressure of 75/50 mm Hg, and a respiratory rate of 20 breaths/min. His oxygen saturation is 80% on 1 L/min of oxygen via nasal cannulae. Physical examination reveals an infant who is beginning to awaken from anesthesia and appears restless. Cardiac examination reveals a prominent first heart sound with a fixed splitting of the second heart sound consistent with his underlying diagnosis. A grade 3 systolic ejection murmur is detected. His breath sounds are shallow, coarse, and diminished bilaterally. Shortly after the examination, the patient becomes acutely agitated, with a rapid decrease in oxygen saturation to 65%. His heart rate is now 145 beats/min, and his blood pressure is 80/55 mm Hg. You rapidly initiate maneuvers to improve his oxygen saturation.

Of the following, the treatment MOST likely to improve his oxygen saturation is

A. albuterol nebulized treatment
B. epinephrine infusion
C. naloxone intravenously
D. sodium bicarbonate bolus intravenously
E. supplemental oxygen administration

Item 95 Preferred Response: E

Neonatal circulation normally makes a transition to an adult circulation during the first 6 to 8 weeks after birth, with marked decreases in pulmonary resistance and pulmonary artery pressures. In some infants, communications between the arterial and venous systems may exist such as persistent ductus arteriosus, aortopulmonary collaterals, atrial septal defects, and ventricular defects which result in left to right blood flow. The resultant increased right-sided blood flow leads to decreased pulmonary compliance, heart failure, and pulmonary hypertension. Pulmonary hypertension can arise from a combination of nonrelaxation of pulmonary smooth muscle after birth, hypoxia-induced pulmonary vasoconstriction, and smooth muscle proliferation. Persistent left to right vascular shunting can eventually result in loss of the normal pulmonary reactivity and irreversible pulmonary hypertension.

Other causes of pulmonary hypertension include pulmonary disease (chronic lung disease, interstitial lung disease), primary pulmonary vascular diseases (idiopathic, thromboembolic), obstructive sleep apnea, and chest wall abnormalities. Cor pulmonale is defined as right-sided heart failure (dilation or hypertrophy) that results from pulmonary hypertension. Primary heart disease (acquired or congenital) is not considered a cause of cor pulmonale.

Treatment of pulmonary hypertension depends on the etiology and symptoms at presentation. Management can include oxygen, diuretics, anticoagulation, and pulmonary vasodilators (nitric oxide, prostanoids, and sildenafil among other agents). Congenital heart lesions are normally surgically repaired before the development of pulmonary pressure. Lung or heart-lung transplantation may be considered in some patients if other measures fail.

The clinical findings in the patient described in the vignette are consistent with an acute elevation of pulmonary artery pressure which can be triggered by several factors, including hypoxia, hypercarbia, and metabolic acidosis. Treatment for this patient would include administration of oxygen and sedation with progression to more aggressive management of his oxygenation and ventilation if the episode does not resolve. Although acidosis can be an important cause of pulmonary hypertension, management of ventilation and prevention of hypercarbia are the first steps after oxygen administration and sedation. The evidence for the effectiveness of sodium bicarbonate for acute treatment of metabolic acidosis is controversial and is often avoided because of potential complications. There is no evidence of bronchospasm on examination to indicate the use of albuterol or narcotic overdose for which naloxone would need to be given to reverse respiratory depression. Finally, the patient does not have evidence of hypotension and the initiation of an epinephrine infusion would raise systemic vascular resistance and potentially worsen the pulmonary hypertension.

PREP Pearls

- Persistent left-to-right vascular shunting can eventually result in irreversible pulmonary hypertension.
- Treatment of pulmonary hypertension can include oxygen, diuretics, anticoagulation, and pulmonary vasodilators (nitric oxide, prostanoids, and sildenafil among other agents).
- Congenital heart lesions are normally surgically repaired before the development of irreversible pulmonary hypertension.

American Board of Pediatrics Content Specifications:

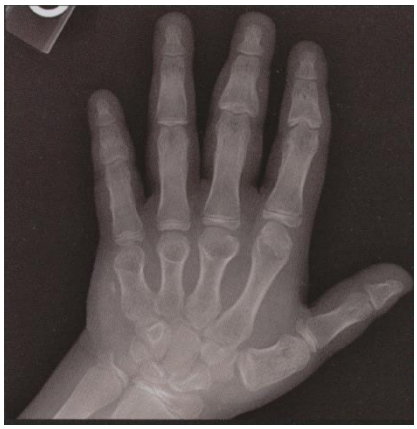
- Know that pulmonary hypertension is potentially reversible
- Know the situations in which pulmonary hypertension and cor pulmonale may occur

Suggested Reading:

- Aschner JL, Poland RL. Sodium bicarbonate: basically useless therapy. Pediatrics. 2008:122:831-835. doi: 10.1542/peds.2007-2400
- Bernstein D. Pulmonary Hypertension. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier, 2011:1600-1602
- Kung GC, Triedman JK. Pathophysiology of left to right shunts. UptoDate. Available online only for subscription

Item 96

A 2-year-old girl has intermittent facial spasms. She has been treated with levothyroxine since the first week after birth, when she was found to have an elevated thyroid-stimulating hormone level of 47 mIU/L on newborn screening. Findings on her physical examination are otherwise normal. Serum calcium level is 4.5 mg/dL (1.13 mmol/L) with a parathyroid hormone level of 103 pg/mL (103 ng/L) (reference range, 10-65 pg/mL [10-65 ng/L]). Radiographic studies reveal shortened metacarpals (Item Q96) and subcutaneous nodules. Her mother is healthy but is also known to have multiple subcutaneous nodules.



Of the following, the MOST LIKELY diagnosis is

A. hypoparathyroidism
B. hypopituitarism
C. multiple endocrine autoimmune syndrome
D. pseudohypoparathyroidism
E. vitamin D resistance

Item 96 Preferred Response: D

The infant described in this vignette has an elevated parathyroid hormone (PTH) level and a low calcium level, indicating resistance to PTH. This state of hormone resistance is called pseudohypoparathyroidism. Children with the most common form of pseudohypoparathyroidism often show signs of resistance to other endocrine hormones as well, such as thyroid-stimulating hormone (TSH), gonadotropins, and growth hormone—releasing hormone (GHRH).

Children with pseudohypoparathyroidism can initially have a normal phenotype, but by 5 years of age they often develop a classic constellation of phenotypic findings termed Albright hereditary osteodystrophy. Albright hereditary osteodystrophy includes: short stature, round facies with a low nasal bridge, obesity, disproportionate shortening of the limbs (particularly brachydactyly of the third, fourth, and fifth metacarpals and first distal phalanx), heterotopic ossification, and intellectual disability. This disorder, caused by a mutation in G-protein signaling, is inherited in an autosomal dominant manner with imprinting (the genetic trait is expressed only if an abnormal allele is transmitted from a specific parent, in this case the mother).

Hypoparathyroidism is not the correct response because with that disease the PTH level is low or not elevated in the face of a low serum calcium level. Hypoparathyroidism can be transient in the newborn period and is seen in infants of diabetic mothers, in premature infants, after birth asphyxia, and in intrauterine growth restriction. Additional causes of hypoparathyroidism are excessive phosphorus intake, hypomagnesemia, and DiGeorge syndrome. Hypocalcemia seen with the autoimmune polyendocrinopathies (also known as autoimmune polyendocrine syndrome type 1 or autoimmune polyendocrinopathy-candidiasis-ectodermal dystrophy [APECED]) also presents with low PTH levels. Hypopituitarism is not directly related to PTH regulation, and when hypothyroidism occurs due to hypopituitarism, there is a low to normal TSH level.

Vitamin D resistance is an autosomal recessive disorder of the vitamin D receptor. Two-thirds of affected children will develop alopecia, resulting from a lack of vitamin D receptor action within keratinocytes. The PTH level will be elevated, but vitamin D resistance does not cause hypothyroidism, as seen in the patient in the vignette.

PREP Pearls

- Hypoparathyroidism is associated with a low PTH level at the time of hypocalcemia.
- With pseudohypoparathyroidism, the PTH level is elevated at the time of hypocalcemia (PTH resistance).
- Pseudohypoparathyroidism patients can have resistance to multiple endocrine hormones and may have the phenotype of Albright hereditary osteodystrophy.

American Board of Pediatrics Content Specification(s):

- Recognize the typical laboratory findings associated with hypoparathyroidism

Suggested Reading:

- Mantovani G. Clinical review: pseudohypoparathyroidism: diagnosis and treatment. J Clin Endocrinol Metab. 2011;96:3020-3030. doi:10.1210/jc.2011-1048
- Pinsker JE, Rogers W, McLean S, Schaefer FV, Fenton C. Pseudohypoparathyroidism type la with congenital hypothyroidism. JPediatr Endocrinol Metab. 2006;19:1049-1052

Item 97

A 16-year-old boy has a 4-week history of progressive coughing spells. He had an upper respiratory infection at the onset of the illness that progressed to increasing coughing episodes with occasional posttussive vomiting. He has had difficulty sleeping over the past week because of the progression of the cough. His family recently moved to your community and is new to your practice. The parents have no record of his immunizations. You are awaiting diagnostic test results.

Of the following agents listed below, the BEST antibiotic for treating this patient is

A. amoxicillin
B. ciprofloxacin
C. doxycycline
D. erythromycin
E. trimethoprim-sulfamethoxazole

Item 97 S Preferred Response: D

For the boy in this vignette, the 3-week history of progressive respiratory illness with paroxysmal cough, especially in the setting of uncertain immunization status, suggests a diagnosis of pertussis. Nasopharyngeal swab for culture on selective media (eg, Reagan Lowe) remains the "gold standard" for diagnosis of pertussis. The specificity of culture is excellent, however the sensitivity is low; a negative test result does not rule out this diagnosis. Polymerase chain reaction (PCR) testing for pertussis appears promising and may be more sensitive than, and as specific as, culture. Presently, PCR testing for pertussis is not approved by the Food and Drug Administration, but the Centers for Disease Control and Prevention does provide guidelines for this procedure.

Given this likely diagnosis, erythromycin or another macrolide would be the antibiotic of choice. Resistance of Bordetella pertussis to macrolides is very rare. Because of the problems with laboratory diagnosis of pertussis and potential delay in getting results, antimicrobial therapy should be initiated before obtaining culture results. Antimicrobial therapy may ameliorate the symptoms of pertussis and help decrease transmission. Penicillins and first-generation cephalosporins such as cephalexin are not effective against B pertussis. Ciprofloxacin has excellent in vitro activity against B pertussis but there are no clinical trial data for this indication. In addition, fluoroquinolones are not approved for routine use in children and would only be recommended for children if an alternative agent were not available.

Although antimicrobial therapy might not have a significant clinical effect on the illness after 3 to 4 weeks of symptoms, treatment is still recommended to decrease transmission of the organism.

Trimethoprim-sulfamethoxazole is not the first-line choice for pertussis but it may have a role as an alternative agent in an individual who cannot tolerate macrolides.

Erythromycin is indicated for infections caused by Mycoplasma pneumonia, Chlamydia trachomatis, and Campylobacter species. It may also be used to treat group A streptococcal pharyngitis in penicillin- and cephalosporin-allergic individuals but macrolide resistance in this setting may be a concern in some areas. Erythromycin is considered an alternative to penicillin for treatment of diphtheria. Finally, erythromycin in a non-antibiotic role may be used to increase gastric emptying in select critically ill patients.

Gastrointestinal symptoms such as abdominal pain, nausea, and anorexia are the most common adverse events associated with erythromycin. Neonatal administration of erythromycin may be associated with an increased risk of hypertrophic pyloric stenosis. Cholestatic hepatitis has been reported with the estolate preparation of erythromycin, and liver functions have been observed to be elevated with erythromycin use. QTc prolongation has also been reported with erythromycin. Hypersensitivity reactions and anaphylaxis have been described rarely. Intravenous erythromycin may be associated with phlebitis at the infusion site. Drug interactions are another potential concern with erythromycin and other drugs metabolized by hepatic cytochrome P450 enzymes.

Examples of such drugs include rifampin, digitoxin, methylprednisolone, benzodiazepines, carbamazepine, warfarin, and zidovudine.

The newer macrolides clarithromycin and azithromycin are frequently used instead of erythromycin because they have a similar antimicrobial spectrum but offer the advantages of decreased frequency of dosing and less gastrointestinal upset. The newer agents also have an increased antimicrobial spectrum including Haemophilus influenza, atypical mycobacteria, and gastrointestinal pathogens.

PREP Pearls

- Erythromycin is indicated for Bordetella pertussis, Mycoplasma pneumonia, Chlamydia trachomatis, and Campylobacter species infections. It is also an acceptable alternative for treatment of group A streptococcal pharyngitis in allergic patients.
- The major adverse reactions associated with erythromycin are primarily related to the gastrointestinal tract.
- Erythromycin should be avoided in infants because of an increased risk of hypertrophic pyloric stenosis.

American Board of Pediatrics Content Specifications:

- Know the appropriate use of erythromycin
- Identify that the major adverse effects of the macrolide antibiotics

Suggested Reading:

- Bradley JS, Byington CL, Shah SS, et al. The management of community-acquired pneumonia in infants and children older than 3 months of age: clinical practice guidelines by the Pediatric Infectious Diseases Society and the Infectious Diseases Society of America. Clin Infect Dis. 2011;53:e25-e76. doi: 10.1093/cid/cir531
- Michelow IC, McCracken GH Jr. Antibacterial therapeutics, macrolides. In: Feigin RD, Cherry JD, Demmler-Harrison GJ, Kaplan SL, eds. Textbook of Pediatric Infectious Diseases. 6th ed. Philadelphia, PA: Elsevier; 2009;3199-3202

Item 98

A 6-year-old boy is seen in the emergency department with fever and knee swelling of 1 day's duration. Physical examination reveals a warm, erythematous, swollen knee with decreased range of motion due to pain. His temperature is 40°C and heart rate is 110 beats/min; the remainder of his physical examination findings and vital signs are within normal limits. A complete blood cell count reveals a white blood cell count of 15,000/µL (15.0×10^9/L) and platelets of 412.0×10^9/µL (412.0×10^9/L). The erythrocyte sedimentation rate is 50 mm/h. Arthrocentesis reveals white synovial fluid, a white blood cell count of 80,000/µL (80.0×10^9/L), and a red blood cell count of 0.1×10^3/µL (0.1×10^9/L). Gram stain and culture results are pending.

Of the following, the BEST antibiotic choice is

A. cefepime
B. cefotaxime
C. doxycycline
D. nafcillin
E. penicillin G

Item 98 Preferred Response: D

The child in this vignette has pyogenic (septic) arthritis, which is a medical emergency. A delay in treatment can result in permanent joint damage. Local symptoms include joint pain, swelling, limp, and refusal to use the affected joint. Systemic features of pyogenic arthritis include fever, malaise, and fussiness. On physical examination, the patient will appear ill with a single warm, erythematous, painful joint with decreased and painful range of motion. Aspiration of joint fluid can help make the diagnosis. Synovial fluid white blood cell counts are often greater than 50,000/µL. Gram stain and culture of the synovial fluid usually yields a causative organism but can be negative in up to 50% of cases. Ultrasonography will reveal a joint effusion, and magnetic resonance imaging can reveal areas of infection and damage in the joint, bone, and adjacent soft tissue. Younger children may have coexisting osteomyelitis. Appropriate management includes evaluation by orthopedics for drainage and debridement. Antibiotic therapy should not be delayed. Treatment of pyogenic arthritis is determined by the likely causative organism, which varies with patient age (Item C98).

The best antibiotic choice in a child older than 5 years of age is nafcillin, since the infection is likely due to Staphylococcus aureus or Streptococcus pyogenes. Cefotaxime might be a good choice in a child younger than 5 years of age if Streptococcus pneumoniae is a potential pathogen, but it should only be used in combination with another antibiotic. Doxycycline would be a good choice for Brucella-induced pyogenic arthritis, which is very unlikely in this patient. Cefepime provides gram-negative coverage, and penicillin G provides group A Streptococcus coverage; therefore, neither would be a good choice for the likely organisms in this case.

PREP Pearls

- Pyogenic arthritis requires prompt treatment to prevent permanent joint damage.
- Pyogenic arthritis should be suspected in any patient who has a single affected joint and fever.
- The recommended antibiotic therapy for pyogenic arthritis varies with age and associated risk factors; naficillin is the best choice for a child greater than 5 years of age.

American Board of Pediatrics Content Specifications

- Know the appropriate antibiotic management of pyogenic arthritis

Suggested Reading:

- Gutierrez K. Bone and joint infections in children. Pediatr Clin N Am. 2005;52(3):779-794. doi:10.1016/j.pc1.2005.02.005
- John J Chandran L. Arthritis in children and adolescents. Pediatr Rev. 2011;32(11):470-480. doi:10.1542/pir.32-11-470

Item 99

In your office, you are seeing a 16-year-old girl who for the past month has had symptoms of depression, including insomnia, irritability, loss of ability to concentrate, decreased physical activity, and worsening of academic performance. She recently started seeing a psychologist who has referred her to you for consideration of starting a medication to treat the depression because of the severity of her symptoms. The patient and her mother are interested in starting a medication today. You decide to start fluoxetine, a selective serotonin reuptake inhibitor.

Of the following, the BEST next step in management is to

A. ask the patient and her family to help monitor for any new irritability or self-harm thoughts
B. instruct the patient she must avoid pregnancy while taking fluoxetine
C. order a complete blood cell count to be performed in 2 months
D. order a serum sodium blood test in 4 weeks
E. schedule the patient for a follow-up in 4 weeks

Item 99 S Preferred Response: A

Approximately 5% of young people who start taking a selective serotonin reuptake inhibitor (SSRI) medication will experience irritability or agitation as an adverse effect during the first month of use. It is essential to screen for this adverse effect, along with the related but less frequent occurrence of new or worsening thoughts of self-harm or suicidality during the first month of administration. The United States Food and Drug Administration (FDA) currently recommends that "all patients being treated with antidepressants for any indication should be monitored appropriately and observed closely for clinical worsening, suicidality, and unusual changes in behavior, especially during the initial few months of a course of drug therapy, or at times of dose changes, either increases or decreases:' It is therefore strongly recommended to have contact with the patient and family (by appointment or by phone) within the first 1 to 2 weeks of starting an SSRI to screen for the occurrence of such adverse effects. Parents should be informed that the beneficial results of SSRIs may not be observed for up to 6 weeks of treatment; 2 weeks is too early to determine if the medication is effective.

Previously, the US Food and Drug Administration (FDA) had recommended that children who receive SSRIs be followed weekly (in person or by telephone) for the first 4 weeks of treatment, biweekly for the second month, and then at least monthly (in person) for continuing treatment: this recommendation was rescinded because of a lack of supporting evidence and because the monitoring plan was unrealistic for prescribers to follow. A clinically reasonable monitoring approach for the girl described in the vignette would be follow-up appointments every 2 weeks during the first month of treatment.

All but one of the SSRIs are listed as pregnancy category C (some animal studies show adverse effects; no controlled studies in humans). Paroxetine is risk category D (positive evidence of risk to human fetus), so alternatives should generally be used instead of paroxetine. Infant "jitteriness" has been reported for the first few days after delivery by a mother using SSRI medications, and prolonged hospitalizations or poor initial feeding have been reported rarely, which are also transient problems. Therefore, there is no indication for patients to avoid pregnancy if they are using any SSRI other than paroxetine. SSRIs have been associated with prolonged bleeding times, primarily in geriatric patients with concomitant administration of nonsteroidal antiinflammatory drugs. SSRIs can prolong bleeding times by their effect on the serotonin signaling process in platelet aggregation, which cannot be detected on a complete blood count. Hyponatremia has been occasionally reported as an SSRI adverse effect, primarily in geriatric patients. Routine monitoring of serum sodium concentration in children is not recommended. Waiting 4 weeks to follow up a patient is too long after the initiation of an SSRI.

PREP Pearls

- Suicidal thoughts could be caused by an SSRI during the first few months of use.
- One to 2 weeks after starting an SSRI, providers need to ask patients if they have any new irritability or agitation or self-harm thoughts.
- SSRIs can occasionally prolong bleeding times.

AAP Mental Health Competency:

- Know how to provide accurate patient/family counseling regarding the use of SSRI medications and risks like side effects and suicidality

Suggested Reading:

- Birmaher B, Brent D; AACAP Work Group on Quality Issues, Bernet W, et al. Practice parameter for the assessment and treatment of children and adolescents with depressive disorders. JAm Acad Child Adolesc Psychiatry. 2007;46:1503-1526
- United States Food and Drug Administration. Revisions to Product Labeling. www.fda.gov

Item 100

You are called to attend the vaginal delivery of a woman who presented completely dilated at 29 weeks of gestation. The newborn emerges limp without any respiratory effort. After gently suctioning and drying the newborn, you begin bag-mask ventilation with 30% oxygen. A pulse oximetry probe is placed on the right hand. The heart rate slowly begins to rise over 100 beats/min after 30 seconds of effective ventilation, and the pulse oximeter reads 90% oxygen saturation. The infant begins to have weak spontaneous respiratory effort associated with deep retractions. A decision is made to intubate, and a 3 min endotracheal tube is placed on the first attempt. At 5 minutes after birth, the newborn continues to require positive pressure ventilation with 30% oxygen.
The newborn is centrally pink with a heart rate of 140 beats/ min and continues with a weak spontaneous respiratory effort while intubated. The newborn has some flexion and grimaces upon intramuscular injection of vitamin K.

Of the following, the MOST appropriate Apgar score to assign at 5 minutes is

A. 5
B. 6
C. 7
D. 8
E. 9

Item 100 TE Preferred Response: B

The appropriate Apgar score to assign to the premature infant in the vignette at 5 minutes after birth is 6. Virginia Apgar developed the Apgar score in 1952 as a method to assess the clinical status of an infant immediately after birth. The Apgar score is composed of 5 components, which include heart rate, respiratory rate, muscle tone, reflex irritability, and color. Each component is assessed at 1 and 5 minutes after birth and assigned a score of 0, 1, or 2 (Item C100, page C-80). Five minutes after birth, the infant in the vignette has a heart rate of more than 100 (2 points for heart rate), weak respiratory effort while receiving positive pressure ventilation (1 point for respiratory effort), some flexion (1 point for muscle tone), a grimace (1 point for reflex irritability), and is centrally pink (1 point for color). The Apgar score that would be assigned to this infant at 5 minutes is 6. Investigators have documented a relationship between a lower Apgar score and lower gestational age. Respiratory effort, muscle tone, and reflex irritability are dependent on the physiological maturity of an infant. These components may be scored lower in the premature infant and are associated with the decreased Apgar score seen with lower gestational age. Other factors such as infection, hypovolemia, hypoxia, maternal medications, birth trauma, or congenital anomalies may also have a negative effect on the Apgar score. A healthy premature infant may receive a low Apgar score without any other contributing factor other than prematurity.

Individual variability exists in the assignment of an Apgar score when an infant requires resuscitation and intubation. It is recommended that an infant who has apnea and requires intubation and ventilation should receive a score of 0 because of the lack of respiratory effort, even if artificial ventilation is effective. If the infant receiving positive pressure ventilation has irregular or shallow spontaneous respirations, he or she should be assigned a score of 1.

Item C100. The Apgar Score

The Apgar Score	0	1	2
Heart rate	Absent	<100 beats per min	>100 beats per min
Respiratory effort	Absent	Weak cry; hypoventilation	Good cry
Muscle Tone	Flaccid	Some flexion	Active motion/well flexed
Reflex irritability	No response	Grimace	Cry/cough/sneeze
Color	Blue/pale	Acrocyanotic	Completely pink

From Warren JB, Phillipi CA. Care of the newborn. *Pediatr Rev.* 2012;33(1):4-18

PREP Pearls

- A healthy premature infant may receive a low Apgar score without any other contributing factor other than prematurity because of decreased respiratory effort, muscle tone, and reflex irritability associated with physiologic immaturity.

American Board of Pediatrics Content Specifications

- Recognize that very-low-birth-weight infants often cannot achieve an Apgar score greater than 6 because they are neurologically immature (eg, hypotonic, blunted response to noxious stimuli)

Suggested Reading:

- American Academy of Pediatrics, Committee on Fetus and Newborn; American College of Obstetricians and Gynecologists, Committee on Obstetric Practice. The Apgar score. Pediatrics. 2006;117:1444-1447. doi: 10.1542/peds.2006-0325
- Hegyi T, Carbone T, Anwar M, et al. The Apgar score and its components in the preterm infant. Pediatrics. 1998;101:77-81. http://pediatrics.aappublications.org/content/101/1/77.full
- Lopriore E, van Burk GF, Walther FL de Beaufort AL Correct use of the Apgar score for resuscitated and intubated newborn infants: questionnaire study. BMJ. 2004;329:143-144. doi: 10.1136/bmj.38117.665197.F7

Item 101

A 5-year-old boy is hospitalized for hypoxia and respiratory distress associated with bacterial pneumonia. He is receiving supplemental oxygen and parenteral antibiotics. On the second hospital day, you are called to his bedside because he is noted to be pale. His oral temperature is 37.9°C, pulse rate is 110 beats/min, respiratory rate is 28 breaths/ min, and blood pressure is 100/55 min Hg. On examination, the child is alert and pale. He has mildly icteric sclerae and a grade 2/6 systolic ejection murmur. The remainder of the physical examination is normal. The following are the results of the child's laboratory tests:

- White blood cell count, 16,500/µL (16.5 x 10^9/L), with 62% polymorphonuclear leukocytes, 30% lymphocytes, 7% monocytes, and 1% eosinophils
- Hemoglobin, 5.0 g/dL (50 g/L)
- Mean corpuscular volume, 80/μm^3 (80 fl,)
- Platelet count, 445 x 10^3/uL (445 x 10^9/L)
- Reticulocyte count, 10% (0.10)
- Direct antiglobulin test (direct Coombs), positive for IgG, negative for C3
- Indirect antiglobulin test (indirect Coombs), negative upon review of his medical record, his hemoglobin on admission was 10.5 g/dL (105 g/L).

Of the following, the MOST likely cause of the acute anemia in this child is

A. aplastic crisis
B. cold agglutinin disease
C. hemolytic uremic syndrome
D. inconclusive because of conflicting Coombs results
E. warm agglutinin autoimmune hemolytic anemia

Item 101 Preferred Response: E

The child described in the vignette has the clinical presentation and laboratory results most consistent with a warm agglutinin autoimmune hemolytic anemia (AIHA). Autoimmune hemolytic anemia in infants and toddlers often occurs after an infection, whereas in adolescents it is more likely to be associated with an underlying systemic disease. Although infections are more commonly associated, there is evidence that antibiotics, such as cephalosporins, can also lead to AIHA. The most common form of primary AIHA in children involves warm-reactive IgG autoantibodies that bind to the red blood cell (RBC) at 37°C and lead to extra-vascular hemolysis. A second form of primary AIHA in children is paroxysmal cold hemoglobinuria. This is common after viral illnesses and is caused by an IgG autoantibody that leads to intravascular hemolysis at cold temperatures but binds complement at 37°C. A third form of primary AIHA is IgM-mediated cold-agglutinin disease, which can cause extravascular or intravascular hemolysis. Although this entity is more common in adults, it can occur in children in association with Mycoplasma infections.

Children with AIHA often present with pallor and weakness due to the anemia, with hemoglobin concentrations as low as 4 to 7 g/dL (40-70 g/L). The spleen may be mildly enlarged and palpable due to an increase in red pulp. A cardiac flow murmur and tachycardia may be present as a result of the high-output state caused by the anemia. The hemolysis can cause an increase in unconjugated bilirubin, leading to jaundice, which is especially noticeable in the sclerae and palms. Dark urine may be present in patients with intravascular hemolysis. Lactate dehydrogenase or aspartate aminotransferase concentrations can be elevated because these are released from the erythrocyte during hemolysis. In warm-reactive AIHA, the peripheral blood smear reveals numerous small spherocytes formed by the ingestion of part of the erythrocyte membrane by the spleen, causing the cells to take on a spherical shape. Reticulocytosis, up to 10% to 30% of the circulating RBC count, is the bone marrow's compensatory response to the shortened erythrocyte life span in the peripheral blood, although reticulocytopenia has been reported in 10% of pediatric patients.

The prognosis for AIHA in children is encouraging, with 77% of children with AIHA having a self-limited course and most children responding well to short-term therapy. The treatment of AIHA depends on the degree of anemia and type of AIHA. Observation alone is reasonable for the patient with mild anemia (hemoglobin concentration >9 g/dL [>90 g/L]) and no evidence of cardiovascular compromise. For the child in the vignette with a severely low hemoglobin con-centration, tachycardia, and a warm IgG autoantibody, the mainstay of treatment is corticosteroids and packed RBC transfusions. Plasmapheresis and immunoglobulin intravenous may be indicated as second-line therapies if corticosteroids and packed RBC transfusions are unsuccessful.

The direct antiglobulin test (DAT, formerly known as the direct Coombs test) detects antibodies or complement proteins bound to the surface of the erythrocytes. The patient's RBCs are washed to remove plasma proteins and then incubated at 37°C with the Coombs reagent, which is an antiserum that binds to human γ-globulin and complement (eg, C3). When the Coombs reagent is added, it binds to the autoantibodies on the RBC

surface, if present, and causes agglutination of the RBCs. Further testing is performed to distinguish between IgG and complement antibodies on the RBC surface. The indirect antiglobulin test (IAT) detects unbound circulating antibodies in the patient's serum. This is useful in pregnant women and before blood transfusions but is not the appropriate test to diagnose AIHA.

The patient described in the vignette does not have laboratory evidence of aplastic crisis because his reticulocyte count and leukocyte count are both elevated and his platelet count is at the high end of the normal range. Cold agglutinin disease is unlikely because it is usually IgM mediated and seen in older patients. Hemolytic uremic syndrome (HUS) can have hemolysis as a feature, but it is usually associated with a gastrointestinal infection with diarrhea, acute renal insufficiency, and thrombocytopenia. Patients with HUS typically do not have a positive DAT result. The DAT and IAT results are not conflicting and support the diagnosis of warm agglutinin AIHA in this patient.

PREP Pearls

- A positive DAT result is consistent with AIHA.
- AIHA is a usually associated with infection or antibiotic use in young children.
- Corticosteroids and transfusion with packed RBCs are the mainstays of treatment for AIHA in children.
- A DAT (Coombs test) is indicated in any patient presenting with a previously undiagnosed hemolytic anemia.

American Board of Pediatrics Content Specifications:

- Understand that direct and indirect Coombs tests are a necessary part of the evaluation of a child with acute-onset anemia

Suggested Reading:

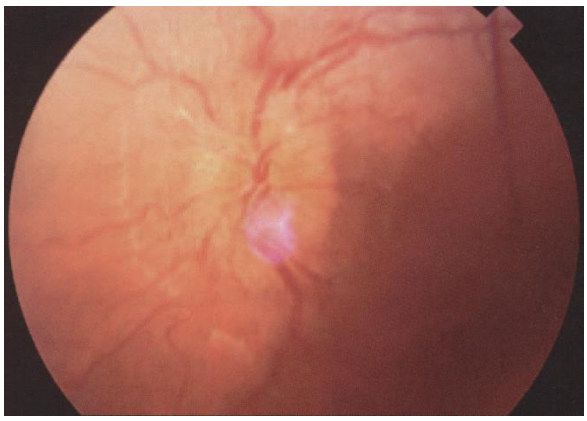
- Garratty G. Drug-induced immune hemolytic anemia. ASH Education Book. 2009;2009:73-79. doi:10.1182/asheducation-2009.1.73
- Michel M. Classification and therapeutic approaches in autoimmune hemolytic anemia: an update. Expert Rev Hematol. 2011;4:607-618. doi:10.1586/EHM.11.60
- Ware RE. Autoimmune hemolytic anemia. In: Nathan and Oski's Hematology of Infancy and Childhood. 7th ed. Philadelphia, PA: Saunders Elsevier; 2009:613-658
- Zantek ND, Koepsell SA, Tharp DR Jr, Cohn CS. The direct antiglobulin test: a critical step in the evaluation of hemolysis. Am I Hematol. 2012;87:707-709. doi:10.1002/ajh.23218

Item 102

A 16-year-old boy has had constant daily headache for 1 month. The headache is all over his head; it comes and goes but never fully resolves. The pain worsens with coughing, sneezing, and laughing. He has mild nausea and photophobia and ringing in his ears. He reports that his vision "grays out" sometimes but he does not have tunnel vision or visual loss. His past medical history is notable for acne, asthma, and attention-deficit/hyperactivity disorder (ADHD). He is currently taking oral isotretinoin for his acne, oral montelukast and inhaled fluticasone for his asthma, and atomoxetine for his ADHD. He also takes vitamin B12 supplements and riboflavin as natural remedies for headache. There is no family history of migraine. On physical examination, his weight is 65 kg, height is 178 cm, and blood pressure is 102/76 mm Hg. His funduscopic examination is shown in Item Q102 (both eyes exhibit similar findings). The remainder of his physical examination findings is normal. Results of magnetic resonance imaging of the brain are normal. Lumbar puncture is performed in the lateral decubitus position with legs extended, and the opening pressure is 340 mm H20. Cerebrospinal fluid protein is 13 mg/dL and glucose is 64 mg/dL, and there are 3 white blood cells/µL and 204 red blood cells/ 4L.

Of the following, the medication MOST likely to cause the boy's symptoms and signs is

A. atomoxetine
B. isotretinoin
C. montelukast
D. riboflavin
E. vitamin B12



ITEM Q102: Funduscopic findings as described for the boy in the vignette.

Item 102 S TE Preferred Response: B

The boy in the vignette has pseudotumor cerebri. This presents with headache, nausea, and vomiting—similar to migraine headaches—but often with a history of transient visual obscurations or "gray outs:' pulse synchronous tinnitus, or worsening headache with Valsalva maneuver (laughing, sneezing, or coughing, for example). The absence of a family history of migraine also suggests that this is not a migraine headache. In pseudotumor cerebri, swollen optic nerves with blurry disc margins are the most important finding. Cranial nerve VI palsy also may be present. On formal visual field testing, a deficit can be detected but this is asymptomatic early on. Risk factors for pseudotumor cerebri include recent weight gain and certain medications. The most common medications are vitamin A derivatives such as isotretinoin and tetracycline antibiotics, such as minocydine and doxycycline. In the absence of known risk factors, pseudotumor cerebri can be termed idiopathic intracranial hypertension.

Diagnosis is based on the history, normal brain imaging, and measurement of an elevated cerebrospinal fluid opening pressure. Typical opening pressure in the pediatric age group is less than 280 mm H2O. Once the diagnosis is made, treatment should be initiated promptly to avoid permanent vision loss. Acetazolamide or topiramate are first-line medications that work by decreasing cerebrospinal fluid production. An ophthalmologist should follow patients with pseudotumor cerebri to ensure improvement in vision and visual fields.

Of the medications listed in the vignette, only isotretinoin is a risk factor for pseudotumor cerebri. In patients with new headaches, it is important to consider pseudotumor cerebri on the differential diagnosis, evaluate any medications the patient is taking, and obtain a recent weight change history.

PREP Pearls

- Risk factors for developing pseudotumor cerebri include weight gain, obesity, and use of common medications such as isotretinoin and tetracycline.
- Initial diagnostic evaluation for pseudotumor cerebri includes neuroimaging followed by lumbar puncture.

American Board of Pediatrics Content Specification(s):

- Know the common causes of pseudotumor cerebri

Suggested Reading:

- Avery RA, Shah SS, Licht DJ, et al. Reference range for cerebrospinal fluid opening pressure in children. N Engl 1 Med. 2010;363(9):891-893. doi: 10.1056/NEJMc1004957
- Mercille G, Ospina LH. Pediatric idiopathic intracranial hypertension: a review. Pediatr Rev. 2007;28(11):e77-86. doi: 10.1542/pir.28-11-e77

Item 103

You are counseling a couple whose 6-month-old son has Down syndrome and is scheduled for a ventricular septal defect (VSD) repair. He is otherwise well and has been gaining weight appropriately while taking digoxin and furosemide for mild congestive heart failure. The parents ask about complications and whether their child's Down syndrome will put him at greater risk than children who undergo VSD repair and do not have Down syndrome.

Of the following, you are MOST likely to tell the parents that their son is at

A. greater risk for anoxic brain injury
B. greater risk for postoperative fatal arrhythmia
C. greater risk for postoperative respiratory complications
D. greater risk for postoperative seizures
E. no greater risk of perioperative complications

Item 103 TE SBP Preferred Response: C

The infant described in the vignette is doing well despite his clinically significant ventricular septal defect (VSD) and diagnosis of Down syndrome. Infants with Down syndrome, as well as infants with other genetic conditions, may be at increased risk for perioperative and postoperative complications, so care must be taken to anticipate potential problems. When compared to control infants who undergo VSD repair, infants with Down syndrome have significantly prolonged postoperative lengths of stay and complications, including a threefold increase in infections, a greater than twofold increase in respiratory complications, a threefold increase in pulmonary hypertension, and a threefold increase in atrioventricular block (but not fatal arrhythmias) that requires a pacemaker. There does not appear to be an increased risk for anoxic brain injury or postoperative seizures.

Infants with certain cytogenetic conditions are at significant risk for concomitant cardiac defects, including Down syndrome (50% risk, often septal defects), trisomy 18, trisomy 13, 22q11 microdeletion (>80% risk of a heart defect, often conotruncal and great artery defects), and Turner syndrome (20%-40% risk, often coarctation of the aorta, aortic stenosis, or bicuspid aortic valve). Also, infants with fetal alcohol syndrome have a significant risk for cardiac anomalies, most often septal defects. Therefore, infants with these diagnoses should undergo echocardiography to rule out a critical cardiac defect.

PREP Pearls

- Infants with Down syndrome undergoing USD repair have a higher risk for postoperative respiratory complications and prolonged length of stay.
- Infants with cytogenetic abnormalities or fetal alcohol syndrome should have echocardiography performed to assess for congenital heart defects.

American Board of Pediatrics Content Specification(s):

- Recognize the increased risk and plan appropriate evaluation of congenital heart disease in a newborn infant with congenital anomalies (eg, trisomy 21, trisomy 18, fetal alcohol syndrome, 22q11 microdeletion, 45,X0)

Suggested Reading:

- Bloemers BLP, van Furth AM, Weijerman ME, et al. Down syndrome, a novel risk factor for respiratory syncytial virus bronchiolitis: a prospective birth-cohort study. Pediatrics. 2007;120(4):e1076-e1081. doi:10.1542/peds.2007-0788
- Botto LD, May K, Fernhoff PM, et al. A population-based study of the 22q11.2 deletion: phenotype, incidence, and contribution to major birth defects in the population. Pediatrics. 2003;112(1):101-107
- Frias JL, Davenport ML; Committee on Genetics, Section on Endocrinology. Health supervision for children with Down syndrome. Pediatrics. 2003;111:692-702. doi:10.1542/peds.111.3.692

- Fudge JC, Li S, Jaggers J, O'Brien SM, et al. Congenital heart surgery outcomes in down syndrome: analysis of a national clinical database. Pediatrics. 2010;126(2):315-322. doi:10.1542/peds.2009-3245
- Health supervision for children with Down Syndrome. Pediatrics. 2011;128(2):393-406. doi:10.1542/peds.2011-1605
- Schieve LA, Boulet SL, Boyle CB, Rasmussen SA, Schendel D. F of children 3 to 17 years of age with Down syndrome in the 19S National Health Interview Survey. Pediatrics. 2009;123(2):e253 doi:10.1542/peds.2008-1440

Item 104

The mother of a 4-year-old boy brings him to your office because he "looks flushed and is acting strangely." There is no history of trauma or recent illness. The boy has been healthy and takes no medications. His symptoms began 1 hour ago. His mother is concerned that he has meningitis because earlier in the morning, another child at his preschool was sent home with fever and headache. The boy has a temperature of 38. VC, heart rate of 156 beats/min, blood pressure of 120/84 mm Hg, and respiratory rate of 28 breaths/min.

The boy appears anxious. He is intermittently picking at his shirt and skin. He looks toward his mother when she speaks to him, but he seems confused and does not respond to questions in a coherent manner. On physical examination, his skin is warm and flushed and mucous membranes are dry. His pupils are both 6 mm in diameter and minimally reactive. You find no signs of traumatic injury or meningismus. His abdomen is soft and nontender, although his bowel sounds are diminished.

As you are interviewing the mother further about the boy's activities prior to symptom onset, his older sister interrupts and states that she saw her brother eating some "candy" out of their mother's purse earlier today. The mother acknowledges that she keeps a variety of over-the-counter medications in her purse to "be ready for emergencies"

Of the following, the medication MOST likely to cause the patient's signs and symptoms is

A. acetaminophen
B. aluminum hydroxide
C. chlorpheniramine
D. loperamide
E. simethicone

Item 104 S Preferred Response: C

The boy in the vignette presents with acute onset of skin flushing and behavioral changes, which began shortly after he ingested an unknown medication (which he thought was candy) from his mother's purse. His symptoms, along with the findings noted on his physical examination– hyperpyrexia, tachycardia, hypertension, dry mucous membranes, flushed skin, mydriasis, decreased bowel sounds, confusion, and apparent hallucinations—are most consistent with ingestion of chlorpheniramine, an antihistamine with anticholinergic properties.

More than 600 compounds, including prescription drugs, over-the-counter medications, and plants, have anticholinergic properties. Examples of classes of medications with anticholinergic properties include antihistamines (eg, chlorpheniramine and diphenhydramine), tricyclic antidepressants, sleep aids, over-the-counter cold preparations, atropine, scopolamine, and contaminated illicit drugs. A number of plants, including jimson weed (Datura stramonium) and deadly nightshade (Atropa belladonna), can also produce anticholinergic toxicity. Anticholinergic toxicity is encountered relatively commonly, and all pediatric practitioners should be able to recognize its signs and symptoms.

Anticholinergic agents are competitive antagonists with acetylcholine at the neuroreceptor site. The major effects of these drugs are on the myocardium, central nervous system (CNS), smooth muscle, and exocrine glands. The classic clinical findings associated with exposure are summarized by a well-known mnemonic that many pediatric practitioners learn during the course of medical training:

- Mad as a hatter (delirium, agitation, and hallucinations, resulting from blockade of muscarinic receptors in the CNS)
- Red as a beet (skin flushing due to cutaneous vasodilation)
- Dry as a bone (anhidrosis, resulting in dry skin)
- Blind as a bat (pupillary dilation with ineffective accommodation)
- Full as a flask (urinary retention)
-

Other clinical features not included in this mnemonic include tachycardia, hypertension, and diminished bowel sounds.

Other conditions that may cause altered mental status should be considered in the differential diagnosis for anti-cholinergic toxicity. Infectious processes, including meningitis, encephalitis, and sepsis, often result in hyperpyrexia, altered mental status, and tachycardia.

Because numerous drug classes and toxins possess anti-cholinergic properties, clinicians must differentiate toxicity of pure anticholinergic agents from poisonings in which the anticholinergic toxidrome is but one piece of the picture. An important example is poisoning by tricyclic antidepressants, which initially cause anticholinergic effects but then lead to cardiac conduction disturbances and profound hypotension through different pharmacologic mechanisms. Toxicity from sympathomimetic agents (such as amphetamines) and aspirin, as well as serotonin syndrome, can cause mental status

changes, tachycardia, and hyperpyrexia in children as well. The absence of dry flushed skin, anhidrosis, and urinary retention helps to distinguish these poisonings from anti-cholinergic toxicity.

Ingestion of acetaminophen would not explain the signs and symptoms displayed by the boy in the vignette. When ingested in small doses, acetaminophen generally causes minimal clinical symptoms. In cases of toxic acetaminophen overdose (>140 mg/kg), early clinical signs are nonspecific and include anorexia, nausea, vomiting, malaise, and diaphoresis.

Ingestion of aluminum hydroxide would be unlikely to result in significant clinical findings in this child. The most commonly reported adverse effects of this over-the-counter agent are gastrointestinal upset and headache.

Loperamide, a widely available over-the-counter agent used to control diarrhea, is chemically similar to opiate receptor agonists, such as diphenoxylate. Potential adverse effects of this agent include sedation, ataxia, bradycardia, hypotension, miosis, constipation, and dry mouth. The occurrence of respiratory depression in pediatric patients after ingestion of loperamide has been reported. Loperamide ingestion would not explain the tachycardia, hypertension, dilated pupils, and flushed skin found in the boy in this vignette.

Simethicone, an antifoaming agent used to decrease abdominal bloating and discomfort caused by excess gas in the stomach, does not cause dangerous adverse effects when ingested in isolation and would not lead to the signs and symptoms displayed by this boy. Child-resistant product packaging, increased parental awareness of potential household toxins, and more sophisticated medical interventions have all contributed to decreased morbidity and mortality from pediatric poisonings in the past few decades. However, poisoning remains an important preventable cause of injury in children and adolescents. Pediatricians should continue to educate parents about the importance of keeping all medications—both prescription and over-the-counter—and potentially toxic household products properly secured and out of the reach of children at all times.

PREP Pearls

- The classic clinical findings associated with anticholinergic exposure are summarized by the mnemonic, "mad as a hatter, red as a beet, dry as a bone, blind as a bat, and full as a flask."
- Other conditions that may cause altered mental status should be considered in the differential diagnosis for anticholinergic toxicity.
- Pediatricians should continue to educate parents about the importance of keeping all medications—both prescription and over-the-counter—and potentially toxic household products secured and out of the reach of children.

American Board of Pediatrics Content Specification(s):

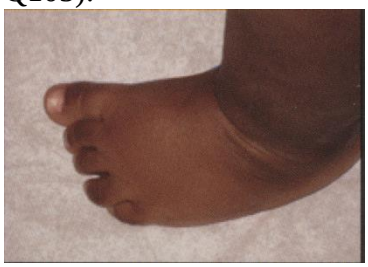
- Recognize the signs and symptoms of anticholinergic drug use

Suggested Reading:

- Osterhoudt KC, Ewald MB, Shannon M, Henretig FM. Toxicologic emergencies. In: Fleisher GR, Ludwig S, eds. Textbook of Pediatric Emergency Medicine. 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2010:1171-1223
- Su M, Goldman M. Anticholinergic poisoning. UptoDate. Available online only for subscription

Item 105

The parents of a 4-month-old male infant are concerned about his left foot. They report that the infant has appeared "pigeon-toed" since birth. On examination, you note medial deviation of the forefoot and the lateral border of the foot is convex. You are able to easily move the foot to a neutral position and abduct the forefoot past the midline (Item Q105).



ITEM Q105: Foot as described for the boy in the vignette.

Of the following, the BEST initial management is

A. immediate referral to orthopedics for probable surgical correction
B. radiologic imaging of the foot
C. serial manipulation and casting
D. stretching maneuvers of the forefoot
E. ultrasonography of the hips

Item 105 Preferred Response: D

The male infant described in the vignette has the classic features of metatarsus adductus, a congenital foot deformity common in newborns and characterized by adduction or medial deviation of the forefoot relative to the hindfoot. The incidence is estimated to be 1 in 1,000 to 5,000 births, and the most common cause is intrauterine molding. The incidence is higher in firstborn children and in twin births. Approximately 50% of affected infants have bilateral involvement. The diagnosis is made by the physical examination. Adduction of the forefoot with the hindfoot remaining in the neutral position creates the convex shape of the lateral border of the foot. A deep medial crease is often present, and the base of the fifth metatarsal appears prominent.

Almost all cases of mild to moderate metatarsus adductus will resolve without orthopedic intervention. The need for treatment is based on the rigidity of the deformity. Flexibility is based on the ability to correct the adduction by providing lateral pressure on the forefoot over the first metatarsal while firmly holding the heel in a neutral position with the other hand. Mildly affected feet that can be overcorrected into abduction with little effort may be observed. Passive stretching exercises are recommended for moderate foot deformities that will passively correct only to the neutral position. The outcome in these patients is excellent. With this minor manipulation that can be performed by parents, most infants correct by 4 to 6 months of age if the condition is diagnosed and treated early.

Surgical correction remains controversial. It may be an option in an older patient (4-6 years of age) who have severe persistent metatarsus adductus and exhibit difficulties with physical function or shoe wear; however, the rate of failure or complications is high. Management with orthotic splints or corrective shoes has been suggested to benefit those with moderate deformities but has not been found to be significantly effective. Radiography is not routinely performed but is indicated in toddlers or older children with persistent deformities. Serial casting is used for patients with more severe or rigid deformities that cannot be passively abducted to the midline. Results are best when treatment begins before 8 months of age. Although historically hip dysplasia had been associated with metatarsus adductus, more recent data have demonstrated no association. The hips should be examined carefully at every health supervision visit until the child is walking well as part of the normal physical examination, but ultrasonography of the hips is not necessary.

PREP Pearls

- The diagnosis of metatarsus adductus, a common congenital foot deformity in newborns, is made by the physical examination.
- The convex lateral border of the foot is due to adduction of the forefoot with the hindfoot remaining in the neutral position.
- The need for treatment is based on the rigidity of the deformity.
- If the forefoot is flexible and can be passively abducted to the midline, massage and stretching are usually sufficient to treat the deformity.

American Board of Pediatrics Content Specification(s):

- Understand that if the forefoot can be abducted past the midline, massage and exercise are usually sufficient to treat metatarsus valgus/varus

Suggested Reading:

- Smith BG. Lower extremity disorders in children and adolescents. Pediatr Rev. 2009;30:287-294. doi:10.1542/pir.30-8-287
- Hosalkar HS. Speigel DA, Davidson RS. Metatarsus adductus. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics 19th ed. Philadelphia, PA: Saunders Elsevier 2011:2335-2336
- McKee-Garrett TM. Lower extremity positional deformations. UptoDate. 2012. Available online only for subscription

Item 106

The parents of a 3-year-old boy would like him tested for allergies. The parents report that the boy has had worsening symptoms of itchy eyes, sneezing fits, and nasal congestion since the family got a new dog 1 year ago. The parents would like the boy tested to determine if they need to give the dog away. They are reluctant to stop the boy's daily antihistamine and are disappointed to learn that skin testing cannot be performed while taking this medication. You decide to obtain blood-specific IgE testing. However, the parents have read on the internet that the "scratch test" is a better test.

Of the following, you are MOST likely to advise the parents that in this situation, blood-specific IgE testing is

A. comparable to skin testing
B. less expensive and better tolerated by children than skin testing
C. more accurate than skin testing
D. the only testing that can be done because he is too young for skin testing
E. a preliminary test and you will obtain skin testing to confirm the results

Item 106 Preferred Response: A

For the child described in the vignette, serum-specific IgE (sIgE) testing is comparable with skin test results in terms of sensitivity and diagnostic properties and the child continues to receive antihistamines. Both the skin prick test (SPT) and sIgE testing are methods of detecting sIgE antibodies. Skin testing, used by allergy specialists, is a bioassay that detects the presence of allergen-specific IgE on cutaneous mast cells. Allergens introduced into the surface of the skin trigger allergen-specific IgE present on sensitized mast cells, causing activation and release of allergic mediators. The clinical result of these cellular events is a positive skin test result, or a transient "wheal and flare" reaction. This reaction consists of a central area of superficial skin edema (wheal) surrounded by erythema (flare) and is measured in 10 to 20 minutes. This pruritic reaction represents the immediate phase of the allergic reaction. Both a positive control of histamine dichloride and a negative control of diluent identical to that of the allergen extracts (usually glycerinated saline) should always be applied to verify that the patient's skin is normally responsive. A positive reaction is defined in 1 of 2 ways: (a) most commonly as a wheal that is equal or larger in size to that of the histamine control (with histamine producing at least a 3-mm wheal diameter) or (b) a wheal diameter 3 mm larger than the negative saline control.

Skin testing is typically well tolerated and can be performed in children of all ages. Several types of medications can interfere with skin testing, particularly H1 antihistamines that can suppress skin reactivity for 1 to 7 days, depending on the specific drug. Hence, skin testing will not be accurate in the child in the vignette currently receiving antihistamines.

A number of enzymatic assays that are based on antiIgE antibodies have replaced the radioallergosorbent test (RAST) that was used in the past. Federally licensed commercial laboratories often use automated systems capable of detecting and quantifying sIgE. Most commonly, laboratory reports are expressed in units reflecting concentrations of sIgE (eg, kU_A/L). Although the 3 commercial detection systems approved by the Food and Drug Administration have excellent performance characteristics (analytical sensitivity, 0.1 kU_A/L), the individual systems appear to detect different populations of IgE antibody or do not measure IgE antibodies with comparable efficiencies. Thus, a result for an allergen in 1 of the 3 test systems may not be equivalent to the same allergen tested in a different system. Despite that variance, in general, both sIgE tests and SPT are sensitive and have similar diagnostic properties (positive predictive accuracy, approximately 50%; negative predictive value, approximately 95%), advantages, and disadvantages. There are minor discrepancies between the tests regarding possible sensitivity to detect specific allergens (eg, SPT is preferred for evaluation of penicillin allergy), probably because different proteins or IgE binding sites are represented.

Advantages of the SPT include immediate results visualized by the patient and family and lower cost compared with sIgE tests. On the other hand, serologic tests are widely available. There is no need to withhold antihistamines before the tests and they can be performed on patients with extensive dermatitis. Disadvantages of sIgE include the need to obtain blood samples, delayed results, and cost.

It is important to be aware that a person with demonstrable IgE to a specific allergen is said to be "sensitized" to that allergen. Both skin testing and sIgE testing are used to demonstrate sensitization. However, a sensitized person is considered "allergic" to the allergen only if he or she has symptoms when exposed to it. This distinction is made because not all sensitized patients will develop actual symptoms on exposure. With some food skin tests, for example, the percentage of people who react to a food for which they have skin-tested positive can be as low as 50%. Thus, food challenge procedures are often necessary to clarify a patient's status. By comparison, the proportion of individuals with positive pollen skin test results who truly react is much higher; therefore, a positive skin test result and a history of symptoms during the appropriate season are sufficient to make the diagnosis. As a result, the confirmation of reactivity is crucial to making an accurate diagnosis and each type of allergy must be approached individually.

PREP Pearls

- Skin prick testing and serum-specific IgE testing are comparable in their diagnostic properties.
- Skin testing can be performed with minimal discomfort, can be readily visualized by patient and family, and is less expensive.
- The sIgE test can be performed regardless of medication intake, dermatitis, and dermatographism but requires a blood sample and results are delayed.
- The clinical history should always be taken into consideration when interpreting the skin prick test or sIgE test to differentiate sensitization from clinical allergy.
- A positive result for the skin test or sIgE test indicates sensitization only. Symptoms on exposure are required to consider a child "allergic."

American Board of Pediatrics Content Specification(s):

- Know that RAST testing correlates closely with results of skin tests

Suggested Reading:

- Bernstein IL, Li JT, Bernstein DI, et al; American Academy of Allergy, Asthma and Immunology; American College of Allergy, Asthma and Immunology. Allergy diagnostic testing: an updated practice parameter. Ann Allergy Asthma Immunol. 2008;100(3 suppl 3):S1-S148
- Sicherer SH, Wood RA; American Academy of Pediatrics Section on Allergy and Immunology. Allergy testing in childhood: using allergen-specific IgE tests. Pediatrics. 2012;129(1);193-197. doi:10.1542/peds.2011-2382

Item 107

A 17-year-old boy has recently been diagnosed as having autosomal dominant polycystic kidney disease. His parents have done some research and are worried about the risk of associated cerebral aneurysms and intracerebral bleeding.

Of the following, you are MOST likely to advise them that the boy

A. can have magnetic resonance angiography–based screening irrespective of his renal function
B. has 50% risk for having cerebral aneurysm
C. has a higher risk for intracerebral bleed if the size of the aneurysm is more than 7 mm
D. will need routine yearly screening for identifying cerebral aneurysms
E. will need surgical correction if an asymptomatic cerebral aneurysm is detected

Item 107 Preferred Response: C

Intracranial bleeding resulting from ruptured cerebral aneurysms is the most serious complication of adult polycystic kidney disease (autosomal dominant polycystic kidney disease [ADPKD]). Prevalence of cerebral aneurysms is lower in younger patients (around 5%) and increases up to 20% in older patients. Patients who have ADPKD and a positive family history of intracranial aneurysms or subarachnoid hemorrhage are at increased risk of developing intracranial aneurysm. Larger aneurysms (>7 mm), poorly controlled hypertension, and age less than 50 years is associated with increased risk for aneurysm rupture. Hypertension is present in 50% to 70% of patients with ADPKD by age 30 years and develops before the deterioration of kidney function.

The role of routine radiologic screening for cerebral aneurysms in asymptomatic patients with ADPKD is unclear. Patients with a (1) history of previous aneurysm rupture, (2) a positive family history of intracerebral bleeding or intracranial aneurysm, (3) an occupation in which loss of consciousness would place them or others at extreme risk, and (4) marked hemodynamic instability and hypertension associated with surgeries are considered at high risk for intracerebral hemorrhage. Routine screening for cerebral aneurysm is indicated in these high-risk populations. Imaging modalities available for screening for cerebral aneurysm include high-resolution computed tomographic angiography or magnetic resonance angiography. Because of the risk for nephrogenic systemic fibrosis among patients with glomerular filtration rates less than 30 mL/min, it is currently recommended that gadolinium-based imaging be avoided in patients with advanced renal failure. Screening recommendations for patients with ADPKD at high risk for intracerebral aneurysm include rescreening every 5 years after the initially negative radiographic studies. Recommendations for patients with known aneurysms or history of intracerebral bleeding are annual screening for 2 to 3 years, and every 2 to 5 years thereafter if the aneurysm is stable.

Surgical treatment of unruptured asymptomatic intracranial aneurysms is controversial. According to the International Study of Unruptured Intracranial Aneurysms, an aneurysm less than 7 mm in size is associated with a low risk for rupture. The decision for surgical intervention for aneurysms is based on the risk for aneurysmal rupture, surgical complications, and patient age. Asymptomatic aneurysms are not routinely surgically corrected because multivariate analyses have shown that the likelihood of a poor postsurgical outcome was equal to or greater than the 5-year likelihood of aneurysm rupture. Therefore, the current recommendations point to a conservative observation and monitoring approach for small aneurysms, especially those located in the anterior cerebral circulation. However, the management of each case needs to be individualized; associated risk factors, presence of comorbid conditions and patient age must be considered, while taking into account the size and location of the aneurysm and surgical risk factors.

PREP Pearls

- Intracranial aneurysm is an extrarenal manifestation of autosomal dominant polycystic kidney disease (ADPKD). The prevalence of intracranial aneurysms is low in children and it increases with age.
- Aneurysms less than 7 mm in diameter, saccular in shape, and present in the anterior cerebral circulation have a low risk for rupture.

American Board of Pediatrics Content Specification(s):

- Know that children with autosomal-dominant polycystic kidney disease may have hypertension
- Know that autosomal-dominant polycystic kidney disease may be associated with intracranial aneurysms

Suggested Reading:

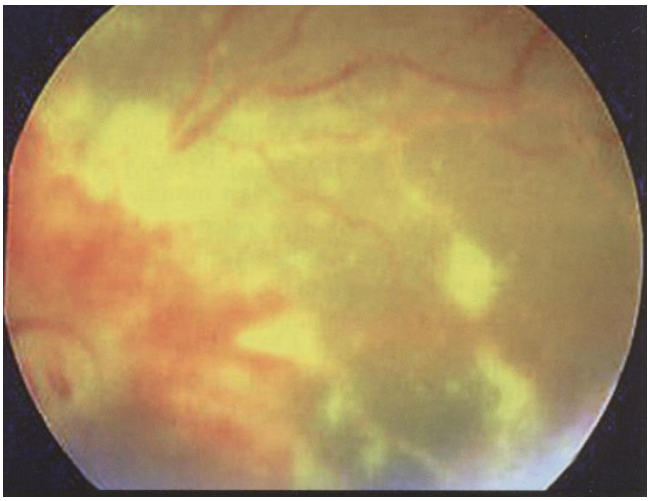
- Bederson JB, Awad IA, Wiebers DO, et al. Recommendations for the management of patients with unruptured intracranial aneurysms: A statement for healthcare professionals from the Stroke Council of the American Heart Association. Circulation. 2000;102(18):2300-2308. doi:10.1161/01.CIR.102.18.2300
- Evner AD. Childhood ADPKD: answers and more questions. Kidney Int.
- 2001;59(5):1979-1980. doi:10.1046/j.1523-1755.2001.0590051979.x
- Grantham JJ. Clinical practice: autosomal dominant polycystic kidney disease. N Engl J Med. 2008; 359(14):1477-1485. doi:10.1056/ NEJMcp0804458
- Takao H, Nojo T. Treatment of unruptured intracranial aneurysms: decision and cost-effectiveness analysis. Radiology. 2007;244(3):755. doi:10.1148/radio1.2443061278
- Wiebers DO, Whisnant JP, Huston J 3rd, et al. Unruptured intracranial aneurysms: natural history, clinical outcome; and risks of surgical and endovascular treatment. Lancet. 2003;362(9378):103-110. doi:10.1016/ S0140-6736(03)13860-3

Item 108

An 18-year-old homeless girl presents to the emergency department because she "cannot see" in her left eye. She complains of slowly progressive decreased visual acuity and the recent development of "floaters." She also reports chronic diarrhea and skin problems. On physical examination, her temperature is 36.5°C, heart rate is 65 beats/min, respiratory rate is 14 breaths/min, and blood pressure is 85/55 min Hg. The examination is notable for a cachectic girl who answers questions appropriately. Funduscopic examination reveals retinitis with extensive areas of hemorrhage and white retinal exudates (Item Q108). Visual acuity is 20/30 in the right eye and 20/400 in the left eye. The oropharyngeal examination reveals white palatal plaques. Auscultation of the lungs reveals coarse breath sounds bilaterally with normal effort. Her abdomen is mildly tender to palpation; the liver and spleen are palpable 3 cm to 4 cm below the costal margins. Neurologic examination is grossly unremarkable except for visual acuity. Examination of the skin reveals multiple erythematous, eroded papules consistent with insect bites that are not healing well.

Of the following, the MOST likely cause of this patient's vision loss is infection with

A. Bartonella henselae
B. Candida albicans
C. cytomegalovirus
D. Epstein–Barr virus
E. Mycobacterium avium

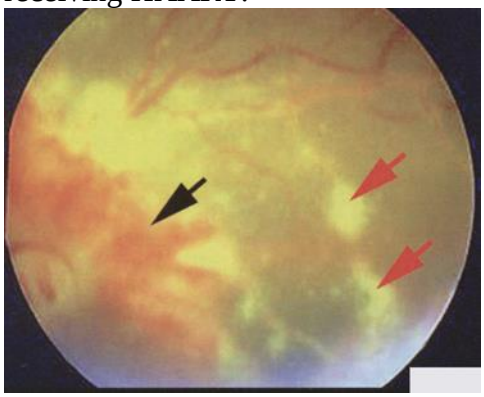


ITEM Q108: Fundoscopic findings as described for the girl in the vignette.

Item 108 Preferred Response: C

The patient described in the vignette presents with painless visual loss associated with cachexia, hypothermia, bradycardia, hypotension, oral thrush, hepatosplenomegaly, chronic diarrhea, and skin problems. An underlying immunodeficiency, such as human immunodeficiency virus (HIV) infection and AIDS, is suspected. Therefore, the most likely cause of this patient's vision loss is cytomegalovirus (CMV) infection.

CMV retinitis occurs in 20% to 40% of patients with AIDS who are not receiving highly active antiretroviral therapy (HAART) and is most common in those patients with CD4 T-lymphocyte counts less than 50/4. It is the most common cause of retinitis in all patients with HIV infection and usually occurs as unilateral disease but will progress to bilateral disease if left untreated. CMV retinitis can cause loss or blurring of vision, blind spots, floaters, and flashing lights (photopsia). Floaters and photopsia are symptomatic predictors of CMV retinitis. Retinal detachment can occur. The diagnosis is made clinically by ophthalmologic examination, which reveals hemorrhage in association with white (or yellow), fluffy retinal lesions, usually close to the retinal vessels (Item C108A). CMV retinitis is associated with little inflammation of the vitreous in patients not receiving HAART.



ITEM C108A: Hemorrhages (black arrow) and exudates (red arrows) in a patient who has cytomegalovirus retinitis and HIV infection

Item C108B. Opportunistic Infection in Patients with Human Immunodeficiency Virus Infection and AIDS

- Aspergillus species
- Candida species
- Coccidioidomycosis
- Cryptococcosis
- Cryptosporidium species
- Cytomegalovirus
- Enteric pathogens, other (eg, Salmonella species)
- Epstein-Barr virus
- Herpes simplex virus
- Histoplasmosis
- Invasive infections due to encapsulated bacteria
- Isospora species
- Mycobacterium avium complex

- Mycobacterium tuberculosis
- Pneumocystis jirovecii
- Toxoplasma gondii
- Varicella-zoster virus

Unlike CMV retinitis, chorioretinitis caused by Candida albicans usually progresses to involve the vitreous. Typical findings include white, infiltrative, moundlike lesions on the retina. In patients with Bartonella henselae (cat scratch disease) infection, only 1% to 2% will develop neuroretinitis. This infection is not thought to be more common in patients with HIV infection and AIDS compared with the general population. Patients usually present with fever, malaise, and acute (unilateral) vision loss from optic nerve edema associated with stellate macular exudates (macular star) on ophthalmologic examination. Other retinal findings can include hemorrhages, multiple lesions deer in the retina, and cotton wool spots. Herpes simplex virus car cause retinitis, especially acute retinal necrosis, but it is less common than CMV retinitis. Epstein-Barr virus and Mycobacterium avium complex are rare causes of retinitis.

In general, in the era of HAART, the frequency of all opportunistic infections (Item C108B) in patients with HIV infection and AIDS has substantially decreased.

PREP Pearls

- CMV is the most common cause of retinitis in patients with HIV infection and AIDS.
- In the era of HAART, the frequency of all opportunistic infections in patients with HIV infection and AIDS has substantially decreased.

American Board of Pediatrics Content Specification(s):

- Recognize the major opportunistic infections seen in patients with HIV/AIDS

Suggested Reading:

- American Academy of Pediatrics. Cytomegalovirus infection In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: 2012:300-305
- American Academy of Pediatrics. Human immunodeficiency virus infection. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL:2012:418-439

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Item 109

An 18-year-old girl is in your office for evaluation of abdominal pain. You diagnose her as having pelvic inflammatory disease (PID). She is using an oral contraceptive but takes the pills erratically, and her partner does not always use condoms. She has no fever or systemic symptoms at this time. You prescribe outpatient antibiotics. You discuss the reasons why adherence with the medication regimen is important. In order to help with adherence, you suggest a number of techniques.

Of the following, the MOST effective strategy to enhance adherence would be to

A. have her mark a calendar each time she takes her pills
B. have her mother supervise medication administration
C. have her place alarm reminders on her cell phone
D. have her take the medicines with meals
E. stress the possible long-term consequences of untreated PID

Item 109 I-C Preferred Response:

The girl described in the vignette is in late adolescence and legally an adult. Therefore, as an otherwise healthy adolescent, she is expected to take more responsibility for her own care and is best served by methods that help with self-monitoring. The alarm on her cellphone is an active reminder system and thus more useful than passive cues, such as taking medications with meals or marking a calendar. She is entitled to confidentiality about both her use of oral contraceptive pills and her treatment for pelvic inflammatory disease. Therefore, unless she indicates that her mother is aware of the situation, it is inappropriate to ask her mother to supervise her medication regimen. For many adolescents, concerns about attempts to restrict autonomy might lead to reluctance to adhere with recommendations.

There are various models that discuss factors that influence adherence to therapy. Among the individual items identified are the patient's perception of the severity of their condition, the need for symptom relief (as with pain), and the benefits of treatment. Adolescents are usually more concerned with the short-term consequences of a condition; discussions about long-term adverse effects may be less relevant and thus unlikely to motivate adherence. In addition, the perception of one's ability to effect change will influence adherence. An adolescent who has a severe chronic illness, especially if cognitively impaired, will have difficulty managing his or her own care and will benefit from social support, especially from parents and family. Therefore, consideration of the patient's developmental level and psychological status, along with health status and the complexity of the specific regimen in question, will need to be considered when developing a plan to help with adherence. From the physician's side, the ability to effectively communicate the rationale for the regimen will help with adherence. In addition, it is important to assess the adolescent's ability to pay for medications and their transportation needs. Using telephone reminders, having close follow-up visits, using rewards for success, and providing counseling are also useful.

PREP Pearls

- The most common reason for failure of therapy is lack of adherence.
- Use of technology (eg, cellphones to create reminders) has been shown to enhance adherence.

American Board of Pediatrics Content Specification(s):

- Identify the features of an illness or a treatment regimen that tend to worsen a patient's adherence to treatment: lack of symptoms, more than one treatment, side effects of treatment, multiple daily medication doses, lack of perceived seriousness of the illness
- Know the behavioral techniques that may enhance patient adherence: medicine calendar, dosing in synchrony with a patient's regular activity

Suggested Reading:

- Arrington-Sander R. In brief: adherence. Pediatr Rev. 2009;30;e9. doi:10.1542/pir.30-2-e9
- Horvath T, Azman H. Kennedy GE, Rutherford GW. Mobile phone text messaging for promoting adherence to antiretroviral therapy in patients with HIV infection. Cochrane Database Syst Rev. 2012;3:CD00956. doi:10.1002/14651858.CD009756
- Perrin JM, Gnanasekaran S, Delahaye J. Psychosocial aspects of chronic health conditions. Pediatr Rev. 2012;33:99-109. doi:10.1542/pir.33-3-99
- Salem NE, Elliott RA, Glazebrook C. A systematic review of adherence-enhancing interventions in adolescents taking long-term medicines.
- J Adolesc Health. 2011;49:455-466. doi:10.1016/j.jadohealth.2011.02.010
- Stein REK. Chronic physical disorders. Pediatr Rev. 1992;13:224-229. doi:10.1542/pir.13-6-224

Item 110

An 8-year-old boy is seen for a health supervision visit. Since his last appointment, he has been well and has had a good appetite and normal development. His only complaint is a recent upper respiratory tract infection for which his parents treated him symptomatically. The boy's parents tell you that he vomited several times last month, and on one occasion, he awoke at night and vomited a large amount of yellow-green, clear fluid. Since then he has been completely asymptomatic. Physical examination demonstrates a well-appearing child, and the remainder of the examination is completely unremarkable.

Of the following, you are MOST likely to recommend

A. abdominal ultrasonography
B. referral for upper gastrointestinal endoscopy
C. routine follow-up in 6 months, or sooner if symptoms recur
D. surgical referral
E. upper gastrointestinal tract radiographic series

Item 110 Preferred Response: E

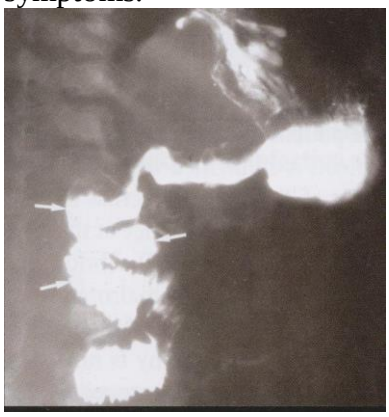
Vomiting is a common symptom associated with both acute and chronic illnesses that affect diverse organ systems. Thus, vomiting may be a consequence of increased intracranial pressure, acute infections, metabolic disease, and functional and anatomical gastrointestinal (GI) disorders. Although most episodes involve the expulsion of gastric secretions plus ingested food and fluids, the vomitus may also be classified as bloody or bilious, depending on the underlying disorder and the frequency and severity of emesis. Accordingly, protracted periods of vomiting and retching may produce fluid that is yellow-tinged (bile refluxate into the stomach) or blood streaked. However, the sudden onset of bilious vomiting must be considered to be an indication (until proven otherwise) of a pathologic condition that causes partial or complete intestinal obstruction beyond the ampulla of Vater.

The boy in the vignette awoke one night and vomited a large amount of yellow-green fluid. Although he is now asymptomatic, such a history should raise concern for GI tract obstruction distal to the ampulla of Vater (the duodenal exit point of the common bile duct), resulting in the reflux and expulsion of bile. Item C110A, page C-88, lists the common causes of bilious vomiting in childhood, subdivided by the typical ages at presentation. Each of the disorders is associated with a partial or total functional or anatomical GI obstruction. In a healthy-appearing child without signs or symptoms of acute illness or a significant medical or surgical history, the most prevalent anatomical abnormality that causes the sudden onset of bilious vomiting is intestinal malrotation, with or without midgut volvulus. The appropriate next step in the evaluation of this patient would be to perform an upper GI tract radiographic series to rule out this diagnosis.

Although malrotation may remain undetected throughout life, symptomatic cases most commonly present during infancy. Bilious vomiting is the predominant symptom in these cases. Beyond the first year of life, the clinical presentation of malrotation is more varied and may include symptoms of intermittent abdominal pain, diarrhea, and vomiting (with or without bile). In the absence of bilious vomiting, the diagnosis is often delayed until a radiographic series is performed, often as a relatively late part of the evaluation. Midgut volvulus may present acutely with abdominal pain, diarrhea, and vomiting and thus may be confused with an episode of gastroenteritis. Because a prolonged period of volvulus may result in life-threatening intestinal ischemia, the diagnosis requires a high index of suspicion.

The upper GI tract contrast study is the imaging study of choice in a stable patient suspected of having intestinal malrotation. The diagnosis is confirmed when the duodenal—jejunal junction (ligament of Treitz) fails to cross the midline. Volvulus often gives a corkscrew appearance at the level of the distal duodenum (Item C110B, page C-88). Abdominal ultrasonography may suggest a diagnosis of malrotation by finding the superior mesenteric vein on the left of the midline rather than the right. However, ultrasonography is not a study of choice in this clinical setting because the procedure has insufficient diagnostic sensitivity for the evaluation of suspected malrotation. Endoscopy is not a routine part of the evaluation for suspected malrotation. Once the diagnosis is

confirmed, surgical correction (regardless of whether the patient is symptomatic) is indicated. However, in a stable, asymptomatic patient, surgical referral should be deferred until after the contrast study is performed. Under no circumstances should the evaluation of a patient with bilious vomiting be delayed because of the absence of acute symptoms.



ITEM C110B: Contrast study showing malrotation. The duodenal sweep does not cross the midline. Arrows show the "corkscrew" appearance of mid-gut volvulus.

Item C110A. Causes of bilious vomiting in childhood

Age	Cause
0 – 3 mo	• Hirschsprung disease
	• Intestinal atresia, stenosis (duodenal, jejunal, ileal)
	• Intestinal malrotation with midgut volvulus
	• Meconium ileus
	• Necrotizing enterocolitis
4 – 12 mo	• Intestinal malrotation with midgut volvulus
	• Intussusception
>12 mo	• Adhesions (postsurgical)
	• Appendicitis
	• Crohn disease
	• Cystic fibrosis (distal intestinal obstruction syndrome)
	• Ileus - Infectious - Metabolic - Postsurgical
	• Incarcerated inguinal hernia
	• Intestinal malrotation *with* or *without* volvulus
	• Intestinal pseudo-obstruction
	• Intestinal stenosis (duodenal, jejunal, ileal)
	• Intussusception
	• Neoplasm (extrinsic or intrinsic bowel obstruction)
	• Superior mesenteric artery syndrome

PREP Pearls

- Bilious vomiting indicates small bowel obstruction until proven otherwise.
- In patients with a history of bilious vomiting, irrespective of their current clinical status, intestinal malrotation must be ruled out.
- In cases of suspected malrotation, the upper GI series is the study of first choice.

American Board of Pediatrics Content Specification(s):

- Understand the significance of bilious vomiting

Suggested Reading:

- Chandran L, Chitkara M. Vomiting in children: reassurance, red flag, or referral? Pediatr Rev. 2008;29:183-192. doi:10.1542/pir.29-6-183
- El-Gohary Y, Alagtal M, Gillick J. Long-term complications following operative intervention for intestinal malrotation: a 10-year review. Pediatr Surg Int. 2010;26:203-206
- Hajivassiliou CA. Intestinal obstruction in neonatal/pediatric surgery. Semin Pediatr Surg. 2003;12:241-253. doi:10.1053/j.sempedsurg.2003.08.005
- Nylund CM, Denson LA, Noel JM. Bacterial enteritis as a risk factor for childhood intussusception: a retrospective cohort study. J Pediatr. 2010;156:761-765. doi:10.1016/j.jpeds.2009.11.026
- Sizemore AW, Rabbani KZ, Ladd A, Applegate KE. Diagnostic performance of the upper gastrointestinal series in the evaluation of children with clinically suspected malrotation. Pediatr Radiol. 2008;38:518-528. doi:10.1007/s00247-008-0762-8

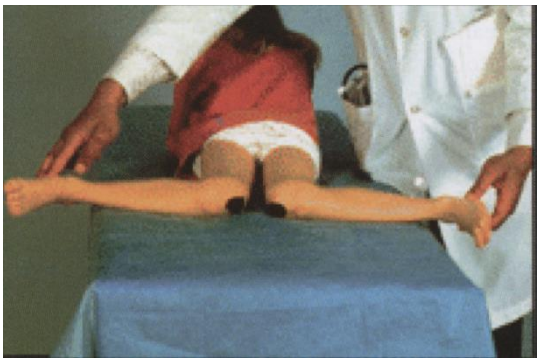
Item 111

Parents bring in their 6-year-old daughter because her toes turn in when she walks. They are concerned because imaging studies have not been performed and treatment has not been prescribed. Examination of this appropriately grown child is notable for moderate intoeing and 90° of internal rotation of both hips when she is in the prone position (Item Q111). There is no evidence of metatarsus adductus or tibial torsion. Aside from the intoeing, she has a normal gait.

Of the following, the MOST appropriate next step in the care of this child is to

A. counsel the family that a substantial portion of children with this condition will require surgical intervention as they grow older
B. obtain a computed tomography scan to define precisely the femoral neck anatomy
C. prescribe twist cables and corrective shoes
D. reassure the parents that this child has a good prognosis for resolution as the child matures
E. refer to orthopedics because the intoeing has not corrected by 6 years of age

ITEM Q111: Findings for the girl described in the vignette.



Item 111 Preferred Response: D

Femoral anteversion occurs when the femoral neck is rotated anteriorly compared with the transcondylar axis of the knee and the long axis of the femur. It is often the result of intrauterine positioning and genetic influences and is twice as frequent in girls as in boys. As seen in the patient in the vignette, common characteristics include medially facing patellae when standing and intoeing, with the patella pointing toward the midline when walking. The running gait of a child with femoral anteversion has been described as having an "egg-beater" or "windmill" appearance with medial rotation of the thighs and outward rotation of the feet. Physical examination for femoral anteversion is conducted with the patient lying prone. With the knees flexed 90°, the lower legs are rotated out, causing the hips to rotate in (Item C111). Children with femoral anteversion have markedly increased internal hip rotation up to 90° compared with the normal 35° to 50°. Examination is sufficient to make the diagnosis, and no imaging studies are necessary unless findings are extreme.

Femoral anteversion is developmentally normal at birth. Typically, the anteversion decreases by 1° to 2° per year until the adult position is achieved at skeletal maturity. Parents most commonly seek care for their child with this condition when the child is between 3 and 6 years old and the normal physiologic external rotation contracture of the hip has resolved. In these children, intoeing may continue to increase until age 6 years and then decrease. The condition is not painful and in most cases does not limit function. In about 80% of affected children, femoral anteversion resolves spontaneously, usually by age 7 years. However, in some children the final outcome is not seen until age 11 years.

In general, referral to an orthopedist is reserved for children who have persistent anteversion beyond age 11 years that causes functional or cosmetic impairment. If anteversion is unilateral, the pediatrician should conduct a thorough physical examination looking for signs of an underlying neurologic condition such as cerebral palsy. Referral to an orthopedic surgeon may be warranted in this setting. For the few patients requiring treatment (ie, those with anteversion greater than 50° as seen on radiographic study), femoral derotational osteotomy is effective but has had, in the past, a high rate of complications. Orthotics, twister cables, splinting, bracing, and physiotherapy are ineffective in changing the course or degree of intoeing.

Despite reassurance by their primary care practitioner, many parents request an orthopedic evaluation for their child with less significant femoral anteversion. In one retrospective study, orthopedic specialists in Scotland found that, of 202 patients referred for intoeing, 86% were discharged after the first visit without a scheduled follow-up and another 5% were discharged within the next 2 years without intervention. Nine children were referred during the study period most commonly because of parental concern. Among parents who completed a questionnaire about their visit, 83% had wanted the referral but a minority specifically asked for it and only 22% felt their child had a major medical problem.

Long-term consequences of femoral anteversion are some-what controversial. Although evidence indicates no substantive link, some practitioners believe there is an increased incidence of osteoarthritis of the hip and knee, slipped capital femoral epiphysis, and knee and patella instability.

PREP Pearls

- Femoral anteversion is diagnosed clinically in a child who has intoeing when there is increased internal hip rotation (>50°) with the child in the prone position.
- Besides intoeing, the examiner notes that the patella points medially when these children walk.
- The normal course of femoral anteversion is spontaneous improvement by the time of skeletal maturity.
- Orthopedic referral is indicated when anteversion persists beyond age 11 years and causes functional or cosmetic impairment or if the anteversion is unilateral.

American Board of Pediatrics Content Specification(s):

- Know that the natural history of femoral anteversion is self-correction
- Know that x-ray studies are not necessary for the diagnosis of femoral anteversion
- Know how to evaluate a child with femoral anteversion

Suggested Reading:

- Blackmur JP, Murray AW. Do children who in-toe need to be referred to an orthopaedic clinic. J Pediatr Orthop B. 2010;19:415-417. doi: 10.1097/ BPB.0b013e3283339067
- Hoekelman RA, Chianese MJ. Foot and leg problems. In: Mclnerny TK, ed. Textbook of Pediatric Care. Elk Grove Village, IL: American Academy of Pediatrics; 2009;Chap 183:1541
- Rosenfeld SB. Approach to the child with in-toeing. UptoDate. 2013. Available online only for subscription

Item 112

You are evaluating a 2-year-old girl whose parents report that she began choking while playing near a table where a box of thumbtacks was located. The choking episode was not improved by her father pounding on her back, but the girl remained conscious while being transported to the emergency department. She has a temperature of 37.0°C, heart rate of 130 beats/min, respiratory rate of 36 breaths/min, and blood pressure of 80/50 mm Hg. Her oxygen saturation is 93% by pulse oximetry on room air. The child is in mild respiratory distress and has bilateral wheezing. Her parents state that she has no history of asthma. You suspect that she has aspirated a thumbtack.

Of the following, the MOST likely complication for this patient would be

A. bronchial stricture
B. bronchiectasis
C. pneumothorax
D. pulmonary infection
E. tracheal perforation

Item 112 S Preferred Response: D

Airway foreign bodies are life-threatening events; they are the fifth leading cause of unintentional injury mortality in children and the first in children less than 1 year of age in the United States. Although the mortality from such foreign bodies has decreased over the years, likely because of improved safety standards and public awareness, there were still more than 3,700 deaths in 2007. Although mortality has decreased, the overall incidence of foreign body aspiration (FBA) has not, necessitating prompt recognition and management. Children younger than 3 years of age (peak incidence between 1 and 2 years of age) are at the highest risk and account for 80% of pediatric FBAs. Various objects are aspirated, one third of which are nuts.

All children who are witnessed to swallow an object or who have a typical clinical presentation (choking episode followed by coughing, wheezing, or inspiratory stridor) should be evaluated for an FBA. Physical findings can include tachypnea, retractions, wheezing, stridor, decreased or unilateral breath sounds, or cyanosis. The foreign body is predominantly located in the main bronchi (70%); the right main bronchus is affected 3 times more often than the left. The trachea, lower bronchi, and larynx are less common sites.

Plain radiographs are usually not diagnostic because less than 15% of foreign bodies are radiopaque. However, they may provide indirect evidence of FBA, such as unilateral hyperinflation, atelectasis, mediastinal shift, or pneumomediastinum. Overall, plain chest radiographs may be normal in up to two-thirds of children who have an FBA. Expiratory radiographs are more sensitive for detection than inspiratory radiographs and fluoroscopy may increase diagnostic sensitivity because of its ability to assess diaphragmatic and mediastinal motion. Bronchoscopy remains the gold standard for both diagnosis and management and should be used to evaluate the airway in all cases of suspected FBA. Removal of the foreign body using rigid bronchoscopy is safe (complication rates of <1%) and has helped reduce mortality. Flexible bronchoscopy may be helpful for diagnosis in equivocal cases but should be not used for removal of the object.

With early diagnosis, complications are rare. Most complications occur in the setting of delayed presentation; pneumonia and atelectasis are most common. Bronchial stricture, pneumothorax, pneumomediastinum, bronchiectasis, and tracheal perforation (because of either aspiration of sharp objects or ongoing infection) are less commonly seen.

PREP Pearls

- Airway foreign bodies are the fifth leading cause of unintentional injury mortality in children and the first in children less than 1 year of age in the United States.
- All children who are witnessed to swallow an object or who have a typical clinical presentation for a foreign body aspiration should undergo evaluation.
- Plain radiographs are usually not diagnostic in foreign body aspiration because less than 15% of foreign bodies are radiopaque.

American Board of Pediatrics Content Specifications:

- Plan the management of a patient with aspiration of a foreign body

Suggested Reading:

- Holinger LD. Foreign bodies of the airway. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier 2011:1453-1454
- Rovin JD, Rogers BM. Pediatric foreign body aspiration. Pediatr Rev. 2000;21:86-90. doi: 10.1542/pir.21-3-86
- Ruiz FE. Airway foreign bodies in children. UptoDate. Available online only for subscription

Item 113

You are called about an 8-year-old boy with ulcerative colitis who developed fever (up to 39.4°C). He is currently at home, and he is still taking oral feedings well. Among his medications, he was taking prednisone daily for 4 months and completed tapering off the medication 1 week ago. A recent morning serum cortisol level was 0.4 µg/dL (11nmol/L) (normal range, 8-19 µg/dL [221-524 nmol/L]). His body surface area is 1 m'. He has an appointment with you tomorrow.

Of the following, the BEST step in the management of this patient is to start 3-times-daily treatment with

A. dexamethasone, 10 mg orally
B. fludrocortisone, 10 mg orally
C. hydrocortisone, 10 mg orally
D. prednisone, 10 mg orally
E. prednisolone, 10 mg orally

Item 113 Preferred Response: C

The child in this vignette has evidence of adrenal insufficiency from long-term steroid use. During times of moderate to severe illness, or if surgery is needed, treatment with stress-doses of steroids is needed to mimic the increased steroid levels normally produced under stress in adrenally sufficient patients.

The steroid of choice for adrenal crisis (of any cause) is hydrocortisone. Hydrocortisone is quick acting and at very high doses saturates all steroid receptors, causing a mineralocorticoid and glucocorticoid effect. Although this patient does not have primary adrenal disease and does not need extra mineralocorticoids, the quick onset of action of hydrocortisone makes it the preferred agent for treatment during stress dosing.
Oral stress doses involve tripling physiologic doses (10 mg/m^2 per day) by giving 30 mg/m^2 per day; thus, in this patient whose body surface area is 1 m^2, hydrocortisone, 10 mg orally three times daily, is the correct answer. Steroid equivalences (glucocorticoid effect) are listed (Item C113).

Item 113. Glucocorticoid Potencies of Commonly Used Steroid Medications

Name	Relative Potency to Cortisol
Hydrocortisone	1
Prednisone	4
Methylprednisolone	5
Dexamethasone	25

According to the relative potency of these steroid medications (Item C113), treatment with any of the glucocorticoids (ie, other than hydrocortisone) at the doses listed in the answer options would far exceed the required steroid levels.

Fludrocortisone, a mineralocorticoid, is not needed because the main issue for oral stress dosing is adding glucocorticoids.

Patients diagnosed as having adrenal insufficiency are often given injectable hydrocortisone for use at home (50-100 mg/m2 intramuscularly). If the patient were to develop emesis and oral intolerance, the family is instructed to give the injectable hydrocortisone at home before immediately coming to the emergency department.

PREP Pearls

- High-dose hydrocortisone is the steroid of choice for stress dosing of steroids.
- Pediatricians should be aware of the relative glucocorticoid potencies of commonly used steroids, especially when prescribed for stress dosing.

American Board of Pediatrics Content Specification(s):

- Know the risks and how to manage a patient who has received long-term corticosteroids and requires surgery
- Know the special management needs of chronically corticosteroid dependent children with respect to stress, surgery, and varicella infection

Suggested Reading:

- Hsu AA, von Elten K, Chan D, et al. Characterization of the cortisol stress response to sedation and anesthesia in children. I Clin Endocrinol Metab. 2012;97:E1830-E1835. doi:10.1210/jc.2012-1499
- Shulman DI, Palmert MR, Kemp SF; Lawson Wilkins Drug and Therapeutics Committee. Adrenal insufficiency: still a cause of morbidity and death in childhood. Pediatrics. 2007;119:e484-e494. doi:10.1542/peds.2006 -1612

Item 114

As part of an evaluation for a small for gestational age (SGA) infant, Toxoplasma serology reveals the presence of IgG and IgM antibodies in the infant's specimen. The baby's weight is at less than the fifth percentile, and weight and head circumference are at the 10th percentile. The remainder of the baby's physical examination is unremarkable, including a normal liver span and normal neurologic examination findings for age. Confirmatory laboratory tests, including cerebrospinal fluid examination, computed tomography scan of the head, and ophthalmologic and audiologic examinations are ordered. In discussing potential for treatment with the parents pending the results of the evaluations, the parents raise concerns regarding the potential toxicities of prolonged sulfa-drug treatment. They ask what their child's long-term risks are if the infant is infected with toxoplasma and not treated.

Of the following, the adverse outcome MOST likely to be seen in this infant is

A. attention-deficit disorder
B. dental deformities
C. hearing loss
D. hepatitis
E. visual loss

Item 114 I-C Preferred Response: E

Toxoplasmosis is one of the conditions associated with intrauterine or so-called TORCH (toxoplasmosis/ Toxo plasma gondii, other infections, rubella, cytomegalovirus herpes simplex virus) infection. These infections may present at birth with multiorgan involvement or be asymptomatic at birth and present with late-onset abnormalities. With intrauterine or congenital toxoplasma infection, acute ocular involvement with chorioretinitis is the most common long-term consequence and can lead to unilateral vision loss. Seventy percent to 90% of congenitally infected infants are asymptomatic at birth. Symptoms of chorioretinitis (blurred vision, eye pain, floaters, or photophobia) may not appear until months to years after birth.

Hearing loss, mental retardation, and learning disabilities have also been associated with congenital Toxoplasma infection but less commonly than visual impairment.
Signs of congenital toxoplasmosis at birth, when they occur may include a maculopapular rash, generalized lymphadenopathy, hepatosplenomegaly, jaundice, pneumonitis, intracerebral calcifications, thrombocytopenia, and petechiae.

Developmental sequelae are also described with other congenital infections. Sensorineural hearing loss is the most common manifestation of congenital cytomegalovirus (CMV) infection. Up to 90% of congenital CMV infections are asymptomatic at birth with hearing loss presenting months to years later. Symptomatic CMV disease at birth may present with intrauterine growth retardation, jaundice, hepatosplenomegaly, purpura, retinitis, intracerebral calcifications, and developmental delay. Congenital syphilis may present with hepatosplenomegaly, snuffles, lymphadenopathy, mucocutaneous lesions, rash, pneumonia, pseudoparalysis, petechiae, and hemolytic anemia at birth or up to age 8 weeks. Asymptomatic infection at birth may be associated with late manifestations involving the nervous system, teeth, bones, joints, eyes (interstitial keratitis), and hearing loss. Finally congenital rubella infection can present with miscarriage, fetal death, or a spectrum of anomalies including cataracts, microphthalmos, congenital heart disease (patent ductus arteriosus or peripheral pulmonic stenosis), hearing loss, microcephaly, mental retardation, hepatosplenomegaly, and dermal erythropoiesis.

PREP Pearls

- Congenital infections may be asymptomatic at birth but present with significant abnormalities months to years later.
- Visual impairment is a major sequela of congenital toxoplasmosis.
- Hearing loss is the major late consequence of congenital cytomegalovirus infection.

American Board of Pediatrics Content Specification(s):

- Recognize the developmental sequelae of congenital infections, eg visual impairment

Suggested Reading:

- American Academy of Pediatrics. Toxoplasma gondii infections. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012: 720-728
- American Academy of Pediatrics. Cytomegalovirus. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:330-335
- American Academy of Pediatrics. Rubella. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:629-634
- American Academy of Pediatrics. Syphilis. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:690-703
- Remington JS, McLeod R, Thulliez P, Desmonts G. Toxoplasmosis. In: Remington JS, Klein JO, Wilson CB, Baker CJ, eds. Infectious Diseases of the Fetus and Newborn Infant. 6th ed. Philadelphia, PA: Elsevier; 2006:947-1091

Item 115

A 9-year-old girl presents to your office for her annual health supervision visit. She has no significant medical history or complaints and is growing normally. Her physical examination, including vital signs, is unremarkable. Her mother was recently diagnosed with systemic lupus erythematous (SLE). Per the mother's request, an antinuclear antibody test was performed on the child, resulting in a low-positive titer.

Of the following, the BEST information you can provide the mother is that her daughter

A. has a confirmed diagnosis of SLE
B. has a high likelihood of developing autoimmune diseases
C. has an infection
D. most likely does not have SLE
E. will develop lupus later in life

Item 115 Preferred Response: D

Antinuclear antibody (ANA) of low titer (l.:80) in asymptomatic individuals with a normal physical examination, such as the girl in the vignette, is not associated with autoimmune disease and is not predictive of developing an autoimmune disease in the future. ANA is positive in up to one-third of the healthy population and may occur in family members of patients with autoimmune disease. A low titer is rarely associated with autoimmune disease. A negative ANA would make the diagnosis of systemic lupus erythematosus (SLE) very unlikely. Patients with active autoimmune disease will typically have a titer of 1:1,280 or higher.

Several diseases can increase the ANA titer, including, but not limited to, autoimmune conditions such as SLE, juvenile idiopathic arthritis, juvenile dermatomyositis, drug-induced lupus, Sjogren syndrome, scleroderma, and systemic sclerosis. The ANA can be elevated with infection, but it is not diagnostic of an infection. Infectious causes of elevated ANA include, but are not limited to, viral infections, tuberculosis, subacute bacterial endocarditis, chronic osteomyelitis, Lyme disease, and malaria. Other conditions that cause ANA positivity include psoriasis, idiopathic thrombocytopenic purpura, autoimmune hepatitis, autoimmune thyroiditis, multiple sclerosis, and type I diabetes mellitus. ANA positivity has also been associated with cancers such as lymphoma and leukemia.

The American College of Rheumatology has a position statement on the methods recommended for testing ANA. Immunofluorescence is the gold standard. Alternative assays that test for fewer autoantigens may not be equivalent to immunofluorescence. Some laboratories are using solid-phase assays for ANA testing that test for 8 to 10 auto-antigens. HEp-2 immunofluorescence tests for over 100 autoantigens. Other methods may result in a false-negative or false-positive test result and should be interpreted with the entire clinical picture in mind.

PREP Pearls

- Low-titer ANA in children with a normal physical examination is not clinically significant.
- The ANA level can be elevated in conditions other than autoimmune disease.
- The gold standard for ANA testing is immunofluorescence.

American Board of Pediatrics Content Specification(s):

- Recognize that a low-titer ANA may be seen in unaffected individuals and family members

Suggested Reading:

- American College of Rheumatology. Position Statement on the Method of Testing for Antinuclear Antibodies. American College of Rheumatology Web site, www.rheumatology.org
- Siegel DM. Antinuclear antibody (ANA) testing. Pediatr Rev. 2003;24(9):320-321. doi:10.1542/pir.24-9-320
- Weiss JE. Pediatric systemic lupus erythematosus: more than a positive antinuclear antibody. Pediatr Rev. 2012;33(2):62-73. doi:10.1542/pir.33-2-62

Item 116

You are seeing a 15-year-old, 55-kg boy in your office for a health supervision visit. He has no significant findings on physical examination. Within your review of adolescent risk behaviors, he revealed that he smokes cigarettes but denied abusing any other substances. One of his parents and several close friends also smoke cigarettes. He says he will usually smoke one-half pack per day, but sometimes will smoke up to one pack per day. The boy states that he sometimes wishes he could quit smoking but so far has not tried to reduce his cigarette use.

Of the following, the BEST next best action would be to

A. ask him to tell you his reasons he wishes to quit smoking
B. ask his parents to put pressure on him to quit smoking
C. prescribe the nicotine transdermal patch
D. prescribe sustained-release bupropion
E. prescribe varenicline

Item 116 I-C Preferred Response: A

This vignette highlights some of the challenges of getting adolescents to quit smoking. Because no single strategy has been shown to be highly effective on its own, tailoring treatment to the adolescent's goals and motivations would be the best place to start. In addition, asking the patient to tell you why he wants to quit completes the clinical history. This may help to create ideas for helping him to quit successfully. Therefore, the next best step for this patient would be to ask him about his reasons for stopping smoking. Getting patients to tell you their reasons for potentially stopping their substance abuse is a key goal in treatment planning and management for most types of substance abuse. The substance abuse therapy approach known as "motivational interviewing" uses this as its central means of intervention. This approach, through various techniques, gets patients to use "change talk" and tell their interviewer their reasons for quitting and the actions they could take in that regard. Evidence supporting the success of motivational interviewing as the sole treatment for adolescent smoking cessation has been mixed, with follow-up quit rates well under 10%. Therefore, this technique should not be the only treatment approach.

Interventions to help parents more skillfully intervene with adolescent substance abuse disorders, including tobacco abuse, have been found to be useful. However, soliciting the patient's reasons to quit should come first. Furthermore, asking the parents to put pressure on their son to quit may backfire, especially if rebellion against authority is a major motivation for smoking. Lastly, because his parents also smoke, it may diminish the validity of any of their exhortations for him to quit.

Nicotine replacement therapy is not helpful in reducing smoking in adolescents, though it can help in reducing immediate cravings. Bupropion has been found to reduce the number of cigarettes used daily by adults, but it has been found to have less consistent benefits in adolescents reducing or quitting smoking. Bupropion also carries with it several adverse effects, such as agitation, irritability, and gastrointestinal distress and as such would not be the first thing to try. Similarly, varenicline has been found to reduce cigarette use in adults, but there are no data to support this use in children; the black box warning about significant neuropsychiatric side effects for varenicline should give one further pause about its use in adolescents. Varenicline therefore would not be a first approach to smoking cessation in adolescents.

PREP Pearls

- There is little benefit of medication treatments for reducing adolescent smoking.
- Bupropion and nicotine replacement have minimal long-term benefits on reducing adolescent smoking.
- Asking patients to describe their own reasons for stopping substance abuse can make subsequent abstinence more likely.

AAP Mental Health Competency:

- Recognize that medication treatments of tobacco abuse are not particularly effective in adolescents

Suggested Reading:

- Hettema JE, Hendricks PS. Motivational interviewing for smoking cessation: a meta-analytic review. I Consult Clin Psycho!. 2010;78(6):868884. doi:10.1037/a0021498
- Kim Y, Myung S-K, Jeon Y-J, et al. Effectiveness of pharmacologic therapy for smoking cessation in adolescent smokers: meta-analysis of randomized controlled trials. Am I Health-Syst Pharm. 2011; 68:219-226. doi:10.21461 ajhp100296
- Muramoto ML, Leischow SJ, Sherrill D, Matthews E, Strayer SJ. Randomized, double-blind, placebo-controlled trial of 2 dosages of sustained-release bupropion for adolescent smoking cessation. Arch Pediatr Adolesc Med. 2007;161(11):1068-1074. doi:10.1001/ archpedi.161.11.1068
- Rollnick S, Miller WR, Butler CC. Motivational Interviewing in Health Care: Helping Patients Change Behavior. New York, NY: Guilford Press; 2007

Item 117

You are assessing a term neonate who was born by vaginal delivery that was complicated by shoulder dystocia and thick meconium. The neonate emerged limp and apneic. No meconium was found below the vocal cords after the initial intubation and suctioning were performed. The neonate required resuscitation with bag-mask ventilation for 2 minutes. Apgar scores assigned were 1 at one minute (1 for heart rate) and 6 at five minutes (2 for heart rate, 2 for respiratory effort, 1 for color, 0 for tone, and 1 for reflex irritability). Examination reveals an alert newborn with mild retractions and decreased tone, with a temperature of 36.7°C, heart rate of 110 beats/min, respiratory rate of 80 breaths/ min, blood pressure of 64/40 min Hg, and oxygen saturation of 95% on room air. An arterial blood gas obtained 20 minutes after delivery reveals a pH of 7.22, $PaCO_2$ of 51 min Hg, PaO_2 of 60 min Hg, bicarbonate of 19 mEq/L (19 mmol/L), and base deficit of -7.

Of the following, the MOST appropriate next step in management for this neonate is

A. intravenous bolus of 10 mL/kg normal saline
B. intravenous infusion of 1 mEq/kg sodium bicarbonate
C. intubation for mechanical ventilation
D. observation of cardiorespiratory status
E. placement in a 100% oxygen hood

Item 117 TE Preferred Response: D

The neonate described in the vignette likely has delayed fetal to neonatal transition and his cardiorespiratory status should continue to be closely observed. Although the neonate has mild respiratory distress, the arterial blood gas reveals adequate oxygenation and ventilation 20 minutes after birth. Normal values for arterial blood gases in the neonate are dependent on the time after birth, with the oxygen pressure (PO_2) increasing and carbon dioxide pressure (PCO_2) decreasing as the pulmonary blood pressure drops and the pulmonary blood flow increases. Normal values for full-term neonates several hours after birth include a PaO_2 ranging between 60 and 90 mm Hg and a $PaCO_2$ ranging between 35 and 45 mm Hg.

Oxygenation is expected to increase after birth because of improvement in pulmonary blood flow as pulmonary resistance decreases. The sixth edition of the Neonatal Resuscitation Program of the American Academy of Pediatrics and American Heart Association has defined goal preductal oxygenation saturations (SpO_2) for neonates in the delivery room, which reflect this transition over time (Item C117).

Item C117. Targeted Preductal Spo_2 After Birth

1 min	60% – 65%
2 min	65% – 70%
3 min	70% – 75%
4 min	75% – 80%
5 min	80% – 85%
10 min	85% – 95%

Reprinted with permission from Kattwinkel J, Perlman JM, Aziz K, et al. Part 15: neonatal resuscitation — 2010 American Heart Association Guidelines for Cardiopulmonary Resuscitation and Emergency Cardiovascular Care. *Circulation*. 2010;122:S909-S919

The actual PO_2 range that is optimal for a specific neonate is contingent on other factors. Full-term infants who have conditions associated with increased pulmonary pressure including persistent pulmonary hypertension or congenital diaphragmatic hernia, may require slightly higher PO_2 values to decrease pulmonary resistance. The goal PO_2 values in premature infants with respiratory distress are often lower, which may decrease the risk of the development of retinopathy of prematurity and chronic lung disease.

Alveolar ventilation is best reflected by PCO_2. Some neonates may hyperventilate to increase minute ventilation to assist with ventilatory impairment or compensate for a metabolic acidosis. Attempts should be made to not artificially drop the PCO_2 values significantly, because hypocapnea is associated with decreased cerebral blood flow and neurologic deficits. Conversely, permissive hypercapnia is now practiced in many neonatal intensive care units in an attempt to decrease the incidence and severity of chronic lung disease.

The infant in the vignette has adequate oxygenation in room air with a PaO_2 of 60 mm Hg and does not require 100% oxygen hood. Intubation for mechanical ventilation is also not needed because the oxygenation and ventilation are adequate. Chest radiography could be considered based on clinical findings. The infant does need to be monitored closely for evidence of worsening respiratory status, at which point further evaluation and

delivery of supplemental oxygen or intubation for mechanical ventilation may be needed. The neonate does not require volume expansion with normal saline because he does not show any evidence of volume loss or hypotension. Although the arterial blood gas does reflect a component of metabolic acidosis, it does not warrant treatment with sodium bicarbonate.

PREP Pearls

- Full-term infants have cyanosis at birth and transition gradually as ventilation increases and pulmonary arterial pressure decreases, taking up to 10 minutes to saturate in the 90% range.

American Board of Pediatrics Content Specification(s):

- Know the normal arterial blood gas values for a newborn infant (p02 60 to 90 mm Hg, pCO2 35 to 45 mm Hg)

Suggested Reading:

- Brouillette RT, Waxman DH. Evaluation of the newborn's blood gas status. Clin Chem. 1997; 43: 215-221
- Kattwinkel J, Perlman JM, Aziz K, et al. Part 15: neonatal resuscitation-2010 American Heart Association Guidelines for Cardiopulmonary Resuscitation and Emergency Cardiovascular Care. Circulation. 2010;122:S909-S919. doi: 10.1161/ CIRCULATIONAHA.110.971119

Item 118

A 16-year-old boy in your care was recently diagnosed with osteosarcoma. His paternal grandfather died of brain cancer at 60 years of age, his father survived adrenocortical carcinoma at 30 years of age, and the father's brother had a soft tissue sarcoma at 35 years of age. The patient's mother is well; no cancer has been seen on her side of the family. The parents ask you about the risk of cancer in their daughter.

Of the following, the MOST appropriate response would be that the

A. cancer predisposition pattern appears to be X-linked, so it is unlikely that the daughter would be at increased risk
B. daughter may have an increased risk of developing breast cancer
C. daughter should receive yearly imaging for brain cancers when she reaches adulthood
D. daughter will likely need a bone marrow transplant in her lifetime
E. family's history is unlikely to be related to any cancer syndrome because the cancer types are different

Item 118 I-C TE Preferred Response: B

The most appropriate response to the family's concerns would be that their daughter may have an increased risk of developing breast cancer. Sarcomas account for less than 1% of cancers in adults and 12.6% of malignant tumors in children younger than 19 years. Therefore, when a child who is diagnosed as having a sarcoma has a family history of multiple family members with cancer, genetic susceptibility to cancer should be considered.

Li-Fraumeni syndrome (LFS) is an inherited cancer predisposition syndrome associated with mutations in the tumor suppressor TP53. The classic criteria for LFS was initially defined by the occurrence of sarcoma in a proband at younger than 45 years with a first-degree relative with any cancer by 45 years of age, plus another first- or second-degree relative with cancer by the age of 45 years or sarcoma at any age. The criteria have been modified over the years for various Li-Fraumeni-like syndromes (LFLSs), which take into account specific types of cancers and different ages at presentation. Among patients who are referred for testing based on criteria for LFS or LFLSs, 29% are found to have a mutation in TP53. Families with cancer predisposition syndromes may present with a variety of cancers, some which may be rare, and often at an earlier age when compared with the general population. In families with LFS, cancers occur at earlier ages with each successive generation as seen in the patient in this vignette. If there is a likelihood of a cancer predisposition syndrome, the family should be referred to a cancer geneticist for testing and counseling.

The most common cancers associated with LFS and LFLSs in infants are leukemia and brain tumors, whereas in older children and adolescents soft tissue sarcomas and osteosarcomas are more likely. The risk of adrenocortical carcinoma remains high from infancy through young adulthood, and any child with this malignant tumor should be evaluated for mutations in TP53. However, in young women with LFS or LFLSs, breast cancer is by far the most common cancer.

Somatic changes in TP53 are the most common gene mutations in human cancers and have been reported to be present in 10% to 60% of cancers. Missense mutations occur most commonly and lead to inactivation of the gene, which then disrupts its antiproliferative and growth-suppressive functions. Some mutations can also cause a gain of function and actually promote tumor development. Overall, in patients with TP53 mutations, the most common cancer reported was breast cancer, followed by sarcomas. Although sarcomas are rare in the general population, in patients with germline TP53 mutations, they account for 25% of cancer diagnoses overall and 37% of cancers in patients younger than 20 years.

The family in the vignette meets the clinical criteria for LFS and should be referred for genetic counseling. The data reveal that multiple types of cancer are found in families with LFS or LFLSs. The inheritance pattern is not X-linked. Although LFS families have an increased risk of brain cancers in infancy (fourth most common cancer in patients with TP53 mutation), breast cancer is far more likely and is the most common cancer in young women with TP53 mutations. Bone marrow transplantation, used as treatment for some

types of high-risk leukemias, is not the most likely outcome for this girl because her risk of breast cancer exceeds her risk of developing leukemia.

PREP Pearls

- Cancer predisposition syndromes should be considered in families with cancers at a young age, relatively rare cancer types, and bilateral/multifocal disease.
- Mutations in TP53, a tumor suppressor gene, are found in 29% of cases of LFS.
- The most common cancers in young adults with TP53 mutations are breast cancer and sarcomas.

American Board of Pediatrics Content Specification(s):

- Recognize that children with a family history of excessive cancers may also be at risk and require screening or evaluation

Suggested Reading:

- Hemel D, Domchek SM. Breast cancer predisposition syndromes. Hematol Oncol Clin N Am. 2010;24:799-814. doi:10.1016/j.hoc.2010.06.004
- Ognjanovic S, Olivier M, Bergemann TL, Hainaut P. Sarcomas in TP53 germline mutation carriers. Cancer. 2012;118:1387-1396. doi:10.1002/ cncr.26390
- Palmero EI, Achatz MI, Ashton-Prolla P, Olivier M, Hainaut P. Tumor protein 53 mutations and inherited cancer: beyond Li-Fraumeni syndrome. Curr Opin Oncol. 2010;22:64-69. doi:10.1097/CC0.0b013e328333bf00
- Tabori U, Malkin D. Risk stratification in cancer predisposition syndromes: lessons learned from novel molecular developments in Li-Fraumeni syndrome. Cancer Res. 2008;68:2053-2057. doi:10.1158/0008- 5472.CAN-07-2091

Item 119

A 3-year-old boy was playing in the park with his family. He ran into his mother's legs, fell to the ground without hitting his head, and started crying. The mother picked him up, noticed he wasn't moving his arms, and brought him to the emergency department. On physical examination, his blood pressure is 108/62 min Hg, heart rate is 110 beats/min, respiratory rate is 40 breaths/min (while crying), and temperature is 37.8°C; there is no nuchal rigidity, meningismus, or tenderness to palpation of the spinous processes, and skin evaluation is normal. His neurologic examination shows an alert, crying toddler; his eyes move conjugately in all directions, and his facial movements are strong and symmetric. There is flaccid paralysis of his upper extremities with normal movements of his lower extremities. Deep tendon reflexes are absent in his arms, he has normal patellar reflexes, and his toes extend on plantar stimulation. The boy's gait is unremarkable.

Of the following, the MOST appropriate next diagnostic test to perform is

A. computed tomography of the brain
B. electromyography and nerve conduction study
C. lumbar puncture
D. magnetic resonance imaging of the brain
E. magnetic resonance imaging of the spine

Item 119 S Preferred Response: E

The boy in the vignette presents with acute flaccid paralysis of his arms after a minor trauma. In conjunction with abnormal response to plantar stimulation, the most likely cause is acute cervical spinal cord dysfunction. This could be the result of transverse myelitis or spinal cord concussion. Other possibilities include ruptured arteriovenous malformation, a spinal cord tumor with acute hemorrhage, or spinal cord infarction. The absence of fever or meningismus makes infection or abscess unlikely. There is no sign of vertebral fracture or other injury on examination. The best imaging choice for acute spinal cord dysfunction in this case is magnetic resonance imaging of the spine.

Acute brain injury typically causes altered mental status, hemiparesis, or hemisensory loss, so results of brain imaging are likely to be normal in this case. Electromyography and nerve conduction study can show injury to the nerves or muscles but are not helpful for evaluating acute spinal cord dysfunction. Lumbar puncture is not the best diagnostic test to evaluate for acute spinal cord dysfunction, unless infection is likely.

Acute spinal cord dysfunction is an emergency. Even in cases with a low expectation of traumatic cause, the patient should be immobilized and transported to the nearest emergency department. High cervical cord lesions can impair the nerves to the diaphragm, so close attention to respiration is important.

PREP Pearls

- Acute weakness of both legs or both arms suggests acute spinal cord dysfunction.
- Acute spinal cord dysfunction is a neurologic emergency.
- Acute bowel or bladder dysfunction can be an indication of spinal cord dysfunction.

American Board of Pediatrics Content Specification(s):

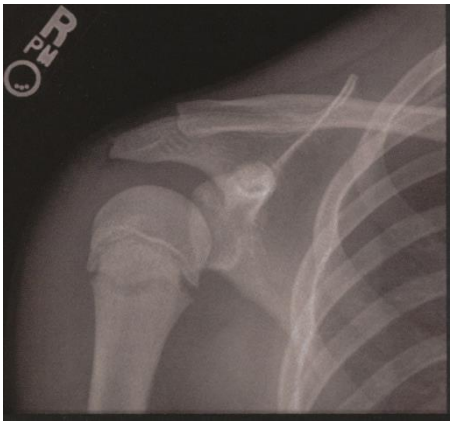
- Plan the initial neurodiagnostic evaluation in a patient with acute cord dysfunction
- Recognize the significance of bladder and bowel dysfunction in spinal cord disease

Suggested Reading:

- Rekate HL. Spinal cord disorders In: Kliegman RM, Stanton BF, St Genie JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders; 2011:2101-2108

Item 120

A 12-year-old boy presents to your office with his parents for evaluation of right shoulder pain. The boy reports that his pain began 3 weeks ago during tennis practice. He plays approximately 10 hours of tennis each week and experiences pain when he serves the ball. On physical examination, you note tenderness over the proximal humerus and pain with resisted shoulder elevation. The remainder of the physical examination is normal. You obtain radiographs; the patient's anteroposterior shoulder radiograph is shown (Q120).



ITEM Q120: Radiograph for the boy described in the vignette.

Of the following, the MOST appropriate statement to include in your discussion with this family is that this child

A. does not have a structural injury and can return to all tennis activities
B. had an injury to the biceps tendon that resulted from a single forceful motion
C. has an injury to the glenoid labrum that resulted from a single forceful motion
D. has an injury to the proximal humerus growth plate that resulted from repetitive overhead motion
E. has an injury to the rotator cuff that resulted from repetitive overhead motion

Item 120 S TE Preferred Response: D

Overuse injuries occur when individuals participate in repetitive physical activity without adequate rest. When an athlete engages in activities involving repetitive motion, the structures of the skeleton may begin to break down; without a sufficient respite from activity, the body cannot adequately repair the damage. The athlete subsequently experiences pain and disability. The pediatric skeleton is especially vulnerable to injury to the physes and apophyses, the cartilaginous growth centers of the bones. The child in the vignette has epiphysiolysis, separation of the epiphysis, involving the proximal humeral physis; this condition is often referred to as "Little League shoulder" because it occurs more commonly in baseball pitchers. Proximal humeral epiphysiolysis generally affects athletes between 11 and 16 years of age who engage in repetitive overhead activities. The diagnosis of Little League shoulder is based on clinical examination findings. Obtaining radiographs, including contralateral views for comparison, may be helpful because the presence of a widened proximal humeral epiphysis on the symptomatic side supports the diagnosis.

The treatment of "Little League" shoulder includes initial rest from overhead activities, physical therapy to strengthen the muscles of the shoulder and upper back, and subsequent gradual return to sports once the athlete is pain free. Counseling athletes, families, and coaches about the importance of learning proper mechanics, the risks of excessive activity, and the need for rest is critical for the prevention of Little League shoulder. For example, USA Baseball has developed recommended numbers of pitches and subsequent rest days for young baseball players; these recommendations are age-specific.

Biceps tendinopathy, which can occur in young athletes, generally results from overuse rather than from a single forceful motion. Rotator cuff tendinopathy and tear are common in adult athletes but rare in the pediatric population. The history and physical examination for the child in the vignette do not suggest shoulder labral pathology, which is most likely to occur as a result of a traumatic shoulder dislocation.

PREP Pearls

- Pediatric athletes are especially vulnerable to overuse injuries involving the physes and apophyses.
- Shoulder injuries such as rotator cuff tendinopathy and labral tears, which are common in adults, are relatively rare in children and adolescents.
- Counseling families about the importance of proper mechanics, the risks of excessive activity, and the benefits of rest following repetitive activity is crucial for the prevention of overuse injuries in young athletes.

American Board of Pediatrics Content Specification(s):

- Understand the importance of growth plate fractures and injuries

Suggested Reading:

- Brenner JS; American Academy of Pediatrics Council on Sports Medicine and Fitness. Overuse injuries, overtraining, and burnout in child and adolescent athletes. Pediatrics. 2007;119(6):1242-1245. doi:10.1542/ peds.2007-0887
- Osbahr DC, Kim HJ, Dugas JR. Little league shoulder. Curr Opin Pediatr. 2010;22(1):35-40. doi:10.1097/M0P.0b013e328334584c
- Stein CJ, Micheli LJ. Overuse injuries in youth sports. Phys Sportsmed. 2010;38(2):102-108. doi:10.3810/psm.2010.06.1787
- Zaremski JL, Krabak BJ. Shoulder injuries in the skeletally immature baseball pitcher and recommendations for the prevention of injury. PM R. 2012;4(7):509-516. doi:10.1016/j.pmrj.2012.04.005

Item 121

You are examining a female neonate born following an uneventful pregnancy to a gravida 3, para 3 woman by repeat cesarean section. The neonate weighs 3.3 kg. A chest radiograph ordered as part of an evaluation for transient tachypnea shows vertebral anomalies and scoliosis. The remainder of the examination is unremarkable. The infant is breastfeeding well and has no other obvious problems.

Of the following, this neonate is MOST at risk for

A. cardiopulmonary compromise
B. a connective tissue disorder
C. a cytogenetic abnormality
D. a genitourinary anomaly
E. a neuromuscular disorder

Item 121 TE Preferred Response: D

The infant described in this vignette has congenital scoliosis secondary to vertebral anomalies. Individuals with congenital scoliosis have high risk for other defects, including an approximate 10% risk of cardiac defects and 25% risk of a concomitant genitourinary malformation. Conversely, more than 50% of patients with esophageal atresia have congenital scoliosis. Workup for such infants should include an echocardiogram, renal ultrasound, and magnetic resonance imaging of the spine to assess for intraspinal anomalies such as lipomas or cord compression resulting from extradural bony or cartilaginous prominences. Unlike scoliosis that develops after infancy, congenital scoliosis with vertebral defects requires extremely close observation and prompt surgical intervention if any progression occurs. Cardiopulmonary compromise is seen in only the most severe cases but can develop over time if the growth of the thoracic cage is compromised. Therefore, early surgical intervention may assist in maximizing spinal growth and preserving alignment while preventing thoracic restriction.

Children with neuromuscular and connective tissue disorders are at increased risk for secondary scoliosis occurring later in childhood. Cytogenetic abnormalities, while often associated with multiple congenital anomalies, are rarely if ever associated with vertebral malformations that result in congenital scoliosis.

PREP Pearls

- Congenital scoliosis is associated with vertebral anomalies as well as increased risks for cardiac and genitourinary defects.
- Early and aggressive surgical intervention for progressive scoliosis may preclude compromised spinal growth and respiratory function down the line.

American Board of Pediatrics Content Specification(s):

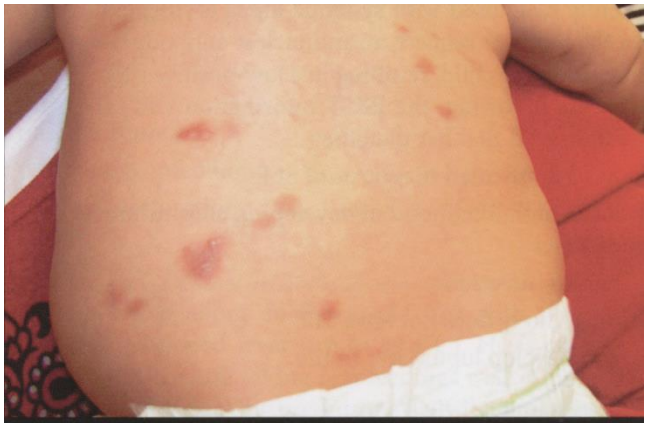
- Know that congenital scoliosis is associated with other congenital abnormalities

Suggested Reading:

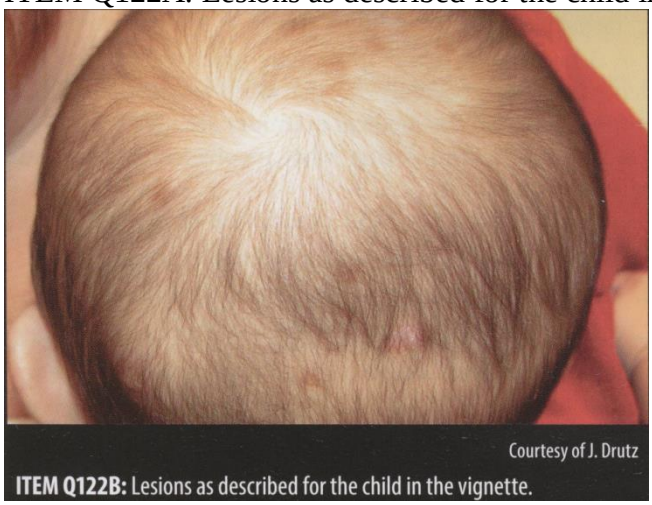
- Scoliosis Research Society. Congenital scoliosis. Scoliosis Research Society Web site.
- Sistonen SJ, Helenius I, Peltonen, Sarna S, Rintala RJ, Pakarinen MP. Natural history of spinal anomalies and scoliosis associated with esophageal atresia. Pediatrics. 2009;124(6):e1198-e1204. doi:10.1542/ peds.2008-3704
- Zieve D, Ogiela D. Scoliosis. PubMed Health Web site

Item 122

During a health supervision visit, the parents of a 6-month-old girl express concern about her "birthmarks" She had a few flat reddish tan lesions on her body that appeared shortly after birth, and she has developed more since that time (Item Q122A). These lesions are present on her trunk, extremities, and scalp (Item Q12213, page Q-31). Her palms and soles are not involved. The parents report that these "birthmarks" sometimes become larger and redder. They attribute this to the girl being overheated or upset. The lesions do not appear to be tender, and the girl is otherwise well. On physical examination, you make the diagnosis by rubbing a lesion on the girl's lower back.



ITEM Q122A: Lesions as described for the child in the vignette.



Courtesy of J. Drutz

ITEM Q122B: Lesions as described for the child in the vignette.

Of the following, the MOST likely diagnosis is

A. bed bug bites
B. cafe au laic macules
C. chronic urticaria
D. erythema nodosum
E. urticaria pigmentosa

Item 122 Preferred Response: E

Urticaria pigmentosa (UP) is the most common form of mastocytosis in childhood. Mastocytosis encompasses a spectrum of disorders characterized by excessive mast cell accumulation that range from solitary cutaneous lesions to diffuse infiltration of skin and other organs. In classic infantile UP, lesions may be present at birth but more often develop in the first postnatal year. New lesions seldom arise after 3 to 4 years of age. Lesions range in size from a few millimeters to several centimeters and may be macular, papular, or nodular. The colors of the lesions range from yellowish tan to brownish red. Stroking of lesions results in rapid appearance of an erythematous wheal and flare (Darier sign) due to local histamine release and is classic for UP, as in the girl in the vignette. The most common symptom is pruritus, which may be triggered by changes in temperature, friction, stress, or ingestion of hot or spicy foods. At times, the lesions may vesiculate when rubbed or when induced by drugs (eg, opiates and nonsteroidal anti-inflammatory drugs) or heat. Sufficient histamine may be released to produce systemic symptoms, most commonly flushing. More serious signs of histamine release, such as hypotension, syncope, tachycardia, wheezing, vomiting, or diarrhea, may occur but are more likely only present in the most severe types of mastocytosis. Dermatographism of intervening normal skin is also common. Most infants with UP do not require treatment other than intermittent symptomatic control with antihistamines. With age, the lesions become more difficult to urticate or blister. Patients are often asymptomatic by 5 years of age and typically have complete resolution of lesions by adolescence.

The classic Darier sign would not be evident in cases of bedbug bites or café au lait macules. Urticarial lesions are well circumscribed, raised, erythematous plaques, often with central pallor. Individual lesions may appear and enlarge in minutes to hours and then disappear completely. Urticaria i s defined as chronic when it has been recurrent for 6 weeks or longer. Erythema nodosum is characterized by the abrupt onset of symmetric, tender, erythematous nodules on the extensor surfaces of the extremities.

PREP Pearls

- A helpful diagnostic sign for urticaria pigmentosa is the Darier sign: rubbing the pigmented lesions results in rapid appearance of an erythematous wheal and flare due to local histamine release.
- At times, the lesions may vesiculate when rubbed or when induced by drugs (eg, opiates and nonsteroidal anti-inflammatory drugs) or heat.
- Most infants with urticaria pigmentosa do not require treatment other than symptomatic control with antihistamines.

American Board of Pediatrics Content Specification(s):

- Understand that a helpful diagnostic sign in urticaria pigmentosa is pigmented lesions that flare after being rubbed

Suggested Reading:

- Morelli JG. Diseases of the Dermis. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders; 2011:2280-2282
- Paller AS, Mancini AJ. Cutaneous tumors and tumor syndromes: Mastocytosis. In: Paller AS, Mancini AJ. Hurwitz Clinical Pediatric Dermatology. 4th ed. Philadelphia, PA: Elsevier; 2011:205-208

Item 123

You are seeing a 14-year-old boy who has asthma. Over each of the past 2 years, he has experienced 3 to 4 exacerbations that required emergency department visits and oral corticosteroid bursts. You prescribed a daily moderate-dose inhaled corticosteroid (ICS), but the boy admits to taking it only when symptoms worsen. He had no asthma exacerbations 2 years ago when he was using daily ICS. He rates his symptoms as "well controlled" on the asthma control test but is not taking part in any physical activity since it triggers his symptoms. He does not feel his lungs "open up" when taking the daily ICS and would rather use albuterol daily, since that seems to make him feel better.

Of the following, you are MOST likely to encourage the boy to

A. limit physical activity and initiate daily montelukast
B. use albuterol as needed and recommend daily peak expiratory flow meter monitoring
C. use the ICS daily and albuterol as needed
D. use the ICS only during worsening symptoms
E. use twice daily scheduled albuterol along with montelukast

Item 123 Preferred Response: C

For the boy described in this vignette, inhaled corticosteroids (ICS) daily will minimize the bronchial inflammation and is most likely to protect him from frequent wheezing episodes.

A primary goal of asthma management is to encourage maintenance of normal activities. Although exercise is a known trigger of asthma, children with well-controlled asthma should be encouraged to participate in regular sports or physical activity of their choice. Once this child's asthma is controlled with the initiation of an effective dose of inhaled steroids, he should be able to engage in appropriate physical activities.
Montelukast is approved for exercise-induced asthma. It is also considered "alternate" therapy for mild persistent (step 2) asthma per the 2007 National Heart, Lung, and Blood Institute (NHLBI) guidelines. This child has moderate persistent asthma (step 3-4) on the basis of the reported number of exacerbations, limitation of physical activity, and probable daily use of albuterol. Montelukast as monotherapy would therefore not be an appropriate choice in this child.

Use of daily albuterol may help the child feel symptomatically better but will not improve the underlying bronchial inflammation and hyperresponsiveness that is causing the symptoms of uncontrolled asthma. In fact, daily scheduled use of albuterol is not recommended by the 2007 NHLBI guidelines because it may be associated with worsening exacerbations. The use of more than one short-acting β-agonist (SABA) canister per month as rescue treatment suggests overreliance on this drug and inadequate control of asthma. Discontinuation of SABAs after chronic use has been associated with decrease in lung function, particularly in patients homozygous for arginine at position 16 (Arg/Arg 16) of the β2-adrenergic receptor compared with those homozygous for glycine (Gly/Gly 16) at this position.

Use of an inhaled steroid on an as-needed basis, when symptoms worsen, may be intuitively considered by patients in their effort to lessen the dose of ICS, or an inhaled steroid may be used during worsening exacerbations in nonadherent patients. However, the use of as-needed ICS has not been well studied. A recent randomized controlled trial explored this question in children and adolescents with mild persistent (step 2) asthma. In the 4 arms of the study, intervention with (a) rescue ICS plus SABA, (b) increased doses of ICS plus SABA in patients on daily low-dose ICS, or (c) daily low dose ICS plus rescue SABA at the earliest signs of asthma worsening was compared with (d) placebo plus rescue SABA. The study found that the most effective strategy to prevent exacerbations was daily use of ICS. Patients in the as-needed ICS group had no significant changes on linear growth compared with placebo. As-needed use of ICS plus SABA appeared to be an effective step-down strategy for well-controlled mild asthma and was better than using rescue albuterol alone. On the basis of this and other studies validating the superiority of daily use of ICS in attaining asthma control, as-needed t se of ICS is not recommended by the 2007 NHLBI guidelines.

PREP Pearls

- Frequent exercise-induced symptoms in a child with asthma may be a sign of uncontrolled airway inflammation.
- Institution of a daily effective dose of controller inhaled corticosteroid therapy will help minimize the airway inflammation and hyper-responsiveness.
- Scheduled daily albuterol is not recommended for individuals with asthma because it has been associated with worsening exacerbations.

American Board of Pediatrics Content Specification(s):

- Know that long-term treatment with inhaled corticosteroids decrease bronchial inflammation and bronchial hyperresponsiveness

Suggested Reading:

- Israel E, Chinchilli VM, Ford JG, et al.; National Heart, Lung, and Blood Institute's Asthma Clinical Research Network. Use of regularly scheduled albuterol treatment in asthma: genotype-stratified, randomised, placebc controlled cross-over trial. Lancet 2004;364(9444):1505-1512
- Martinez FD, Chinchilli VM, Morgan WJ, et al. Use of beclomethasone dipropionate as rescue treatment for children with mild persistent asthma
- (TREXA): a randomised, double-blind, placebo-controlled trial. Lancet. A
- 2011;377(9766): 650-657. doi:10.1016/S0140-6736(10)62145-9
- National Asthma Education and Prevention Program. Expert Panel Report 3 (EPR-3): guidelines for the diagnosis and management of asthma—summary report 2007 [published correction appears in J Allergy Clin Immunol. 2008;121(6):1330]. JAllergy Clin Immunol. 2007;120(suppl 5):S94-S138. doi:10.1016/j.jaci.2007.09.029
- Spitzer WO, Suissa S, Ernst P, et al. The use of beta-agonists and the risk of death and near death from asthma. N Engl J Med. 1992;326(8):501-50t doi:10.1056/NEJM199202203260801

Item 124

The parents of a 10-year-old girl present to your office for a second opinion. The girl has been having asymptomatic microscopic hematuria for the last 18 months. Her records show a normal physical examination on regularly documented visits, normal renal ultrasonography, and a normal serum chemistry (most recently checked 1 month ago). She has multiple urine analyses showing 4+ blood by urine test strips and 20 to 50 red blood cells per high-power field (HPF). There is no family history of autoimmune diseases or renal failure.

Her physical examination reveals that her temperature is 37.9°C, heart rate is 76 beats/min, respiratory rate is 17 breaths/min, and blood pressure is 110/60 min Hg. She has normal growth parameters and a normal physical examination. Her urinalysis demonstrates a specific gravity of 1.035, a pH of 6.0, 4+ blood, and no leukocyte esterase, protein, or nitrites. Her urine microscopy shows 50 to 100 red blood cells/HPF, less than 5 white blood cells/HPF, and no crystals or bacteria.

Of the following, you are MOST likely to inform the parents that

A. asymptomatic microscopic hematuria is uncommon in schoolchildren
B. persistent microscopic hematuria indicates significant risk for renal failure in the future
C. proteinuria would increase her risk for progressive renal disease
D. she will need a renal biopsy for evaluation of her hematuria
E. she will need a urology referral

Item 124 Preferred Response: C

Isolated asymptomatic microscopic hematuria (>5 RBC/ high-power field) is relatively common in school-aged children, with population-based studies estimating its prevalence at 3% to 4% in a single urine sample test. Persistent asymptomatic microscopic hematuria (hematuria present on repeat testing after 6 months) and asymptomatic microscopic hematuria along with proteinuria are less commonly seen. However, such patients are at increased risk for renal disease and therefore need a detailed evaluation by a pediatric nephrologist. Urology referral for invasive evaluation such as cystoscopy for asymptomatic microscopic hematuria is rarely indicated in pediatric patients.

Evaluation of persistent hematuria includes testing for renal function (serum creatinine, serum urea nitrogen), complete blood count with platelets, and immune-mediated kidney injury as seen in glomerulonephritis (complements, antinuclear antibody, anti-double stranded DNA, erythrocyte sedimentation rate). Urine tests in these patients include urinalysis with microscopy, urine protein-creatinine ratio, and urine calcium-creatinine ratio. Renal imaging on renal-bladder ultrasonography is recommended to evaluate other potential causes such as renal cysts, stones, or masses.

A renal biopsy usually is not indicated for patients with isolated microscopic hematuria. A biopsy is considered in patients with microscopic hematuria in association with proteinuria, elevated serum creatinine, or a positive family history of renal failure at a young age.

Renal biopsies in patients with isolated microscopic hematuria have shown normal findings, thin basement membrane disease, or IgA nephropathy. Thin basement membrane or benign familial hematuria is a benign genetic condition associated with isolated thinning of the glomerular basement membrane and is one of the most common causes of isolated asymptomatic hematuria (prevalence 1%-10% of the population). A family history of microscopic hematuria, absence of renal disease, or positive test result for hematuria in an asymptomatic parent is suggestive of thin basement membrane disease. These patients do not need treatment because thin basement membrane is nonprogressive. However, affected patients need monitoring for the development of hypertension, renal failure, or proteinuria, which may point to progressive renal disease. IgA nephropathy, Alport syndrome, nephrolithiasis, or glomerulonephritis have been reported in association with thin basement membrane. Patients with asymptomatic microscopic hematuria and IgA nephropathy have minimal risk for progressive renal injury in the absence of significant proteinuria or recurrent episodes of gross hematuria. Other common causes of microscopic hematuria include idiopathic hypercalciuria and sickle cell disease/trait.

Patients with persistent asymptomatic microscopic hematuria have a low risk for systemic disease or abnormalities of the urinary tract. However, these patients need to be monitored periodically for the development of proteinuria, hypertension, deteriorating renal function, or signs and symptoms of generalized illness, which may be indicative of progressive renal pathology requiring detailed evaluation.

PREP Pearls

- Isolated asymptomatic microscopic hematuria (>5 RBC/high power field) is relatively common in school-aged children with hematuria, and such patients have a low risk for systemic disease or abnormalities of the urinary tract.
- Thin basement membrane or benign familial hematuria is a benign genetic condition and is one of the most common causes for isolated asymptomatic hematuria.
- Patients with persistent microscopic hematuria (hematuria present for >6 months) need to be monitored periodically for the development of proteinuria, hypertension, or deteriorating renal function. This may be indicative of progressive renal pathology requiring detailed evaluation.

American Board of Pediatrics Content Specification(s):

- Recognize the differential diagnosis and prognosis of patients with persistent microscopic hematuria with and without persistent proteinuria

Suggested Reading:

- Kashtan CE. Familial hematuria. Pediatr Nephrol. 2009;24:1951-1958. doi: 10.1007/s00467-007-0622-z
- Massengill SF. Hematuria. Pediatr Rev. 2008;29:342-348. doi: 10.1542/ pir.29-10-342.
- Reidy KJ, Rio MD. Hematuria In: Mclnerny TK, Adam HM, Campbell DE, Kamat DM, Kelleher KJ, eds. American Academy of Pediatrics Textbook of Pediatric Care. 19th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2009;chap 188:1566-1570

Item 125

A 10-month-old male infant presents with a several-month history of poor weight gain, fevers, persistent upper respiratory tract infections, recurrent diarrhea, and persistent diaper candidiasis. His maternal grandmother is his caregiver and reports that the patient's mother recently died from "an infection. " On physical examination, he is afebrile and vital signs are normal for age. The boy's head circumference and length are at the 20th percentile, but weight is at less than the 5th percentile for age. He has nasal congestion, clear rhinorrhea, oral thrush, and dull tympanic membranes. Auscultation of the lungs reveals coarse breath sounds bilaterally with normal effort. The results of the cardiovascular examination are normal. The abdomen is soft, with the liver and spleen palpated 4 cm and 3 cm below the costal margins, respectively. The patient has normal genitalia with diaper dermatitis consistent with candidiasis. You palpate lymph nodes of less than 0.5 cm in the neck and groin. The peripheral white blood cell count is 4,000/µL (4.0×10^9/L), with 80% polymorphonuclear neutrophils, 10% lymphocytes, and 10% monocytes. His hemoglobin level is 10 g/dL (100 g/L) and platelet count is 280×10^3/µL (280×10^9/L). You are concerned that the patient has an immunodeficiency.

Of the following, the test MOST likely to establish the patient's diagnosis is a(n)

A. antigen detection assay
B. complement activity test
C. enzyme immunoassay
D. polymerase chain reaction assay
E. viral culture

Item 125 Preferred Response: D

The infant's history of numerous infections (including persistent candidiasis) and poor growth, hepatosplenomegaly, and lymphopenia suggest an immunodeficiency most likely due to human immunodeficiency virus (HIV) infection, since his mother recently died from "an infection:' The test most likely to establish the diagnosis is an HIV DNA polymerase chain reaction (PCR) test. This is the preferred test for diagnosing HIV infection in infants and children younger than 18 months who may still have circulating maternal antibodies against HIV. A single HIV DNA PCR test is 95% sensitive for detecting HIV infection in children 1 to 36 months of age. It is 30% to 40% sensitive when performed in infected newborns within the first 48 hours of life. At 2 weeks of life, the sensitivity of the PCR test improves to 93%. Therefore, in infants exposed to HIV, HIV DNA PCR testing is recommended at 14 to 21 days of age, repeated at 1 to 2 months of age if the result is initially negative, and again at 4 to 6 months of age for diagnosing infection.

The p24 antigen detection assay is less sensitive than the HIV DNA PCR test, so it is not recommended routinely. Enzyme immunoassay (HIV antibody test) is the preferred screening test for detecting HIV in children older than 18 months, when maternal anti-HIV antibodies would no longer be present to cause false-positive results. This screening test is sensitive and specific, and positive test results are confirmed by Western blot analysis. HIV isolation in viral culture is expensive, not widely available, time-consuming (results take up to 28 days), and less sensitive than the HIV DNA PCR. A complement activity test can detect terminal complement defects (primary immunodeficiency) in patients with recurrent meningitis caused by meningococcus or pneumococcus, but this test is not helpful in diagnosing HIV.

PREP Pearls

- The most appropriate screening test for HIV infection in children younger than 18 months is the HIV DNA PCR.
- The most appropriate screening test for HIV infection in children older than 18 months is an HIV enzyme immunoassay (antibody test).

American Board of Pediatrics Content Specification(s):

- Know the most appropriate screening test (ie, HIV antibody titer) for HIV infection in children older than 18 months of age

Suggested Reading:

- American Academy of Pediatrics. Human immunodeficiency virus infection. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:418-439
- Schwarzwald H. Diagnostic testing for HIV infection in infants and children younger than 18 months. UptoDate. Available online only for subscription

Item 126

A 13-year-old boy presents for a routine health supervision visit. He is very concerned about a lump on his chest that appears to be increasing in size. You have not seen him in 2 years, and your last note indicates that the results of his examination were normal. He appears well, has normal vital signs, and has a body mass index of 25. He is very embarrassed and reluctantly allows you to examine him. You find a slightly tender, rubbery mass under his right areola that measures approximately 2 cm in diameter. The remainder of his examination, including a genital examination, is unremarkable. He is at sexual maturity rating 2/3.

Of the following, the MOST appropriate next step in this boy's management is to

A. discuss normal development
B. obtain an endocrinology consult
C. obtain ultrasonography of the mass
D. refer him to a plastic surgeon for excision of the mass
E. suggest that treatment for the mass is weight loss

Item 126 Preferred Response: A

The boy in the vignette presents with gynecomastia as evidenced by the presence of a firm rubbery mass under the nipple-areolar complex. In males, during early puberty, the ratio of estrogen to testosterone is increased, and up to 70% of males in sexual maturity rating (SMR) stage 2 of pubertal development (testicular volume of 5-10 mL) have some breast enlargement on examination that may be tender as a result of edema and inflammation. Although initially unilateral, the other breast enlarges in up to 75% of cases. In most males, such breast tissue is no longer palpable after 18 months (1-3 years) because puberty progresses and androgen concentrations increase. Therefore, discussing normal developmental changes and reassuring this adolescent is all that is required at this time. If the mass does not regress after 2 years or by the end of pubertal development, especially if it causes emotional concerns, referral to a plastic surgeon may be considered.

Weight loss is useful only in pseudogynecomastia (also known as lipomastia). This boy is not obese but rather is at the lower limit of the overweight category, with a body mass index of 25; weight loss would not affect his breast size. Ultrasonography of the mass would only be warranted if a pathologic condition is suspected, as with rapid enlargement or a size greater than 4 cm. Local masses, such as hemangiomas, lymphangiomas, lipomas, and neurofibromas, are usually unilateral, not circular, and not directly beneath the areola. Breast cancer in a male adolescent is extremely rare, and the mass would more likely be hard and fixed rather than rubbery and movable. Referral to an endocrinologist should be considered if the history or physical examination suggests a hormonal cause. Estrogen excess may result from exogenous administration (eg, various teas and creams) or endogenous production as in testicular and adrenal tumors and chronic kidney and liver disease. Androgen insufficiency that leads to gynecomastia is seen in certain genetic conditions (eg, Klinefelter syndrome and androgen insensitivity, an X-linked disorder) and in the case of testicular failure (eg, anorchia and primary hypogonadism). A large number of medications and street drugs have been implicated in male breast enlargement due to either an estrogen (stimulatory) or an antiandrogen effect. Some examples are neurologic or psychiatric medications (eg, diazepam and risperidone), antiandrogens (eg, ketoconazole and spironolactone), cardiovascular medications (eg, calcium channel Mockers), antiulcer drugs (eg, omeprazole), antineoplastic medications (eg, methotrexate), antimicrobials (eg, isoniazid), and a host of other exogenous hormones and products (eg, vaginal creams, licorice, and teas). Among the street drugs that have been implicated are anabolic steroids, marijuana, amphetamines, and alcohol.

PREP Pearls

- Male breast enlargement in early puberty is most often caused by physiologic hormonal changes.
- A comprehensive history and physical examination are usually the extent of the evaluation required for male breast enlargement in early puberty.
- Reassurance and follow-up to ensure regression of male breast enlargement in early puberty is usually sufficient.

American Board of Pediatrics Content Specification(s):

- Know the etiology and management of gynecomastia in boys

Suggested Reading:

- Diamantopoulos S, Bac, Y. Gynecomastia and premature thelarche: a guide for practitioners. Pediatr Rev. 2007;28:e57-e68. doi: 10.1542/pir.28-9-e57
- Joffe A. Gynecomastia. In: Neinstein L, Gordon CM, Katzman DK, Rosen DS, Woods ER, eds. Adolescent Health Care: A Practical Guide. 5th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2008:180-184
- Ma NS, Geffner ME. Gynecomastia in prepubertal and pubertal men. Carr Opin Pediatr. 2008;20:465-470. doi: I0.I097/MOP.Ob013e328305e415

Item 127

An 18-year-old, African-American young man comes to your office with a recent history of weight loss. He was well until 1 month ago, when he returned from a camping trip with his friends. During the trip, they ran out of bottled water and drank from a mountain stream near their campsite. Also while camping, he developed an infected cut on his arm and was prescribed clindamycin at a local emergency department. Since returning home, he has been complaining of vague, generalized abdominal discomfort that is associated with 2 to 3 semiformed stools per day. His bowel movements do not contain visible blood or mucus. He denies vomiting, fevers, or joint pains. He does not like dairy products and tries to avoid them. Review of his past records indicates that he has lost 1.5 kg of body weight since his last office visit 6 months ago.

Of the following, the organism MOST likely responsible for this young man's symptoms is

A. Blastocystis hominis
B. Campylobacter jejuni
C. Clostridium difficile
D. Cryptosporidium species
E. Giardia intestinalis

Item 127 Preferred Response: E

The young man in the vignette describes a 1 -month history of loose stools and a 1.5-kg weight loss. These symptoms began shortly after he returned home from a camping trip. In a previously well patient, the presentation of chronic (>2 weeks' duration) diarrhea without gross bleeding can herald either a new-onset disease state or an acquired infection (Item C127, page C-99). Because he drank unpurified water during his camping trip, an infectious origin is most likely, and, in this clinical setting, the most common infecting agent is the parasitic organism Giardia intestinalis (formerly G lamblia). Giardia is one of the most common gastrointestinal parasites, infecting more than 20,000 individuals in the United States annually. Infection is limited to the gastrointestinal tract, where the organism attaches to the intestinal mucosal surface, leading to diarrhea and, in many cases, nutrient malabsorption. Waterborne transmission is the most frequent known mode of spread. Infection is especially prevalent in children younger than 5 years. However, giardiasis may affect any individual who consumes water from contaminated sources (especially streams, lakes, and contaminated wells). Giardia may also colonize domestic and farm animals; however, the role of animal-to-human spread is unclear.

Symptoms of Giardia infection are almost exclusively limited to the gastrointestinal tract. Thus, diarrhea is present in 90% of patients, and abdominal pain, bloating, and flatulence are reported in more than 70%. Anorexia, nausea, and malaise are common symptoms, and weight loss is reported in more than 50% of infected patients. Extraintestinal manifestations are rare and include allergic symptoms, such as urticaria, erythema multiforme, bronchospasm, and reactive arthritis. These manifestations of infection are thought to occur as the consequence of host immune activation.

The young man in the vignette also presents with several "confounding variables" that should be considered. The patient's avoidance of dairy products suggests late-onset, primary lactose intolerance, present in 75% of African Americans (ethnic Chinese have the highest prevalence of lactose intolerance, approaching 95%).This genetically programmed loss of lactase enzyme usually develops toward the end of the first decade of life. However, in lactase-sufficient patients, secondary lactase deficiency may occur in up to 20% of individuals infected with Giardia. His use of clindamycin should alert his physician to the possibility of antibiotic-associated diarrhea caused by Clostridium difficile. Because his symptoms predated the antibiotic prescription, infection with this organism is unlikely. Although gross rectal bleeding is not a universal symptom in C difficile infection, it is the most common associated finding, particularly beyond infancy. Blastocystis hominis is a ubiquitous parasite found in the feces of many healthy, asymptomatic individuals. Its role in disease pathogenesis is unclear at present.

Campylobacter species represent the most common cause of infectious diarrhea in the United States. However, lower GI bleeding, a common symptom of this typically foodborne infection, is absent in this case. The protozoal organism Cryptosporidium parvum was once thought to infect primarily individuals with immune deficiency (especially those infected with human immunodeficiency virus); however, epidemics in healthy hosts have been reported as a result of drinking water from a contaminated

source. Patients typically present with a watery diarrhea, and in the setting described in the vignette, Giardia infection is far more likely.

PREP Pearls

- Giardia intestinalis (formerly G lamblia) is a common acquired cause of carbohydrate and fat malabsorption, usually acquired from contaminated water, often by drinking from streams and lakes.
- In otherwise well, non-white patients with new onset diarrhea, lactose intolerance should be considered.
- Ethnic Chinese have the highest prevalence of lactose intolerance, approaching 95%.

American Board of Pediatrics Content Specification(s):

- Know the clinical manifestations of Giardia lamblia (giardiasis)

Item C127. Major Causes of Chronic Diarrhea in Childhood

Agent	Common Sources
Infectious Causes	
Aeromonas species	Contaminated water
Campylobacter species	Raw poultry, unpasteurized milk or apple juice, farm animals, birds, domestic animals
Clostridium difficile	Antibiotic use, community acquired, nosocomial spread
Cryptosporidium parvum	Petting zoos, contaminated water
Giardia intestinalis (formerly *G lamblia*)	Contaminated lakes and streams, fecal–oral
Other parasitic infections:	
• *Blastocystis hominis*	• Pathogenicity is controversial
• *Cyclospora cayetanensis*	• Unpasteurized apple juice, imported raspberries
• *Entamoeba histolytica*	• Fecal–oral
• *Isospora belli*	• Fecal–oral
• *Strongyloides stercoralis*	• Fecal–oral
• *Yersinia enterocolitica*	• Raw, contaminated pork products
Sprue (tropical)	Travel to endemic area
Noninfectious Causes	
Celiac disease	Irritable bowel syndrome
Chronic nonspecific diarrhea	Lactose intolerance
	Laxative abuse
Congenital chloride diarrhea*	Immunoglobulin A deficiency (G *lamblia* infection)
Congenital enteropathies (including tufting enteropathy and microvillus membrane inclusion disease)*	Other immune deficiencies
	Sucrase-isomaltase deficiency*
Excessive sorbitol intake	
Food allergy	
Food poisoning	
Glucose-galactose malabsorption*	
Glucoamylase-maltase deficiency*	
Inflammatory bowel disease	

*Congenital disorders with onset of diarrhea during infancy.

Suggested Reading:

- Bailey JM, Erramouspe J. Nitazoxanide treatment for giardiasis and cryptosporidiosis in children. Ann Pharmacother. 2004;38:634-640. doi:10.1345/aph.1D451. http://www.ncbi.nlm.nih.gov/pubmed/14990779
- Huang DB, White AC. An updated review on Cryptosporidium and Giardia. Gastroenterol Clin North Am. 2006;35:291-314. doi:10.1016/j. gtc.2006.03.006
- John CC. Giardiasis and balantidiasis. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:1180-1183
- Keating JP. Chronic diarrhea. Pediatr Rev. 2005;26:5-14. doi:10.1542/ pir.26-1-5
- Pickering LK. Giardia lamblia (Giardiasis). In: Long SS. Principles and Practice of Pediatric Infectious Diseases. 3rd ed. Philadelphia, PA: Churchill Livingstone Elsevier Inc; 2008:1241-1245

Item 128

A 3-month-old infant from your practice has died suddenly and unexpectedly in the sleep environment. Law enforcement and the medical examiner are investigating the death. You are considering what involvement you should have with this family.

Of the following, the BEST description of your role is to

A. arrange a meeting only if the parents request it
B. arrange to meet with the family and surviving siblings
C. discuss grief issues only if the parents report a concern
D. leave the medical examiner to review the autopsy and investigation with the family
E. provide medical records to law enforcement but avoid discussing the case with the family

Item 128 P TE I-C SBP Preferred Response: B

Sudden unexpected infant death (SUID) is an overwhelming event, requiring a wide range of coping skills within families. The pediatrician can play an important role helping families deal with the community and legal response to the tragic event as well as their own grief. Because of the unexpected nature of the death and the differential diagnosis that includes child abuse, the death must be scrutinized with a scene investigation and autopsy. In areas in which these deaths are rare, the pediatrician may need to advocate for an appropriate investigation. Despite the involvement of law enforcement, the pediatrician should remain a resource for families to help them understand the processes in place and to review the results once they become available. Although many medical examiners meet with families to review results, families may feel that discussing the findings with a trusted professional such as the family pediatrician is valuable. For this reason, many experts recommend that the pediatrician schedule a meeting with the family within a few weeks after the infant's death. In addition, the physician can use this opportunity to refer the family to community resources including family support groups such as First Candle/SIDS Alliance (www.firstcandle.org). Beyond the immediate response to SUID, the pediatrician should remain an important support for families. If there are surviving siblings, their own as well as their parents' grief responses should be assessed at subsequent well child and sick visits, and bereavement referrals made as indicated for the siblings and the parents.

In addition to the official legal investigation, many communities have recently instituted child fatality review programs that may request information from the pediatrician. Confidentiality restrictions vary among jurisdictions, so the practitioner should consult with the local program to determine the statutes regulating release of information in their community. The purpose of these programs is to develop a greater understanding of the causes of child mortality and to promote community responses that would decrease the burden of child deaths.

PREP Pearls

- Many experts recommend that the family pediatrician schedule an appointment with the family a few weeks after a sudden unexpected infant death to review the findings of the investigation and provide referrals to support groups or bereavement resources.
- Future health supervision visits provide an opportunity to discuss grief responses in the surviving siblings and parents.
- Pediatricians may be contacted by their local child fatality review program for information to aid in public health responses to sudden unexpected infant death.

American Board of Pediatrics Content Specification(s):

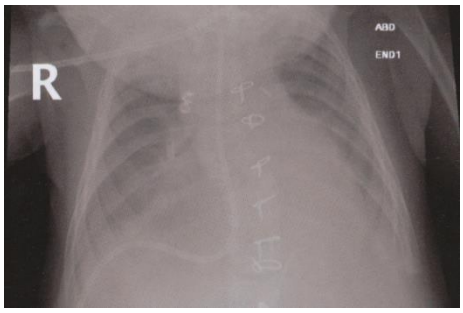
- Recognize the importance of physician review of case with parents after SIDS has occurred (including risk of SIDS in siblings)

Suggested Reading:

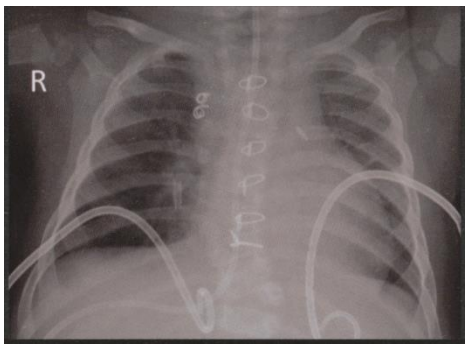
- Linebarger JS, Sahler OJ, Egan KA. Coping with death. Pediatr Rev. 2009;30:350-355. doi: 10.1542/pir.30-9-350
- Moon RY, Fu LY. Sudden infant death syndrome. Pediatr Rev. 2007;28:209214. doi:10.1542/pir.33-7-314

Item 129

While at the hospital, you visit the family of a 6-weekold boy who has been recovering in the cardiac intensive care unit after undergoing a stage I Norwood procedure for hypoplastic left heart syndrome. The surgery and postoperative course have gone well, and the patient is being fed standard formula through a nasoenteric tube. The bedside nurse informs you that the infant has developed large bilateral pleural effusions (Item Q129A) this morning and has had bilateral thoracostomy tubes placed (Item Q129B) with a significant amount of milky white fluid that tests positive



ITEM Q129A: *Anterior-posterior radiograph of the chest of the patient in the vignette demonstrating bilateral diffuse haziness and blunting of the costophrenic angles consistent with pleural effusions*.



ITEM Q129B: *Anterior-posterior radiograph of the chest of the patient in the vignette following bilateral throacostomy tube placement*.

for a large amount of triglycerides and a predominance of lymphocytes.
Of the following, the MOST likely diagnosis in this patient is

A. chylothorax
B. empyema
C. heart failure
D. hemothorax
E. lymphoma

Item 129 TE Preferred Response: A

The infant described in the vignette has chylothorax, a leak of chyle into the pleural space, which most often results from an injury to the thoracic duct during cardiac surgery (50% of cases). Less common causes include chest injuries, intrathoracic malignancies, lymphangiomatosis, infections, and thrombosis of the thoracic duct or subclavian vein. Chylous fluids are a characteristic milky white and contain high concentrations of triglycerides (generally >110 mg/dL [1.24 mmol/L). The electrolyte composition is similar to plasma, and cell counts will reveal a lymphocytic predominance. Of note, chyle is rich in immunoglobulins so patients with ongoing chylous effusions may need supplementation with immune globulin intravenous.

Chylothoraces are difficult to manage: the first steps are drainage, usually with the use of thoracostomy tubes, and treating the underlying condition if possible. Instituting a low-fat diet decreases the flow of chyle through the thoracic duct, potentially allowing spontaneous closure of a defect in the duct. Excluding long-chain triglycerides from the diet prevents their conversion into monoglycerides and free fatty acids which are normally transported as chylomicrons to the intestinal lymph ducts and, ultimately, the thoracic duct. The diet should be supplemented with medium-chain triglycerides that are absorbed directly to the liver via the portal vein and bypass the thoracic duct. Some advocate eliminating all enteral feeds but conclusive evidence supporting this recommendation is lacking. Eliminating all enteral feeds necessitates the use of total parenteral nutrition, an intervention that is invasive, expensive, and challenging for children and families.

For children who do not respond to drainage and dietary measures after several weeks, other options include the use of chemical pleurodesis with an agent such as doxycycline. Pleurodesis is very painful and has been shown to be more successful with nonoperative causes of chylothorax. Thoracic duct ligation is performed when the chylous effusion remains high-volume despite drainage and dietary measures. Infusion of somatostatin or octreotide (somatostatin analogue) are thought to decrease the flow of lymph but are used less commonly because of concerns about their effectiveness and potential adverse effects. Pleuroperitoneal and pleurovenous shunts are used in patients who have chronic effusions that are unresponsive to other treatments.

For the patient described in the vignette, the pleural fluid composition is not consistent with empyema or hemothorax. Although congestive heart failure and lymphoma can cause chylous effusions, it would be an unusual cause in this age group and the recent cardiac surgery is a much more likely etiology.

PREP Pearls

- Chylothorax most often results from an injury to the thoracic duct during cardiac surgery.
- Chyle has a characteristic milky white appearance, high concentrations of triglycerides, and electrolyte composition similar to plasma, and a predominance.
- Chylothoraces are difficult to manage and often require a combination of diet, medical, and surgical management.

American Board of Pediatrics Content Specification(s):

- Know the characteristics of pleural fluid due to chylothorax

Suggested Reading:

- Efrati 0, Barak A. Pleural effusions in the pediatric population. Pediatr Rev. 2002:23:417-426. doi:10.1542/pir.23-12-417.
- Heffner JE. Management of chylothorax. UptoDate. Available online only for subscription
- Heffner JE. Etiology, clinical presentation, and diagnosis of chylothorax. UptoDate. Available online only for subscription
- Winnie GB, Loddrg SV. Chylothorax. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:1514-1515

Item 130

An 11-year-old girl is seen for evaluation of short stature. Her past medical history is significant for swelling of her right foot, noted at birth, that never resolved. She also had a lisp that required speech therapy, and pressure equalization tubes placed 4 times for recurrent otitis media. Her height is at the first percentile, well below her midparental target height at the 90th percentile. Physical examination reveals a high-arched palate, bilateral short fourth metacarpals, and moderate edema of the right foot.

Of the following, the MOST likely diagnosis is

A. Noonan syndrome
B. pseudohypoparathyroidism
C. Russell-Silver syndrome
D. Turner syndrome
E. Williams syndrome

Item 130 Preferred Response: D

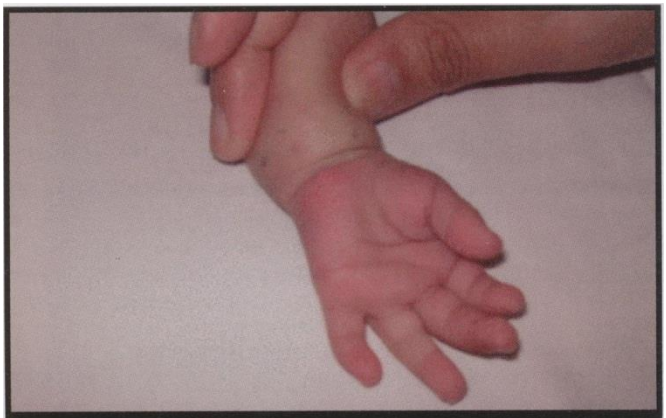
Girls with Turner syndrome, such as the girl in the vignette, have characteristic physical features coupled with complete or partial absence of the second sex chromosome. Guidelines from the Turner Syndrome Study Group recommend considering the diagnosis of Turner syndrome in any female with unexplained growth failure, pubertal delay, or any constellation of the clinical findings shown (Item C130A).
The girl described in this vignette had lymphedema of one foot, a high arched palate, recurrent otitis media, short metacarpals, and poor linear growth. The most common clinical feature seen in Turner syndrome is poor linear growth (short stature). A karyotype should be considered in a female with abnormal linear growth and otherwise normal screening test results, even with normal physical examination findings and no other phenotypic findings of Turner syndrome.

Noonan syndrome is characterized by proportionate postnatal short stature, dysmorphic facial features, chest deformities, and congenital heart disease (most commonly pulmonary valve stenosis and hypertrophic cardiomyopathy). Developmental delay, cryptorchidism, and clotting disorders are also common. This condition is autosomal dominant. Both males and females can have Noonan syndrome.

Children with pseudohypoparathyroidism can also have short metacarpals. However, this is a nonspecific finding, and none of the other characteristic hormonal or phenotypic findings of pseudohypoparathyroidism are present in this girl. In addition to resistance to parathyroid hormone and other endocrine hormones, children with pseudohypoparathyroidism can have round facies with a low nasal bridge, obesity, disproportionate shortening of the limbs (particularly brachydactyly of third, fourth, and fifth metacarpals and first distal phalanx), heterotopic ossification, and mental retardation.

Russell-Silver syndrome is characterized by short stature of prenatal onset; however, phenotypic findings include small triangular facies, downturned corners of the mouth, and micrognathia. Short, inward curved fifth fingers (clinodactyly) as shown (Item C130B, page C-102) are a common finding in Russell-Silver syndrome and are distinctly different from the short metacarpals found in Turner syndrome.

Although both Williams and Turner syndromes are characterized by short stature, Williams syndrome has classic facial features that include medial flaring of the eyebrows, depressed nasal bridge, anteverted nares, long philtrum, and prominent lips. A stellate pattern of the iris is unique to Williams syndrome. Hypercalcemia can also be present.



Courtesy of Prasad D, Navarrette V, Naganathan S. Visual Diagnosis: Infant with growth failure, body asymmetry and dysmorphic features. *Pediatr Rev*: 2013: e17 -e21

ITEM C130B: Clinodactly as seen in patients with Russell Silver syndrome.

Item C130A. Clinical Findings in Girls with Turner Syndrome

Clinical findings	
Unexplained growth failure	— Short stature with growth velocity less than the 10th percentile for age
Pubertal delay	— Markedly elevated levels of follicle-stimulating hormone
Cardiac anomalies	— Especially coarctation of the aorta or hypoplastic left heart
Characteristic facies	— Low hairline, low-set ears, small mandible
Nails	— Hypoplasia, hyperconvex and uplifted
Skeletal	— Congenital hip dislocation, cubitus valgus, short fourth metacarpal, scoliosis
Skin	— Widespread nipples, shield chest, edema of the hands or feet, nuchal folds, multiple pigmented nevi
Other	— Learning disabilities, social awkwardness, type 2 diabetes mellitus, inflammatory bowel disease, high-arched palate or chronic otitis media

PREP Pearls

- The most common clinical feature seen in Turner syndrome is poor linear growth (short stature).
- Turner syndrome can be present even with normal physical examination findings and no phenotypic findings other than poor growth or delayed puberty.

American Board of Pediatrics Content Specification(s):

- Recognize the signs and symptoms of gonadal dysgenesis (Turner syndrome)

Suggested Reading:

- Bacino CA, Lee B. Sex Chromosome Aneuploidy. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:408-410
- Bondy CA; Turner Syndrome Study Group. Care of girls and women with Turner syndrome: a guideline of the Turner Syndrome Study Group. I Clin Endocrinol Metab. 2007;92:10-25
- Pinsker JE. Clinical review: Turner syndrome: updating the paradigm of clinical care. I Clin Endocrinol Metab. 2012;97:E994-E1003. doi:10.1210/jc.2012-1245

Item 131

A 4-month-old infant is seen in the emergency department with a 2-week history of progressive coughing associated with duskiness. Nasopharyngeal swab is positive for pertussis. The nurse who was caring for the child prior to the diagnosis was not wearing a mask during extensive contact with the infant. She reports receiving a booster dose of diphtheria-pertussis-tetanus vaccine 6 years ago.

Of the following, the MOST appropriate treatment for this nurse is

A. azithromycin for 5 days
B. a booster dose of diphtheria-pertussis-tetanus vaccine
C. erythromycin for 7 days
D. no treatment necessary
E. trimethoprim-sulfamethoxazole for 7 days

Item 131 S TE Preferred Response: A

Antibiotic prophylaxis is recommended for all health care personnel who have unprotected exposure to pertussis, regardless of prior immunization with the diphtheria, tetanus, and pertussis (dTap) vaccine, if they are likely to expose a patient at high risk of severe pertussis. Of the antibiotics listed in the vignette, azithromycin for 5 days would be the appropriate choice.

Erythromycin is also an acceptable agent for prophylaxis against pertussis, but 14 days of therapy is the recommended duration. Although erythromycin is a cheaper agent with narrower antimicrobial spectrum compared with azithromycin, many clinicians prefer azithromycin in this setting because of the shorter dosing schedule and lower incidence of gastrointestinal upset.

Trimethoprim-sulfamethoxazole is considered an alternative agent for treatment or prevention of pertussis in patients older than 2 months who cannot tolerate macrolides, but studies evaluating trimethoprim-sulfamethoxazole in cases of pertussis are limited. A second dose of dTap is not recommended at present except for mothers during pregnancy.

PREP Pearls

- Antibiotic prophylaxis is recommended regardless of vaccine status after significant exposure to pertussis.
- A macrolide is the preferred agent for treatment and prevention of pertussis.
- Trimethoprim—sulfamethoxazole is considered an alternative for treatment and prevention of pertussis in the individual unable to tolerate macrolides.

American Board of Pediatrics Content Specification(s):

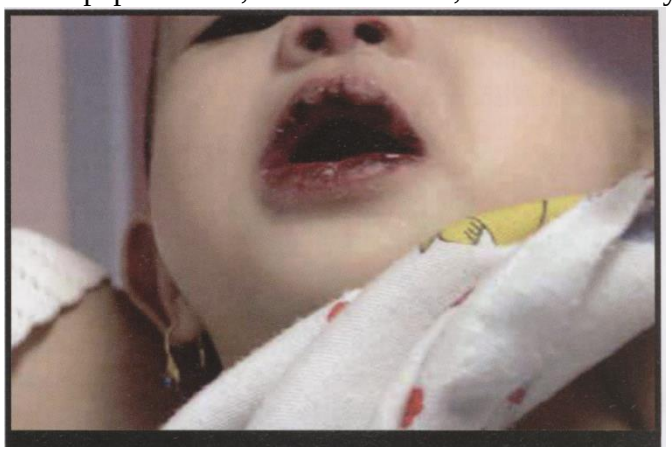
- Plan the use of chemoprophylaxis for the contacts of patients who have pertussis

Suggested Reading:

- American Academy of Pediatrics. Pertussis (whooping cough). In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:553-566
  Tiwari T, Murphy TV, Moran J. Recommended antimicrobial agents for treatment and postexposure prophylaxis of pertussis: 2005 CDC guidelines. MMWR Recomm Rep. 2005;54(RR14):1-16

Item 132

A 2-year-old girl presents to your office after 5 days of fever (up to 38.3°C). Three days ago, she was seen in the emergency department and started on amoxicillin for the diagnosis of otitis media. On physical examination, the patient is irritable and has injection of the conjunctiva bilaterally without dis-charge, dry cracked lips, a red tongue, a fine papular rash, and a unilateral, 2-cm cervical lymph node (Item Q132).



ITEM Q132: *Findings for the child described in the vignette.*

The following are the results of laboratory tests for the girl:

- White blood cell count, 16,000/µL (16 x 10^9/L), with 55% polymorphonuclear leukocytes, 40% lymphocytes, and 5% monocytes
- Platelets, 340 x 10^3/µL (340 x 10^9/L)
- Erythrocyte sedimentation rate, 45 mm/h,
- C-reactive protein, 30.1 mg/L (287.6 nmol/L)
- Aspartate transaminase, 200 U/L
- Alanine transaminase, 235 U/L

Of the following, the MOST likely diagnosis is

A. herpes gingivostomatitis
B. Kawasaki disease
C. pharyngoconjunctival fever
D. Stevens-Johnson syndrome
E. toxic shock syndrome

Item 132 Preferred Response: B

The child described in the vignette has Kawasaki disease (KD), the second most common vasculitis in childhood. There is no specific test that diagnoses KD. It is a clinical diagnosis based on history and physical examination criteria. Diagnostic criteria for KD include fever that persists for 5 days plus 4 of the following: bilateral conjunctival injection without exudate, changes of the lips and oral cavity, cervical lymphadenopathy greater than 1.5 cm in diameter, polymorphous exanthem, or changes in the peripheral extremities or the perineal area. The clinical course of KD is triphasic, with an acute febrile stage that can last 2 weeks if untreated, a subacute phase lasting 2 to 4 weeks, and a convalescent phase that can last months. Laboratory studies can support the diagnosis in patients that do not fulfill the diagnostic criteria. Acute phase reactants such as erythrocyte sedimentation rate (ESR) and C-reactive protein are commonly elevated. These may be normal in the early febrile period but are usually increased if repeated later in the disease course. Leukocytosis is common and is characterized by a neutrophil predominance. A normochromic normocytic anemia may be present in patients with prolonged inflammation, and thrombocytosis can be seen in the second or third week of disease. Leukopenia and thrombocytopenia are uncommon. Often, serum transaminase levels are mildly elevated, and hypoalbuminemia is common. Hyperbilirubinemia and cerebrospinal fluid pleocytosis may be present. Patients who have KD often have a sterile pyuria.

If KD is untreated, the symptoms may resolve; however, therapy is important in the prevention of coronary artery aneurysms. Kawasaki disease is treated with high-dose immune globulin intravenous (IGIV) and high-dose aspirin during the acute febrile phase with elevated inflammatory markers. The patient is switched to low-dose aspirin once becoming afebrile with normalization of the acute phase reactants. Occasionally, a second administration of IGIV or systemic corticosteroid therapy is used. If the patient has refractory KD, other therapy may be required. Of note, administration of IGIV will prolong ESR, thereafter making ESR of questionable relevance in determining disease activity.

This patient's symptoms include mucocutaneous changes, conjuctival injection, 5 days of fever with elevated ESR and C-reactive protein, white blood cell, and platelet levels, and liver transaminases that are consistent with KD. The physical findings in this case are not consistent with the other diagnostic choices. Herpes gingivostomatitis can cause a moderate leukocytosis, and a disseminated herpes infection can cause elevated liver enzymes, thrombocytopenia, and coagulation abnormalities. Pharyngoconjuctival fever can present with conjuctivitis and elevated acute-phase reactants but would not present with mucocutaneous changes and would not likely affect the liver enzymes. Stevens—Johnson syndrome presents with nonspecific laboratory abnormalities that include elevated ESR, leukocytosis, elevated liver transaminases, and decreased albumin. Toxic shock syndrome presents with persistent fever, increased white blood cell count, and increased acute phase reactants.

PREP Pearls

- Kawasaki disease is a clinical diagnosis; no laboratory test is diagnostic.
- Laboratory abnormalities in Kawasaki disease include leukocytosis, thrombocytosis, elevated acute phase reactants, elevated liver transaminases, and sterile pyuria.
- Several conditions, including specific infections, can present similarly to Kawasaki disease.

American Board of Pediatrics Content Specification(s):

- Know the laboratory abnormalities seen in Kawasaki disease

Suggested Reading:

- Fimbres A, Shulman S. Kawasaki disease. Pediatr Rev. 2008;29(9):308-315. doi:10.1542/pir.29-9-308 Weiss P. Pediatric vasculitis. Pediatr Clin N Am. 2012;59(2):407-423. doi:10.1016/j.pc1.2012.03.013

Item 133

A 17-year-old girl is brought to your office by her mother because of insomnia and mood problems. For the past month, she has had difficulty falling asleep (getting no more than 4-5 hours of sleep each night) and has watched television in her room while awake. She has decreased energy, poor concentration, sad mood, and less frequent contact with her friends. She no longer wants to play on the volleyball team. She denies having made any current or previous plans to hurt herself but has had occasions when she wished she were dead. There have been no acute stressors. Two years ago, she was briefly hospitalized in a psychiatric unit for manic behaviors, but she recovered quickly. She has received care from a psychiatrist who prescribed an antipsychotic and counseling from another therapist. However, 6 months ago she stopped seeing these providers and discontinued her medication because she was feeling fine. Her mother has made an appointment for the girl to meet with her therapist next week, but because of limited access, it will be at least 2 months until she can see the psychiatrist.

Of the following, the BEST next step in management is to

A. prescribe fluoxetine and arrange to see her back in 1 week
B. prescribe risperidone, and arrange to see her back in 1 week
C. prescribe valproic acid, and arrange to see her back in 1 week
D. recommend sleep hygiene improvements and arrange to see her back in 1 week
E. send her to the emergency department to be evaluated for a psychiatric hospitalization

Item 133 Preferred Response: D

The patient described in the vignette is experiencing a current episode of major depression and has a history of bipolar disorder type 1 (she was once hospitalized because of mania). Among the options listed, helping her to improve her sleep habits would be the most appropriate next step. For patients with poor sleep, the use of stimulating electronic devices overnight (text messaging, watching television, and playing video games) will actually worsen and prolong the sleep problem. If this patient were to stop watching TV late at night, along with implementing other good sleep habits, it would likely reduce or resolve the insomnia. Eliminating insomnia will help resolve her depression and reduce the likelihood of her switching back to having a manic episode. Periods of insomnia often precede and trigger manic episodes in patients who have bipolar disorder.

Fluoxetine is a first-line medication for treating adolescent depression, but it should be avoided in the presence of bipolar disorder. Selective serotonin reuptake inhibitors (SSRIs) have been found to psychiatrically destabilize many patients who have bipolar disorder and they increase the likelihood of a manic switch. Risperidone has been found to be very helpful in reducing manic symptoms in adolescent patients who have bipolar disorder, but it has not been found to treat bipolar depression. Patients already taking an antimanic medication like risperidone are sometimes offered antidepressant medications like SSRIs with the idea that the antimanic agent will prevent manic switching, but this strategy should generally only be prescribed by mental health specialists. Valproic acid is a moderately beneficial antimanic agent in adolescents, but it lacks antidepressant properties. Although both valproic acid and risperidone have sedation as an adverse effect, prescribing them for use at bedtime as a sleep aide in patients' bipolar disorder is not indicated before implementing good sleep habits. There is a good chance that improved sleep hygiene will solve her insomnia, and thus it is not currently advisable to expose her to the side effects of valproic acid or risperidone. Psychiatric hospitalization might be appropriate for depressed patients with active thoughts of committing suicide or for patients with bipolar disorder currently in a state of mania. However, the girl in the vignette does not have any signs of requiring a psychiatric hospitalization at this time.

The best medication treatment for bipolar depression in adolescents is still unclear. Many specialists will prescribe lithium for this indication, some will prescribe lamotrigine and another mood stabilizer, and as noted before, some will prescribe SSRIs for a limited time while another mood stabilizing agent is simultaneously used.

PREP Pearls

- Avoid giving SSRIs to patients who have bipolar disorder.
- Ensure good sleep occurs as a key part of bipolar depression treatment.
- Ensure good sleep hygiene habits are used as the first-line intervention for insomnia.

AAP Mental Health Competency:

- Know the evidence based treatments of bipolar disorder in children

Suggested Reading:

- McClellan J, Kowatch R, Findling RL; Work Group on Quality Issues. Practice parameter for the assessment and treatment of children and adolescents with bipolar disorder. J Am Acad Child Adolesc Psychiatry. 2007;46:107-125. doi:10.1097/O1.chi.0000242240.69678.c4
- Mindell J, Owens J. A Clinical Guide To Pediatric Sleep: Diagnosis and Management of Sleep Problems. 2nd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2009

Item 134

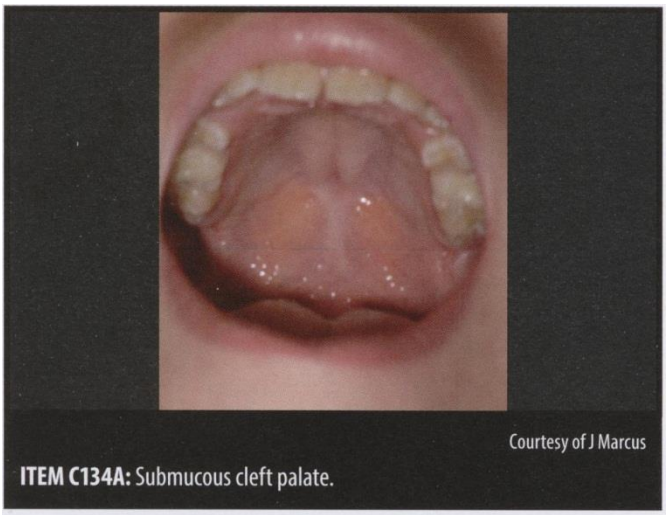
A mother well known to your practice brings her five-day-old infant in for assessment of jitters. The mother is a reliable historian and relates that the infant was born at term weighing 3,000 grams after an uncomplicated pregnancy and discharged home 2 days after delivery. The infant has been breastfeeding exclusively, with 6 wet diapers and 2 stools in the past twenty-four hours. The jitters started eight hours ago and are worsening. Currently the infant weights 2,800 grams. Physical examination reveals an irritable infant with subtle jitters and tremors of all extremities, slightly low set ears, wide-set eyes, and a bifid uvula.

Of the following, the screening study MOST likely to reveal the underlying cause of the infant's jitters is a (n)

A. blood glucose level
B. CT scan of the head
C. electroencephalogram
D. serum calcium
E. urine toxicology screening

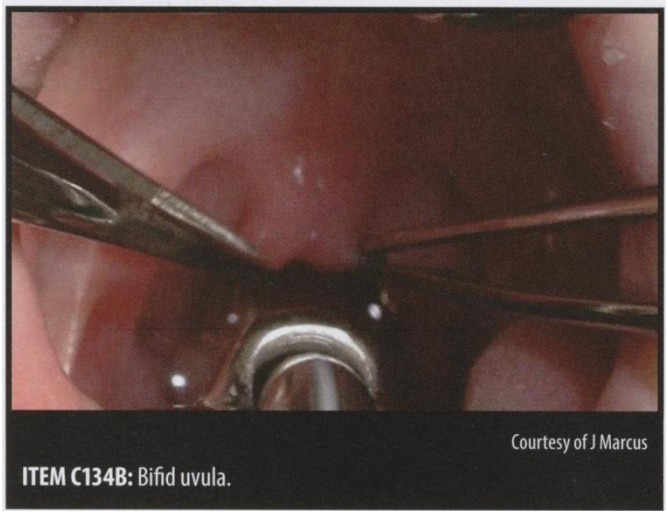
Item 134 **Preferred Response: D**

The infant described in the vignette has the physical stigmata of DiGeorge syndrome (DGS) and is most likely to have jitters from hypocalcemia. DiGeorge syndrome or anomaly is caused by abnormal development of the third and fourth pharyngeal pouches leading to conotruncal or cardiac abnormalities, abnormal facies, thymic hypoplasia, cleft palate, and hypocalcemia (CATCH). The hypocalcemia is the result of developmental defects in the parathyroid glands. DiGeorge syndrome is associated with a number of causes, with more than 80% associated with the 22q11 deletion (CATCH 22). It also has great variability in its expressivity. A submucosal cleft palate (Item C134A) or bifid uvula (Item C134B) should prompt a clinician to consider further investigation of DGS or the closely related velocardiofacial syndrome (VCFS). Mildly affected individuals with DGS or VCFS may be missed in the neonatal period and present as an older child with nasal speech, learning disabilities, or psychiatric illness.



Courtesy of J Marcus

ITEM C134A: Submucous cleft palate.



Courtesy of J Marcus

ITEM C134B: Bifid uvula.

Neonatal hypocalcemia is broken down by the timing of onset into early (birth to 4 days) and late (5 to 10 days). Etiologic factors in early neonatal hypocalcemia include prematurity, maternal diabetes, perinatal asphyxia, and intrauterine growth retardation. Late neonatal hypocalcemia may be seen with hyperphosphatemia, hypomagnesemia, hypoparathyroidism, maternal vitamin D deficiency, and infantile osteopetrosis. The hypocalcemia associated with DGS is late-onset, presenting with jitters, tetany, or seizures. The underlying cause is absent or hypoplastic parathyroid glands leading to hypoparathyroidism. Hypoglycemia must be considered in any infant with the jitters, and typically presents within the first 24 hours of birth. Infants who are small for gestational age and/or feeding poorly may exhibit issues related to hypoglycemia after that period. Although blood glucose screening should be performed, it is unlikely to reveal hypoglycemia in the infant in the vignette because of the lack of evidence of dehydration or poor feeding. Central nervous system (CNS) abnormalities that are either congenital or traumatic in origin may manifest with irritability and jitters. Jitters and tremors may also reflect seizure activity, which may be

due to an underlying CNS disorder or occur in isolation. In this infant with such classic features of DGS, it is unlikely that either electroencephalography or computed tomography scan of the head would reveal the underlying cause of the jitters. Neonatal withdrawal syndrome may manifest up to 1 week after birth and should also be considered in any infant presenting with jitters. Urine toxicology screening is unlikely to contribute to the diagnosis because this infant has no history of prenatal drug exposure.

PREP Pearls

- Early hypocalcemia (birth to 4 days) in the neonate is associated with prematurity, maternal diabetes, perinatal asphyxia, and intrauterine growth retardation.
- Late neonatal hypocalcemia (5 to 10 days) may be seen with hyperphosphatemia, hypomagnesaemia, hypoparathyroidism, maternal vitamin D deficiency, and infantile osteopetrosis.
- The underlying diagnosis of DiGeorge syndrome or velocardiofacial syndrome should be considered in infants with a submucous cleft palate or a bifid uvula.

American Board of Pediatrics Content Specification(s):

- Know the causes of hypocalcemia in a neonate

Suggested Reading:

- Abrams SA. Neonatal hypocalcemia. UptoDate. Available online only for subscription
- Robin NH, Shprintzen RJ. Defining the clinical spectrum of deletion 22q11.2. J Pediatr. 2005;147:90-96. doi:10.1016/j.jpeds.2005.03.007
- Thomas TC, Smith JM, White PC, Adhikari S. Transient neonatal hypocalcemia: presentation and outcomes. Pediatrics. 2012;129:e1461-e1467. doi:10.1542/peds.2011-2659.
- Zhou P, Markowitz M. Hypocalcemia in infants and children. Pediatr Rev. 2009;30:190-192. doi:10.1542/pir.30-5-190

Item 135

A 2-year-old girl with no significant past medical history is brought to your office for pallor. The mother reports that the child has had fever and upper respiratory symptoms for 5 days, which are now improving. Her axillary temperature is 36.8°C, pulse rate is 140 beats/min, respiratory rate is 24 breaths/min, and blood pressure is 100/60 mm Hg. On examination, the child is alert and in no apparent distress. She has anicteric sclerae and no hepatosplenomegaly or lymphadenopathy. The remainder of the physical examination is normal. The results of the child's laboratory tests are shown:

- White blood cell count, 5,500/µL (5.5×10^9/L), with 30% polymorphonuclear leukocytes, 64% lymphocytes, 5% monocytes, and 1% eosinophils
- Hemoglobin, 4.5 g/dL (45 g/L)
- Mean corpuscular volume, 80/µm3 (80 fL)
- Platelet count, 255×10^3/µL (255×10^9/L)
- Reticulocyte count, 1.5% (0.015)

The child is transfused with packed red blood cells.

Of the following, the MOST appropriate next step in management would be to

A. determine the human leukocyte antigen types of family members for bone marrow transplantation
B. follow complete blood cell count and reticulocyte count weekly
C. obtain bone marrow studies
D. order hemoglobin electrophoresis
E. order iron studies

Item 135 Preferred Response: B

The girl described in the vignette has a presentation consistent with transient erythroblastopenia of childhood (TEC). Disorders of decreased erythrocyte production, known as pure red cell aplasia (PRCA), usually present with reticulocytopenia and a macrocytic, normochromic anemia. These can be divided into congenital and acquired disorders. Diamond-Blackfan anemia (DBA) is a congenital PRCA that presents in infancy or early childhood with progressive macrocytic anemia, markedly decreased or absent erythroid precursors in the bone marrow, congenital anomalies (in 50% of patients), and increased risk of malignant tumors. In familial cases, it appears to follow an autosomal dominant inheritance pattern, although there can be variability in the severity of the disease within the same family. The diagnostic criteria for classic DBA are as follows:

- Age younger than 1 year
- Macrocytic anemia with no other cytopenias
- Reticulocytopenia
- Bone marrow with decreased erythroid precursors but normal marrow cellularity

Diamond-Blackfan anemia is caused by genetic mutations in ribosome synthesis. Mutations in the gene encoding ribosomal protein 19 (RPS19) are found in 25% of patients. Mutations in genes encoding the large ribosomal subunit (eg, RPL11) and the small ribosomal subunit (eg, RPS24) have also been reported. Mutations in RPL5 or RPL11 have been associated with somatic mutations, such as cleft palate. Because of the high incidence of cardiac and renal anomalies, cardiac and renal imaging is recommended in the evaluation of DBA patients. Laboratory features that distinguish DBA from TEC include elevated hemoglobin F concentration at diagnosis, elevated erythrocyte adenosine deaminase (eADA) activity, and the presence of the i RBC antigen (Item C135). Patients with DBA are treated with corticosteroids and red blood cell transfusions. Approximately 40% of patients are dependent on steroid therapy, 40% are dependent on blood transfusions, and 20% undergo remission by 25 years of age. Patients who are refractory to steroids may require hematopoietic stem cell transplantation. Acquired PRCA can occur from infections, drugs, and autoimmune diseases, but the most common cause in children is TEC. Although the cause of TEC is not entirely clear, in half of the cases, there is a history of a preceding viral illness within 2 to 3 months before presentation. TEC is self-limited and occurs in otherwise healthy, normal children. The anemia seen in TEC is slightly less severe, and there may be mild abnormalities in the other cell lines. In a child with clinical features and laboratory data suggestive of TEC (Item C135), bone marrow examination may not be necessary. TEC usually resolves spontaneously in 1 to 2 months, although some patients may require packed red blood cell transfusions if the anemia is severe enough to cause cardiorespiratory compromise.

The child described in the vignette who was previously healthy and presents with anemia at 2 years old is more likely to have TEC than DBA. In the absence of other cytopenias or symptoms concerning for a bone marrow disorder, a bone marrow examination would not be indicated. Because DBA is less likely in this patient, checking the eADA level, ordering a RPS19 gene analysis, and preparing for bone marrow transplantation would

not be warranted. For a patient with TEC, the appropriate management would be to follow the complete blood cell count and reticulocyte count closely (eg, weekly) until the condition resolves over the next several months. As the MCV is in the normal range, iron deficiency is unlikely to be the cause of the severe anemia seen in the child described in this vignette.

Item C135. Clinical and Laboratory Findings in Diamond-Blackfan Anemia and Transient Erythroblastopenia of Childhood

	Diamond–Blackfan Anemia	Transient Erythroblastopenia of Childhood
Age at diagnosis	<1 year in 90% cases	1–4 years in 80% cases
Congenital anomalies	35% have at least one anomaly	Absent
White blood cell count	Normal	Normal
Platelet count	Normal	Normal
Hemoglobin at diagnosis	2–6 g/dL (20-60 g/L)	3–9 g/dL (30-90 g/L)
Mean corpuscular volume at diagnosis	Increased in 30% cases	Normal
Mean corpuscular volume during recovery	Increased in 100% cases	Increased
Mean corpuscular volume during remission	Increased in 100% cases	Normal
Hemoglobin F at diagnosis	Increased in 100% cases	Normal
Hemoglobin F during recovery	Increased in 100% cases	Increased
Hemoglobin F during remission	Increased in 85% cases	Normal
Erythrocyte adenosine deaminase activity	Elevated	Normal
i RBC antigen	Present	Absent

PREP Pearls

- The diagnostic criteria for classic DBA are as follows:
- Age younger than 1 year
- Macrocytic anemia with no other cytopenias
- Reticulocytopenia
- Bone marrow with decreased erythroid precursors but normal marrow cellularity
- TEC can be distinguished from DBA in most cases by clinical and laboratory features.
- TEC is self-limited and occurs in otherwise healthy children, often after a viral illness.

American Board of Pediatrics Content Specification(s):

- Distinguish between the clinical characteristics of Diamond-Blackfan syndrome and transient erythroblastopenia of childhood

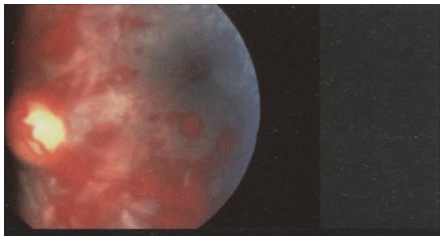
Suggested Reading:

- Sandoval C. Anemia in children due to decreased red blood cell production. UptoDate. Available online only for subscription Shaw J, Meeder R. Transient

erythroblastopenia of childhood in siblings: case report and review of the literature. J Pediatr Hematol Oncol. 2007;29:659-660. doi:10.1097/MPH.0b013e31814684e9 Vlachos A, Muir E. How I treat Diamond-Blackfan anemia. Blood. 2010;116:3715-3723. doi:10.1182/blood-2010-02-251090

Item 136

A 9-month-old boy has a generalized tonic–clonic seizure. He has not had a fever and his parents report no trauma. The boy was born at 26 weeks' estimated gestational age and spent 4 months in a neonatal intensive care unit. He had intraventricular hemorrhages and now is blind. He has spastic quadriparetic cerebral palsy but has had no seizures until now. On physical examination, his temperature is 37.2°C, blood pressure is 92/66 mm Hg, heart rate is 98 beats/min, and respiratory rate is 24 breaths/min. He has constant, conjugate roving eye movements. You attempt a funduscopic examination and see glimpses of his retina as shown in Item Q136. His general examination results are normal and his neurologic examination is notable for spastic quadriparesis. Computed tomography of the head without contrast reveals a subdural hematoma.



ITEM Q136: Funduscopic findings as described for the infant in the vignette.

Of the following, the MOST likely cause of the subdural hematoma is

A. abusive head trauma
B. arteriovenous malformation
C. brain atrophy from prematurity
D. new onset seizure
E. prior intraventricular hemorrhage

Item 136 S TE Preferred Response: A

The most likely cause of a subdural hematoma in an infant with retinal hemorrhages is abusive head trauma. Although his history of prematurity puts the infant in the vignette at risk of developing seizures, the presence of the subdural hematoma with retinal hemorrhages makes the subdural hematoma the most likely cause of the seizure.

For the infant in this vignette, young age and history of disability are risk factors for abusive head trauma. Additional risk factors such as intimate partner violence and poverty may be elicited on history. A detailed physical examination, including an ophthalmologic examination may reveal retinal hemorrhages, bruising, marks, or injuries suggestive of abuse. In addition to further evaluation, this infant needs close clinical monitoring for sequelae of intracranial injury. The absence of papilledema on ophthalmologic examination does not rule out increased intracranial pressure, especially early in the clinical course. Neurosurgical consultation would be helpful.

Arteriovenous malformations, when they rupture, typically cause intraparenchymal hemorrhage, not subdural hematomas. Brain atrophy from prematurity can predispose an infant to the development of a subdural hematoma, but this would not explain the retinal hemorrhages. Seizures alone do not usually cause subdural hematomas. Intraventricular hemorrhage secondary to prematurity can take months to years to resolve radiographically; however, the imaging finding is of residual blood products in the ventricles, not subdural hematoma.

PREP Pearls

- It is more likely for subdural hematomas to cause seizures, than for seizures to cause subdural hematomas.
- New-onset seizure in an infant can be the first sign of abusive head trauma.
- Examination of the retina is an important part of the evaluation in any child with seizures.

American Board of Pediatrics Content Specification(s):

- Understand the value of retinal examination in a patient with seizures or in a coma
- Know that papilledema may not be present initially and may develop later in the course of intracranial hypertension

Suggested Reading:

- Dubowitz H, Lane WG. Abused and neglected children In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:135-147
- Tung GA, Kumar M, Richardson RC, Jenny C, Brown WD. Comparison of accidental and nonaccidental traumatic head injury in children on noncontrast computed tomography. Pediatrics. 2006;118(2):626-633. doi: 10.1542/peds.2006-0130.

Item 137

An 11-year-old boy is seen for a routine health supervision visit. He plays basketball for his junior high school and is on a travel team. He would like to begin lifting weights to improve his performance during the basketball season. His past medical history is remarkable for mild asthma. You do not note any abnormalities on physical examination. He is at sexual maturity rating 2.

Of the following, the MOST appropriate statement to include in your discussions with this family is

A. children younger than 14 years old do not have the balance and coordination required for weight training
B. he can begin a program using heavy resistance but should only perform 3 to 5 repetitions for each muscle group
C. he should begin strength training by performing exercises with no little or no load
D. lifting weights in skeletally immature patients is contraindicated due to the risk of damage to the physes
E. strength training is likely to lead to a significant increase in muscle hypertrophy at his age

Item 137 S Preferred Response: C

Parents often ask pediatricians whether strength training is appropriate for children and adolescents. Well-designed resistance training programs can be both safe and beneficial for young athletes. Before initiating a strength-training program, young athletes should have a medical evaluation performed by a primary care physician. This allows the physician to determine whether there are any conditions that warrant additional evaluation before engaging in resistance training, such as severe hypertension or history of heart disease, and affords an opportunity to discuss strategies for preventing injury.

Most injuries that occur as a result of resistance training are associated with inadequate supervision or poor technique. Children should begin a training program by learning proper form using little or no weight. Once children master the technique for an exercise and can perform 8 to 15 repetitions with good form, they can begin to gradually increase load. An analysis of "weightlifting" injuries presenting to emergency departments in the United States demonstrated that most injuries resulted from accidents while handling the weights rather than from the actual strength training. To minimize the risks of injury, children and adolescents should work with coaches or personal trainers who have certification in pediatric strength training.

Children can begin resistance training during middle childhood, potentially as early as 6 to 8 years of age, as long as they have the balance and posture necessary to perform exercises with good form. While cases of growth plate injury due to weightlifting have been reported in the medical literature, these incidents have generally occurred in the setting of inadequate supervision or poor technique. In preadolescents, participation in strength-training programs will lead to improved strength by facilitating the recruitment of motor neurons but will not result in muscle hypertrophy.

PREP Pearls

- Most strength-training injuries in children and adolescents occur as a result of improper form or inadequate supervision.
- Children as young as 6 to 8 years old can benefit from a resistance training program.
- Children should begin strength training by learning proper form with little or no weight.

American Board of Pediatrics Content Specification(s):

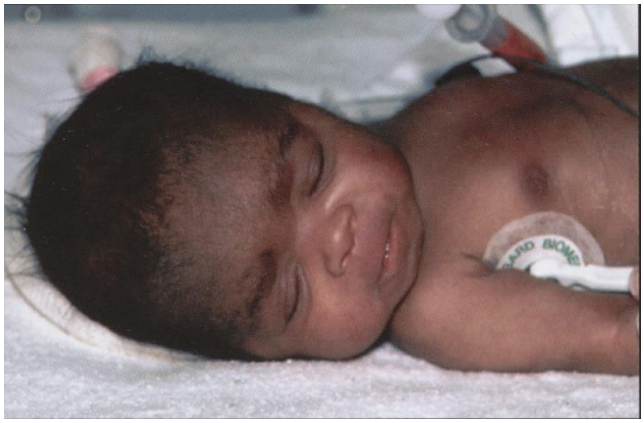
- Identify risks in a conditioning program for junior high school athletes at the beginning of the sports season

Suggested Reading

- American Academy of Pediatrics Council on Sports Medicine and Fitness; McCambridge TM, Stricker PR. Strength training by children and adolescents. Pediatrics. 2008;121(4):835-840. doi:10.1542/peds.2007-3790
- Faigenbaum AD, Myer GD. Resistance training among young
- athletes: safety, efficacy, and injury prevention effects. Br I Sports Med. 2010;44(1):56-63. doi:10.1136/bjsm.2009.068098
- Myer GD, Quatman CE, Khoury J, Wall EL Hewett TE. Youth versus adult "weightlifting" injuries presenting to United States emergency rooms: accidental versus nonaccidental injury mechanisms. I Strength Cond Research. 2009;23(7):2054-2060. doi:10.1519/JSC.0b013e3181b86712

Item 138

You are called in to see a female neonate born at term to a 34-year-old gravida 2, para 2 woman following a reportedly uneventful pregnancy, although the mother had only 2 prenatal visits at 30 and 36 weeks' gestation. The neonate's birth weight is 2.3 kg (below the 5th percentile), and her Apgar scores were 7 and 9 at one and five minutes, respectively. On examination, her head circumference is at the fifth percentile. Facial features are seen in Item Q138. Examination is also remarkable for bilateral radioulnar synostosis, small distal phalanges, fifth-fingernail hypoplasia, and a soft systolic cardiac murmur. Echocardiogram demonstrates a small muscular ventricular septal defect.



ITEM Q138: Image of the neonate described in the vignette.

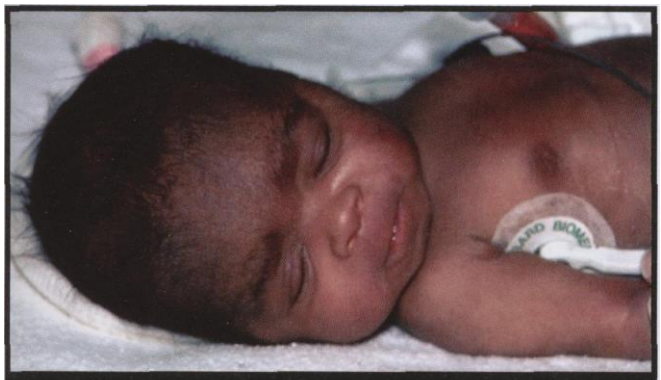
Of the following, the MOST likely diagnosis for this infant is

A. Angelman syndrome
B. fetal alcohol syndrome
C. trisomy 21
D. velocardiofacial syndrome
E. Williams syndrome

Item 138 Preferred Response: B

The infant described in the vignette has clinical features suggestive of fetal alcohol syndrome (FAS). In addition to the classic facial features of short palpebrae, long smooth philtrum, and thin upper lip (Item C138), she also has radioulnar synostosis and a ventricular septal defect (VSD), which are consistent with the suspected diagnosis. Because these clinical features may be seen in children with no prenatal exposure to alcohol, in order to confirm this diagnosis, an accurate and honest prenatal history regarding maternal alcohol consumption needs to be obtained.

Other common features of FAS include a "hockey stick" upper palmar crease, "railroad track" upper helix of the ear, ptosis, strabismus, hypoplastic nails, short fifth digits, fifth-finger clinodactyly, and camptodactyly. Relatively common birth defects include cardiac defects, radioulnar synostosis, vertebral segmentation defects, renal anomalies, optic nerve hypoplasia, hearing loss, and pectus deformities. Most children with FAS will have prenatal or postnatal growth retardation and ultimately will develop microcephaly (with head circumference <10th percentile for age). Developmental delays or cognitive deficits will be noted over time. Children with FAS commonly demonstrate marked impairment in carrying out complex tasks, higher-level receptive and expressive language delays, and behavioral difficulties.



Courtesy of M Rimsza

ITEM C138: A smooth philtrum, narrow upper lip, and hirsutism may be observed in infants who have fetal alcohol syndrome.

Newborns with Angelman syndrome or Williams syndrome may not exhibit any abnormalities at birth but may have mild prenatal growth deficiency. Both of these conditions have facial and clinical features that evolve over time, which makes an early diagnosis less likely. Children with Angelman syndrome typically have postnatal development of microcephaly and onset of seizures, with very limited expressive language and later appearance of prognathism. Radioulnar synostosis is not seen in any of the other syndromes listed, whereas VSDs may be seen with some frequency in children with Down syndrome and velocardiofacial syndrome. Children with Williams syndrome are much more likely to have supravalvular aortic stenosis, although VSDs may also occur. Facial features associated with Williams syndrome that become more apparent over time include medial eyebrow flare, epicanthal folds, periorbital fullness, stellate iris pattern, anteverted nares, and full lips. Individuals with velocardiofacial syndrome have a high risk for cleft palate and abnormalities of the great vessels and, over time, may be

noted to have a broad nasal root, long face, postnatal microcephaly, and slender fingers. The typical facial features in children with Down syndrome include a flat facial profile, epicanthal folds, upslanting palpebrae, Brushfield spots on the iris, small ears, and a protruding tongue.

PREP Pearls

- Infants with fetal alcohol syndrome have typical facial features at birth and are at increased risk for cardiac defects (especially ventricular septal defects) as well as limb anomalies such as radioulnar synostosis.

American Board of Pediatrics Content Specification(s):

- Recognize the physical features of fetal alcohol syndrome, and manage appropriately

Suggested Reading:

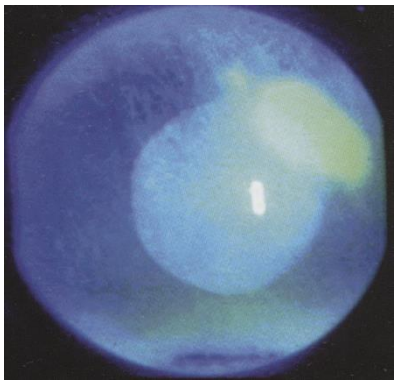
- Astley SJ. Comparison of the 4-Digit Diagnostic Code and the Hoyme Diagnostic Guidelines for Fetal Alcohol Spectrum Disorders. Pediatrics. 2006;118(4):1532-1545. doi:10.1542/peds.2006-0577
- Gahagan S, Sharpe TT, Brimacombe M, et al. Pediatricians' Knowledge, Training, and Experience in the Care of Children With Fetal Alcohol Syndrome. Pediatrics. 2006;118(3):e657-e668. doi:10.1542/peds.2005-0516
- Godel J; Canadian Paediatric Society. Canadian Paediatric Society statement: fetal alcohol syndrome. Paediatr Child Health. 2002;7(3):161-174
- Hoyme HE, May PA, Kalberg WO, et al. A practical clinical approach to diagnosis of fetal alcohol spectrum disorders: clarification of the 1996 Institute of Medicine criteria. Pediatrics. 2005;115(1):39-47. doi:10.1542/ peds.2004-0259
- Landgren M, Svensson L, Stromland K, Gronlund MA. Prenatal alcohol exposure and neurodevelopmental disorders in children adopted from Eastern Europe. Pediatrics. 2010;125(5):e1178-e1175. doi:10.1542/ peds.2009-0712

Item 139

The mother of a healthy 11-month-old girl brings her to your office for evaluation because of "nonstop crying" for the past few hours. She has had no recent fever or other signs of illness. She began crying abruptly 3 hours ago while her mother was taking her for a walk in the stroller to "enjoy the nice morning breeze:' During the walk, the girl began crying and rubbing her eyes and has continued crying inconsolably since that time. The girl is nontoxic appearing, but she is crying loudly and cannot be consoled by her mother. She is afebrile. You find no abnormalities on physical examination other than injection of both conjunctivae and mild erythema of the right upper eyelid without swelling. The girl is producing profuse clear tears from both eyes. You perform fluorescein staining of both eyes and view them with a Wood lamp. Examination of the left eye is unremarkable. A photograph of the child's right eye is shown (Item Q139). No foreign bodies are seen on examination, including eversion of both upper eyelids. You administer a dose of oral acetaminophen to help relieve the girl's discomfort.

Of the following, the MOST appropriate home management in addition to acetaminophen for discomfort is

A. application of a pressure patch to the affected eye for at least 48 hours
B. application of tetracaine ophthalmic drops to the affected eye as needed for pain
C. application of topical antibiotic ointment to the affected eye
D. application of topical corticosteroid drops to the affected eye
E. follow-up examination by an ophthalmologist within the next 24 hours



ITEM Q139: Wood lamp examination for the girl described in the vignette.

Item 139 Preferred Response: C

The girl described in the vignette has a corneal abrasion. The most appropriate home management plan is application of a topical antibiotic ointment to the affected eye.

Corneal abrasions occur commonly in children of all ages. Children may incur corneal abrasions unintentionally during play or from contact of a projectile with the cornea. Children who use contact lenses may develop abrasions due to damaged or poorly fitting lenses, as well as from wearing lenses overnight.

The cornea is one of the most richly innervated tissues of the body. Pain from a corneal abrasion may range from minimal to severe, depending on the size of the defect. Symptoms may include eye discomfort (which generally begins acutely), photophobia, blurry vision, foreign-body sensation, and watery eye discharge. Children who cannot verbalize the symptom of eye pain may present because of nonspecific, persistent irritability. Eyelid edema, tearing, conjunctival injection, and refusal to open the affected eye are common associated signs.

Application of a drop of a topical anesthetic, such as 0.5% tetracaine solution, may have both diagnostic and transient therapeutic value for children with suspected corneal abrasions. Children who become more comfortable after the application of anesthetic drops most likely have an injury to the ocular surface (conjunctiva or cornea) that is causing their pain. The analgesia provided by topical anesthetics often facilitates the eye examination, making it much more comfortable for affected patients to open their eyes and cooperate with examination.

Although larger corneal abrasions may be visible with the naked eye, fluorescein dye is needed in many cases to identify and define the size of abrasions. When used, fluorescein dye is instilled into the affected eye, and a Wood or cobalt blue lamp is subsequently used to illuminate it. The dye will stain the exposed corneal stroma but not the intact corneal epithelium. Corneal abrasions will stain a vibrant yellow color when present. Whenever corneal abrasions are found, the conjunctival culdesac and upper tarsus should be examined to rule out the presence of retained ocular foreign bodies, which can continue to abrade the cornea. This is facilitated through eversion of the upper eyelid over a cotton-tip applicator or fingertip.

Uncomplicated corneal abrasions heal rapidly, often within 24 to 72 hours, regardless of therapy. Topical antibiotics are generally recommended in affected children to prevent superinfection. Ointments are preferred over drops because of their persistent lubricating effect and because they typically cause less stinging. Aminoglycosides should be avoided, except in cases of contact lens-related abrasions because they are generally more toxic to the corneal epithelium and may delay healing.

Several controlled studies have demonstrated no benefit from pressure patching in speeding the rate of healing or increasing comfort in patients with corneal abrasions. Furthermore, children are often irritated by the placement of pressure patches and attempt to pull them off. It is generally recommended that corneal abrasions in children be treated

without eye patching because of the lack of proven benefit, the possibility of improper patching, and the potential for patient discomfort and inconvenience. Exceptions to this rule include children with large abrasions occupying more than half of the corneal surface, since pressure patching may result in greater pain relief.

Although topical anesthetics, such as 0.5% tetracaine solution, can be useful in temporarily relieving pain in children presenting with corneal abrasions in the acute care setting, home use of these agents is not recommended based on current evidence that suggests that topical anesthetics can interfere with corneal epithelial healing, especially with repeated applications. In addition, inappropriate use of these agents at home by patients or caregivers for persistent eye pain creates a risk of corneal ulceration, perforation, scarring, and even vision loss. Systemic analgesics, including acetaminophen and opioids (depending on the degree of the child's discomfort), can help to relieve the pain and distress that result from a corneal abrasion. Nonsteroidal antiinflammatory ophthalmic solutions, such as ketorolac and diclofenac, may also provide some pain relief without adversely affecting healing

Topical corticosteroid preparations are contraindicated for patients with corneal abrasions. These preparations can delay healing of the epithelium and increase the risk of superinfection.

Ophthalmologic follow-up is not required for small corneal abrasions that heal as expected during a 48- to 72-hour period. Corneal abrasions that are particularly large and those involving the visual axis should be seen within 24 hours after the injury. Children with persistent pain or foreign-body sensation for more than 2 to 3 days after sustaining a corneal abrasion, as well as those with increasing pain or eye redness, should be referred for ophthalmologic care. In addition, any child who wears contact lenses or who has a history of ocular herpes should be referred urgently for ophthalmology consultation. Children with corneal abrasions who wear contact lenses should be advised to remove their lenses immediately and leave them out until they are cleared by an ophthalmologist to resume use.

PREP Pearls

- Symptoms of a corneal abrasion may include eye discomfort, photophobia, blurry vision, foreign-body sensation, and watery eye discharge. Nonverbal children may present with nonspecific, persistent irritability.
- Topical corticosteroid preparations are contraindicated for patients with corneal abrasions.
- Ophthalmologic follow-up is not required for small corneal abrasions that heal as expected during a 48- to 72-hour period. Corneal abrasions that are particularly large, involve the visual axis, or occur in a child who uses contact lenses should be evaluated by an ophthalmologist within 24 hours after the injury.

American Board of Pediatrics Content Specification(s):

- Recognize the clinical presentations of corneal abrasion, including irritability in young infants
- Know the appropriate management and follow-up evaluation of corneal abrasions

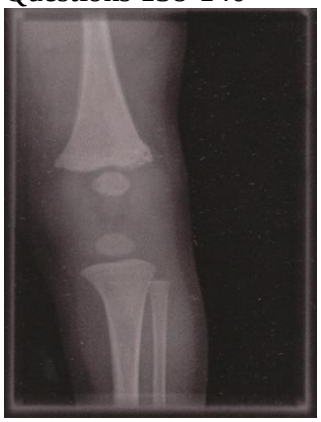
Suggested Reading:

- Jacobs DS. Corneal abrasions and corneal foreign bodies. UptoDate. Available online only for subscription
- Levin AV. Eye trauma. In: Fleisher GR, Ludwig S, eds. Textbook of Pediatric Emergency Medicine. 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2010:1448-1458
- Sands Braverman R. Eye. In: Hay WW, Levin MJ, Sondheimer JM, Deterding RR, eds. Current Diagnosis and Treatment: Pediatrics. 20th ed. New York, NY: McGraw-Hill; 2011
- Stout AU. Corneal abrasions. Pediatr Rev. 2006;27:433-434. doi:10.1542/pir.27-11-433

Item 140

A 4-month-old boy is brought to the emergency department by his parents for evaluation of fussiness. For the past 2 days he has been more irritable than usual. The mother noticed some swelling of the right thigh that she attributed to an insect bite, but he has been otherwise well. He is afebrile and has been feeding normally. He was born by elective Cesarean section for twin gestation at 37 weeks estimated gestational age. He has been growing and developing appropriately, but he is thinner than his twin. On physical examination, you note that he cries when you lie him down and when you remove the diaper. You order radiographs of the lower extremities (Item Q140, page Q-36).

Questions 138-140



ITEM Q140: Radiograph for the child in the vignette.

Of the following, the MOST appropriate next step in this infant's management is

A. evaluation for nonaccidental trauma
B. evaluation for septic arthritis
C. initiation of vitamin D supplementation
D. referral to genetics
E. repeating the newborn screen

Item 140 S I-C TE SBP Preferred Response: A

Many children who have fractures from inflicted trauma come to medical attention because of vague symptoms, such as those seen in the infant boy in the vignette. Therefore, careful questioning regarding any falls or injuries, observation of the parent-child interaction, and a complete physical examination that includes the skin are important. Infants may present with irritability and limitation of spontaneous movement of the injured limb. As in this case, careful palpation of the entire limb may help locate the point of maximal tenderness and direct the plan for radiographs. The radiographic finding of a metaphyseal "chip" fracture should prompt further evaluation for intentional trauma immediately. The fracture typically occurs when the extremity is pulled or twisted forcibly.

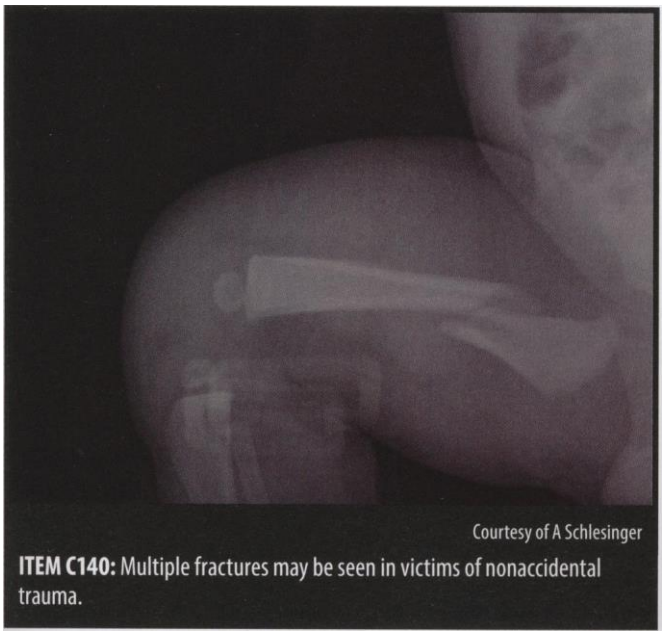
Most fractures inflicted by intentional trauma occur during infancy and early childhood, with most occurring in children younger than 1 year of age. Among patients with a single fracture, the femur is the most common location, followed by the humerus and the skull. Approximately half of patients will have more than one fracture, as seen in this child (Item C140).

The medical evaluation of suspected child abuse begins with a detailed account of the mechanism of injury from the parents or caregivers. The evaluator should ask about the initial position of the child, the dynamics of the fall, and the final position of the child. Aspects of the history that increase the level of suspicion for inflicted injuries include the following: the history is inconsistent with the injury, the history is vague or the injury is not witnessed, the history varies in repeated versions or among caregivers, the injury is attributed to the actions of young siblings, the history is inconsistent with the developmental stage of the child, or the history is implausible.

On physical examination, the entire surface of the skin should be examined and any skin lesions (eg, burns, scars, and bruises) should be documented. Bruising may or may not be present at the site of fractures. The absence of bruising does not help differentiate inflicted trauma from an underlying bone disorder. Findings that raise suspicion of child abuse include the following: injuries that are not consistent with the history; different types of injuries coexisting or in various stages of healing; evidence of poor caretaking or under nutrition; certain patterns of injuries (bruising in a child that is not ambulating, cigarette burns, belt strap or hand prints, injury to the genitalia, or bruising to the pinna, neck, or abdomen). A detailed neurologic examination should be performed. In the young child, any sudden onset of altered mental status that is not attributable to medical illness should raise concern for abuse.

The behavior of the parents and the interaction among family members should be observed. Certain behaviors may increase the level of suspicion, such as arguing or violence, aloofness or lack of emotion, and delay in seeking medical care. Screening for maternal depression and family support is an important preventive measure that should be performed by all primary care physicians. Many family stressors, including multiple or premature births and infantile colic, predispose infants to abuse, as was found in the case of the boy in the vignette.

Orthogonal radiographs (anteroposterior and lateral) should be obtained of all areas of swelling, bone tenderness, deformity, limited range of motion, or history of trauma. Skeletal survey is the method of choice for global skeletal imaging in cases of suspected child abuse and is mandatory for all children younger than 2 years in whom intentional trauma is suspected. Skeletal survey is usually not necessary in children older than 5 years, but it should be considered in children with intellectual disabilities that preclude them from giving a history or indicating areas of trauma or pain. Although there are fracture patterns that are suggestive of inflicted injury, none are absolutely pathognomonic of abuse. Any fracture in an infant should raise concern for abuse. Other types of fractures that should alert the clinician include the following: metaphyseal corner, or "chip:' fractures; femoral fractures in a nonambulatory child; transverse fractures of long bones; rib fractures; multiple fractures; fractures in various stages of healing; skull fractures in a child younger than 18 months; and bilateral long bone fractures.



Courtesy of A Schlesinger

ITEM C140: Multiple fractures may be seen in victims of nonaccidental trauma.

When evaluating the child suspected of being physically abused, it is important to consider other conditions that may present with similar findings. Septic arthritis should be considered when a joint is swollen, erythematous, or warm to touch and the child appears ill. The boy in the vignette had none of these features.
Laboratory evaluation should be tailored to the findings on physical examination, the specific history of the injury, and family history. Radiographic findings of vitamin D deficiency rickets include low bone density, widening of the growth plate, and metaphyseal cupping. Osteogenesis imperfecta and metabolic bone disease should be considered in the differential diagnosis in infants who present with multiple fractures in various stages of healing; however, referral to genetics or repeating the newborn screen would occur after complete evaluation and treatment of the child's urgent medical needs. Consultation with a multidisciplinary child abuse team is recommended when available. In many parts of the world, suspected cases of child abuse must be reported to the appropriate government authorities.

PREP Pearls

- The metaphyseal chip, or corner fracture is highly suspicious for intentional trauma.

- Most fractures that occur as the result of intentional trauma occur during early childhood.
- Careful history and physical examination, plus consultation with a multidisciplinary child abuse team and reporting of suspected abuse, are warranted for specific patterns of injuries.

American Board of Pediatrics Content Specification(s):

- Recognize the chip fracture of metaphysic as commonly due to wrenching or pulling injuries

Suggested Reading:

- Endom EE. Physical abuse in children: diagnostic evaluation and management. UptoDate. Available online only for subscription. Scherl SA, Endom EE. Orthopedic aspects of child abuse. UptoDate. Available online only for subscription
- Wells L, Sehgal K, Dormans JP. Common fractures. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Philadelphia, PA: Elsevier Saunders; 2011:2387-2393

Item 141

The mother of a 7-month-old infant is frustrated that the infant's atopic dermatitis is not getting better. He is awake "all night" scratching and is irritable and fussy. She has been giving him diphenhydramine every 8 hours and applying hypoallergenic moisturizer and a topical corticosteroid cream twice a day. The infant was breastfed until 3 months ago and then switched to a cow milk-based formula. On physical examination, you notice that he has dry, erythematous papules and patches, with excoriation marks on his face, neck, antecubital fossae, popliteal fossae, and back. He has normal growth parameters.

Of the following, the MOST appropriate next step in this infant's management is to recommend

A. discontinuing diphenhydramine and switching him to daily loratadine
B. eliminating cow milk, egg, soy, and wheat from his diet
C. introducing cow milk on a trial basis to see if the rash worsens
D. switching to hypoallergenic formula and a diet of only rice and chicken
E. testing for pertinent, potential food allergen triggers

Item 141 S Preferred Response: E

The child described in the vignette should be tested for pertinent potential food allergen triggers. Up to 40% of children younger than 5 years with moderate to severe atopic dermatitis (AD) will have IgE-mediated food allergy (FA). The data are conflicting on whether food allergy can exacerbate AD partly because well-designed relevant food allergen avoidance trials have rarely been conducted. An expert panel recently published National Institute of Allergy and Infectious Diseases food allergy guidelines suggesting that children younger than 5 years with moderate to severe AD be considered for FA evaluation for milk, egg, peanut, wheat, and soy if at least 1 of the following conditions is met: (a) persistent AD despite optimized management and topical therapy or (b) reliable history of an immediate reaction after ingestion of a specific food. On the basis of the history, it is possible that this child has a milk allergy exacerbating the eczema.

Although cow milk, egg, soy, and wheat are common food allergens, random elimination of the foods from the diet can result in nutritional imbalances in the critical period of growth and development and is not recommended. Switching to a hypoallergenic formula with chicken and rice only for supplementation is not recommended, even on a trial basis, owing to similar concerns regarding optimization of growth and maintenance of nutritional balance.

Introduction of whole cow milk is recommended at 1 year of age by the American Academy of Pediatrics. Introducing whole milk on a trial basis at home in this child with suspected milk allergy, unsupervised by an expert trained in managing severe anaphylactic reactions, is risky. It is preferable to first test for possible allergy to suspected foods, such as milk, using blood or skin testing. If the testing result is negative or inconclusive, an oral challenge performed under the care of an FA expert may be considered in order to determine the relationship of milk allergy to eczema.

Pruritus is a key feature of AD and has a significant impact on sleep and quality of life. Allokinesis, a phenomenon in which a normally innocuous stimulus induces itch, is common in AD. Sweating, sudden changes in temperature, clothing changes, and direct contact with wool can lead to intense itching. Injury to the skin from scratching stimulates a vicious itch-scratch cycle. Itch control in eczema is therefore extremely challenging. Antihistamines are widely used to treat pruritus, although the evidence supporting their use is relatively weak since no large, randomized, placebo-controlled trials with definitive conclusions have been performed. Nevertheless, first-generation antihistamines, such as diphenhydramine and hydroxyzine, are most effective in controlling the itch owing to the antihistamine effect as well as the sedative action. Non-sedating preparations such as fexofenadine or loratadine may help, especially when there is an urticarial component and for patients in whom the sedating effect of first-generation antihistamines is undesirable (eg, school-age children). Doses higher than normal may be necessary.

PREP Pearls

- Children younger than 5 years with moderate to severe atopic dermatitis may be considered for evaluation of food allergy to milk, egg, peanut, wheat, and soy, if they have persistent atopic dermatitis despite optimized management, or they have a reliable history of an immediate reaction after ingestion of a specific food.

American Board of Pediatrics Content Specification(s):

- Understand the relationship of eczema and food allergies, and how to evaluate a patient with both

Suggested Reading:

- Eigenmann PA, Sicherer SH, Borkowski TA, Cohen BA, Sampson HA. Prevalence of IgE-mediated food allergy among children with atopic dermatitis. Pediatrics. 1998 Mar;101(3):E8

Item 142

You are evaluating a 50-hour-old term female neonate. The 2.9-kg neonate was born to a 34-year-old gravida 2, para 2 woman by emergent cesarean section for bradycardia and cord prolapse. The newborn emerged limp and apneic with an initial heart rate of 0. A full resuscitation was required, including intubation with positive pressure ventilation, compressions, and one dose of epinephrine. A heart rate was detected at 3 minutes after birth, and the infant began to have respiratory effort at 7 minutes.

The neonate's current vital signs include a weight of 3.0 kg, temperature of 37°C, heart rate of 140 beats/min, respiratory rate of 47 breaths/min, blood pressure of 120/70 mm Hg, and oxygen saturation of 97% by pulse oximetry on room air. She has taken nothing by mouth while receiving intravenous fluids. She has a urine output of 10 mL in the last 8 hours. Physical examination of the infant reveals a comfortable, pink newborn infant, clear and equal lung sounds bilaterally, no murmur, and right flank mass.

The following are notable results from her blood test and urinalysis:
Complete blood cell count:

- White blood cell count, 20,000/µL (20 x 10^9/L)
- Electrolytes, normal
- Hemoglobin, 8.9 g/dL (89 g/L)
- Hematocrit, 27% (0.27)
- Platelet count, 13 x 10^3/µL (13 x 10^9/L)
- Blood urea nitrogen, 28 mg/dL (10.0 mmol/L)
- creatinine, 1.9 mg/dL (168 µmol/L)

Urine test strip and microscopy:

- Specific gravity, 1.015
- pH, 5.5
- 4+ blood
- 3+ protein
- Red blood cells, 40 to 50 per high-power field

Of the following, the MOST accurate interpretation of the patient's serum creatinine, urine output, and flank mass is

A. acute renal failure, normal urine output, renal vein thrombosis
B. acute renal failure, normal urine output, sepsis
C. acute renal failure, oliguria, renal vein thrombosis
D. maternal serum creatinine, oliguria, neonatal renal mass
E. maternal serum creatinine, oliguria, sepsis

Item 142 Preferred Response: C

Neonatal serum creatinine concentration usually <1.0 mg/dL [88.4 µmol/L] is reflective of maternal serum creatinine concentration. In a full-term neonate, the serum creatinine concentration normalizes in 7 to 10 days whereas in a preterm infant, it may take up to 1 month to normalize. A serum creatinine concentration higher than 1.5 mg/dL (133 µmol/L) usually indicates acute kidney injury (also known as acute renal failure), as seen in the newborn described in the vignette.

For further management of neonatal acute kidney injury, it is important to evaluate the urine output. Placement of a urinary catheter is useful in evaluating neonates with oliguria/anuria as the catheter bypasses the urethral/bladder outlet obstruction (eg, posterior urethral valves in boys) and also provides an accurate estimate of urine output which is helpful in both evaluation and management. Oliguria/anuria is defined as no urine output by 48 hours of age or a urine output less than 1 mL/kg per hour. The patient in the vignette has a urine output of 0.4 mL/kg per hour and hence has oliguria. Initial evaluation in an infant with anuria/oliguria should include a detailed history and physical examination. The obstetric history should focus on presence or absence of oligohydramnios or polyhydramnios on prenatal ultrasonography, drug exposure, and family history of renal failure. Physical examination should evaluate genitalia, abdomen (flank mass or palpable bladder), and signs of oligohydramnios/Potter sequence. Initial evaluation will include electrolytes, serum urea nitrogen, creatinine, urinalysis, and renal ultrasonography.

Renal vein thrombosis is the most common cause of non-catheter-associated thrombosis in the newborn. Prematurity, perinatal asphyxia, shock, dehydration, sepsis, polycythemia, cyanotic congenital heart disease, and maternal diabetes have been associated with an increased risk for renal vein thrombosis. Flank mass, thrombocytopenia, and hematuria are the classic features associated with renal vein thrombosis. Renal vein thrombosis may also be associated with elevated blood pressure, laboratory features of disseminated intra-vascular coagulation, and leukocytosis. Ultrasonography of the kidney is the imaging modality of choice for diagnosing renal vein thrombosis in a newborn. In the early stages of thrombosis, the kidneys appear swollen and echogenic. This gradually evolves into loss of corticomedullary differentiation followed by scarring and decrease in renal size. Color Doppler examination on renal ultrasonography will show absent intrarenal and renal venous flow in the early stages of thrombosis.

Sepsis associated with acute kidney injury is unlikely in a well-appearing newborn with stable vital signs and evidence of good peripheral circulation.

PREP Pearls

- Serum creatinine higher than 1.5 mg/dL (132.6 µmol/L) is usually indicative of acute kidney injury in a newborn.
- Oligoanuria in a newborn is defined as no urine output for 48 hours or a urine output less than 1 mL/kg per hour.
- Flank mass, hematuria, and thrombocytopenia are suggestive of renal vein thrombosis in a newborn. These patients often present with associated acute kidney injury and/or hypertension.

American Board of Pediatrics Content Specification(s):

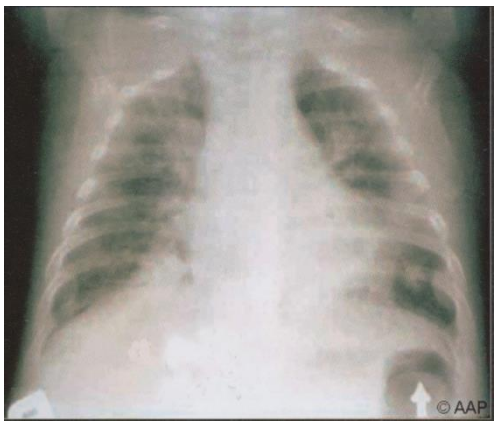
- Plan the evaluation of an infant with anuria more than 48 hours after birth

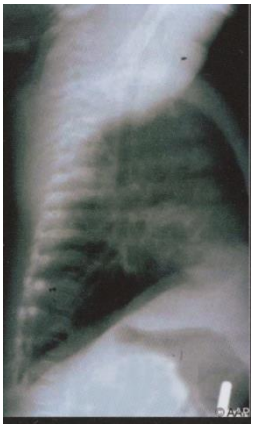
Suggested Reading:

- Chua AN, Sarwal MM. Acute renal failure management in the neonate. NeoReviews. 2005;6:e369-e376. doi: 10.1542/neo.6-8-e369
- Lau KK, Stoffman JM, Williams S, et al. Neonatal renal vein thrombosis: review of the English-language literature between 1992-2006. Pediatrics. 2007;120(5):e1278-1284. doi: 10.1542/peds.2007-0510

Item 143

A 4-week-old former term infant in foster care presents to your office with "fast breathing" and cough of 1 week's duration. No information is available regarding the birth mother other than that she had no prenatal care. The results of recent polymerase chain reaction tests for human immunodeficiency virus were negative. On physical examination, the temperature is 37°C, heart rate is 135 beats/min, respiratory rate is 65 breaths/min, and blood pressure is 70/50 mm Hg. Oxygen saturation is 91 % on room air. The infant is well developed and well nourished. The anterior fontanelle is soft, open and flat. There is nasal congestion with mild nasal flaring but no discharge. The conjunctivae, oropharynx, and tympanic membranes are normal. The patient has moderate subcostal retractions, and there are diffuse rales on auscultation. During the examination, the infant has a repetitive staccato cough. The remainder of the physical examination is normal. Chest radiographs are obtained (Item Q143, page Q-37).





ITEM Q143: Chest radiographs for the infant in the vignette.

Of the following, the MOST appropriate initial antibiotic therapy for this infant should be

A. ceftriaxone
B. doxycycline
C. erythromycin
D. ofloxacin
E. trimethoprim–sulfamethoxazole

Item 143 Preferred Response: C

The infant described in the vignette has an afebrile pneumonia that developed in the first weeks after birth and is characterized by tachypnea and a staccato cough accompanied by nasal congestion. The chest radiograph reveals hyperinflation with bilateral, symmetrical, interstitial infiltrates. Infection with Chlamydia trachomatis, the most common cause of sexually transmitted infection in the United States, is likely based on the timing of the illness, the clinical presentation, and the lack of prenatal care. Therefore, the most appropriate antibiotic therapy is erythromycin. Erythromycin base or ethylsuccinate is administered orally at a dose of 50 mg/kg per day divided every 6 hours for 14 days. A second course of therapy may be required in up to 20% of patients. Although there has been an association between the administration of oral erythromycin and the development of infantile hypertrophic pyloric stenosis in infants younger than 6 weeks, erythromycin is recommended for the treatment of C trachomatis because a causal relationship has not been confirmed and alternative therapies are not well studied. The risk of pyloric stenosis after treatment with other macrolides (eg, azithromycin) is unknown.

Chlamydia trachomatis pneumonia occurs in 5% to 30% of infants born to mothers with cervical infection and usually develops between 2 and 19 weeks after birth. The onset of illness is insidious, and infants usually are afebrile. Physical examination findings may include tachypnea, nasal congestion, otitis media, rates on auscultation of the lungs, a staccato cough that can be paroxysmal, and a palpable liver and spleen due to hyperinflation of the lungs. Wheezing is uncommon.

For infants outside the neonatal period with C trachomatis infection who do not tolerate erythromycin, an oral sulfonamide (eg, trimethoprim-sulfamethoxazole) may be used for treatment. For adolescents or adults with uncomplicated anogenital tract infection, oral doxycycline for 7 days is recommended.

Ceftriaxone, intramuscular or intravenous, would be the recommended treatment for gonococcal infection in newborns: one dose only for treatment of ophthalmia neonatorum or 7 days for disseminated disease. However, cefotaxime is preferred over ceftriaxone for the treatment of disseminated gonococcal infection in neonates with hyperbilirubinemia because ceftriaxone has been reported to displace bilirubin from albumin-binding sites. Oral ofloxacin can be used in patients older than 17 years for the treatment of nongonococcal cervicitis or urethritis or for pelvic inflammatory disease if the prevalence of quinolone-resistant gonococcal organisms in the community is low.

PREP Pearls

- Physical examination findings in infants with C trachomatis pneumonia may include tachypnea, nasal congestion, otitis media, rales on auscultation of the lungs, a staccato cough that can be paroxysmal, and a palpable liver and spleen due to hyperinflation of the lungs.
- Erythromycin is the recommended treatment for C trachomatis infection in infants.
- There is an association between the administration of oral erythromycin and the development of infantile hypertrophic pyloric stenosis in infants younger than 6 weeks, but no causal relationship has been confirmed.

American Board of Pediatrics Content Specification(s):

- Know the clinical manifestations of Chlamydia trachomatis pneumonia in young infants

Suggested Reading:

- American Academy of Pediatrics. Chlamydia trachomatis. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:276-281
- Pammi M, Hammerschlaghtt M. Chlamydia trachomatis infections in the newborn. UptoDate. Available online for subscription only.

Item 144

A 15-year-old girl fainted at school and is brought to the emergency department. She said she was in class when she arose from her desk, felt lightheaded, and blacked out. Presently, she reports feeling well and wants to go home. She did not eat breakfast this morning. There is no significant past medical history, she takes no medications, and there is no family history of cardiac problems or epilepsy. On further questioning about her eating habits, you learn that she has lost 30 pounds in the last 2 months. She reports that she has been exercising to "tone up" and is eating more "healthy" foods. Her last menstrual period was 2 months ago, and she is not sexually active. On physical examination, you note a body mass index of 18, blood pressure of 104/68 mm Hg, and a heart rate of 48 beats/min. When she stands from a seated position, her heart rate increases to 78 beats/min and her blood pressure is 90/58 mm Hg. A pregnancy test result is negative.

Of the following, the MOST appropriate next step in the management of this girl's condition is to

- admit her for further management
- advise her to stop exercising for a month
- discharge to home after intravenous fluid administration
- refer her to a dietitian
- refer her to a neurologist

Item 144 TE SBP Preferred Response: A

The girl described in the vignette most likely has an eating disorder and fits the category of unspecified feeding or eating disorder because her BMI is still between 10-25 percentile for age, and she has not explicitly stated having an intense fear of gaining weight. She has lost a significant amount of weight in a short period of time, denies the severity of the situation, and her request to go home may stem from her fear of being forced to eat and gain weight, if admitted. She meets the medical criteria for admission, with a resting heart rate of less than 50 beats/min during the day and signs of orthostatic changes (ie, blood pressure change of >10 mm Hg and pulse rate change of >20 beats/min). Other criteria often used for admission are a heart rate of less than 40/min during the night; systolic blood pressure less than 90 mm Hg; hypothermia (body temperature <96°F); cardiac arrhythmias; severe malnutrition with a weight less than 75% of ideal body weight for age, sex, and stature; acute weight decrease; refusal of food; or electrolyte disturbances. All these changes reflect the severity of malnutrition and the need for stabilization. In addition, if there are any concerns about mental health status (suicidal ideation, a plan, or an attempt) or if the adolescent has been followed up as an outpatient but is not responding to treatment, she or he should not be sent home.

The immediate goal for admission is to correct metabolic abnormalities and the malnourished state while being careful to avoid the refeeding syndrome (as a result of further depletion of already low levels of potassium, phosphate and magnesium as insulin secretion increases with reintroduction of carbohydrates). The need to correct unhealthy patterns of eating, promote weight gain, and evaluate for and treat psychiatric comorbidities should be addressed next along with developing plans for close follow-up on discharge. This treatment plan is best accomplished with a team approach from medical specialists, mental health personnel, and nutritionists.

Rehydration, if needed for the girl in the vignette, can probably be performed safely by the oral route. She needs to be placed on bed rest until she is medically stable and showing consistent weight gain. After discharge it would be advisable to restrict exercise until continued weight gain is documented because often adolescents will eat while hospitalized to speed up the discharge but start to restrict soon after. There is no indication for a neurologic consultation at this time because her syncope is most likely secondary to postural hypotension.

PREP Pearls

- Most adolescents who have eating disorders do not meet all the Diagnostic and Statistical Manual of Mental Disorders criteria for an anorexia diagnosis and fall into the Unspecified Feeding or Eating Disorder category.
- There are accepted criteria available to guide the need for admission: resting heart rate less than 50 beats/min during the day and signs of orthostatic changes; heart rate of less than 40 beats/min during the night; systolic blood pressure less than 90 mm Hg; hypothermia (body temperature <96°F); cardiac arrhythmias; severe malnutrition with a weight less than 75% of ideal body weight for age, sex, and stature; acute weight decline; refusal of food; or electrolyte disturbances.

- Refeeding syndrome results when phosphorus levels are too low.

American Board of Pediatrics Content Specification(s):

- Know the indications for the hospitalization of adolescents with anorexia nervosa

Suggested Reading:

- American Psychiatric Association. Diagnostic and Statistical. Manual of Mental Disorders Fifth ed. Washington, DC: American Psychiatric Association; 2013:329-354
- Breuner CC. Complementary, holistic, and integrative medicine: eating disorders. Pediatr Rev. 2010;31:e75-e82. doi:10.1542/pir.31-10-e75
- Fisher M. Treatment of eating disorders in children, adolescents, and young adults. Pediatr Rev. 2006;27:5-16. doi:10.1542/pir.27-1-5
- Goldstein MA, Dechant EJ, Beresin EV. Eating disorders. Pediatr Rev. 2011;32:508-521. doi:10.1542/pir.32-12-508
- Le Grange D, Doyle PM, Swanson SA, Ludwig K, Glunz C, Kreipe RE. Calculation of expected body weight in adolescents with eating disorders. Pediatrics. 2012;129(2):e438-e446. doi:10.1542/peds.2011-1676

Item 145

An 8-year-old boy presents with rectal bleeding. The child was well until this evening, when he passed a normal-appearing stool followed by a large amount of blood per rectum. The blood appeared red to burgundy in color, and passage was not associated with complaints of abdominal pain, vomiting, diarrhea, or fever. On physical examination, the child appears somewhat pale and anxious but not in distress. His heart rate is 110 beats/min, respiratory rate is 18 breaths/ min, and blood pressure is 90/55 mm/Hg. The remainder of the examination is unremarkable. Initial laboratory studies indicate the following:

- Hemoglobin, 10.2 g/dL (102 g/L)
- White blood cell count, 5,500/µL (5.5×10^9/L)
- Platelets, 180×10^3/µL (180×10^9/L)
- Prothrombin time, 12.0 s
- Partial thromboplastin time, 33.0 s
- International normalized ratio, 1.1
- Aspartate aminotransferase, 80 U/L; reference range, <_50 U/L
- Alanine aminotransferase, 30 U/L; reference range, <_40 U/L
- Albumin, 4.2 g/dL (42 g/L)

Of the following, you are MOST likely to order an additional test that will seek to identify the presence of

A. colonic polyp(s)
B. heterotopic gastric mucosa
C. intestinal duplication
D. intussusception
E. vascular malformation

Item 145 Preferred Response: B

Rectal bleeding is a relatively common symptom in infants, children, and adolescents. Both the common and uncommon causes of rectal bleeding are listed in Item C145. In many instances, the causes are benign, blood loss is insignificant, and bleeding is either self-limited or responsive to simple interventions (eg, bright red blood on the toilet tissue after A hard bowel movement, lymphonodular hyperplasia, milk protein proctocolitis, and many infectious causes). Bleeding of sufficient quantity to result in hemodynamic instability is a rare phenomenon. When patients present with a large volume blood loss, the initial goal of management must be to assess and stabilize cardiovascular status. The boy in the vignette presents with signs and symptoms of significant bleeding, marked by anemia and tachycardia. His bleeding is reported to be painless and red to burgundy in color, suggesting a bleeding source in the distal small bowel or the proximal colon. Because fresh blood quickly changes color to brown in an acid environment and intestinal bacteria oxidize hemoglobin to hematin, resulting in a tarry stool (melena), rectal passage of red blood rarely originates from the upper gastrointestinal (GI) tract, unless the rate of bleeding is brisk (eg, bleeding from esophageal or gastric varices). Nevertheless, when blood loss is significant and the source is unknown, a nasogastric tube should always be placed to determine whether a lesion is in the upper GI tract (ie, proximal to the ligament of Treitz). For the boy in the vignette, his age and the passage of a large volume of red to burgundy blood suggests that the most likely cause is a Meckel diverticulum.

Meckel diverticulum results from the incomplete obliteration of the omphalomesenteric (ie, vitelline) duct in utero. It is the most common congenital abnormality of the small intestine and has been reported in 0.2% to 4% of autopsy specimens. The diverticulum occurs on the antimesenteric border of the ileum, usually 40 to 60 cm proximal to the ileocecal valve. On average, the diverticulum is 3 cm long and 2 cm wide. More than half contain ectopic mucosa. Although various heterotopic tissues have been found to line the diverticulum, the most common type of ectopic tissue (found in 62% of cases in one study) is gastric mucosa.

Despite the fact that the Meckel diverticulum is a common anomaly, complications are uncommon, only occurring in approximately 5% of cases. In adults, obstruction and inflammation (similar to diverticulitis) are the most common presenting signs. In contrast, the classic presentation in children, most frequently occurring between 2 and 8 years of age, is painless rectal bleeding. Lower GI tract bleeding occurs with a typical acute onset that is secondary to hemorrhage from peptic ulceration. This bleeding occurs when acid secreted by heterotopic gastric mucosa damages adjacent small intestinal tissue, which may result in erosion of a blood vessel. Obstruction as a presenting sign of a Meckel diverticulum is less common in the pediatric age group. When it occurs, obstruction is the consequence of an omphalomesenteric band or internal herniation through, or volvulus around, omphalomesenteric duct remnants. In some cases, the diverticulum acts as a lead point for an ileocolic or ileoileal intussusception.

In the evaluation of a suspected Meckel diverticulum, routine laboratory studies (complete blood cell count, electrolytes, creatinine, liver function, and coagulation

profile) may be required for patient management but are not helpful in establishing a diagnosis. Importantly, however, one study reported that children with bleeding from this anomaly presented with a hemoglobin level of less than 8.8 g/dL (88 g/L) in more than 60% of cases. When a patient has GI bleeding suggestive of Meckel diverticulum, the diagnostic study of choice is a technetium Tc 99m pertechnetate scintiscan (the so-called Meckel scan). Because the pertechnetate is taken up by gastric mucosa, this study will demonstrate heterotopic tissue lining the diverticulum. False-positive results may occur wherever ectopic gastric mucosa is present, including in duodenal ulcer disease and in some cases of intestinal duplication.

Although Meckel diverticulum is the most common cause of painless, brisk GI bleeding in early childhood, it is certainly not the only cause. Bleeding from benign juvenile, inflammatory polyps is common in children 2 to 4 years of age; however, in this instance, bleeding is rarely massive and usually presents with chronic blood loss. Bleeding is a common late finding in intussusception. However, this diagnosis would be unusual at 8 years of age, and it is typically associated with signs of pain or obstruction. Vascular malformations and intestinal duplications may also present with acute, massive GI bleeding. However, a Meckel diverticulum is a much more common diagnosis in childhood.

Item C145. Causes of Rectal Bleeding in Childhood

Age, y	Common Causes	Uncommon Causes
0-1	Anal fissure Milk protein colitis Necrotizing enterocolitis Swallowed blood	Hirschsprung enterocolitis Infectious enterocolitis Intestinal duplication (ileal most common) Meckel diverticulum Vascular lesions
>1	Anal fissure Infectious enterocolitis *Campylobacter* species *Salmonella* species *Shigella* species Inflammatory bowel disease (>4 y) Intussusception Juvenile polyp Lymphonodular hyperplasia Meckel diverticulum	Anastomotic ulcer (postsurgical bowel resection) Colonic or rectal varices Hemolytic-uremic syndrome (*Escherichia coli* 0157:H7) Hemorrhoids Henoch-Schönlein purpura Inflammatory bowel disease (<4 y) Intestinal duplication Pseudomembranous enterocolitis (*Clostridium difficile*) Rectal trauma Sexual abuse Solitary ulcer of the rectum Vascular malformations

PREP Pearls

- The hallmark of Meckel diverticulum is hemodynamically significant, painless rectal bleeding.
- Dark red or purple rectal bleeding suggests a right colon or small bowel source.
- Meckel diverticulum bleeding occurs only in patients in whom the diverticulum contains gastric mucosa.

American Board of Pediatrics Content Specification(s):

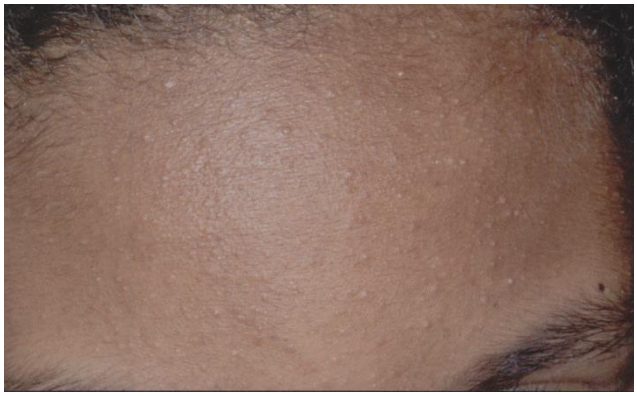
- Know the signs and symptoms of Meckel diverticulum

Suggested Reading:

- Elsayes KM, Menias CO, Harvin HJ, Francis IR. Imaging manifestations of Meckel's diverticulum. AIR Am J Roentgenol. 2007;189:81-88. doi:10.2214/AJR.06.1257
- Park JJ, Wolff BG, Tollefson MK. Meckel diverticulum: the Mayo Clinic experience with 1476 patients (1950-2002). Ann Surg. 2005;241:529-533. doi:10.1097/01.sla.0000154270.14308.5f
- Sagar J, Kumar V, Shah DK. Meckel's diverticulum: a systematic review. J R Soc Med. 2006;99:501-555. DOI: 10.1258/jrsm.99.10.501
- Squires RH Jr. Gastrointestinal bleeding. Pediatr Rev. 1999;20:95-101. doi: 10.1542/pir.20-3-95
- Stone PA, Hofeldt MJ, Campbell JE. Meckel diverticulum: ten-year experience in adults. South Med J. 2004;97:1038-1041
- St-Vil D, Brandt ML, Panic S, Bensoussan AL, Blanchard H. Meckel's diverticulum in children: a 20-year review. J Pediatr Surg. 1991;26:1289-
- 1292. doi:10.1016/0022-3468(91)90601-0

Item 146

A 13-year-old girl is requesting treatment for acne. She has no underlying medical conditions and has normal growth. She had menarche at 11 years of age and has a regular menstrual pattern. She is not sexually active. Skin examination shows acne located only on the forehead. She has multiple closed and a few open comedones (Item Q146).



ITEM Q146: Closed comedones as described for the girl in the vignette.

Of the following, the PREFERRED initial treatment for this patient is

A. oral contraceptive pill
B. oral doxycycline
C. topical clindamycin
D. topical retinoid
E. topical salicylic acid

Item 146 S Preferred Response: D

Although there is no uniform method for categorizing acne, the location and nature of the lesions are important considerations when deciding how to treat the condition. In one staging system, mild disease involves about one quarter of the face, with few to several papules or pustules but no nodules or scarring. Moderate disease affects one half of the face, with several to many papules and pustules, few to several nodules, and few scars. Severe disease involves three quarters of the face with many papules, pustules, and nodules as well as scarring. Patients such as the one in the vignette with mild disease are treated topically, whereas those with moderate and severe disease may require at least limited courses of systemic antibiotics or oral retinoids.

Treatment of acne is directed at limiting formation of microcomedones (the initial lesion leading to comedones) and preventing inflammation related to the presence of Propionibacterium acnes. Topical retinoids including tretinoin, adapalene, tazarotene, and isotretinoin (not available in the United States) act on both these issues by reducing obstruction in follicles, and are considered first-line treatment for most acne by the Global Alliance to Improve Outcomes in Acne. Benzoyl peroxide also can be used as primary therapy for acne. It is a potent bactericidal agent, and P acnes cannot develop resistance to it. The topical antibiotics clindamycin and erythromycin have been shown to be effective and well tolerated, but P acnes readily develops resistance, and thus these agents should not be used as monotherapy. Combining topical antibiotics with benzoyl peroxide helps eliminate antibiotic resistance and improves efficacy, with the combination being more effective than either component alone. Other topical therapies, including salicylic acid, are less effective than retinoids or benzoyl peroxide and thus are not usually considered first-line therapy. Salicylic acid does have a role for patients with mild disease who are unable to tolerate the irritation associated with topical retinoids or benzoyl peroxide.

Systemic antibiotics, primarily doxycycline and minocycline, are frequently used for moderate to severe acne. As with topical antibiotics, oral therapy is also associated with increasing antibiotic resistance. Therefore, the duration of treatment should be limited, and concurrent topical therapy may be of some benefit. Once inflammation has improved with oral antibiotics, the patient should be moved to topical therapy with benzoyl peroxide and/or retinoids to prevent recurrence of inflammation. For severe cases or when the response to oral antibiotics is inadequate, oral isotretinoin can be prescribed. Because of the side effects, including teratogenicity and possible mood disorders, oral isotretinoin should only be prescribed by practitioners with special training using a risk management program called iPLEDGE (www.ipledgeprogram.com).

Therapy using estrogen-containing contraceptive agents can be a useful adjunct for girls, particularly those requiring hormone therapy for contraception or to control menorrhagia. However, it is not first-line therapy, particularly for girls with mild disease such as the patient in the vignette. Currently the Food and Drug Administration approves contraceptives containing either norgestimate or norethindrone acetate with ethinyl estradiol for the treatment of acne, but it is likely that most estrogen-containing oral

contraceptives will be effective. Spironolactone, which blocks androgen receptors, also can be used in treating acne, but it is not a first-line drug for this indication.

PREP Pearls

- Topical retinoids or benzoyl peroxide are first-line therapy for mild to moderate acne.
- P acnes readily develops antibiotic resistance, so topical antibiotics should be combined with benzoyl peroxide, and oral antibiotics should only be used for a limited duration.
- Oral isotretinoin should be prescribed by practitioners who have been trained in risk management regarding the potential side effects of the medication.

American Board of Pediatrics Content Specification:

- Plan for the treatment of acne vulgaris with first-line topical medications, retinoic acid, and benzoyl peroxide

Suggested Reading:

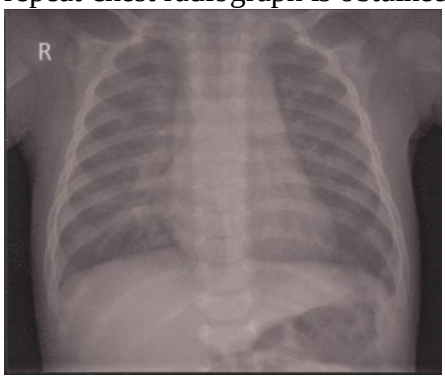
- Krowchuk D. Managing adolescent acne: a guide for pediatricians. Pediatr Rev. 2005;26:250-261. doi: 10.1542/pir.26-7-250
- Strauss JS, Krowchuk DP, Leyden JJ, et al; American Academy of Dermatology/ American Academy of Dermatology Association.
- Guidelines of care for acne vulgaris management. J Am Acad Dermatol. 2007;56:651-663. doi:10.1016/j.jaad.2006.08.048
- Zaenglein AL, Thiboutot DM. Expert committee recommendations for acne management. Pediatrics. 2006;118:1188. doi: 10.1542/peds.2005-2022

Item 147

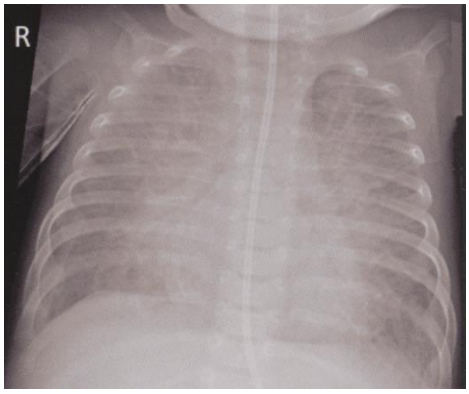
A 3-month-old boy was brought into the emergency department because of decreased activity and poor feeding for the past several weeks. The parents state that their child has not had a fever, has not been hospitalized previously, and has been followed for poor weight gain. They report that he has had an intermittent mild cough but no nasal discharge.

The boy's temperature is 37.0°C, heart rate is 150 beats/ min, respiratory rate is 45 breaths/min, and blood pressure is 65/40 mm Hg. His peripheral pulses are palpable but diminished. He appears in moderate respiratory distress with coarse bilateral breath sounds. A chest radiograph is obtained (Item Q147A).

The emergency department staff obtains laboratory studies and blood cultures and administers a total of 40 mL/kg of isotonic fluid and antibiotics. The infant becomes less responsive, with a heart rate of 170 beats/min, respiratory rate of 60 breaths/min, and blood pressure of 50/28 mm Hg, and his peripheral pulses are now difficult to palpate. A repeat chest radiograph is obtained (Item Q147B).



ITEM Q147A: Initial chest radiographic findings for the boy in the vignette.



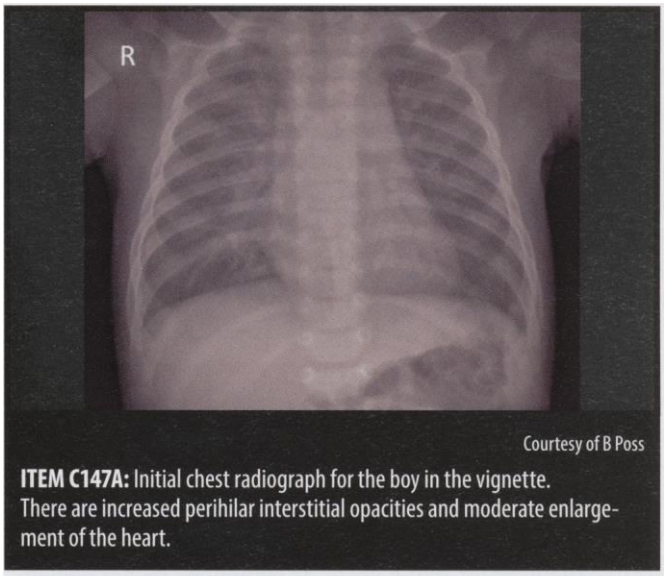
ITEM Q147B: Findings for the boy in the vignette on repeat chest radiography.

Of the following, the MOST likely cause of this boy's shock is

A. bacterial pneumonia
B. congestive heart failure
C. hypovolemia
D. sepsis
E. viral bronchiolitis

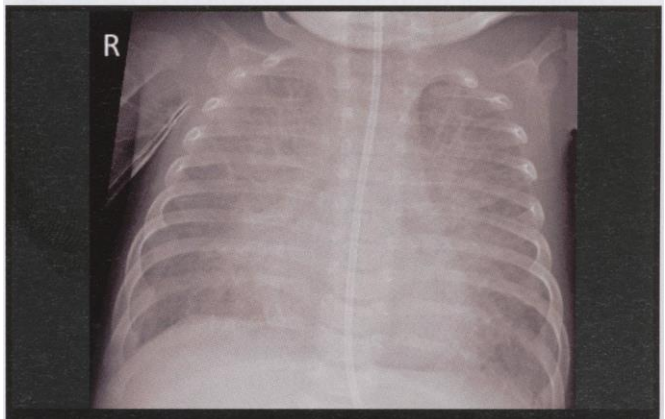
Item 147 Preferred Response: B

The child described in the vignette is in shock, as evidenced by tachycardia and poor perfusion. Shock is defined as the inadequate delivery of oxygen to meet the metabolic demand of tissues and can be divided into several types: cardiogenic, hypovolemic, and septic, as well as the less common anaphylactic and neurogenic. The child in the vignette most likely has cardiogenic shock secondary to congestive heart failure with marked deterioration after large volume fluid resuscitation. Symptoms of congestive heart failure (CHF) can include tachypnea, poor feeding, and decreased activity as reported for the child in the vignette.



Courtesy of B Poss

ITEM C147A: Initial chest radiograph for the boy in the vignette. There are increased perihilar interstitial opacities and moderate enlargement of the heart.



Courtesy of B Poss

ITEM C147B Chest radiograph following fluid administration: there is cardiomegaly and increased interstitial markings suggestive of congestive heart failure.

The boy's initial chest radiograph (Item C147A) shows no evidence of pulmonary consolidation which, coupled with an absence of fever, argues against a diagnosis of bacterial pneumonia or sepsis. The presence of perihilar opacities can be consistent with either mild pulmonary edema (as seen in CHF) or bronchiolitis. In addition, his mild cardiomegaly, especially in consideration of the large degree of inspiration, should be noted. Following aggressive resuscitation with fluid, he deteriorates clinically, and radiography documents marked cardiomegaly and pulmonary edema (Item C147B), which is highly suggestive of cardiac disease. This marked deterioration with fluid administration excludes hypovolemia as a cause of shock and makes viral bronchiolitis much less likely than CHF. In addition, the prolonged clinical history makes viral bronchiolitis unlikely.

Initial treatment of cardiogenic shock is respiratory stabilization, diuretic administration, and initiation of inotropic medications to improve cardiac function. Once stabilized, treatment of CHF will normally consist of a combination of pharmacologic and nutritional interventions. Affected children grow poorly because of reduced caloric intake and unmet increases in metabolic requirements and will often need fortified breast milk or formulas (older children will need age-appropriate nutritional supplementation). The goals of pharmacologic treatment are preload and after-load reduction, with diuretics used for preload reduction and angiotensin-converting

enzyme inhibitors for afterload reduction. Digoxin is still used in some patients whereas β-blockers are less commonly used than in adult patients.

PREP Pearls

- Shock is defined as the inadequate delivery of oxygen to meet the metabolic demand of tissues.
- A worsening of the patient's clinical condition after large volume resuscitation should prompt consideration of cardiac failure.
- Initial treatment of cardiogenic shock is respiratory stabilization, diuretic administration, and initiation of inotropic medications to improve cardiac function.

American Board of Pediatrics Content Specification(s):

- Plan the acute treatment of congestive heart failure in a child or adolescent

Suggested Reading:

- Macicek SM, Macias CG, Jefferies JL, Kim JJ, Price JF. Acute heart failure syndromes in the pediatric emergency department. Pediatrics. doi: 10.1542/peds.2008-2198
- Madriago E, Silberbach M. Heart failure in infants and children. Pediatr Rev. 2010;31:4-12. doi: 10.1542/10.1542/pir.31-1-4
- McKiernan CA, Lieberman SA. Circulatory shock in children: an overview. Pediatr Rev. 2005;26:451-460. doi: 10.1542/pir.26-12-451
- Turner DA, Cheifetz IM. Shock. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:305-314

Item 148

A 17-year-old girl has poorly controlled type 1 diabetes mellitus since 5 years of age. Her blood pressure is now elevated at 132/82 mm Hg. Her most recent hemoglobin A1c was 11.5%. Her other annual screening tests, including urine microalbumin, thyroid function testing, and lipid panel, yield normal results.

Of the following, the intervention MOST likely to reduce long-term renal complications in this child is

A. angiotensin-converting enzyme inhibitor therapy
B. atorvastatin therapy
C. increased physical activity
D. long-term blood glucose control
E. low-salt diet

Item 148 Preferred Response: D

The adolescent patient in the vignette has had poorly controlled diabetes for many years, as evidenced by her elevated hemoglobin A1c level and elevated blood pressure. As is true with almost all complications of diabetes mellitus, the degree of glucose control and duration of diabetes mellitus are linked to long-term complications. In long-term (20-year) follow-up of patients with type 1 diabetes mellitus, extensive clinical trials have shown that the glomerular filtration rate (and overall kidney function) is improved in patients treated with intensive insulin therapy earlier in their disease and correlates with the degree of glucose control.

Angiotensin-converting enzyme (ACE) inhibitor therapy is warranted for persistent elevation of blood pressure or if microalbuminuria develops. Similar to 3-hydroxy-3-methyl-glutaryl coenzyme A reductase inhibitors (eg, atorvastatin) that are used in the treatment of hyperlipidemia, ACE inhibitors are teratogens (Class D and Class X agents in pregnancy, respectively) and should only be used with appropriate counseling in female adolescents. Initiating ACE inhibitor therapy is still not the best answer because normalizing glucose levels to appropriate targets is most important.
Increasing physical activity and initiating a low-salt diet will improve glucose control and possibly help to improve blood pressure, but the effect on long-term prognosis is much less than more stringent control of glucose levels.

PREP Pearls

- Tight glucose control improves long-term outcomes in patients with type 1 diabetes mellitus.
- Although adjuvant treatments are important to prescribe when . indicated (eg, ACE inhibitors, statins), pediatricians must be a' of their adverse effects and must still stress control of glucose levels as the primary method to prevent complications.

American Board of Pediatrics Content Specification(s):

- Know the long-term complications of type 1 diabetes

Suggested Reading:

- DCCT/EDIC Research Group, de Boer IH, Sun W, Cleary PA, et al. Intensive diabetes therapy and glomerular filtration rate in type 1 diabetes. N Engl J Med. 2011;365:2366-2376. doi:10.1056/NEJMoa1111732
- Vergouwe Y, Soedamah-Muthu SS, Zgibor J, Chaturvedi N, Forsblom C, Snell-Bergeon JK, Maahs DM, Groop PH, Rewers M, Orchard TJ, Fuller JH, Moons KG. Progression to microalbuminuria in type 1 diabetes: development and validation of a prediction rule. Diabetologia. 2010 Feb;53(2):254-62. doi: 10.1007/s00125-009-1585-3

Item 149

An 8-year-old boy presents to your office with a 2-day history of sore throat, headache, and temperature up to 39.1°C. A rapid test result for Group A Streptococcus is positive. The boy has a past history of an anaphylactic reaction to amoxicillin.

Of the following, the BEST choice of treatment for this patient is

A. azithromycin for 5 days
B. cephalexin for 10 days
C. ciprofloxacin for 10 days
D. doxycycline for 10 days
E. trimethoprim-sulfamethoxazole for 10 days

Item 149 S Preferred Response: A

Penicillin (or amoxicillin) remains the drug of choice for the treatment of group A streptococcal (GAS) pharyngitis. In the face of a history of allergy to penicillins, characterized by a delayed reaction, a first-generation cephalosporin (eg, cephalexin) becomes the best choice for treatment of this condition. When the patient has a history of anaphylaxis or an immediate type 1 hypersensitivity reaction such as urticaria after exposure to penicillin, a non-beta lactam agent is preferred.

Of the choices listed, azithromycin for 5 days is the best option. The dosage of azithromycin for GAS infection is 12 mg/kg per day; not 10 mg/kg on day 1 and 5 mg/kg on days 2 to 5 as recommended for community-acquired pneumonia. If macrolide resistance is prevalent in a given region, clindamycin would be the drug of choice in the penicillin-allergic patient.

Although GAS isolates are generally sensitive to in vitro ciprofloxacin, the drug does not have an indication for GAS pharyngitis. Fluoroquinolones are not recommended for use in children unless they are the best available drug, which is not the case in the vignette. Doxycycline and trimethoprim-sulfamethoxazole do not have significant activity against GAS and hence would not be appropriate choices even in the face of a penicillin allergy.

PREP Pearls

- Penicillin or amoxicillin remains the drug of choice for treatment of group A streptococcal pharyngitis.
- For most penicillin-allergic patients, a first-generation cephalosporin is the best choice for treating GAS pharyngitis. In the face of a history of anaphylaxis or type 1 hypersensitivity reaction to penicillin a macrolide is a recommended alternative for treating GAS pharyngitis.

American Board of Pediatrics Content Specification(s):

- Plan the treatment of group A streptococcal infection

Suggested Reading:

- American Academy of Pediatrics. Group A streptococcal infections. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:668-680
- Shulman ST, Bisno AL, Clegg HW, et al. Clinical practice guideline for the diagnosis and management of group A streptococcal pharyngitis: 2012 update by the Infectious Diseases Society of America. Clin Infect Dis. 2012;55(10):e86-e102. doi:10.1093/cid/cis629

Item 150

A 15-year-old boy presents to your office with knees that have been swollen and painful for the last 6 weeks. He has morning stiffness daily that lasts 2 hours. The review of systems is positive for weight loss of 2.3 kg, occasional loose stools, and vague abdominal pain for the last 6 months. Physical examination reveals warmth, swelling, and pain with range of motion of the knees bilaterally. There is diffuse abdominal tenderness without guarding or rebound. The following are the results of the boy's laboratory studies:

- White blood cell count, 8,900/µL (8.9×10^9/L)
- Erythrocyte sedimentation rate, 79 mm/h
- Hemoglobin, 8.4 g/dL (84 g/L)
- Hematocrit, 26% (0.26)
- Platelets, 348×10^3/µL (348×10^9/L)
- Mean corpuscular volume, 75 μm^3 (75 fL)

His stool is negative for occult blood and ova and parasites. Of the following, the study MOST likely to identify the cause of this child's symptoms is

A. antinuclear antibody
B. arthrocentesis
C. colonoscopy
D. upper gastrointestinal endoscopy
E. upper gastrointestinal series with small-bowel follow-through

Item 150 Preferred Response: E

The patient described in the vignette has symptoms, physical examination findings, and laboratory findings consistent with inflammatory bowel disease (IBD). The constellation of diarrhea, weight loss, anemia, and arthritis suggest a diagnosis of IBD.

There are two patterns of arthritis associated with IBD. Most often, patients present with polyarticular peripheral arthritis involving large joints, usually of the lower extremities. Occasionally, the upper extremities and small joints can be involved. Usually, these episodes of arthritis last 1 to 2 weeks and correlate with disease activity of the IBD. Rarely, the arthritis can persist, but joint damage is unusual. This pattern of arthritis is often associated with erythema nodosum. The second pattern of arthritis associated with IBD is a spondyloarthropathy affecting the axial skeleton and the sacroiliac joints. Often this is associated with human leukocyte antigen B27 (HLA-B27) positivity. The spondyloarthropathy of IBD, which is not related to the disease activity, can be clinically silent initially and usually presents in the third decade of life.

Occasionally, the arthropathy of IBD can precede the gastrointestinal symptoms; therefore, all patients with arthritis should be screened for gastrointestinal symptoms. Of the choices, an upper gastrointestinal series with small-bowel follow-through is the best next step. Although colonoscopy and upper gastrointestinal endoscopy can be used to diagnose inflammatory bowel disease with confirming biopsy, the risk of performing these tests may outweigh the diagnostic benefit in a patient with actively inflamed bowel. Arthrocentesis would likely show an inflammatory synovial fluid in this case but would not yield a definitive diagnosis. Antinuclear antibody is not specific to IBD and would not yield a definitive diagnosis.

PREP Pearls

- Arthritis can precede the gastrointestinal symptoms in inflammatory bowel disease.
- The polyarticular arthritis seen in IBD is usually correlated with disease activity and associated with erythema nodosum.
- HLA-B27—associated arthritis that occurs in IBD is usually clinically silent initially and presents in the third decade of life.

American Board of Pediatrics Content Specification(s):

- Recognize that arthritis may occur in patients with inflammatory bowel disease

Suggested Reading:

- Glick S, Carvalho R. Inflammatory bowel disease. Pediatr Rev. 2011;32(0:14-25. doi:10.1542/pir.32-1-14
- Kim S, Ferry G. Inflammatory bowel diseases in children. Curr Probl Pediatr. 2002;32:103-132. doi:10.1067/mps.2002.122638

Item 151

You are evaluating a 7-year-old girl whose mother reports that she is having difficulties in "keeping up" with her classmates. In kindergarten, she struggled with coloring and drawing during art projects; lagged behind other students in learning her alphabet, counting, and identifying colors; and produced schoolwork described as "sloppy." Her first-grade teacher finds the girl to be socially appropriate but reports that she struggles with reading skills and math. She has a number of close friends at school and in the neighborhood. According to her mother, the child appears to understand concepts and retain them for a few weeks but then forgets them and has to start over. She often tries to guess at words without sounding them out. Results of a physical examination and vision screening are within normal limits. When you talk with the girl, she is appropriately social and maintains good eye contact, but she exhibits some language problems. Specifically, she sometimes substitutes b sounds for d sounds.

Of the following, the MOST appropriate evaluation for this child is

A. developmental behavioral pediatric evaluation
B. neurologic evaluation
C. neuropsychiatric evaluation
D. occupational therapy evaluation
E. physical therapy evaluation

Item 151 SBP Preferred Response: C

The girl described in the vignette has difficulty with reading, math, fine motor skills, and retention of learned concepts. The most likely reason for her problems is a learning disability, or a mild global cognitive impairment, that may not have been initially detected because her good social skills made her appear less impaired. It is common for such problems to become more apparent as academic demands increase. The next best step in her evaluation would be a neuropsychiatric evaluation which will assess the domains of cognitive, attention, memory, processing, and academic information. This evaluation would be performed by a neuropsychologist. Results will reveal her cognitive strengths and weaknesses, and help define the most effective educational approach.

A similar but related evaluation would be a "psychoeducational" evaluation performed by a school psychologist or a child clinical psychologist. This evaluation is less comprehensive than a neuropsychiatric evaluation. Either a psycho-educational evaluation or a neuropsychiatric evaluation could delineate the type of learning difficulties seen in most children. However, more detailed neuropsychiatric testing may be needed to investigate more complex scenarios, such as an acquired brain function abnormality from illness or injury. For this girl, either approach would likely be beneficial.

An assessment by a developmental behavioral pediatrician would likely help to reconfirm the presence and general extent of her impairments. However, such an evaluation would not include the detailed educational plan or detailed assessments of cognition that will be needed to help this child in school. A neurologic evaluation would not be indicated at this time because of the absence of any specific neurologic signs or symptoms other than a mild fine motor skill delay. An occupational therapist might be able to provide some strategies for addressing her fine motor skill problems but would not address her learning difficulties. Similarly, a physical therapist might be useful if it was discovered that she had a treatable musculoskeletal problem related to her fine motor difficulties, but again, this would not address her primary learning and academic problems.

PREP Pearls

- Both neuropsychiatric and psychoeducational evaluations can delineate a child's specific learning disabilities and help define appropriate educational intervention strategies.
- Learning disabilities may not become apparent until academic demands in school increase.

AAP Mental Health Competency:

- Recognize what knowledge is gained from ordering "neuropsychiatric testing"

Suggested Reading:

- Handler SM, Fierson WM, et. al. Joint technical report—learning disabilities, dyslexia, and vision. Pediatrics. 2011;127(3):e818-e856. doi:10.1542/peds.2010-3670

- Silver CH, Blackburn LB, Arffa S, American Academy of Pediatrics Section on Ophthalmology and Council on Children with Disabilities, American Academy of Ophthalmology, American Association for Pediatric Ophthalmology and Strabismus, and American Association of Certified Orthoptists. The importance of neuropsychological assessment for the evaluation of childhood learning disorders: NAN Policy and Planning Committee. Arch ClinNeuropsychol. 2006;21:741-744

Item 152

You are called by a family who has just learned that an infant will become available for adoption. The mother is a late entrant into prenatal care. Results of maternal screening studies, including hepatitis B, human immunodeficiency virus, and rapid plasma reagin, are negative. Urine toxicology result is positive for marijuana and cocaine at the most recent prenatal visit at 30 weeks of gestation. The family is delighted about the upcoming adoption but asks if the illicit substances used by the mother during pregnancy may affect the infant.

Of the following, the MOST appropriate response is that the infant is at increased risk of

A. congenital heart disease
B. failure to thrive
C. moderate intellectual disability
D. neonatal drug withdrawal
E. preterm birth

Item 152 Preferred Response: E

Fetal cocaine exposure is associated with preterm birth, low birthweight, and small-for-gestational age size. This relationship exists after controlling for maternal confounders, including cigarette smoking, other drug exposures, lower socioeconomic status, and inadequate prenatal care. Cocaine exposure has been associated with a decrease in all fetal growth measurements, which worsens with advancing gestational age. Maternal cocaine use is postulated to have vasoconstrictive effects that contribute to placental insufficiency, infarction, and/or abruption. These events may contribute to the preterm birth, low birthweight, and small-for-gestational age size associated with infants exposed to cocaine prenatally.

No drug withdrawal syndrome has been formally described for cocaine. Cocaine is a central nervous system stimulant and may be found in the urine sample of an exposed infant up to 1 week after delivery. Although some infants exposed to prenatal cocaine will exhibit tremors and irritability 2 to 3 days after delivery, studies have not supported either a drug withdrawal syndrome or drug toxicity syndrome.

The effects of fetal cocaine exposure on the developing brain remain unclear. A recent review by Ackerman, et al, summarized existing data on school-aged children prenatally exposed to cocaine. After adjusting for environmental influences, exposed children have sustained attention and behavioral self-regulation deficits. Growth, cognitive ability, academic achievement, and language appear to be minimally affected. Brain imaging studies suggest minor effects on structure and function, but the study numbers are limited.

Cocaine has not been associated with major teratogenic malformations of the cardiac or genitourinary system. Although infants exposed to cocaine may be small for gestational age or of low birthweight, they typically catch up within 6 months and demonstrate small to no growth differences at school age. When environmental risk factors are removed, cognitive ability appears to be normal. Prenatal identification and support of cocaine-dependent mothers, with continued postnatal education and support may have a positive effect on the long-term development of at-risk infants.

PREP Pearls

- Fetal cocaine exposure is associated with preterm birth, low birthweight, and small-for-gestational age size.
- No drug withdrawal syndrome has been formally described for cocaine.

American Board of Pediatrics Content Specification(s):

- Know the association between the maternal use of cocaine and any fetal abnormalities and/or neonatal withdrawal syndrome

Suggested Reading

- Ackerman JP, Riggins T, Black MM. A review of the effects of prenatal cocaine exposure among school-aged children. Pediatrics. 2010;125:554565. doi:10.1542/peds.2009-0637. http://pediatrics.aappublications.org/content/125/3/554.full
- Behnke M, Smith VC; Committee on Substance Abuse, Committee on Fetus and Newborn. Prenatal substance abuse: short- and long-term effects on the exposed fetus. Pediatrics. 2013;131(3):e1009-e1024. doi:10.1542/peds.2012-3931
- Gouin K, Murphy K, Shah PS, et al. Effects of cocaine use during pregnancy on low birthweight and preterm birth: systematic review and metaanalyses. Am J Obstet Gynecol. 2011;204:340e1-e12. doi:10.1016/j. ajog.2010.11.013
- Hudak ML, Tan RC, the Committee on Drugs, the Committee on Fetus and Newborn. Neonatal drug withdrawal. Pediatrics. 2012;129:e540-e560. doi:10.1542/peds.2011-3212

Item 153

A 4-year-old boy is brought to the emergency department by his mother for easy bruising and recurrent nosebleeds. She reports that he was healthy until 2 weeks ago, when he had an upper respiratory illness. She denies fevers, weight loss, or change in activity for the boy but says she noticed large bruises all over his body over the past few days. Last night, he had 3 episodes of epistaxis, each lasting 10 minutes. His oral temperature is 36.2°C, pulse rate is 88 beats/ min, respiratory rate is 20 breaths/min, and blood pressure is 100/65 min Hg. On physical examination, he is alert and active. There are petechiae on his face and bruises of varying stages on his legs, arms, and back. There are several purpura on his oral mucosa. The remainder of the physical examination is normal. The following complete blood cell count values are obtained:

- White blood cell count, 8,500/µL (8.5×10^9/L) with 33% polymorphonuclear leukocytes, 57% lymphocytes, 7% monocytes, and 3% eosinophils
- Hemoglobin, 11.0 g/dL (110 g/L)
- Mean corpuscular volume, 75/μm^3 (75 fL)
- Platelet count, 3×10^3/µL (3×10^9/L)

Of the following, the MOST appropriate treatment for this patient is

A. γ-globulin intravenous
B. plasmapheresis
C. splenectomy
D. thrombopoietin receptor agonist
E. washed maternal platelets

Item 153 Preferred Response: A

The child described in the vignette has a presentation consistent with acute immune/idiopathic thrombocytopenia purpura (ITP). Immune/idiopathic thrombocytopenia purpura is characterized mainly by increased destruction of platelets via autoantibodies to platelet membrane glycoproteins. Additionally, emerging evidence suggests that a relative decrease in bone marrow production of platelet precursors may also play a role in ITP. The annual incidence is 1 in 10,000 children, making ITP the most common autoimmune cytopenia. In young children, ITP is usually associated with a preceding illness or vaccination, although direct causality has not been proven. The peak age at diagnosis is 2 to 6 years. Approximately 80% of children present with severe thrombocytopenia with platelet counts less than 20 x 103/4 (20 x 109/L). The white blood cell count and hemoglobin concentration are usually normal at presentation, although anemia is seen in 15% of cases, largely due to mucosal bleeding. Most patients are otherwise healthy and present suddenly with petechiae and bruising. Petechiae and oral bleeding are seen in up to 33% of cases, hematuria and gastrointestinal bleeding in up to 10% of cases, and intracranial hemorrhage in less than 1% of cases. Splenomegaly has been reported in 10% of cases of childhood ITP.

The diagnosis of ITP is made by history, physical examination, and a complete blood cell count. A review of the peripheral blood smear is necessary in all cases of suspected ITP. However, according to the 2011 American Society of Hematology (ASH) Evidence-Based Practice Guideline for Immune Thrombocytopenia, a bone marrow examination is not routinely recommended for patients who have a presentation typical for ITP (ie, isolated thrombocytopenia without other cytopenias, no adenopathy, and no bone pain).

Acute ITP most often occurs in children younger than 10 years and is benign and self-limited. Large studies of children with ITP show that 76% of children have complete remission within 6 months from the time of diagnosis. Another 37% of the remaining patients will eventually have complete remission. Chronic ITP, which occurs in 15% to 20% of children with ITP, is defined by the persistence of thrombocytopenia for more than 6 or 12 months, depending on consensus group. Infants and adolescents are more likely to have chronic ITP associated with other immune disorders, such as common variable immune deficiency or a rheumatologic disorder.

The need to treat children with ITP is based on the severity of bleeding symptoms. The goal of therapy in ITP is to achieve a platelet count that allows adequate hemostasis, rather than a normal platelet count. There is no evidence that treatment will change the natural course of the disease or decrease the incidence of intracranial hemorrhage. The ASH guideline recommends that children with no bleeding or mild bleeding (ie, bruising and petechiae) be managed with observation alone, regardless of the platelet count. For children with more significant bleeding symptoms, first-line treatment options are a single dose of γ-globulin intravenous (IVIG), 1 g/kg, or a short course of corticosteroids. IVIG is useful if a more rapid increase in platelet count is necessary. Therefore, the child in the vignette who is having recurrent epistaxis should receive one of these first-line therapies.

Plasmapheresis is not indicated as first-line therapy in acute childhood ITP. It is a blood purification technique to remove large-molecular-weight substances from the plasma, such as immune complexes, cryoglobulins, myeloma light chains, endotoxin, and cholesterol-containing lipoproteins. Although it has a role in thrombotic thrombocytopenic purpura (more commonly seen in adults), there are only rare case reports of its use in refractory adult ITP with variable success.

Splenectomy can be effective therapy for chronic or refractory ITP, with a success rate of 70% to 80%. However, given the high incidence of complete remission in children within 12 months of diagnosis, splenectomy should be reserved for chronic severe ITP.

Thrombopoietin receptor agonists (eg, romiplostim and eltrombopag) are approved by the Food and Drug Administration for chronic ITP in adults who have not responded to IVIG, corticosteroids, or splenectomy. There are clinical trials investigating the use of these agents in pediatrics, but there is no consensus recommendation for its use in children.

Washed maternal platelets are recommended in the treatment of neonatal alloimmune thrombocytopenia, a condition in which there is maternal immunization against fetal platelet antigens inherited from the father. Platelet transfusions are not effective in ITP because the circulating autoantibodies are likely to consume the transfused platelets.

PREP Pearls

- Immune/idiopathic thrombocytopenia purpura (ITP) is the most common immune-mediated cytopenia in young children.
- The need to treat children with ITP is based on the severity of bleeding symptom. The goal of therapy is to achieve a platelet count that allows adequate hemostasis, rather than a normal platelet count.
- 76% of children have complete remission within 6 months from the time of diagnosis.

American Board of Pediatrics Content Specification(s):

- Plan the appropriate management of a patient with ITP

Suggested Reading:

- Consolini D. Thrombocytopenia in infants and children. Pediatr Rev. 2011;32:135-151. doi:10.1542/pir.32-4-135
- Imbach P, Crowther M. Thrombopoietin-receptor agonists for primary immune thrombocytopenia. N Engl J Med. 2011;365:734-741. doi:10.1056/ NEJMct1014202
- Neunert C, Lim W, Crowther M, Cohen A, Solberg L, Crowther MA. The American Society of Hematology 2011 evidence-based practice guideline for immune thrombocytopenia. Blood. 2011;117:4190-4207. doi:10.1182/ blood-2010-08-302984
- Wilson DB. Acquired platelet defects. In: Nathan and Oski's Hematology of Infancy and Childhood. 7th ed. Philadelphia, PA: Saunders Elsevier; 2009:1553-1590

Item 154

A 7-year-old girl has attention-deficit/hyperactivity disorder (ADHD) that has not been treated with medication. She has been homeschooled, but her parents are planning to enroll her in public school next month. The girl tells you that she is worried about making friends in her new school. After discussion with parents about management options for ADHD, you prescribe methylphenidate. Two weeks later, she develops eye-rolling episodes that last seconds at a time. She can transiently suppress the movements, but then they come back in a flurry. She tells you she has a sense of relief after she does them. There is no alteration of consciousness with the episodes of eye rolling. She has never had a seizure.

Of the following, the MOST likely diagnosis is

A. absence seizure
B. motor tic
C. somatoform disorder
D. stimulant-induced movement disorder
E. Tourette syndrome

Item 154 Preferred Response: B

The girl described in the vignette has a motor tic. Motor tics are repetitive, stereotyped movements, usually of the eyes, face, head, or upper extremities. Blinking, eye-rolling, and grimacing are common motor tics. Vocal tics can include coughing, snorting, or throat clearing. Tics are associated with an internal urge to perform the movement and a sense of relief afterwards. They can be transiently suppressed and like most movement disorders, abate during sleep. Tics are a movement disorder thought to originate in the basal ganglia. They can increase in frequency or intensity in situations of anxiety, fatigue, or sometimes, when a stimulant medication is introduced in a person who has an underlying tic disorder.

It is important to counsel families that stimulants may exacerbate tics but do not cause them. Children with tics may have comorbid attention disorders or obsessive-compulsive disorder, and these should be treated if they have a significant negative effect on the child.

Some medications can directly cause movement disorders. Antidopaminergic medications such as antipsychotics (eg, haloperidol, risperidone) or some antiemetics (eg, metoclopramide, promethazine) are examples. Antidopaminergics can cause acute dystonia, which can be treated with an antihistamine such as diphenhydramine. Rarely, patients can develop a tardive dyskinesia because of antidopaminergic medications. Tardive dyskinesia is an involuntary, repetitive, twisting, or writhing movement that can affect the face, mouth, jaw, tongue, trunk, or limbs. It is very difficult to treat.

Absence seizures are characterized by a brief alteration of consciousness, lasting 5 to 10 seconds, often with eyelid fluttering, then an immediate return to normal consciousness. Somatoform disorder presents with varied symptoms that often evolve until the diagnosis is made and treatment is started. As stated before, stimulants may lead to exacerbation of an underlying tic disorder, but they do not induce tics in a child who does not have them. Tourette syndrome is characterized by persistent motor and vocal tics in a child, lasting more than a year. The girl in the vignette has had tics for only 2 weeks, so her tics do not meet the criteria for this diagnosis yet.

PREP Pearls

- Stimulants do not cause tics, although they may exacerbate them.
- Antidopaminergics can cause acute dystonia or chronic tardive dyskinesia.

American Board of Pediatrics Content Specification(s):

- Know which drugs can cause movement disorders

Suggested Reading:

- Augustine EF, Mink JW. Movement disorders. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:2053-2061
- Ryan CA, Gosselin GJ, DeMaso DR. Habit and tic disorders. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:75-77e1

Item 155

A 15-year-old female soccer player presents to your office for evaluation 1 day after an ankle injury. She reports an inversion injury that occurred while landing from a jump. Following the injury, she was able to bear weight. However, she has had a limp since the injury occurred. She is currently able to ambulate with minimal pain. On physical examination, you note moderate swelling and bruising over the lateral malleolus and tenderness just anterior and inferior to the lateral malleolus. Ankle range of motion is limited in all directions because of pain.

Of the following, the MOST appropriate next step in management is

A. begin proprioceptive training exercises in anticipation of return to soccer
B. cast or walker boot immobilization
C. crutch ambulation with no weight-bearing
D. obtain magnetic resonance imaging of the ankle
E. use of an air-stirrup brace and active range-of-motion exercises

Item 155 Preferred response: E

The girl described in the vignette suffered an acute inversion injury and has tenderness anterior and inferior to the lateral malleolus. History and physical examination strongly suggest a sprain of the anterior talofibular ligament (ATFL), the most commonly sprained ankle ligament. The majority of ankle sprains involve the lateral ankle ligaments, including the ATFL. Injuries to the syndesmosis (high ankle sprains) and medial ankle ligament complex (deltoid ligament sprains) occur less frequently than lateral sprains but tend to result in a more prolonged recovery. Ankle sprains are the most common sports injury, with an estimated 600,000 people seeking treatment in US emergency departments each year. The peak incidence of ankle sprains occurs in individuals between 15 and 19 years of age. Basketball, football, and soccer are the sports that most commonly result in ankle sprain.

Initial management of ankle sprains should include rest, ice, compression, and elevation (RICE). Early weight-bearing and range-of-motion exercises appear to shorten the recovery and result in earlier return to activity. Crutch ambulation and/or immobilization of the ankle are indicated only for individuals with more severe sprains who cannot tolerate weight bearing because of pain. The patient in the vignette does not have significant pain with weight bearing and should be able to ambulate with an air-stirrup brace. This type of brace allows an individual to plantarflex and dorsiflex the ankle with ambulation while providing stability and preventing reinjury. Once the patient has regained normal or near-normal range of motion she can progress with more advanced rehabilitation, including strengthening exercises and proprioceptive training. Patients who fail to complete a comprehensive ankle rehabilitation program are at higher risk for persistent pain and functional limitations. For patients with lateral ligament injuries, radiographs of the ankle should be obtained in the presence of significant bony tenderness or inability to bear weight following injury. Magnetic resonance imaging is not indicated in patients with a history and physical examination indicative of an acute lateral ankle ligament sprain. Referral to a sports medicine provider should be considered if the patient has continued pain or instability despite an adequate course of ankle rehabilitation.

PREP Pearls

- Lateral ankle ligament injuries are among the most common sports injuries.
- When tolerated, early ambulation and early mobilization of the ankle decrease recovery time in patients who have lateral ankle ligament sprains.

American Board of Pediatrics Content Specification(s):

- Plan the treatment of uncomplicated ankle injuries
- Know when to treat or refer an athlete with an uncomplicated ankle injury

Suggested Reading:

- Kerkhoffs GM, Rowe BH, Assendelft WJ, Kelly K, Struijs PA, van Dijk CN. Immobilisation and functional treatment for acute lateral ankle ligament injuries in adults. Cochrane Database Syst Rev. 2002(3):CD003762. doi:10.1002/14651858.CD003762
- Seah R, Mani-Babu S. Managing ankle sprains in primary care: what is best practice? a systematic review of the last 10 years of evidence. Br Med Bull. 2011;97:105-135. doi: 10.1093/bmbildq028
- Waterman BR, Owens BD, Davey S, Zacchilli MA, Belmont PJ, Jr. The epidemiology of ankle sprains in the United States. J Bone Joint Surg Am. 2010;92(13):2279-2284. doi:10.2106/JBJS.I.01537

Item 156

A 1-day-old female infant was noted to have lethargy, diaphoresis, and jitteriness. Within 10 minutes, the infant had a brief seizure, and her initial glucose measurement was 25 mg/dL (1.39 mmol/L) prior to receiving a bolus of intravenous (IV) glucose. Initial electrolyte measurements were within normal limits except for the hypoglycemia, and her mental status improved shortly after the glucose infusion. She was transferred to the intensive care nursery for further management. She was born after a pregnancy complicated only by gestational diabetes that was diagnosed in the late second trimester; maternal glucose was managed with diet alone. She was born at term, weighed 3 kg, and did well initially in the regular nursery. Upon weaning her IV glucose infusion, her heel stick glucose level fell again to less than 40 mg/dL (2.2 mmol/L), and a continuous IV glucose infusion was restarted. Blood drawn at the time of her second hypoglycemic episode was sent for glucose, insulin, ketones, and free fatty acid levels. Serum glucose level was 37 mg/dL (2.051 mmol/L), insulin was 15 µlU/mL (104 pmol/L) (normal range, 5 to 20 µlU/mL [35-139 pmol/L]), ketones were absent, and free fatty acids were within normal limits. Since that time she has required continuous IV glucose to maintain euglycemia.

Of the following, the MOST likely diagnosis for this infant according to her presentation and laboratory findings is

A. Beckwith-Wiedemann syndrome
B. cortisol deficiency
C. a fatty acid oxidation defect
D. hypoglycemia secondary to maternal diabetes
E. hyperinsulinism

Item 156 S Preferred Response: E

The infant described in this vignette has a typical presentation for hyperinsulinism, which includes hypoglycemia in the absence of ketones or reducing substances and a normal insulin value that is inappropriate for the degree of hypoglycemia. A plasma insulin level greater than 13 [μIU/mL (90 pmol/L) with a glucose level of less than 40 mg/dL (2.2 mmol/L) confirms this diagnosis. Persistent hyperinsulinism as seen in this infant who requires continuous intravenous glucose to maintain euglycemia may be caused by either a pancreatic islet cell adenoma or focal islet cell hyperplasia.

Infants with hyperinsulinism are more susceptible to neurologic deficits due to the hypoglycemia because insulin inhibits lipolysis and ketogenesis that can create alternate fuel sources for the central nervous system when glucose is unavailable. Such patients may require high glucose infusion rates to maintain blood sugars above 60 mg/dL (3.3 mmol/L). The first line of treatment includes one or more of the following medications: diazoxide, octreotide, and calcium channel blockers. Only if medical therapy fails should partial pancreatectomy be considered. However, close to 80% of infants who have persistent hyperinsulinemic hypoglycemia do not respond to medical therapy alone.

Beckwith-Wiedemann syndrome (BWS) may be associated with hypoglycemia in the newborn period secondary to hyperinsulinism but is generally seen in infants who are large for gestational age and have other physical characteristics, such as macroglossia and linear creases on the ear lobe with or without an omphalocele. Hypoglycemia in BWS is treated with hydrocortisone, which is usually only required until a few months of age. Inappropriately elevated insulin levels in the face of hypoglycemia would not be seen with cortisol deficiency or a fatty acid oxidation defect. Although hypoglycemia secondary to maternal diabetes would also present in the early neonatal period, it would be transient, usually resolve in 2 to 4 days, and generally not require continuous intravenous glucose infusions. Hypoglycemia secondary to maternal diabetes is frequently associated with macrosomia, which is not seen in the infant in the vignette.

PREP Pearls

- Although persistent hyperinsulinemic hypoglycemia is a relatively rare cause of hypoglycemia in the neonate, recognition and prompt treatment are essential to preclude neurologic sequelae.
- The diagnosis of hyperinsulinemic hypoglycemia can be made if during an episode of hypoglycemia the simultaneous insulin level is greater than 13 μIU/mL.

American Board of Pediatrics Content Specification(s):

- Recognize the signs and symptoms of hyperinsulinism

Suggested Reading:

- de Vroede M, Bursgaard K, Dunne MJ, Groenendaal F. Laparoscopic diagnosis and cure of hyperinsulinism in two cases of focal adenomatous hyperplasia in infancy. Pediatrics. 2004;114(4):e520-e522. doi:10.1542/peds.2003-1180-L
- Kelly A, Tang R, Becker S, Stanley CA. Poor specificity of low growth hormone and cortisol levels during fasting hypoglycemia for the diagnoses of growth hormone deficiency and adrenal insufficiency. Pediatrics. 2008;122;e522-e528. doi:10.1542/peds.2008-0806
- Menni F, de Lonlay P, Sevin C, et al. Neurologic outcomes of 90 neonates and infants with persistent hyperinsulinemic hypoglycemia. Pediatrics. 2001;107(3):476-479. doi:10.1542/peds.107.3.476
- Rotenstein D, Servin S, Walsh T. Palliative treatment of hyperinsulinism with cyproheptadine and diazoxide. Pediatrics. 1992;90(2):212-215

Item 157

You are serving as a camp physician when you are called to evaluate a 12-year-old boy who was bitten on the right forearm by a snake as he was gathering firewood. One of the camp counselors saw the snake and identified it as a copperhead.
On examination, the boy is anxious but in no acute distress. He complains of right forearm pain and nausea, but he is not vomiting. He denies dizziness or paresthesias. You note 2 puncture wounds on his right forearm, just distal to the elbow. There is moderate swelling and erythema surrounding these wounds.
The camp counselor has already called emergency medical services and cleansed the boy's wound. You begin preparing the boy for ground transport to a hospital located 40 minutes away.

Of the following, the MOST appropriate next step in management is

A. administration of oral diphenhydramine
B. application of a tourniquet just proximal to the wound
C. application of ice packs to the site of the wound
D. incision and mechanical suctioning of the wound
E. splinting of the affected extremity in a position of comfort

Item 157 S Preferred Response: E

The camper in the vignette requires transport to a hospital for emergency medical care after being bitten by a snake identified as a copperhead. Of the options given, splinting of the affected extremity in a position of comfort is the most appropriate next step in management.

In North America, approximately 2,500 children experience a poisonous snake bite annually. Most (>95%) of these bites are from members of the Crotalidae family of snakes, which includes rattlesnakes, copperheads, and water moccasins. Because children have smaller limbs and less subcutaneous tissue, the clinical effects of envenomation by poisonous snakes are generally more severe than those seen in adults. All pediatric care practitioners should understand the potential clinical manifestations of snake envenomation and the initial steps in management.

Determining whether a snake is venomous can be difficult, particularly in an emergency situation. Venomous snakes in the United Stated typically have triangular-shaped heads, elliptical-shaped pupils, and hollow, retractable fangs. In contrast, nonvenomous snakes characteristically have rounded heads, round pupils, and lack fangs. Because misidentification can have potentially serious outcomes, expert consultation should be sought for all patients with possible snake envenomation, and close clinical observation is recommended. Attempts to identify the snake should never put the patient or caregivers at risk of further injury and should never delay transport of the patient to a medical facility Digital photographs taken at a safe distance may be useful.

A wide spectrum of clinical findings may result from snake envenomation in children, including the following:

- Significant pain from the moment of envenomation
- Local tissue damage, including progressive soft tissue swelling and ecchymosis
- Nonspecific systemic symptoms, including nausea, vomiting, diarrhea, weakness, light-headedness, diaphoresis, and chills
- Coagulopathy
- Rhabdomyolysis, with potential for acute renal injury
- Increased vascular permeability, with resultant tachycardia and hypotension
- Neurologic sequelae, including altered mental status, seizures, metallic taste sensation, paresthesias, and muscle fasciculations

Definitive treatment of any child with a suspected Crotalidae bite should be based on symptoms and clinical findings. The following strategies should be used, however, in the initial prehospital management of all children with suspected snake envenomation:

- The child should be moved to safety and kept warm, calm, and still.
- The child's injured body part(s) should be immobilized in a functional position and kept at the level of the heart if possible.
- Any constrictive clothing or jewelry should be removed from the affected extremity.
- The bite wound should be cleansed.

- The child should be transported in a supine position to the nearest medical facility as soon as possible, preferably via emergency medical services.

Diphenhydramine has no proven benefit in the management of snake bites and could confound the child's clinical assessment due to its sedating properties. Therefore, administration of oral diphenhydramine would not be a recommended step in management of this patient.

Traditional management techniques, including placement of tourniquets proximal to the bite wound, pressure immobilization, application of ice to the bite, incision and oral suction of the bite wound, and mechanical suctioning, have not been reported to be effective and are not recommended. These "remedies" may actually cause harm to snake bite patients and should therefore be avoided.

Analgesics and supportive care, including intravenous fluids, should be provided for affected children as required. Tetanus status should be assessed and updated. Administration of antivenom may be required in some patients based on the severity of their symptoms, clinical manifestations, and laboratory findings. Antivenom therapy is most effective when given within 6 hours of envenomation. Consultation with a medical toxicologist or other physician(s) experienced in managing poisonous snake bites is recommended before antivenom administration. Telephone consultation with a medical toxicologist is always available in the United States through a regional poison control center, which can be accessed by calling 1-800-222-1222.

PREP Pearls

- Any child who is bitten by a potentially venomous snake should be moved to safety and kept as warm, still, and calm as possible.
- The child's injured body parts should be immobilized in a functional position and kept at the level of the heart if possible. Any constrictive clothing or jewelry should be removed.
- Traditional management techniques, including placement of tourniquets proximal to the bite wound, pressure immobilization, application of ice to the bite, incision and oral suction of the bite wound, and mechanical suctioning, are not recommended.

American Board of Pediatrics Content Specification(s):

- Plan the management of a snake bite

Suggested Reading:

- Bond GR. Snake, spider, and scorpion envenomation in North America. Pediatr Rev. 1999;20147. doi:10.1542/pir.20-5-147
- Cheng AC, Seifert SA. Management of Crotalinae (rattlesnake, water moccasin [cottonmouth], or copperhead) bites in the United States. UptoDate. Available online only for subscription

Item 158

The parents of a 4-year-old girl request that you complete a preschool medical form. They report that she has some problems with articulation and they hope she will qualify for speech therapy through the school. She has been healthy and her physical examination is completely unremarkable. You are not able to understand all of her speech perfectly. The child is very cooperative and your nurse attempts routine screening audiometry. However, despite the nurse's best efforts, the results are equivocal.

Of the following, the BEST next step in the evaluation of the child's hearing is

A. assessment for hearing aid placement
B. reassurance
C. referral to an audiologist
D. referral to an otolaryngologist
E. sedated auditory brainstem evoked potentials

Item 158 TE SBP Preferred Response: C

Mild hearing loss can be a silent handicap, and hearing loss that is not detected or managed appropriately may result in speech, language, or cognitive delays. Therefore, any patient with equivocal or failed office audiometry should be referred for formal audiology assessment. Reassurance is not appropriate management. An audiologist will perform more sophisticated testing in a soundproof environment to determine the specifics of type and degree of hearing loss. Appropriate testing is determined by the age and ability of the patient, availability of equipment and environment, and skill of the test administrator. Behavioral testing, used for those who may not be able to cooperate to the level needed to accomplish pure tone or speech audiometry, would be the next step for the girl in the vignette. In conditioned play audiometry, used to evaluate hearing in children between the ages of 30 months and 5 years, the child is taught to perform a simple task each time a sound is heard. Visual reinforcement audiometry is used to evaluate younger children, 6 months to 3 years of age, by visually rewarding with lighted or animated toys each time the child turns his or her head toward the sound source.

Screening audiometry is routinely performed in physician offices at health supervision visits for children 3 to 4 years and older. The goal is to identify individuals who may have developed hearing impairment that is likely to interfere with communication and educational achievement. Pure tone audiometry, speech audiometry, or otoacoustic emission (OAE) testing may be performed in a quiet room by trained office personnel. Pure tone audiometry measures the ability to hear pure tones of various frequencies as a function of the intensity measured in decibels. It involves determining the softest decibel threshold at which the patient can hear a sound 50% of the time for each tested frequency. Normal hearing threshold is 0 to 20 dB. Speech audiometry is similar to the pure tone air conduction thresholds in that it measures the lowest decibel that patients can identify or repeat words across frequencies. Accurate results for both of these tests depend on cooperation from the patient and a quiet testing environment, so the busy office setting may limit the ability to perform well in some patients. Otoacoustic emission testing measures the presence and strength of low-intensity sound produced by the cochlea in response to an acoustic stimulus. Testing does not require a behavioral response from the child, so it can be used in children of all ages, including newborns. An abnormal OAE test result may be due to cochlear dysfunction or conductive hearing loss.

In 2008, the US Preventive Services Task Force (USPSTF) recommended universal newborn hearing screening, with the hope of improving early detection of congenital hearing loss and timely intervention. The auditory brainstem response (ABR) test, OAE test, or both may be used for 1- or 2-stage hearing screening in newborns. The USPSTF recommendations include the following: screening all newborns before 1 month of age, audiologic assessment by 3 months of age of all infants who fail their newborn screening test, and individualized intervention by 6 months of age for those with significant hearing impairment. In addition to these efforts to improve identification and treatment of congenital hearing loss, it remains important to perform hearing screens at regular intervals for those infants who are at risk for progressive or late-onset hearing loss. Risk factors include in utero infection, such as cytomegalovirus, herpes simplex virus, Toxoplasma, rubella virus, or Treponema (syphilis); neonatal hyperbilirubinemia;

postnatal bacterial meningitis; craniofacial abnormalities; head trauma; prolonged exposure to aminoglycosides; or known syndrome, family history, or stigmata of conditions associated with hearing loss.

The ABR test uses click and tone burst stimuli to evoke electroencephalographic waveform responses from the auditory pathway as recorded using electrodes placed on the head and earlobes of the child. The test may be completed on a patient of any age. It is most often used in newborns, young infants, or older patients who are not able to cooperate with behavioral testing because of intellectual disability. Because movement can disrupt the ABR results, sedation may be required for more accurate testing. The practitioner must take this into consideration when deciding whether to use this technique.

Referral to an otolaryngologist or for hearing aid evaluation would be warranted only after documentation of hearing loss by audiologic evaluation.

PREP Pearls

- The accuracy of screening audiometry depends on cooperation from the patient and a quiet testing environment.
- Any patient with equivocal or failed office audiometry should be reevaluated or referred for formal audiology assessment.
- Mild to moderate hearing loss is subtle and may result in speech, language, or cognitive delays if not detected or managed appropriately.

American Board of Pediatrics Content Specification(s):

- Know the limitations of screening audiometry

Suggested Reading:

- Adcock LM, Freysdottir DF. Screening the newborn for hearing loss. UptoDate. Available online only for subscription
- Gifford KA, Holmes MG, Bernstein HH. Hearing loss in children. Pediatr Rev. 2009;30:207-216. doi:10.1021/bi00145a016
- Gregg RB, Wiorek LS, Arvedson JC. Pediatric audiology: a review. Pediatr Rev. 2004;25:224-233
- Smith RJH, Gooi A. Evaluation of hearing impairment in children. UptoDate. Available online only for subscription
- Sokol J, Hyde M. Hearing screening. Pediatr Rev. 2002;23:155-162. doi: 10.1542/pir.23-5-155

Item 159

A 2-year-old girl presented to the urgent care center for rash and swelling of her lips. While visiting a neighbor, the girl took a few bites of a peanut butter cracker and started coughing. The mother initially thought that her daughter had choked on the cracker but then noticed that the girl had developed hives on her face. The mother wiped her daughter's face and mouth and gave her a teaspoon of diphenhydramine. The hives began to fade, but the girl then developed lip swelling and a raspy voice. She became progressively irritable and now appears to be breathing fast, although she is also crying.

Of the following, the MOST appropriate step in this girl's Management is to administer

A. injectable corticosteroid
B. injectable epinephrine
C. nebulized albuterol
D. ranitidine and diphenhydramine
E. scheduled diphenhydramine

Item 159 S Preferred Response: B

The child described in the vignette is having an anaphylactic reaction to peanut butter and the most appropriate treatment is immediate administration of injectable epinephrine.

Anaphylaxis is a progressive, potentially life-threatening reaction involving multiple organs. It presents as one of 3 clinical scenarios: (1) acute onset (minutes to hours) with involvement of the skin and/or mucosal tissue and at least 1 of the following: respiratory compromise, reduced blood pressure, or symptoms of end-organ dysfunction; (2) two or more of the following that occur rapidly after exposure to a likely allergen for that patient: involvement of the skin/mucosal tissue, respiratory compromise, reduced blood pressure, or associated symptoms and/or persistent gastrointestinal symptoms; or (3) reduced blood pressure after exposure to a known allergen (Item C159A, page C-124). The time-line of resolution of an anaphylactic reaction may range from minutes to hours (uniphasic); there may be recurrence after an initial improvement of symptoms, usually approximately 6 to 8 hours after the first reaction (biphasic); or the resolution may be protracted. Signs and symptoms of anaphylaxis and frequency of their occurrence are summarized in Item C159B (page C-125).

The evidence-based approach to diagnosis and management of anaphylaxis has been well delineated in the practice parameters of the national allergy organizations and the World Allergy Organization guidelines. In addition, guidelines published by the National Institute of Allergy and Infectious Diseases provide instruction for the diagnosis and management of food-induced anaphylaxis. Epinephrine is the drug of choice and should be administered promptly at the onset of symptoms suggestive of anaphylaxis. Epinephrine is life-saving for many reasons: alpha-adrenergic vasoconstrictor effects in most body organ systems, ability to prevent and relieve airway obstruction, β-adrenergic inotropic and chronotropic properties on the heart, and ability to decrease mediator release from mast cells.

Second-line treatment options in the outpatient setting include β2-agonists, antihistamines, and glucocorticoids. Antihistamines (both H1 and H2 blockers) are second-line medications that can be given after epinephrine administration for control of cutaneous and cardiovascular manifestations. A combination of diphenhydramine and ranitidine has been shown to be more effective than diphenhydramine alone. However, these agents have a much slower onset of action than epinephrine and should never be used alone in the treatment of anaphylaxis. Glucocorticoids are not helpful acutely but have the potential to mitigate/prevent the biphasic reaction and recurrent/protracted anaphylaxis.

Nebulized albuterol can improve bronchospasm but will not alter the course of the reaction. The consensus of experts is that anaphylaxis treatment in order of importance is epinephrine, patient position, oxygen, intravenous fluids, nebulized therapy, vasopressors, antihistamines, corticosteroids, and other agents.

PREP Pearls

- Epinephrine is the treatment of choice in anaphylaxis because of its chronotropic and inotropic actions on the cardiovascular system, its ability to relieve bronchospasm, and its ability to mitigate degranulation of mast cells in response to an allergen challenge.
- Second-line treatment options in the outpatient setting include β2-agonists, antihistamines, glucocorticoids, and combinations of the above medications.

American Board of Pediatrics Content Specification(s):

- Know the signs and symptoms of anaphylaxis

Suggested Reading:

- Dinakar C. Anaphylaxis in children: current understanding and key issues in diagnosis and treatment. Curr Allergy Asthma Rep. 2012;12(6):641-649. doi:10.1007/s11882-012-0284-1
- Lieberman P, Nicklas RA, Oppenheimer J, et al. The diagnosis and management of anaphylaxis practice parameter: 2010 update [published correction appears in J Allergy Clin Immunol. 2010;126(6):1104]. I Allergy Clin Immunol. 2010;126(3):477-480.e1-42. doi:10.1016/j.jaci.2010.06.022
- Sampson HA, Mufioz-Furlong A, Campbell RL, et al. Second symposium on the definition and management of anaphylaxis: summary report-Second National Institute of Allergy and Infectious Disease/Food Allergy and Anaphylaxis Network symposium. J Allergy Clin Immunol. 2006;117(2):391-397. doi:10.1016/j.jaci.2005.12.1303
- Sponsored Expert Panel; Boyce JA, Assa'ad A, Burks AW, et al. Guidelines for the diagnosis and management of food allergy in the United States: report of the NIAID-Sponsored Expert Panel. J Allergy Clin Immunol. 2010;126(6 suppl):S1-S58. doi:10.1016/j.jaci.2010.10.007
- Simons FE, Ardusso LR, Bilo MB, et al; World Allergy Organization. World Allergy Organization anaphylaxis guidelines: summary. j Allergy Clin Immunol. 2011;127(3):587-593.e1-22. doi:10.1016/j.jaci.2011.01.038

Item C159B. Frequency of Occurrence of Signs and Symptoms of Anaphylaxis *†

Signs and Symptoms	Percent
Cutaneous	
Urticaria and angioedema	85-90
Flushing	45-55
Pruritus without rash	2-5
Respiratory	
Dyspnea, wheeze	45-50
Upper airway angioedema	50-60
Rhinitis	15-20
Dizziness, syncope, hypotension	30-35
Abdominal	
Nausea, vomiting, diarrhea, cramping pain	25-30
Miscellaneous	
Headache	5-8
Substernal pain	4-6
Seizure	1-2

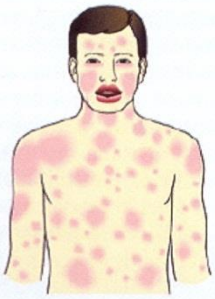
* Percentages are approximations.

† Children may have a lower frequency of cutaneous symptoms in anaphylaxis.

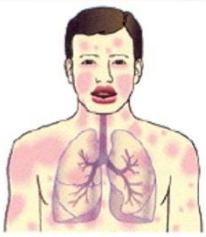
Reprinted with permission from Lieberman, et al. The diagnosis and management of anaphylaxis practice parameter: 2010 update. *J Allergy Clin Immunol*. 2010:126:480.e35

Anaphylaxis is highly likely when any one of the following three criteria is fulfilled:

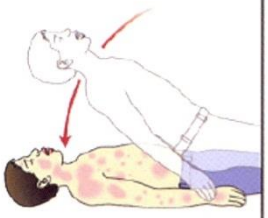
1 Sudden onset of an illness (minutes to several hours), with involvement of the skin, mucosal tissue, or both (e.g. generalized hives, itching or flushing, swollen lips-tongue-uvula)



AND AT LEAST ONE OF THE FOLLOWING:

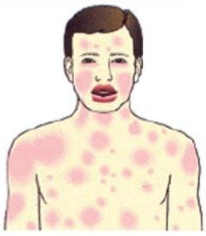


Sudden respiratory symptoms and signs (e.g. shortness of breath, wheeze, cough, stridor, hypoxemia)

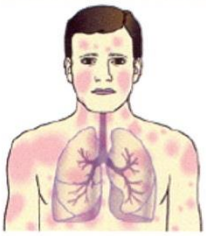


Sudden reduced BP or symptoms of end-organ dysfunction (e.g. hypotonia [collapse], incontinence)

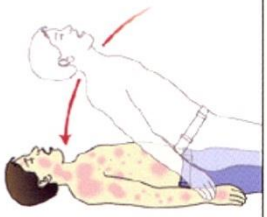
OR **2** Two or more of the following that occur suddenly after exposure to a ***likely allergen or other trigger**** ***for that patient*** (minutes to several hours):



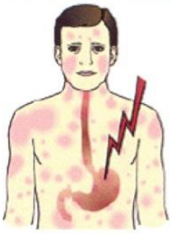
Sudden skin or mucosal symptoms and signs (e.g. generalized hives, itch-flush, swollen lips-tongue-uvula)



Sudden respiratory symptoms and signs (e.g. shortness of breath, wheeze, cough, stridor, hypoxemia)



Sudden reduced BP or symptoms of end-organ dysfunction (e.g. hypotonia [collapse], incontinence)



Sudden gastrointestinal symptoms (e.g. crampy abdominal pain, vomiting)

OR **3** Reduced blood pressure (BP) after exposure to a ***known allergen*** ** ***for that patient*** (minutes to several hours):

Infants and children: low systolic BP (age-specific) or greater than 30% decrease in systolic BP***

Adults: systolic BP of less than 90 mm Hg or greater than 30% decrease from that person's baseline

* For example, immunologic but IgE-independent, or non-immunologic (direct mast cell activation)

** For example, after an insect sting, reduced blood pressure might be the only manifestation of anaphylaxis; or, after allergen immunotherapy, generalized hives might be the only initial manifestation of anaphylaxis.

*** Low systolic blood pressure for children is defined as less than 70 mm Hg from 1 month to 1 year, less than (70 mm Hg + [2 x age]) from 1 to 10 years, and less than 90 mm Hg from 11 to 17 years. Normal heart rate ranges from 80-140 beats/minute at age 1-2 years; from 80-120 beats/minute at age 3 years; and from 70-115 beats/minute after age 3 years. In infants and children, respiratory compromise is more likely than hypotension or shock, and shock is more likely to be manifest initially by tachycardia than by hypotension.

Reprinted with permission from Simons FE, Ardusso LR, Bilò MB, et al; World Allergy Organization Anaphylaxis Guidelines: Summary. *J Allergy Clin immunol*. 2011;127(3):587-593.e1-22

ITEM C159A: Criteria for the diagnosis of anaphylaxis.

Item 160

A 10-year-old boy presents to your office with the chief complaint of having cola-colored urine for one day. His review of systems is significant only for a mild sore throat without fever 4 weeks ago. On physical examination, the boy has normal growth parameters. He has a respiratory rate to of 18 breaths/min, heart rate of 94 beats/min, and blood pressure of 138/90 mm Hg. The remainder of the physical examination findings is normal. A urine test strip analysis demonstrates a specific gravity of 1.015, pH of 5.5, 3+ blood, 2+ leukocyte esterase, and no protein or nitrites.

Of the following, the test MOST likely to offer both diagnostic and prognostic information about this boy's condition is

A. antistreptolysin O titer
B. C3 measurement
C. renal ultrasonography
D. urine culture
E. urine microscopy

Item 160 TE Preferred Response: B

The patient described in the vignette has clinical features of acute glomerulonephritis (GN; cola-colored urine and hypertension). Serum chemistries will likely reveal azotemia and dyselectrolytemia depending on the severity of renal failure. In addition to standard renal function analysis, further characterization of acute GN requires complement evaluation. Based on the complement levels, patients are categorized as having hypocomplementemic GN (associated with a low C3) or normocomplementemic GN (associated with a normal C3) (Item C160). Hypocomplementemic GN can be further characterized with additional historic and laboratory information. Hypocomplementemia usually resolves within 8 weeks in postinfectious GN (PIGN). Persistently low C3 level is associated with an increased risk for membranoproliferative GN and possibly a poorer prognosis.

Postinfectious GN is characterized by an immune-complex-mediated nephritis after an infectious process. Post-streptococcal GN (PSGN) exclusively relates to a group A, (β-hemolytic streptococcal infection. Therefore ASO titers will be elevated only when PIGN associated with strep infection. However, in PIGN secondary to nonstreptococcal infections, ASO titers will be normal. Initial urine microscopy shows hematuria, pyuria (glomerular inflammation), and RBC casts. Subsequent urine microscopy in patients with classic PIGN may show persistent microscopic hematuria (which may persist for up to 1 to 3 years in some cases). This has not been associated with worse prognosis in the patients.

Renal ultrasonography is usually suggestive of medical renal disease (enlarged echogenic kidneys) as may be seen in any form of acute renal injury. It does not distinguish between different forms of acute nephritis. Urine culture is not indicated in a patient with acute glomerulonephritis.

Item C160. Complement Levels and Different Causes of Acute Nephritis

Low C3 GN	Normal C3 GN
Postinfectious/Poststreptococcal	Immunoglobulin A GN
Membranoproliferative GN (more common)	Henoch–Schönlein purpura nephritis
Lupus nephritis	ANCA-associated GN
Shunt nephritis	Alport syndrome
Subacute bacterial endocarditis	Membranoproliferative GN (1/3 of cases)

Abbreviations: ANCA, Antineutrophil cytoplasmic antibodies; GN, glomerulonephritis.

PREP Pearls

- Acute glomerulonephritis is characterized by the triad of cola-colored urine, hypertension, and azotemia on serum chemistry. These patients may or may not have proteinuria with or without hypoalbuminemia.
- Serum complement evaluation is important for characterizing patients with acute nephritis. Patients with nephritis may have hypocomplementemic (associated with a low C3) or normocomplementemic (associated with a normal C3) glomerulonephritis.

American Board of Pediatrics Content Specification(s):

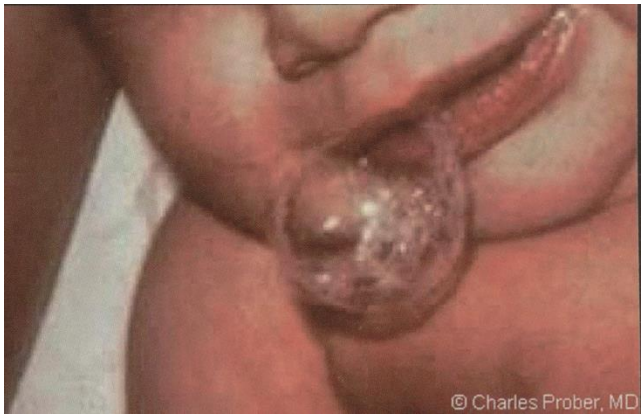
- Know the laboratory evaluation of acute post-streptococcal nephritis

Suggested Reading:

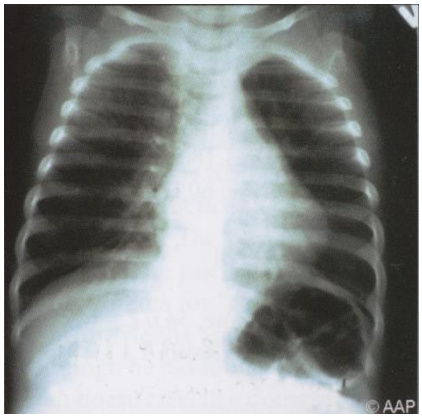
- Eison TM, Ault BH, Jones DP, Chesney RW, Wyatt RJ. Post-streptococcal acute glomerulonephritis in children: clinical features and pathogenesis. Pediatr Nephrol. 2011;26(2):165-180. doi:10.1007/s00467-010-1554-6
- Massengill SF. Hematuria. Pediatr Rev. 2008;29:342-348. doi:10.1542/pir.29-10-342
- Simckes AM, Spitzer A. Poststreptococcal acute glomerulonephritis. Pediatr Rev. 1995;16(7):278. doi:10.1542/pir

Item 161

A 6-week-old infant is seen in the emergency department with congestion, cough, and gagging that is resulting in facial cyanosis. The infant was born at term to a healthy 25-year-old mother with appropriate prenatal care and negative prenatal laboratory studies for infection. The patient's grandmother also has an illness characterized by cough. On physical examination, the infant's temperature is 37°C, heart rate is 135 beats/min, respiratory rate is 50 breaths/min, and blood pressure is 70/50 mm Hg. The infant is well developed and well nourished. She has episodes of coughing and gagging as you examine her, and the oxygen saturation decreases from 95% to 88% on room air during these episodes. She has thick oral secretions (Item Q161A). The anterior fontanelle is soft, open, and flat. There is nasal congestion, but the conjunctivae, oropharynx, and tympanic membranes are unremarkable. The patient has coarse breaths sounds on auscultation, but there are no retractions. The remainder of the physical examination is unremarkable. The peripheral white blood cell count is 55,000/µL (55.0 x 10^9/L), with 20% polymorphonuclear neutrophils and 80% lymphocytes. The patient's hemoglobin is 10 g/dL (100 g/L) and the platelet count is 260 x 10^3/µL (260 x 10^9/L). A chest radiograph is obtained (Item Q161B).



ITEM Q161A: Thick oral secretions as described for the infant in the vignette.



ITEM Q161B: Chest radiograph for the infant in the vignette.

Of the following, the test MOST likely to establish the patient's diagnosis is a

A. bacterial culture on routine media
B. direct fluorescent antibody test
C. polymerase chain reaction test
D. test for specific immunoglobulin M
E. viral culture using specific media

Item 161 Preferred Response: C

The infant described in the vignette has a respiratory illness characterized by cough, thick oral secretions, gagging, and facial cyanosis, as well as hyperinflation and infiltrate apparent on chest radiograph. These findings, in addition to the marked peripheral lymphocytosis and a grandmother who also is ill (source of infection), strongly suggest Bordetella pertussis as the cause of infection. The test most likely to establish the patient's diagnosis is a polymerase chain reaction (PCR) assay.

The PCR assay is increasingly used for the detection of pertussis because of excellent sensitivity and increasing availability. Although bacterial culture is considered the standard criterion for diagnosing pertussis, culture confirmation of illness can be difficult because (1) the organism is fastidious and requires special transport media and culture conditions (bacterial culture on routine media is not sufficient), (2) antibiotic therapy initiated in the patient can interfere with growth of the organism in culture, (3) previous immunization can result in a negative culture result, and (4) the result of a culture performed beyond 2 weeks into the illness can be negative. Overall, the sensitivity of bacterial culture is 20% to 60%. However, both PCR assay and bacterial culture are tests recommended by the Centers for Disease Control and Prevention (CDC) for the diagnosis of pertussis. Isolating B pertussis in culture allows for strain identification and testing for antimicrobial susceptibility, which can be especially useful in the setting of a pertussis outbreak. Growth in bacterial culture typically takes at least 1 week. Viral culture will not isolate the organism.

Specimens from the respiratory epithelium of the posterior oropharynx should be collected by aspiration (rarely available) or swab. Dacron swabs (with a flexible shaft) are preferred. Both rayon and cotton swabs contain substances that kill B pertussis, and calcium alginate interferes with the PCR assay.

There are no serologic tests for pertussis licensed by the Food and Drug Administration. However, commercial tests for serum IgG antibody to pertussis toxin exist. In addition, serologic testing is available through the CDC. Provided a patient has not been immunized against pertussis within the prior 2 years, a single elevated serum pertussis toxin IgG level weeks into illness can suggest recent infection. In addition, an increase in IgG titer between acute and convalescent sera can be used for diagnosis. Pertussis toxin IgM assays and direct fluorescent antibody testing are not recommended because they lack sensitivity and specificity.

PREP Pearls

- PCR testing is recommended for diagnosing pertussis because of its high sensitivity.
- Both PCR testing and bacterial culture are recommended by the CDC for diagnosing pertussis.
- Dacron swabs are recommended for obtaining a specimen from the posterior oropharynx for pertussis testing.

American Board of Pediatrics Content Specification(s):

- Know the diagnostic tests available for pertussis: isolation, polymerase chain reaction, serology, direct fluorescent antibody

Suggested Reading:

- American Academy of Pediatrics. Pertussis (whooping cough). In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:553-566
  Centers for Disease Control and Prevention. Best practices for health care professionals on the use of polymerase chain reaction for diagnosing pertussis
- Centers for Disease Control and Prevention. Pertussis (whooping cough), www.cdc.gov/pertussis

Item 162

A 16-year-old girl has been having unprotected sexual intercourse. When you discuss the need for contraception, the girl says she has no concerns about becoming pregnant. She has been taking care of her 17-year-old sister's child and believes that she knows "what it takes to be a mother," but she says she wants to complete her high school education. The medical student shadowing you asks what factor is most positively correlated with high school graduation for teen mothers.

Of the following, you are MOST likely to tell the medical student that the factor is:

A. having been raised in a large family
B. having parents with a lower educational level
C. the presence of reading material in the home
D. the teenage mother being unemployed
E. white race

Item 162 Preferred Response: C

Studies indicate that students who became parents as adolescents finish fewer years of formal education and have lower-level jobs as adults than those who do not become parents during adolescence. High school completion correlates closely with the presence of reading material in the home. Other factors associated with high school completion include black race (compared with white race), employment of the adolescent's mother, being raised in a smaller family, and having parents with higher educational levels.

A pregnancy during the adolescent years is associated with an increased risk of future poverty, marital instability, and additional pregnancies; as well as multiple psychosocial problems. It is controversial as to whether psychosocial problems predate the pregnancy or are a consequence of it. Teenage mothers, compared with older mothers, are more likely to have been raised in a less advantaged social environment with a single parent in a low-wage job and have low career aspirations themselves. Their mothers and sisters are also more likely to have borne children when they were adolescents. Disadvantage as a consequence of the pregnancy, however, is suggested by a study in Sweden, with a more homogenous population of higher socioeconomic status, which found that those who become mothers in their adolescent years also had a less favorable outcome than those who do not. As noted, a major risk factor associated with poor outcomes for any adolescent is interruption in education. This can be remediated and underscores the importance of enabling teenagers who want to carry the pregnancy to term to enroll in programs that help them get an education.

PREP Pearls

- Adolescent motherhood increases the risk for future poverty and social instability.
- High-school completion improves the outcome for mother and child.
- Encouraging reading at an early age may increase the odds of high-school completion.

American Board of Pediatrics Content Specification(s):

- Recognize the social, economic, and educational problems associated with teenage pregnancy

Suggested Reading:

- Dehlendorf C, Marchi K, Vittinghoff E, Braveman P. Sociocultural determinants of teenage childbearing among Latinas in California. Maternal Child Health J. 2010;14:194-201
- Gueorguieva RV, Carter RL, Ariet A, Roth J, Mahan CS, Resnick MB. Effect of teenage pregnancy on educational disabilities in kindergarten. Am J Epidemiol. 2001;154:212-220. doi:10.1093/aje/154.3.212
- Klein JD, American Academy of Pediatrics Committee on Adolescence. Adolescent pregnancy: current trends and issues. Pediatrics. 2005;116: 281286. doi:10.1542/peds. 2005-0999
- Sipsma HL, Ickovics JR, Lewis JB, Ethier KA, Kershaw TS. Adolescent pregnancy desire and pregnancy incidence. Womens Health Issues. 2011;21:110-116. doi:10.1016/j.whi.2010.09.004
- Stevens-Simon C, Lowy R. Teenage childbearing: an adaptive strategy for the socioeconomically disadvantaged or a strategy for adapting to socioeconomic disadvantage? Arch Pediatr Adolesc Med. 1995;149:912-915. doi:10.1001/archpedi.1995

Item 163

A 10-month-old boy presents to your office because of poor growth. He was born at 37 weeks' gestation and had a birth weight of 2,800 g. No problems were noted during the neonatal period, and early growth and development were normal. However, at the age of about 5 months, he was treated for purulent otitis media. Since that time, he has been treated for a second episode of otitis media and for lobar pneumonia. His parents note that he has 2 to 3 large stools per day and has failed to gain weight over the past few months. His developmental milestones are normal for age and family history is noncontributory. Physical examination of the alert, well-hydrated infant shows a weight of 8.2 kg, a length of 73 cm, a palpable liver edge 2.0 cm below the right costal margin, and several bruises on his extremities. Initial laboratory study results include the following:

- Hemoglobin, 9.8 g/dL(98 g/L)
- White blood cell count, 3,200/µL, with 15% neutrophils, 80% lymphocytes, and 5% monocytes
- Platelet count, 80 x 10^3/µL (80 x 10^9/L)
- Alanine aminotransferase, 60 U/L; reference range, <40 U/L
- Albumin, 3.8 g/dL (38 g/L)
- Prothrombin time, 12.5 s
- Partial thromboplastin time, 34.0 s
- International Normalized Ratio, 1.1

Of the following, the test that is MOST likely to suggest the correct diagnosis is

A. abdominal ultrasonography
B. liver-spleen scan
C. small-bowel radiographic series
D. sweat chloride
E. tissue transglutaminase antibody

Item 163 Preferred Response: A

The boy described in the vignette presents with poor growth and weight gain, a history of multiple episodes of otitis media, and a recent history of several large stools per day. His physical findings include hepatomegaly and extremity bruising, and laboratory studies indicate anemia, neutropenia, a low platelet count, and a mild increase in alanine aminotransferase. These data strongly suggest a clinical condition characterized by nutrient malabsorption and abnormal hematopoiesis. The disorder that manifests these findings is Shwachman-Diamond syndrome (SDS), a rare autosomal recessive disorder that is the second most com-mon cause of exocrine pancreatic insufficiency in childhood, after cystic fibrosis (CF). Shwachman-Diamond syndrome is also characterized by bone marrow dysfunction, skeletal abnormalities, and an increased leukemia risk. These children often present in early infancy with steatorrhea, growth failure, deficiencies of fat-soluble vitamins A, D, E, and K (a likely contributor to bruising in the present case), and symptoms arising from bone marrow dysfunction. However, SDS (unlike CF) is not associated with abnormalities in pancreatic ductular morphologic features or function. In fact, lipase excretion increases with age, and older patients often experience normalization of pancreatic function and fat absorption. Thus, in later childhood and adolescence, approximately 50% of patients who have SDS eventually do not require enzyme replacement therapy. Up to 90% of affected patients manifest specific genetic mutations at chromosomal locus 7q11. In patients with suspected cases of SDS, initial evaluation should include imaging studies, such as abdominal ultrasonography, computed tomography, or magnetic resonance imaging. These tests often confirm extensive fatty replacement of the pancreas, particularly in children older than 1 year of age.

Pancreatic insufficiency is defined as the loss of pancreatic exocrine function, resulting in reduced digestive enzyme output and the consequent failure of adequate intraluminal nutrient hydrolysis and absorption. Causes of pancreatic insufficiency in children are listed in Item C163, page C-128. With the notable exception of CF, most of these conditions are rare disorders. Nevertheless, an evaluation of exocrine pancreatic function should be considered for all patients who present with signs and symptoms suggestive of maldigestion or malabsorption.

In the evaluation of intestinal maldigestion or malabsorption, estimates of steatorrhea may be made by a quantitative fecal fat determination, which can only be determined from a 72-hour stool collection. This test is reserved for infants older than 6 months because of the normal state of relative pancreatic insufficiency in younger infants. Total stool output is collected in a dry, preweighed vessel in a 72-hour period, during which time the patient consumes a diet containing 3 g/kg of fat per day to a maximum of 100 g/d. A coefficient of fat absorption (total fat excreted divided by total fat intake) of less than 0.93 indicates malabsorption. The criterion standard for evaluation of exocrine pancreatic function is the determination of enzyme output after intravenous secretin stimulation. However, this study is cumbersome, involving endoscopic intubation of the duodenum, and difficult to perform accurately. Other tests are more widely used to identify the malabsorptive state, and several screening studies have been used to assess exocrine pancreatic function. The serum immunoreactive trypsinogen value is low in most cases of pancreatic insufficiency, including SDS, but is markedly elevated in most

infants who have CF, in whom exocrine pancreatic insufficiency is the consequence of pancreatic ductular obstruction, leading to "reflux" of enzyme into the bloodstream. In older CF patients, in whom progressive fibrosis and acinar cell destruction of the exocrine pancreas have occurred, serum trypsinogen concentrations are well below normal in 95% of cases. More recently, assay of fecal elastase has been used to evaluate pancreatic function. Pancreatic elastase is excreted in stool and is not affected by bacterial degradation. Exocrine insufficiency is suggested by a fecal elastase concentration of less than 200 .µg/g of stool. However, although the test is highly sensitive, it has decreased specificity when evaluating exocrine pancreatic insufficiency in children.

In evaluating a child like the one in the vignette, a sweat chloride test should be considered. In this case, however, the findings of bone marrow dysfunction and recurrent, purulent infections clearly rule against a diagnosis of CF. Neither a radionuclide liver-spleen scan nor a small bowel radiographic series would be helpful in diagnosing SDS, although the latter study may show evidence of bowel wall edema in patients with intestinal malabsorptive states (eg, celiac disease). In celiac disease, the clinical expression of disease includes growth failure and signs of fat soluble vitamin deficiency. However, bone marrow dysfunction and recurrent infections are not typical of this disorder; therefore, a tissue transglutaminase antibody assay result would be negative in this case.

Item C163. Conditions Associated With Pancreatic Insufficiency

- Cystic fibrosis
- Chronic pancreatitis
  - Autosomal-dominant hereditary pancreatitis
  - Anatomic causes
    - Annular pancreas
    - Pancreas divisum
  - Drug-induced (multiple agents)
  - Trauma
  - Metabolic disorders
  - α1-antitrypsin deficiency
  - Fatty acid oxidation defects
  - Lipoprotein lipase gene defects
  - Mitochondria! respiratory chain disorders
  - Organic acidemias
- Johanson-Blizzard syndrome
- Pearson syndrome
- Pancreatic agenesis or hypoplasia
- Shwachman-Diamond syndrome

PREP Pearls

- After cystic fibrosis, Shwachman syndrome is the most common cause of pancreatic insufficiency; 90% of patients demonstrate a mutation on chromosome 7 distinct from the Δ-F508 mutation seen in CF.
- Fatty infiltration of the pancreas, seen on ultrasonography, is characteristic of Shwachman syndrome.
- Patients with Shwachman syndrome have a heightened risk for leukemia.
- Shwachman syndrome should always be ruled out in patients with signs of malnutrition and bone marrow dysfunction.

American Board of Pediatrics Content Specification(s):

- Recognize that Shwachman syndrome is a cause of pancreatic insufficiency

Suggested Reading:

- Belamarich PF. Recognizing and diagnosing pancreatic insufficiency in infants. Pediatr Rev. 2002;23:69-70. doi: 10.1542/pir.23-2-69
- Burroughs L, Woolfrey A, Shimamura A. Shwachman-Diamond syndrome: a review of the clinical presentation, molecular pathogenesis, diagnosis, and treatment. Hematol Oncol Clin North Am. 2009;23:233-248. doi:10.1016/j.hoc.2009.01.007
- Ip WF, Dupuis A, Ellis L, et al. Serum pancreatic enzymes define the pancreatic phenotype in patients with Shwachman-Diamond syndrome. J Pediatr. 2002;141:259-265. doi:10.1067/mpd.2002.125849
- Mack DR, Forstner GG, Wilschanski M, Freedman MH, Durie PR. Shwachman syndrome: exocrine pancreatic dysfunction and variable phenotypic expression. Gastroenterology. 1996;111:1593-1602
- Shwachman H, Diamond L, Oski F, Khaw KT. The syndrome of pancreatic insufficiency and bone marrow dysfunction. J Pediatr. 1964;65:645-663. doi:10.1016/50022-3476(64)80150-5

Item 164

An 8-year-old boy has developed severe right ear pain. He has not had fever or respiratory symptoms. He has been taking swimming lessons daily for the last 2 weeks. One year ago he had tympanostomy tubes placed, but he has not seen a doctor since then and it is unknown if the tubes remain in place. Physical examination is notable for pain when the right pinna is pulled, concentric edema of the external ear canal, and yellow drainage in the canal. The tympanic membrane is obscured by the swelling and discharge, and when you attempt to clean the canal, the child experiences so much discomfort that you are compelled to stop.

Of the following, the MOST appropriate treatment for this child is

A. oral amoxicillin-clavulanate
B. topical 2% acetic acid
C. topical antipyrine-benzocaine
D. topical ofloxacin
E. topical polymyxin B, neomycin, and hydrocortisone

Item 164 Preferred Response: D

Otitis externa (OE), an inflammatory disease of the external auditory canal, is associated with infection with Pseudomonas aeruginosa and Staphylococcus aureus (most typically) and Candida (rarely). The patient may complain of tenderness, itching, discharge, or a sensation of fullness in the ear and occasionally decreased hearing. History often reveals exposure to warm, damp conditions; swimming is the classic predisposing event. Additional inciting factors include foreign bodies in the ear including hearing aids, dermatitis, and infection with herpes simplex or varicella. The most consistent physical finding is tenderness with manipulation of the tragus or pinna. Additional signs include edema of the ear canal and discharge or debris which together frequently obscure the tympanic membrane. The patient may also have peri- and preauricular lymphadenopathy, and rarely he/she will experience cranial nerve palsies or vertigo. Otitis media with tympanic perforation and discharge may resemble the findings of OE, but patients with this condition, unlike those with OE, often have unprovoked ear pain and fever, and they do not have tenderness with manipulation of the pinna or tragus. A patent tympanostomy tube also can be associated with drainage, and it may be difficult to differentiate OE from chronic otorrhea in this setting. However, the patient in the vignette had no prior episodes of ear drainage and his current symptoms are more typical of OE.

Treatment for OE consists of 3 potential categories of topical medications: antibiotics, steroids, and pH-lowering agents. Previously the agent of choice was polymyxin B/ neomycin/hydrocortisone with a demonstrated cure rate of approximately 90%. However, this agent has the potential for ototoxicity when in direct contact with middle ear structures, hypersensitivity to neomycin, pain with installation, and increasing bacterial resistance. The current preferred antibacterial topical agents are fluoroquinolones (ciprofloxacin or ofloxacin). A Cochrane review found that although either type of topical antibiotic is likely to result in similar cure rates, fluoroquinolones have several advantages over polymyxin B/neomycin/hydrocortisone. They are applied just twice daily, have a neutral pH that limits pain with application, and have not been associated with high levels of hypersensitivity. They are not associated with ototoxicity, so they can be used even if there is a question of tympanic membrane patency, as seen in the patient in the vignette.

Evidence on the efficacy of topical steroids for OE is limited, with 1 study showing a minimally earlier resolution of symptoms when steroids were used. Agents such as acetic acid, isopropyl alcohol, and boric acid work by lowering the environmental pH, which limits bacterial growth. The Cochrane review found that these agents were equally effective compared with antibiotic/steroid drops unless symptoms required treatment for more than 1 week; in that case antibiotic/steroid combinations were clearly superior. Use of these pH-lowering agents may cause potential ototoxicity when the tympanic membrane is disrupted.

Another treatment that may be considered is mechanically cleaning the ear canal. This is a labor-intensive therapy, is not readily available in most primary care offices, is potentially quite painful, and its efficacy has not been studied.

PREP Pearls

- Otitis externa is an infection most commonly caused by P aeruginosa and S aureus; a less common cause is Candida.
- Topical fluoroquinolones are the current treatment of choice because of absence of ototoxicity, low rate of hypersensitivity reactions, decreased pain during administration, and less frequent dosing intervals.
- Evidence for or against the use of topical steroids in the treatment of otitis externa is limited.

American Board of Pediatrics Content Specification(s):

- Know the treatment of swimmer's ear

Suggested Reading:

- Bradley JS, Jackson MA. Clinical report: the use of systemic and topical fluoroquinolones. Pediatrics. 2011;128:4e1034-e1045. doi: 10.1542/peds.2011-1496
- Kaushik V, Malik T, Saeed SR. Interventions for acute otitis externa. Cochrane Database Syst Rev. 2010;1. DOI:10.1002/14651858.CD004740.pub2 Stone KE. Otitis externa. Pediatr Rev. 2007;28:77-78. doi: 10.1542/pir.28-2-77

Item 165

A 3-year-old girl is seen in the emergency department 30 minutes after ingesting a large amount of her father's propranolol that was prescribed for the treatment of hypertension. You are working with a group of medical students who ask you what symptoms the patient may exhibit.

Of the following, the MOST likely symptom that would be seen is

A. hyperglycemia
B. hypertension
C. seizures
D. tachycardia
E. tachypnea

Item 165 Preferred Response: C

Beta-blocking agents are used to treat various adult and pediatric medical conditions, including hypertension, heart failure, ischemic heart disease, dysrhythmias, migraine headaches, and thyrotoxicosis. Cardiac side effects that can be seen at therapeutic doses are hypotension, sinus bradycardia, or heart block. Noncardiac effects can include increased airway resistance, hypoglycemia, and hyperkalemia. Especially in pediatric patients, these findings may be accompanied by respiratory depression and changes in mental status, including coma, delirium, and seizures.

Symptoms vary depending on the β-blocker ingested; propranolol is particularly likely to cause central nervous system symptoms including seizures. The onset of symptoms generally is within 2 hours of ingestion and almost always within .6 hours unless the drug is a sustained-release formulation. As a result, patients with a known or suspected ingestion of an overdose should be observed for 6 hours (24 hours for a sustained-release formulation). Symptomatic patients should be admitted to an intensive care unit for monitoring and treatment.

PREP Pearls

- β -blocker overdose can cause hypotension, sinus bradycardia, heart block, increased airway resistance, respiratory depression, hypoglycemia, and hyperkalemia.
- β -blocker overdose can produce changes in mental status, including coma, delirium, and seizures.
- Symptoms of (β -blocker overdose are almost always seen by 6 hours unless the drug is a sustained-release formulation.

American Board of Pediatrics Content Specification(s):

- Recognize common side effects of beta-blocking drugs

Suggested Reading:

- Lemkin E, Barrueto F. Beta blocker poisoning. UptoDate. Available online only for subscription
- O'Donnell KA, Ewald MB. Poisonings. In: Kliegman RM, Stanton BMD, St Geme J, Schor N, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Elsevier Saunders; 2011: 261
- Podrid PJ. Major side effects of beta blockers. UptoDate. Available online only for subscription

Item 166

An 18-month-old boy is brought to your office because his parents are concerned about bowed legs. History reveals he was late in learning to walk independently (just starting at 15 months). Laboratory testing reveals the following:

- Alkaline phosphatase, 862 U/L; normal range, 150-420 U/L
- 25-hydroxyvitamin D, 31 ng/mL (77 nmol/L); normal range, 20-50 ng/mL (50 to 125 nmol/L)
- Parathyroid hormone, 15 pg/mL (15 ng/L); normal range, 10-65 pg/mL (10 to 65 ng/L)
- Serum calcium, 8.7 mg/dL (2.17 mmol/L)
- Serum phosphorus, 1.8 mg/dL (0.58 mmol/L).

A radiograph of the legs is shown below (Item Q166).

ITEM Q166: Radiograph of the lower extremities for the patient in the vignette.

Of the following, the MOST likely diagnosis is

A. fibrous dysplasia
B. hypophosphatasia
C. hypophosphatemic rickets
D. osteogenesis imperfecta type I
E. vitamin D deficiency rickets

Item 166 Preferred Response: C

The child described in this vignette has hypophosphatemic rickets. There are multiple heritable forms of hypophosphatemic rickets, but the most commonly encountered is X-linked hypophosphatemic rickets (XLH) due to a mutation in PHEX. This mutation leads to elevated levels of fibroblast growth factor 23 (FGF23), a hormone that causes renal phosphate wasting.

Because this genetic defect causes low phosphorous levels and not low calcium levels, parathyroid hormone (PTH) release is not stimulated, and PTH levels are typically normal or only slightly elevated, as seen in the child in this vignette. This is in contrast to vitamin D deficiency rickets, where PTH is usually quite elevated.

Children with XLH may develop recurrent dental abscesses, joint pain, and enthesopathy (disorders of muscle or ligament attachment) due to calcification of tendons and ligaments. Treatment is with phosphorous replacement and calcitriol (1,25-dihydroxyvitamin D). Linear growth and bony deformities improve with therapy but do not typically normalize completely. Surgery may be indicated for correction of bowing after medical therapy is maximized. Care should be directed by a pediatric endocrinologist with experience in managing this condition.

None of the other answer choices are consistent with a child who has a low phosphorus level but otherwise normal PTH and 25-hydroxyvitamin D levels.
Fibrous dysplasia refers to bone lesions that produce FGF23 and lead to hypophosphatemia. Fibrous dysplasia can be seen in isolation or in the setting of McCune-Albright syndrome. These patients typically present with precocious puberty and have characteristic café au lait macules. They do not present with clinical signs of rickets.

Hypophosphatasia is characterized by extremely low alkaline phosphatase levels due to a defect in alkaline phosphatase. The child in this vignette has an elevated alkaline phosphatase level, consistent with increased bone turnover.
Osteogenesis imperfecta typically presents with recurrent fractures. Markers of bone turnover, calcium, and phosphorus are normal.

Item C166. Summary of Biochemical Changes Seen in Vitamin D Deficiency vs X-Linked Hypophosphatemic Rickets

Analyte	Vitamin D Deficiency (Early)	Vitamin D Deficiency (Late)	XLH
Serum calcium	Normal or low	Low	Normal
Serum phosphorus	Normal or low	Low	Very low
Serum alkaline phosphatase	Elevated	Very elevated	Elevated
Serum PTH	Elevated	Very elevated	Normal
25-hydroxyvitamin D	Low	Very low	Normal
1,25-Vitamin D	Variable	Variable	Normal or low

Abbreviations: PTH, parathyroid hormone; XLH, X-linked hypophosphatemic rickets

Vitamin D deficiency is characterized by low 25-hydroxyvitamin D levels. The 25-hydroxyvitamin D level measured in the child in this vignette was normal. Importantly,

levels of 1,25-vitamin D should never be used to assess for vitamin D deficiency because they are often quite variable. Instead, measurement of 25-hydroxyvitamin D is the appropriate test. The laboratory changes seen in XLH and vitamin D deficiency are compared in the Item C166, page C-130.

PREP Pearls

- In X-linked hypophosphatemic rickets, the serum phosphorus level will be very low, but the PTH level is generally normal or mildly elevated.
- Measurement of 25-hydroxyvitamin D (the storage form of vitamin D) is the definitive test for diagnosing vitamin D deficiency.

American Board of Pediatrics Content Specification(s):

- Recognize the typical clinical and laboratory findings associated with familial hypophosphatemic rickets

Suggested Reading:

- Baroncelli GI, Bertelloni S, Sodini F, et al. Genetic advances, biochemical and clinical features and critical approach to treatment of patients with X-linked hypophosphatemic rickets. Pediatr Endocrinol Rev. 2004;1(4):361-379
- Carpenter TO, Imel EA, Holm IA, Jan de Beur SM, Insogna KL. A clinician's guide to X-linked hypophosphatemia. I Bone Miner Res. 2011;26(7):1381-1388. doi: 10.1002/jbmr.340
- Sperling M. Pediatric Endocrinology. 3rd ed. Philadelphia, PA: Saunders; 2008:645-654.

Item 167

An obstetrician notes vesicular lesions on the cervix of a woman in labor. He believes the lesions are consistent with herpes simplex virus (HSV) infection. She has a history of previous HSV genital infection. She has had rupture of membranes for approximately 8 hours. An emergency Caesarian section is performed. The neonate's birth weight is 2.76 kg and Apgar scores are 8 at one minute and 9 at five minutes.

Of the following, the BEST option to monitor this infant is to obtain

A. surface viral cultures at birth and initiate acyclovir therapy pending culture results
B. surface viral cultures at 24 to 36 hours after delivery and then initiate acyclovir therapy pending culture results
C. surface viral cultures at birth and observe the baby without antiviral therapy
D. surface viral cultures at 24 to 36 hours after birth and observe the baby without acyclovir therapy pending cultures
E. viral cultures of vesicular lesions if they appear, and initiate acyclovir therapy if the baby is clinically ill

Item 167 TE S Preferred Response: D

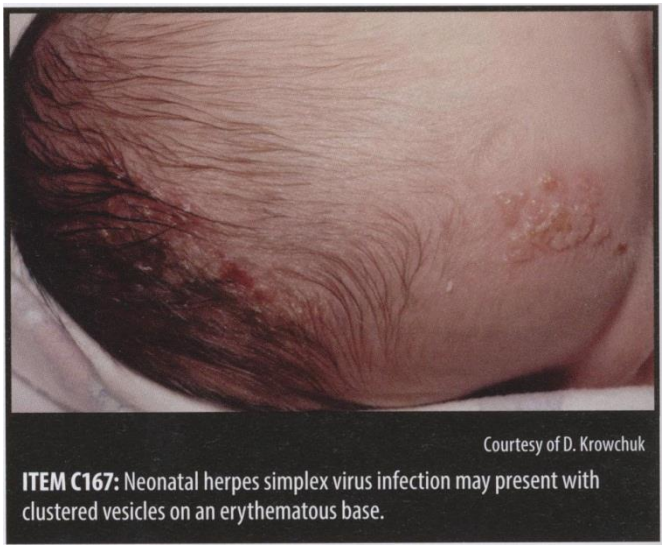
Neonatal herpes simplex virus (HSV) infection occurs in 1 in 3,200 live births and may be associated with significant morbidity and mortality. The risk of transmission of HSV to a full-term newborn exposed to recurrent active HSV genital lesions at delivery with 4 or more hours of rupture of membranes and delivered via vaginal or cesarean delivery as described in the vignette is low (<3%). Surface viral cultures including swabs of the mouth, nasopharynx, conjunctivae, and rectum obtained 24 to 36 hours after delivery may be used to determine if the infant was infected before developing symptoms; this will enable early institution of antiviral therapy with acyclovir if the cultures are positive. If the infant looks well in a low-risk setting such as that described in the vignette, observation without initiating acyclovir while awaiting the results of the cultures is most appropriate.

Surface viral cultures obtained at birth or within the first 24 hours after birth may only reflect contamination during delivery but not true infection of the infant. Such cultures would not be useful in determining the need for treatment of this infant. If the infant appears ill and/or develops signs or symptoms of HSV infection including vesicular lesions (Item C167), a sepsis like picture, and/or central nervous system involvement, in the face of potential HSV exposure, acyclovir therapy should be initiated immediately before obtaining results of viral cultures.

The greatest risk of HSV transmission to the infant occurs if the mother has active primary genital infection at the time of delivery (25%-60%). Infants born prematurely are at increased risk for HSV infection if exposed. Primary cesarean section before, or less than 4 hours after, rupture of membranes appears to be protective against perinatal HSV transmission.

PREP Pearls

- Neonatal herpes simplex virus (HSV) infection is most common when an infant is delivered vaginally and mother has active primary HSV infection.
- Cesarean delivery with less than 4 hours of rupture of membranes protects the infant from HSV exposure.
- If true neonatal HSV infection is suspected, acyclovir therapy should be initiated before obtaining results of viral cultures.



Courtesy of D. Krowchuk

ITEM C167: Neonatal herpes simplex virus infection may present with clustered vesicles on an erythematous base.

American Board of Pediatrics Content Specification(s):

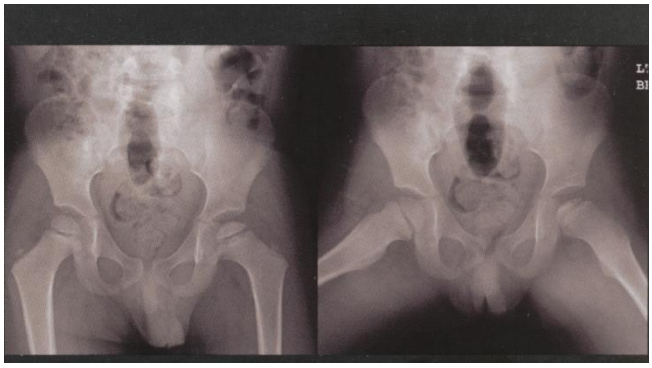
- Know that newborn infants of mothers with primary herpes infections are more likely to be infected than infants born to mothers with recurrent genital herpes simplex infections
- Plan the appropriate management of a neonatal herpes simplex infection, including timing (ie, immediately, even before test results are available)

Suggested Reading:

- American Academy of Pediatrics. Herpes simplex. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:398-408
- Long SS, Pool TE, Vodzak J, Daskalaki I, Gould JM. Herpes simplex virus infection in young infants during 2 decades of empiric acyclovir therapy. Pediatr Infect Dis. 2011;30:556-561. doi:10.1097/INF.0b013e31820e3398
- Shah SS, Aronson PL, Mohamad Z, Lorch SA. Delayed acyclovir therapy and death among neonates with herpes simplex infection. Pediatrics. 2011;128(6):1153-1160. doi:10.1542/peds.2011-0177

Item 168

A 4-year-old boy presents to the emergency department with worsening left hip pain with associated limp. The hip pain has become more consistent over the last 2 weeks. There is no history of fever or trauma. Physical examination reveals guarding and limited external rotation of the left hip. The remainder of the examination, including temperature and vital signs, is within normal limits. You obtain plain radiographs of the hip (Item Q168).



ITEM Q168: Radiographs for the boy in the vignette

Of the following, the MOST likely diagnosis is

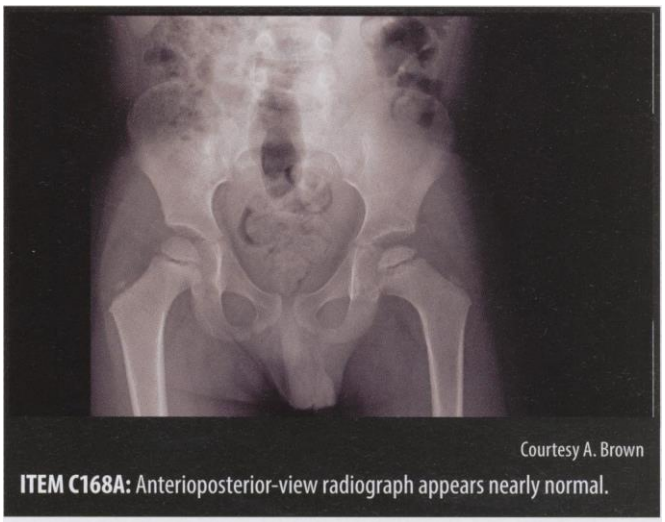
A. developmental dysplasia of the hip
B. hemophilia B
C. Legg-Calve-Perthes disease
D. slipped capital femoral epiphysis
E. toxic synovitis

Item 168 Preferred Response: C

The patient described in the vignette has radiographic findings diagnostic of Legg-Calve-Perthes disease. It is important to note that the anterioposterior view radiograph appears nearly normal (C168A); however, the obvious abnormality of the left femoral head is apparent on the frog-leg view (C168B). The frog-leg view is important when evaluating hip pain. There is no evidence of slipped capital femoral epiphysis or developmental hip dysplasia on his radiographs. There is no evidence of fluid in the hip joint by plain film radiograph, and there is no evidence of bruising or preceding trauma that would suggest hemophilia as the cause of his joint pain. While toxic synovitis can cause hip pain and decreased range of motion, the evidence of avascular necrosis at the femoral epiphysis rules this out. Legg-Calve-Perthes disease can be a late complication of reactive arthritis. The differential diagnosis in a patient with a painful hip is dependent upon the sex and age of the child (Item C168C, page C-132).

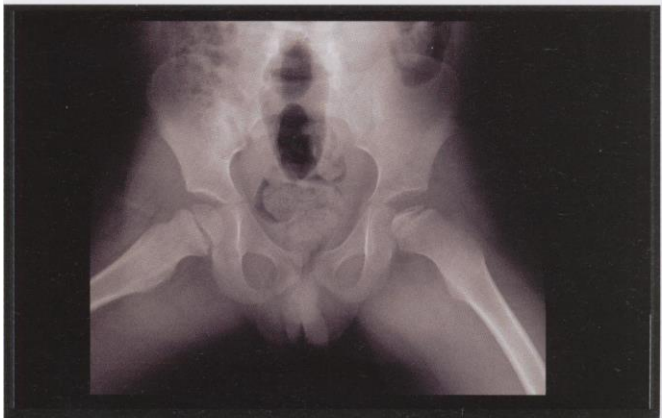
PREP Pearls

- The differential diagnosis for joint pain varies by age, sex, and type of activity.
- Obtaining the appropriate radiographic views of joints is important in making the correct diagnosis.
- Legg-Calve-Perthes disease can be a late complication of reactive arthritis.



Courtesy A. Brown

ITEM C168A: Anterioposterior-view radiograph appears nearly normal.



Courtesy A. Brown

ITEM C168B: Frog-leg view depicts the left femoral head abnormality not apparent on the anterioposterior-view.

American Board of Pediatrics Content Specification(s):

- Know the differential diagnosis of a painful hip varies according to patient age and gender

Suggested Reading:

- Berard R. Approach to the child with joint inflammation. Pediatr Clin N Am. 2012;59(2):245-262. doi:10.1016/j.pc1.2012.03.003
- Sherry D. Limb pain in childhood. Pediatr Rev. 1990;12(2):39-46. doi:10.1542/pir.12-2-39

Item C168C. Differential Diagnosis in a Patient With a Painful Hip

Category	Causes of Hip Pain in Children	Most common age at presentation	Sex or activity predominance
Hematologic disorders	Hemophilia	Any	Male
	Sickle cell disease	Any	None
Infection	Lyme disease	5–14 years	None
	Myositis	Second to third decade of life	Male
	Osteomyelitis	1–5 years	Male
	Septic arthritis	70% younger than 4 years Peak age 6-24 months	Male:Female, 2:1
	Psoas muscle abcess	Adolescence	None
Inflammation	Juvenile idiopathic arthritis	Peak age 2-4 years and adolescence	Slight female
	Rheumatic fever	5–15 years	None
	Transient synovitis	Most common cause of hip pain at age 3–6 years	Male
Miscellaneous	Benign joint hypermobility	Peak age 3–10 years	Female:Male, 2:1
	Developmental dysplasia of the hip	Usually at birth, but can be delayed	none
	Legg–Calvé–Perthes disease	3–12 years Peak age 5–7 years	Male:Female, 3:1
	Slipped capital fermoral epiphysis	Peaks at onset of adolescent growth spurt Boys 12–16 years Girls 10–14 years	Male:Female, 2:1
Neoplasm	Leukemia	Peak age 2–4 years	None
	Metastatic cancer	Variable	Variable
	Osteogenic sarcoma	9–19 years	Slight male
	Ewing sarcoma	Adolescents	Slight male
Trauma	Hip or pelvic fracture	Avulsion fractures: adolescents playing sports	None
		Femoral neck stress fractures: endurance athletes	Female
	Non-accidental trauma	94% of fractures before age 3 years	None
	Overuse injury	Snapping hip syndrome	Dancers, runners
		Acetabular labrum tears	Athletes
	Toddler's fracture	9–36 months	None

Item 169

During a health supervision visit, the mother of a 14-year-old boy remarks that her son is eating less than usual and having difficulty falling asleep at night. She says that over the past month he has been much less physically active, more irritable and moody, and resistant to waking up and getting out of bed each morning. She further reports that he refuses to do his schoolwork or practice the piano and that he yells at her to leave him alone. The boy does not feel there is a problem. He states that he just feels "down" at times and is frustrated with his parents for "getting on my case." He still gets along well with friends and remains on the freshman soccer team. Although he has found school to be a little more difficult and often doesn't feel like doing all of his homework, he is maintaining a C average in ninth grade. His grades were generally at an A and B level up through eighth grade. On physical examination, he is a well-appearing boy with no abnormal findings. A review of systems yields no additional symptoms.

Of the following, the MOST appropriate next step is to

A. begin a well-monitored trial of a selective serotonin reuptake inhibitor medication
B. counsel the mother that this is just an expected phase of adolescent development and should self-resolve
C. recommend family therapy to address his oppositional behavior as a potential symptom of familial dysfunction
D. recommend that the mother develop a reward system to reinforce performance of required routines and schedules
E. refer the boy to a mental health professional experienced with adolescents

Item 169 Preferred Response: E

The adolescent described in the vignette is most likely suffering from major depression. He has decreased appetite, poor sleep, and irritable mood, which indicate a discrete change from his baseline functioning. He also has symptoms of diminished interest in activities he previously enjoyed and a decreased energy level, which make a diagnosis of depression more likely; however, the vignette is less definite on these points. A definitive diagnosis of major depression requires that at least 4 symptoms of depression be present for 2 weeks or more, or 5 symptoms if mood is only irritable rather than clearly depressed. The symptoms of major depression, as defined in the Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition, can be remembered with the mnemonic SIGECAPS:

- Sleep
- Loss of interest
- New guilt or hopelessness
- Loss of energy
- Poor concentration
- Change in appetite
- Psychomotor retardation
- Suicidality

The differential diagnosis includes an unrecognized substance abuse problem or an as yet unidentified psychosocial stressor causing these problems as an adjustment disorder.

The most appropriate next step among the choices listed would be to refer him to a mental health specialist (ie, a psychiatrist or psychotherapist), who can screen him further for substance abuse or contributing psychosocial stressors and help determine whether he has major depression. The same person can then initiate the first recommended steps in treatment: psychoeducation about the disorder and psychotherapy, preferably cognitive behavioral therapy.

A well-monitored trial of a selective serotonin reuptake inhibitor (SSRI) would be appropriate if the boy is confirmed to have moderate to severe major depression, and is either going to simultaneously initiate psychotherapy or is engaged in psychotherapy but is failing to make progress. Treatment with an SSRI alone is less preferred. Counseling the mother that her son is going through an expected adolescent phase of development which should self-resolve can significantly delay the initiation of appropriate interventions. Untreated major depression is reported to take 6 to 12 months to resolve in adolescents.

Although it is true that oppositional behaviors can be a sign of family dysfunction, recommending that the family in the vignette enter family therapy misses the opportunity to intervene directly with the current problem and is likely to be received poorly by a family who may feel blamed for their child's difficulties.

Instituting a reward system for completing homework and participating in parent desired activities like piano practice is a reasonable approach to try to increase this young man's

compliance with expectations, but it fails to address the root cause of the difficulties, which is likely to be major depression.

PREP Pearls

- It is important to confirm that major depression is present before considering initiating medication.
- Psychotherapy is the first-line intervention for major depression, with or without the addition of SSRI medication.
- Watchful waiting in the case of major depression can lead to prolonged dysfunction.

AAP Mental Health Competency:

- Know the role of counseling in depression treatment

Suggested Reading:

- Birmaher B, Brent D; AACAP Work Group on Quality Issues, et al. Practice parameter for the assessment and treatment of children and adolescents with depressive disorders. J Am Acad Child Adolesc Psychiatry. 2007;46:1503-1526. doi:10.1097/chi.0b013e318145aelc
- Carlat DJ. The psychiatric review of symptoms: a screening tool for family physicians. Am Fam Phys. 1998;58(7):1617-1624

Item 170

You are called by the normal nursery because an infant has failed the congenital heart disease screening testing per-formed 3 days after birth. On room air, the oxygen saturation value on the right hand (preductal) is 93% and the oxygen saturation value on the right foot (postductal) is 85%. Vital signs include a temperature of 37°C, heart rate of 150 beats/ min, and respiratory rate of 70 breaths/min. The blood pressure readings are 70/40 mm Hg in the right arm and 55/30 mm Hg in the right leg. Examination reveals a soft 1/6 systolic murmur at the left lower sternal border, clear lungs, weak femoral pulses bilaterally, and a capillary refill of 4 to 5 seconds in the lower extremities. Arterial blood gas includes a pH of 7.25, PCO_2 of 39 mm Hg, PO_2 of 45 mm Hg, bicarbonate of 17 mEq/L (17 mmol/L), and base deficit of -9.

Of the following, the MOST appropriate next step in this infant's management is to

A. infuse sodium bicarbonate
B. initiate prostaglandin E,
C. place an endotracheal tube
D. provide 40% hood oxygen
E. start dopamine

Item 170 S SBP TE Preferred Response: B

Differential preductal and postductal values of oxygenation saturation and blood pressure suggest coarctation of the aorta, which requires the initiation of prostaglandin El (PGE1). Neonates with coarctation of the aorta depend on a patent ductus arteriosus to allow blood flow into the descending aorta. Open at birth, the smooth muscle in the ductus arteriosus begins to constrict in response to the increased oxygenation levels after delivery. It may be functionally closed as early as 12 to 24 hours after delivery or persist up to a week. Neonates with coarctation may have no murmur or visible cyanosis to suggest underlying congenital heart disease. A pulse oximetry reading below 95% on routine congenital heart disease screening at discharge may be the first sign of underlying congenital heart disease. A difference of more than 10 points between the preductal and postductal systolic blood pressures strongly suggests a coarctation or interruption of the aorta. The neonate described in the vignette is demonstrating a metabolic acidosis with compensatory tachypnea, which suggests ongoing hypoperfusion. Initiation of PGE1 to maintain ductal patency is appropriate while awaiting echocardiographic confirmation of the diagnosis.

Critical congenital heart disease (CCHD) refers to cardiac defects that require treatment in the neonatal period. Transposition of the great arteries, coarctation of the aorta, interrupted aortic arch, and hypoplastic left heart syndrome are types of CCHD that depend on the patency of the ductus arteriosus and pose a unique diagnostic challenge for the clinician. The ductus arteriosus may not close until a neonate has been discharged home from the normal nursery, delaying the development of the clinical findings of tachypnea, decreased perfusion, and heart failure that would lead to further investigation. It has been estimated that in the past 30% of neonates with CCHD may have been discharged from the hospital undiagnosed.

In 2011, the Secretary of the US Department of Health and Human Services recommended that all neonates be screened for CCHD using pulse oximetry before being discharged from the hospital. Seven forms of CCHD are identified as the primary targets of the screening, including hypoplastic left heart syndrome, pulmonary atresia, transposition of the great vessels, truncus arteriosus, tricuspid atresia, tetralogy of Fallot, and total anomalous pulmonary venous return. Coarctation of the aorta is identified as a secondary target because it is a type of CCHD that may not be identified consistently with oximetry screening. Congenital heart disease screening is performed by measuring oxygen saturation in the right hand (preductal) and 1 foot (postductal) more than 24 hours after birth or as late as possible if the neonate is being discharged early. An oxygen saturation level of greater than or equal to 95% in either extremity with an absolute difference between upper and lower extremities of less than or equal to 3% is considered a negative screening result. Infants with screening values below 90% are immediately referred to a pediatric cardiologist. Neonates with pulse oximetry readings between 90% and 94% undergo repeat screening within 12 hours of the initial screening.

Although the neonate in the vignette has metabolic acidosis, correction with sodium bicarbonate is not the initial step in management. The acidosis may improve after initiation of PGE1 and subsequent improved perfusion because of reopening of the ductus

arteriosus. The neonate should not be given hood oxygen, because the preductal saturation values are acceptable and increased oxygen levels in the blood may lead to further constriction of the ductus arteriosus. The neonate has decreased blood pressure in the lower extremity because of the coarctation, which requires treatment with PGE1 rather than dopamine. Intubation is not required because the neonate's oxygenation and ventilation levels are adequate. The clinician will need to monitor the infant closely for apnea after the initiation of PGE1 and be prepared to intubate at that time.

PREP Pearls

- Transposition of the great arteries, coarctation of the aorta, interrupted aortic arch, and hypoplastic left heart syndrome are types of critical congenital heart disease that are dependent on the patency of the ductus arteriosus.
- In 2011, the Secretary of the US Department of Health and Human Services recommended that all neonates be screened for critical congenital heart disease using pulse oximetry before being discharge from the hospital.

American Board of Pediatrics Content Specification(s):

- Know the importance of patent ductus arteriosus in the presentation of hypoplastic left heart syndrome and in coarctation of the aorta

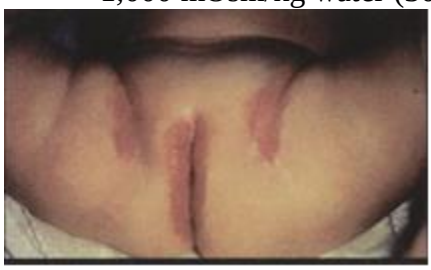
Suggested Reading:

- Centers for Disease Control and Prevention. Newborn screening for critical congenital heart disease: potential roles of birth defects surveillance programs - United States, 2010-2011. MMWR Morb Mortal Wkly Rep. 2012;61:849-853
- Hoffman JIE. Is it time for routine neonatal screening by pulse oximetry. Neonatology. 2011;99:1-9. doi: 10.1159/000311216
- Mahle WT, Martin GR, Beekman RH, Morrow WR, Section on Cardiology and Cardiac Surgery Executive Committee. Endorsement of health and human services recommendation for pulse oximetry screening for critical congenital heart disease. Pediatrics. 2012;129:190-192. doi: 10.1542/ peds.2011-3211

Item 171

A 15-month-old girl is seen for persistent diaper rash. Despite treatment with zinc oxide cream and topical antifungal cream for over a month, there has been no improvement. The mom also reports that she wants to drink all day long and has doubled the number of wet diapers per day. In the office, she is afebrile with stable vital signs. She is at the 50th percentile for weight and height. On physical examination, there are crusted erythematous and brown papules covering the groin and involving the intertriginous areas (Item Q171). A similar rash is seen on areas of the posterior scalp and axillae. The affected area of the scalp is scaly and seborrheic. The remainder of his physical examination is normal. Initial laboratory test results are as follows:

- Serum sodium, 146 mEq/L (146 mmol/L); normal range, 136 to 145 mEq/L (136-145 mmol/L)
- Serum osmolality, 301 mOsm/kg water (301 mmol/kg
- water); normal range, 275 to 295 mOsm/kg water (275-295 mmol/kg water)
- Urine specific gravity, 1.001; normal range, 1.001 to 1.035
- Urine osmolality, 95 mOsm/kg water (95 mmol/kg water); normal range, 300 to 1,000 mOsm/kg water (300-1,000 mmol/kg water)



Q171: Rash as described for the girl in the vignette.

Of the following, the MOST likely finding in this patient would be

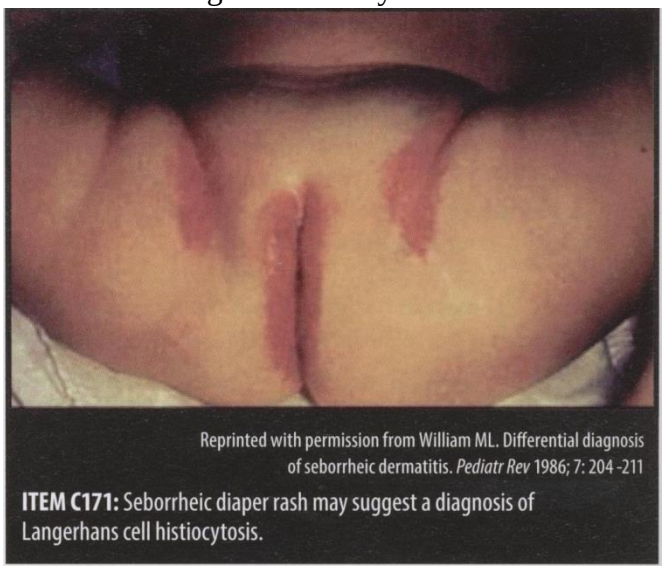
A. enzyme-linked immunoassay testing positive for human immunodeficiency virus
B. herpes simplex virus from the culture of the papular fluid
C. hyphae on potassium hydroxide (KOH) preparation of papular fluid
D. lytic lesion on radiograph of the skull
E. serum glucose greater than 300 mg/dL (16.7 mmol/L)

Item 171 Preferred Response: D

The child described in the vignette with a persistent scalp and diaper rash (Item C171) and symptoms of diabetes insipidus may have Langerhans cell histiocytosis (LCH); therefore, the most likely additional finding would be lytic lesions seen on a skull radiograph.

Histiocytes are cells of the mononuclear phagocyte system. The histiocytoses are rare conditions caused by the abnormal proliferation or functioning of these cells. The most common of these is LCH. The diagnosis of LCH is based on histologic and immunohistochemical criteria and clinical features. The histopathologic features are granuloma containing inflammatory cells and abnormal Langerhans cells. Results of immunohistochemical analysis are positive for CD1a and CD207 (correlates with Birbeck granules). In LCH, cells accumulate and release inflammatory chemokines, which can lead to stimulation of other inflammatory cells and eventually create a "cytokine storm:' These cytokines contribute to the clinical symptoms. Langerhans cell histiocytosis can occur as a single system or multisystem disease. Single system disease can affect bone (in one or multiple sites) or skin. Multisystem disease is categorized into low-risk and high-risk.

Although LCH can present at any age, the systemic life-threatening form usually occurs in children younger than 4 years. Adults with LCH often have only skin involvement. Skin involvement occurs in 50% of patients and most often appears seborrheic. It can also manifest as papules, vesicles, nodules, and purpuric nodules. Infants with skin-only LCH may progress to multisystem disease, which can be fatal. Any child with seborrheic dermatitis or diaper dermatitis that is persistent should be evaluated for LCH. In children, bone is the most commonly affected organ. The characteristic finding is a lytic lesion in the skull or vertebral collapse. Those with a single bone lesion have an excellent prognosis. Central nervous system and endocrine involvement can also occur. Diabetes insipidus occurs in 24% of patients with LCH due to hypothalamic-pituitary axis disease. Additional findings suggestive of LCH include chronic draining otitis media or chronic mastoiditis.



Reprinted with permission from William ML. Differential diagnosis of seborrheic dermatitis. *Pediatr Rev* 1986; 7: 204 -211

ITEM C171: Seborrheic diaper rash may suggest a diagnosis of Langerhans cell histiocytosis.

Multisystem LCH is divided into high-risk and low-risk based on the risk of mortality from disease. High-risk LCH includes patients with disease in 2 or more organs, including at least one of the at-risk organs liver, spleen, lung, and blood (ie, cytopenia). Low-risk LCH is defined as multisystem disease from 2 or more organs but not involving any of the at-risk organs.

Treatment for LCH involves surgical excision, steroid injection, chemotherapy, and radiation, depending on the site of the lesion and how widespread it is.

The child in the vignette has a recurrent seborrheic rash and diabetes insipidus. The next test to obtain would be radiographs of the entire skeletal system, including the skull looking for the characteristic lytic lesions. Additional evaluation includes a complete chemistry profile, complete blood cell count, urinalysis (as done for the child in the vignette), coagulation studies, and radiographic evaluation of any involved organs. Infections with human immunodeficiency virus, herpes simplex virus, or fungus are not usually associated with diabetes insipidus and are not seen more frequently in LCH. A very high serum glucose level would be seen in patients with diabetes mellitus but is not associated with LCH.

PREP Pearls

- LCH is the most common form of histiocytosis, and can occur as a single-organ or multisystem disease.
- Common symptoms of LCH in children are recurrent rash, diabetes insipidus, and bone lesions.
- Any child with severe seborrheic dermatitis that does not respond to treatment should be evaluated for LCH.

American Board of Pediatrics Content Specification(s):

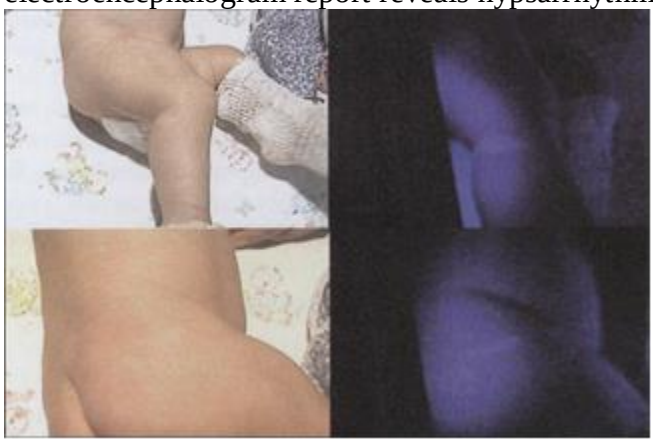
- Recognize the clinical manifestations of the histiocytosis syndromes of childhood

Suggested Reading:

- Filipovich A, McClain K, Grom A. Histiocytic disorders: recent insights into pathophysiology and practical guidelines. Biol Blood Marrow Transplant. 2010;16(1 suppl):S82-S89. doi:10.1016/j.bbmt.2009.11.014
- Haupt R, Minkov M, Astigarra I, et al; Euro Histio Network. Langerhans cell histiocytosis (LCH): guidelines for diagnosis, clinical work-up, and treatment for patients till the age of 18 years. Pediatr Blood Cancer. 2013;60:175-184. doi:10.1002/pbc.24367
- Weitzman S, Egeler RM. Langerhans cell histiocytosis: update for the pediatrician. Curr Opin Pediatr. 2008;20:23-29. doi:10.1097/MOP.0b013e3282f45ba4

Item 172

A 9-month-old female infant develops stereotyped spells of eye rolling, head drop, and truncal flexion. They last 5 seconds each and come in clusters of 10 to 15, typically when she is falling asleep or waking up. She cries after the cluster of spells ends but then returns to normal. She is otherwise healthy. Her physical examination shows an occipital frontal circumference at the 50th percentile. She has several hypopigmented macules that are observed more clearly during a Wood lamp examination (Item Q172). The remainder of her general and neurologic examination findings is normal. Routine electroencephalogram report reveals hypsarrhythmia.



ITEM Q172: Findings as exhibited by the girl in the vignette.

Of the following, the MOST likely diagnosis is

A. ataxia telangiectasia
B. hypomelanosis of Ito
C. neurofibromatosis type 1
D. Sturge-Weber syndrome
E. tuberous sclerosis

Item 172 TE Preferred Response: E

The infant described in the vignette presents with infantile spasms. On examination, she is found to have hypopigmented skin lesions, suggesting a diagnosis of tuberous sclerosis. Other clinical manifestations of tuberous sclerosis usually do not appear until older ages. These include facial angiofibromas that can appear at about 4 years of age and periungual fibromas that appear in adolescence.

This infant should be referred to a pediatric neurologist for management of infantile spasms. She will also need a magnetic resonance image of the brain, with and without contrast, to evaluate for the typical findings of cortical tubers, radial glial bands, and subependymal nodules. Occasionally, the subependymal nodules can transform into subependymal giant cell astrocytomas, which can cause obstructive hydrocephalus. Infants and children with tuberous sclerosis should be clinically monitored for abnormal head growth and signs of increased intracranial pressure. Other manifestations of tuberous sclerosis include cardiac rhabdomyomas (especially in neonates), renal angiomyolipomas, autism spectrum disorder, and epilepsy.

It is important to recognize the characteristic skin findings in neurocutaneous syndromes. Ataxia telangiectasia presents with ataxia in young toddlers; telangiectasis of the sclera and face develop as early as 5 years of age. Hypomelanosis of Ito presents with whorls of hypopigmented and hyperpigmented skin, and some but not all patients have intellectual disability. Neurofibromatosis type 1 is characterized by café au lait spots, and later, axillary and inguinal freckling. Sturge-Weber syndrome is characterized by facial angiomatosis (also known as a port wine stain) of the forehead and upper eyelid.

PREP Pearls

- Hypopigmented macules are associated with tuberous sclerosis, and hyperpigmented café au lait spots are associated with neurofibromatosis type 1.
- Infantile spasms may be the first recognized clinical manifestation of tuberous sclerosis. They occur in up to 65%-70% of patients with tuberous sclerosis and frequently indicate a poor neurodevelopmental outcome.

American Board of Pediatrics Content Specification(s):

- Recognize the clinical manifestations of tuberous sclerosis, and manage appropriately

Suggested Reading:

- Sahin M. Neurocutaneous syndromes.. In: Kliegman RM, Stanton BMD, St Geme J, Schor N, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Elsevier Saunders; 2011:2046-2053

Item 173

A 14-year-old girl presents to your office with her father the day after sustaining an injury to her left knee. She reports experiencing a sudden sharp pain and feeling a "pop" in the knee while landing from a jump during volleyball practice. She has been able to bear weight but reports feeling unsteady with ambulation. She developed significant knee swelling almost immediately after the injury. On physical examination, you note a large effusion and decreased range of motion of the left knee; she has no tenderness to palpation. When you flex the knee to 30° and pull the tibia anteriorly while holding the femur stable, you note about 1.5 cm of anterior motion of the tibia. The remainder of the physical examination is normal.

Of the following, the MOST likely diagnosis is

A. anterior cruciate ligament tear
B. medial collateral ligament tear
C. medial meniscus tear
D. patellar subluxation
E. patellar tendon rupture

Item 173 Preferred Response: A

The girl described in the vignette reports acute onset of knee pain and feeling a "pop" while landing from a jump; following her injury, she developed significant swelling. This patient provides a classic description of a noncontact anterior cruciate ligament (ACL) tear. More than 70% of ACL tears do not involve contact; these injuries generally occur with a pivot or twist, landing from a jump or sudden deceleration. Most affected patients recall feeling or hearing a "pop" at the time of injury. Athletes often note significant swelling due to hemarthrosis within several hours following an ACL tear.

The Lachman maneuver is used to assess for ACL laxity: with the patient supine and the knee flexed to 30°, the examiner holds the femur and attempts to pull the tibia forward. Increased anterior translation of the tibia relative to the femur compared with the uninjured side suggests disruption of the ACL. In the absence of associated injuries, many individuals with ACL tears do not have any tenderness on examination.

Medial collateral ligament (MCL) sprains tend to occur with valgus stress (a force to the outside of the knee) while the foot is planted. An athlete with an MCL tear generally presents with medial knee pain and swelling and has tenderness along the medial aspect of the knee. The valgus stress test is used to assess for MCL sprain: the examiner places the heel of the hand over the lateral aspect of the knee and applies a medially directed force with the joint flexed to 30°; pain indicates stretch or partial tear of the MCL, whereas laxity is generally present with a complete tear. Lateral collateral ligament injuries occur with varus force to the knee; these injuries are rare and tend to be associated with injuries to other structures.

The posterior cruciate ligament (PCL) prevents the tibia from moving posterior relative to the femur. Hyperextension of the knee and direct posterior force to the tibia are the most common mechanisms of PCL tear. The posterior drawer test is used to assess for PCL laxity: with the patient supine and the knee flexed enough to allow the foot to rest flat on the examination table, the examiner stabilizes the foot and pushes the tibia posteriorly. In a patient with a PCL tear, the examiner will note excessive posterior translation of the tibia relative to the femur.

Patients who suffer an acute meniscal tear generally present with effusion, tenderness along the affected joint line, and a positive McMurray test result. The McMurray test result is positive if a patient has pain or if the examiner notes a palpable click with valgus stress to the knee, external rotation of the tibia, and passive motion of the knee from extreme flexion to extension.

The clinical presentation for a patellar subluxation or dislocation can be similar to that of an ACL tear. Patients often report acute onset of pain with a twisting motion, a popping sensation, and subsequent swelling. On physical examination, patients generally have tenderness around the kneecap, especially over the medial patellar facet. Patellar tendon rupture is rare in adolescents. Affected individuals are unable to actively extend the knee, and the patella retracts so that it is higher on the affected side.

PREP Pearls

- Most ACL tears result from a noncontact injury mechanism, such as a pivot or twisting motion or landing from a jump.
- In the absence of associated injuries, many individuals with ACL tears do not have tenderness on examination.
- The clinical presentation for an acute patellar dislocation and an ACL tear can be similar.

American Board of Pediatrics Content Specification(s):

- Understand that the presence or character of pain may be the dominant indicator of type and severity of ligament injuries

Suggested Reading:

- LaBella C, Carl R. Preventing knee ligament injuries in young athletes. Pediatr Ann. 2010;39(11):714-720. doi:10.3928/00904481-20101013-10
- Sarwark JF, LaBella CR, eds. Pediatric Orthopaedics and Sports Injuries: A Quick Reference Guide. Elk Grove Village, IL: American Academy of Pediatrics; 2010

Item 174

A 13-month-old boy is seen in the emergency department (ED) for the third time with a history of decreased oral intake, lethargy, and irritability. Symptoms have been present for 4 hours since awakening at 6:00 AM; his last formula feed prior to this morning was at 11:00 PM last night. His first visit to the ED at 5 months of age resulted in admission to the hospital with concurrent symptoms of an upper respiratory infection without fever; he also had hypoglycemia that resolved spontaneously. The result of a sepsis workup was negative (normal complete blood cell count as well as negative blood, cerebral spinal fluid, and urine cultures), and he received 3 days of antibiotics until the cultures were negative. His second visit to the ED took place at 8 months of age, when he had a mild bout of acute gastroenteritis. He was also hypoglycemic at that time but responded well to intravenous glucose, a single bolus of normal saline, and oral rehydration; he was discharged after 6 hours of observation in the ED, during which time he had frequent fingerstick glucose levels that were within normal limits.
Results of a basic chemistry panel are within normal limits except for a glucose level of 27 mg/dL (1.5 mmol/L). There is moderate elevation in his transaminases, a mildly elevated ammonia level, and negative urinary ketones. His examination is significant for mild hepatomegaly with no fever or other systemic symptoms.

Of the following, the study MOST likely to be helpful in identifying a cause for this child's symptoms is

A. a cortisol level
B. a liver biopsy
C. plasma carnitine levels
D. serum immunoglobulins
E. urine organic acids

Item 174 Preferred Response: C

The child described in the vignette has some of the typical presenting signs of systemic primary carnitine deficiency (PCD). Children with PCD generally present between the ages of 3 months and 2 years with hypoketotic hypoglycemia characterized by poor oral intake, lethargy, and irritability. Primary carnitine deficiency results in defective long-chain fatty acid transport into the mitochondria, which leads to decreased energy production. Symptoms are often triggered by an intercurrent illness such as an upper respiratory infection or acute gastroenteritis. Symptoms are also often apparent after longer than usual periods of fasting. In addition to low carnitine levels, other laboratory findings may include mildly elevated transaminases and mildly elevated ammonia levels. Hepatomegaly may also be noted on examination in some patients. Primary carnitine deficiency is inherited as an autosomal recessive condition and may be detected in some infants on expanded newborn screening. Diagnostic testing for PCD includes a skin biopsy for a fibroblast assay that assesses carnitine transport or a molecular genetic study on a DNA sample that looks for mutations in SLC22A5 on chromosome 5. Untreated, PCD can result in symptomatic hypoglycemia and heart and skeletal muscle dysfunction. Supplementation with levocarnitine can restore plasma carnitine levels and prevent long-term, life-threatening sequelae.

Although this is a boy, he is rather old to present with congenital adrenal hyperplasia (CAH) with a secondary cortisol deficiency. Classic CAH due to 21-hydroxylase deficiency would generally present with hyponatremia secondary to a salt-losing crisis with or without hypotension and elevations in 17-hydroxyprogesterone but would not present with hypoglycemia. A liver biopsy might be indicated if a glycogen storage disease or tyrosinemia was suspected. However, glycogen storage disease type 1 (Von Gierke disease) would typically present much earlier with persistent and intractable hypoglycemia, significant hepatomegaly and renomegaly, and secondary lactic acidosis, hyperlipidemia, and hyperuricemia. A normal blood chemistry panel is inconsistent with this diagnosis. Tyrosinemia type 1 (T1) would also present in the first few months of life with failure to thrive, jaundice, Fanconi syndrome, and signs of liver failure but would not present with hypoglycemia. Plasma amino acids would demonstrate elevations in tyrosine and methionine, with excretion of succinylacetone in the urine being pathognomonic for T1. Liver findings might include nodular cirrhosis, but early identification of T1 through newborn screening and prompt treatment with a metabolic pathway blocker known as NTBC has shown promise in slowing down or preventing liver failure in patients with T1.

The 2 previous infections seen in the child in the vignette are not unusual for a child his age and would not raise suspicions about an immunodeficiency. Therefore, immunoglobulins would not be indicated. A child with an organic acidemia may also have a mildly elevated ammonia level but should also have evidence of a metabolic acidosis with a significant anion gap on blood chemistries. While the history is certainly consistent with an inborn error of metabolism and a metabolic workup, including urine organic acids, is in order, his history and presentation are most consistent with PCD. Other metabolic tests to consider while awaiting the carnitine levels should include plasma amino acids, lactate, pyruvate, and urinary reducing substances.

PREP Pearls

- Recurrent hypoglycemia associated with intercurrent illness in the absence of metabolic acidosis or other electrolyte imbalances is suggestive of carnitine deficiency.
- Untreated carnitine deficiency can result in irreversible cardiomyopathy and skeletal muscle dysfunction.

American Board of Pediatrics Content Specification(s):

- Recognize the laboratory findings in a patient who has a disorder of carnitine metabolism

Suggested Reading:

- El-Hattah AW. Systemic primary carnitine deficiency. In: Pagon RA, Bird TD, Dolan CR, eds. GeneReviews. Seattle, Washington: University of Washington; 2013
- Ficicioglu C, an Haack K. Failure to thrive: when to suspect inborn errors of metabolism. Pediatrics. 2009;124(3):972-979
- Gessner BD, Gillingham MB, Birch S, Wood T, Koeller DM.
- Evidence for an association between infant mortality and a carnitine palmitoyltransferase 1A genetic variant. Pediatrics. 2010;126(6):945-951. doi:10.1542/peds.2010-0687
- Helton E, Darragh R, Francis P, et al. Metabolic aspects of myocardial disease and a role for L-carnitine in the treatment of childhood cardiomyopathy. Pediatrics. 2000;105(6):1260-1270

Item 175

A 2-year-old girl with no significant past medical history is brought to your office for the second visit this week because of a cough and intermittent wheezing that started 1 week ago. A review of her chart shows that your partner prescribed inhaled albuterol for wheezing at her initial visit 4 days ago. Her symptoms have not improved. The girl's mother brought her for reevaluation today because her cough seems to be worsening, and this morning she coughed up a small amount of blood.
Physical examination reveals a well-nourished, interactive girl who is coughing frequently and has wheezing localized to the right lower lung field. The mother asks you if her daughter's symptoms could indicate a food allergy, since her cough began shortly after she ate peanuts for the first time.

Of the following, the study MOST likely to confirm the patient's diagnosis is

A. bronchoscopy
B. pulmonary function testing
C. skin testing for peanut allergy
D. sputum culture
E. sweat chloride test

Item 175 S I-C Preferred Response: A

The girl described in the vignette presents with a history and clinical findings that strongly suggest the diagnosis of foreign-body aspiration. Of the diagnostic studies listed, bronchoscopy is most likely to confirm her diagnosis. Furthermore, rigid bronchoscopy is the standard of care for treatment of foreign body aspiration.

Foreign-body aspiration should be suspected in any patient presenting with respiratory distress and/or wheezing of sudden onset, even in the absence of a clear history of a choking episode. A detailed history regarding the circumstances surrounding the onset of wheezing is very important. Although a history of choking is highly suggestive of foreign-body aspiration, it is not witnessed or recalled in all cases. Foreign-body aspiration is most common in children younger than 3 years. The diagnosis may be delayed in some children because of negative (or misinterpreted) findings on the history, examination, and chest radiography. The classic triad of coughing, wheezing, and decreased breath sounds is present in only approximately 40% of cases.

Wheezing is a common presenting symptom of respiratory disease in children. Although most instances of wheezing arise from infectious causes or asthma, it is crucial for pediatric practitioners to understand the extensive differential diagnosis for wheezing and to appreciate that "all that wheezes is not asthma:' Although the differential diagnosis of recurrent wheezing is broad, distinguishing between diffuse and localized wheezing can be helpful in pinpointing the underlying cause. Diseases causing generalized wheezing include asthma, bronchiolitis, cystic fibrosis, anaphylaxis with pulmonary involvement, and gastric esophageal reflux. Localized wheezing may be caused by foreign-body aspiration, bronchomalacia, or extrinsic compression by a vascular anomaly or mass, along with other less common causes.

Physical examination can provide clues to the diagnosis of foreign-body aspiration. Foreign-body aspiration should be suspected in any child with a unilateral monophonic wheeze or asymmetric breath sounds. Regional variation in air entry is an important clue to the diagnosis. In children, an aspirated foreign body may lodge in any bronchus, without predilection to the right side. In some cases, an aspirated foreign body may trigger a generalized irritant response, resulting in diffuse polyphonic wheezes. Other associated symptoms may include persistent cough, hemoptysis, dyspnea, choking, fever, and even cyanosis. If diagnosis is delayed, patients with foreign-body aspiration may develop pneumonia. In contrast to children presenting with asthma symptoms, the symptoms of children with foreign-body aspiration generally do not improve significantly with bronchodilator therapy. It is crucial for clinicians to recognize that if a patient does not improve as expected with the treatment provided for a certain condition, alternative differential diagnoses should be considered and investigated.

A prompt and aggressive workup is justified in children suspected of having an aspirated airway foreign body. Evaluations that may be useful include chest radiographs, airway fluoroscopy, and bronchoscopy. Chest radiographs of children with foreign-body aspiration may show normal findings-because most foreign bodies aspirated by children are radiolucent-but may detect unilateral hyperexpansion (Item 175), atelectasis, or lobar

infiltrate. Diagnostic imaging should consist of anteroposterior and lateral radiographs of the chest and the entire neck. Inspiratory and expiratory chest radiographs may reveal air trapping on the side of the foreign body. Left and right lateral decubitus films may be useful in infants and younger children who cannot cooperate with inspiratory and expiratory films. The side of the foreign body will not deflate when placed in the dependent position. Rigid bronchoscopy is the procedure of choice from both a diagnostic and therapeutic perspective when an aspirated foreign body is highly suspected. It should be performed if there is strong clinical suspicion of foreign-body aspiration, regardless of radiographic findings.

Pulmonary function tests can be useful as a diagnostic tool for children with suspected asthma. The girl in this vignette has no prior history of respiratory symptoms, has focal rather than diffuse wheezing on examination, and did not improve with bronchodilator (albuterol) treatments; therefore, the diagnosis of asthma is much less likely and should prompt her pediatrician to consider alternative differential diagnoses for her symptoms. Skin testing for peanut allergy would not help in identifying the correct diagnosis in this patient. Although her symptoms began after she ingested peanuts, foreign-body aspiration of peanut pieces is much more likely than peanut allergy in this case with localized wheeze and the absence of other symptoms of food allergy, including urticaria, angioedema, stridor, pruritus, hypotension, and vomiting.

Sputum culture, which may be helpful in identifying an infectious cause in patients with pneumonia, would not be useful in pinpointing the underlying clinical problem in this patient. Although children with pneumonia can certainly present with focal findings on lung examination, the patient in this vignette has been afebrile and otherwise healthy. Her cough most likely results from pulmonary irritation by the aspirated foreign body rather than from an infectious cause. Pneumonia, however, may arise as a complication of aspirated foreign bodies.

A sweat chloride test to diagnose cystic fibrosis would not be indicated for the girl in the vignette. She is well nourished with no prior history of cough, gastrointestinal symptoms, or common sinopulmonary infections in her first 2 years of life, making cystic fibrosis highly unlikely. Furthermore, patients with cystic fibrosis would be expected to have diffuse rather than localized findings on lung examination.

Prevention is the key to minimizing the significant morbidity and even mortality in children that may arise when foreign objects obstruct the airway. The American Academy of Pediatrics recommends that anticipatory guidance to prevent choking or foreign-body aspiration be provided to caregivers beginning at 6 months of age. At this age, children begin to develop the fine motor dexterity to pick up small objects and place them into their mouths. Important aspects of parental education include the following points:

- Hard or round foods should not be offered to children younger than 4 years; these foods include (but are not limited to) hot dogs and sausages, chunks of meat, grapes, raisins, apple chunks, nuts, seeds, peanuts, popcorn, raw carrots, and hard candy.

- Infants should be fed solid food only by adults and only when sitting upright; adults should supervise all meals for young children.
- Children should be taught to chew their food well; talking, playing, running, crying, and laughing while eating should be discouraged.
- Chewable medications should be given only after the age of 3 years (when molars are present).
- Toys with small parts should be avoided, and other small household items should be kept out of reach of infants and young children (including coins and batteries). Marbles, small rubber balls, and latex balloons should be mentioned specifically because aspiration of these items may be fatal.
- The age recommendations on toy packages should be followed.
- The practice of using the mouth to hold school supplies or other small objects should be discouraged.
- The actions of older children, who may give younger siblings dangerous objects or leave these objects within their reach, should be considered.
- Parents, teachers, child care providers, and others who care for children should be encouraged to take a course in basic life support and choking first aid.

PREP Pearls

- Foreign-body aspiration should be suspected in any patient presenting with respiratory distress or wheezing of sudden onset even in the absence of a clear history of a choking episode.
- Foreign-body aspiration should be suspected in any child with a unilateral monophonic wheeze or asymmetric breath sounds.
- Whenever a patient does not respond as expected to the treatment provided for a given condition, clinicians should be prompted to consider alternative differential diagnoses.
- Anticipatory guidance to prevent choking or foreign-body aspiration should be provided to caregivers beginning when their child is 6 months of age.

American Board of Pediatrics Content Specification(s):

- Distinguish between asthma and foreign body aspiration

Suggested Reading:

- Fakhoury K. Wheezing illnesses other than asthma in children. UptoDate. Available online only for subscription
- Roven JD, Rogers BM. Pediatric foreign body aspiration. Pediatr Rev. 2000;22:86. doi:10.1542/pir.21-3-86
- Ruiz FE. Airway foreign bodies in children. UptoDate. Available online only for subscription
- Wang VI. Wheezing. In: Fleisher GR, Ludwig S, eds. Textbook of Pediatric Emergency Medicine. 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2010:635-642

Item 176

The mother of a healthy, 30-month-old girl is concerned because her daughter has begun to run on her toes. She is able to walk with a normal heel–toe gait at times but often walks "on her tip toes.' The birth history is unremarkable, and her developmental milestones have been age appropriate. Results of her physical examination, including the full neuromuscular examination, are normal.

Of the following, your BEST recommendation today is

A. bracing of the lower extremities
B. genetic testing for muscular dystrophy
C. magnetic resonance imaging of the spine
D. reassurance that this will likely resolve by 4 years of age
E. referral for botulinum toxin injections

Item 176 Preferred Response: D

The young age of the girl in this vignette, the parents' report that toe walking is only intermittent, and the normal birth and developmental history lead to the most likely diagnosis of idiopathic toe walking. Importantly, the musculoskeletal and neurologic examination findings are normal. In this case, toe walking most likely reflects a normal stage in gait development, and the family can be reassured that it will spontaneously resolve by 4 to 5 years of age. Toe walking is more likely to persist beyond 5 years of age in children with neurocognitive disorders.

Children without underlying medical conditions who walk on their toes beyond 3 years of age are referred to as idiopathic toe walkers. Toddlers may normally experiment with different walking techniques. The heel-toe gait pattern is typically established by 4 years of age. Initially, patients with idiopathic toe walking have no specific abnormalities on static examination, only on observation of gait. If toe walking persists after 7 to 8 years of age, heel cord contractures and calf hypertrophy develop. Compensatory external tibial torsion may occur in the long term. Stretching exercises performed by a physical therapist or the parents to ensure the child's heel cords remain supple are the first mode of treatment.

A thorough history and physical examination will help to rule out other causes of toe walking, including neurologic disorders, such as cerebral palsy, muscular dystrophy, tethered spinal cord, or autism. Further evaluation, such as genetic testing for muscular dystrophy or magnetic resonance imaging of the spine, should be performed based on the history and physical examination. Bracing of the lower extremities may be used to prevent heel cord contractures in older or more impaired children. Interventions, such as serial casting, botulinum toxin injections, or surgery for gastrocnemius or heel-cord lengthening, are sometimes needed in refractory cases.

PREP Pearls

- Toe walking may be a normal stage in gait development, especially in children younger than 4 years.
- Pathologic causes of toe walking, such as neuromuscular disorders, should be ruled out by a thorough history and physical examination.
- Toe walking that persists beyond 5 years of age is more common in children with neurocognitive disorders.

American Board of Pediatrics Content Specification(s):

- Know that toe-walking may be a normal stage in gait development or may reflect underlying pathologic conditions such as neuromuscular disease

Suggested Reading:

- Engstrom P, Tedroff K. The prevalence and course of idiopathic toe-walking in 5-year-old children. Pediatrics. 2012;130:279-284. doi:10.1542/ peds.2012-0225
- Smith BG. Lower extremity disorders in children and adolescents. Pediatr Rev. 2009;30:287-294. doi:10.1542/pir.30-8-287

Item 177

The mother of a 15-year-old boy reports that ever since her son joined the freshman wrestling team, he has been having worsening exercise-induced shortness of breath and wheezing. He has been using albuterol before activity as a preventative measure but has still been having breakthrough symptoms. It has become much worse this fall season since he began performing his conditioning regimen at the local park. He now requires albuterol on an average of 3 nights a week. The mother is concerned that he started panting, coughing, and wheezing in the middle of a wrestling tournament last weekend and had to stop and take 2 puffs of albuterol with a spacer.

Of the following, the advice you are MOST likely to provide is that the boy should

A. be evaluated for gastroesophageal reflux
B. be evaluated for poorly controlled asthma
C. be evaluated for vocal cord dysfunction
D. initiate a conditioning regimen to get back into shape
E. initiate a warm-up regimen prior to exercise

Item 177 I-C Preferred Response: B

The adolescent described in the vignette should be evaluated for poorly controlled asthma. He has breakthrough symptoms of exercise-induced bronchospasm (EIB) requiring rescue short-acting beta agonist (SABA), such as albuterol, despite prophylactic SABA use. In addition, he requires rescue SABA 3 or more nights per week and is experiencing limitations to the quality of his life. On the basis of the 2007 National Heart, Lung, and Blood Institute guidelines for asthma, these symptoms indicate lack of adequate asthma control. Although he does not appear to be deconditioned since he has been participating in a conditioning regimen, it is possible that he could benefit from further cardiovascular conditioning. Nevertheless, his symptoms of wheezing and shortness of breath, responsive to albuterol, suggest reversible bronchospasm as the cause of his symptoms.

Exercise-induced bronchospasm is defined as transient narrowing of the lower airway following vigorous exercise that may appear with or without clinically recognized asthma. Typical symptoms of shortness of breath, chest tightness, and cough begin 3 to 5 minutes after the onset of vigorous or strenuous exercise, peak within 10 to 15 minutes, and resolve by 60 minutes. The EIB is thought to result from changes in airway physiology triggered by the respiratory water loss that occurs at high ventilation rates, associated with airway cooling and dehydration, which lead to increased osmolarity of the airway surface. This induces degranulation of airway mast cells with release of chemical mediators that stimulate bronchoconstriction. The EIB is usually measured as a reduction in forced expiratory volume 1 second after exercise (decrease of 10%-15% of the pre-exercise value). This adolescent did not have hoarseness or stridor that would be associated with paradoxical vocal cord dysfunction or exercise-induced laryngeal dysfunction. Children with vocal cord dysfunction also show inadequate response to SABA and have flattening of their inspiratory curve on the flow-volume loop during an exacerbation.

The combination of general measures and pharmacologic intervention can prevent EIB in almost all asthmatics. A major goal is to ensure that exercise is not avoided by patients with EIB. Asthmatics should exercise as much as desired and should be encouraged by the fact that athletes have won Olympic medals and played professional sports, in spite of symptomatic asthma.

Improving a patient's cardiovascular fitness reduces the minute ventilation required for a given level of exercise, thereby decreasing the stimulus for bronchoconstriction. Similarly, bronchoconstriction is lessened when the inspired gas is warmer and more humid. Patients should be instructed to breathe through a loosely fitting scarf or mask when exercising in cold, dry conditions. Since environmental allergens may trigger attacks, activities may need to be avoided, or performed at other venues, during high-pollen or poor air quality days. Pre-exercise warm-up may be helpful in reducing the severity of EIB. Pre-exercise warm-up should be done at 60% to 80% maximum heart rate to provide partial attenuation of EIB; this refractory period typically may last from 1 to 3 hours and occasionally 4 hours. However, this may not alleviate the need for

medications such as SABA. Albuterol plus a warm-up gives better protection than the warm-up or albuterol alone.

Asymptomatic gastroesophageal reflux disease (GERD) was hypothesized by clinicians in the past to be associated with uncontrolled asthma, and patients were often given anti-reflux medication. However, recent randomized controlled trials have shown that treatment of presumed asymptomatic reflux ("silent reflux") with proton pump inhibitors in adults and children with moderate to severe persistent asthma does not improve asthma control. The association between symptomatic GERD and EIB is controversial, and although there are reports of exertional GERD in healthy individuals, most studies have demonstrated no significant correlations.

PREP Pearls

- Exercise-induced bronchospasm (EIB) may be a sign of poorly controlled asthma.
- Exercise-induced bronchospasm should not limit the child's ability to participate in physical activities as long as the appropriate types of physical activities and management are recommended.
- Treatment of EIB includes conditioning regimens, pre-exercise warm-up routines, ensuring adequate hydration, monitoring pollen and air quality triggers, and pharmacologic therapies.

American Board of Pediatrics Content Specification(s):

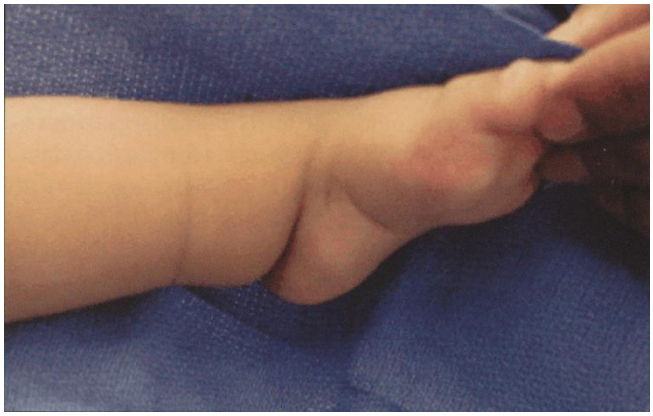
- Know that exercised-induced asthma may be a sign of poorly controlled asthma

Suggested Reading:

- Maturo S, Hill C, Bunting G, et al. Pediatric paradoxical vocal-fold motion: presentation and natural history. Pediatrics. 2011;128(6):e1443-e1449. doi:10.1542/peds.2011-1003
- Miller MG, Weiler JM, Baker R, Collins J, D'Alonzo G. National Athletic Trainers' Association position statement: management of asthma in athletes. J Athl Train. 2005;40(3):224-245
- Weiler JM, Anderson SD, Randolph C, et al; American Academy of Allergy, Asthma and Immunology; American College of Allergy, Asthma and Immunology; Joint Council of Allergy, Asthma and Immunology. Pathogenesis, prevalence, diagnosis, and management of exercise-induced bronchoconstriction: a practice parameter. Ann Allergy Asthma Immunol. 2010;105(6 suppl):S1-S47. doi:10.1016/j.anai.2010.09.021

Item 178

The newborn infant of a 28-year-old primigravida has severe respiratory distress and is noted to have pseudoepicanthus, flattened ears and nose, and bilateral club feet (Item Q 178). There is no family history of renal failure. Abdominal ultrasonography reveals bilaterally enlarged echogenic kidneys with poor corticomedullary differentiation; there are no cysts or hydronephrosis. The liver, spleen, pancreas, and gall bladder are reported to be normal.



ITEM Q178: Deformity of the foot as described for the infant in the vignette.

Of the following, the MOST likely cause of this neonate's condition is

A. autosomal recessive polycystic kidney disease
B. bilateral Wilms tumor
C. multicystic dysplastic kidney
D. nephronophthisis
E. renal vein thrombosis

Item 178 Preferred Response: A

The newborn described in the vignette has Potter syndrome associated with autosomal recessive polycystic kidney disease (ARPKD). Potter syndrome/Potter sequence or oligohydramnios sequence is the characteristic phenotypic features in the neonate caused by oligohydramnios. The characteristic features include pulmonary hypoplasia (respiratory distress in the newborn), facial appearance (pseudo-epicanthus, flattened ears and nose, recessed chin), and limb abnormalities (club feet and hip dislocation).

Classic Potter syndrome has been described in association with bilateral renal agenesis. Potter syndrome, as in the child in the vignette, occurs in association with ARPKD. In patients with ARPKD, ultrasonography demonstrates that the kidneys are large and echogenic, with decreased corticomedullary differentiation, in which macrocysts are not typically seen. Hepatomegaly and increased echogenicity are usually seen in severe cases of ARPKD and indicate the development of congenital hepatic fibrosis. Liver involvement is almost always seen in patients with ARPKD as malformation of the developing biliary ducts. Initially the liver functions are normal, with most patients developing portal hypertension, liver enlargement, and injury over time.

Autosomal dominant polycystic kidney disease (ADPKD) is considered an adult-onset disease, with clinical manifestations developing later in life. Patients with ADPKD have renal macrocysts that are visible on ultrasonographic imaging of the kidneys. However, with improvement in ultrasonographic techniques, ADPKD is increasingly being reported in the pediatric population including fetuses and neonates. Oligohydramnios along with the Potter phenotype has been described in association with ADPKD; however, this presentation is very rare. Patients with ADPKD usually have a positive family history of renal failure. Patients with multicystic dysplastic kidney (MCDK) are usually asymptomatic. MCDK is usually suspected based on renal abnormalities detected on antenatal ultrasonography or in neonates with abdominal mass on examination. Classic findings on renal ultrasonography include multiple noncommunicating cysts with intervening dysplastic renal tissue. The contralateral normal kidney has increased risk for congenital renal anomalies such as vesicoureteral reflux.

Nephronophthisis is a genetically heterogeneous disorder characterized by autosomal recessive inheritance, reduced urinary concentration, and chronic tubulointerstitial nephritis. These patients have polyuria, a bland urinary sediment on presentation, and slow progression to renal failure and end-stage renal disease. Based on the median age at onset of end-stage renal failure, nephronophthisis may be infantile (1 year), juvenile (13 years; most common form), or adolescent (19 years). Renal ultrasonography shows normal or slight decrease in renal size with increased echogenicity.

Renal vein thrombosis is the most common cause of non-catheter-associated thrombosis in the newborn. Prematurity, perinatal asphyxia, shock, dehydration, sepsis, polycythemia, cyanotic congenital heart disease, and maternal diabetes have been associated with an increased risk for renal vein thrombosis. Flank mass, thrombocytopenia, and hematuria are the classic features associated with renal vein thrombosis. Ultrasonography of the kidney is the imaging modality of choice for

diagnosing renal vein thrombosis in a newborn. In the early stages of thrombosis, the kidneys appear swollen and echogenic and this gradually evolves into loss of corticomedullary differentiation followed by scarring and decrease in renal size. Color Doppler examination on renal ultrasonography will show absent intrarenal and renal venous flow in the early stages of thrombosis.

The classic presentation of Wilms tumor is abdominal swelling with or without associated symptoms. Patients with Wilms tumor rarely present in the neonatal period. Neonatal presentation though rare has been reported in association with nonimmune hydrops, is usually unilateral, and is not usually associated with the multiple congenital malformation syndromes described in association with Wilms tumor.

PREP Pearls

- Potter syndrome/Potter sequence/oligohydramnios sequence is the characteristic phenotypic features in the neonate caused by oligohydramnios. The characteristic features include pulmonary hypoplasia (respiratory distress in the newborn), facial appearance (pseudoepicanthus, flattened ears and nose, recessed chin), and limb abnormalities (club feet and hip dislocation).
- Potter syndrome type 1 occurs in association with ARPKD.
- On ultrasonography, patients with ARPKD have enlarged echogenic kidneys without obvious renal cysts whereas ADPKD is associated with macrocysts.

American Board of Pediatrics Content Specification(s):

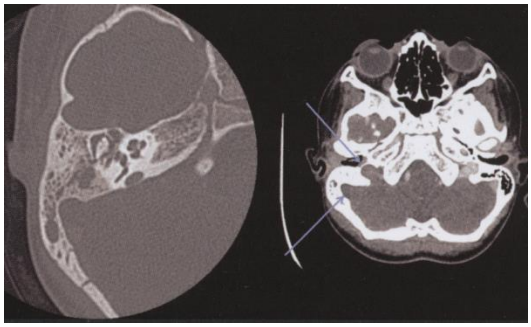
- Know the clinical presentation of autosomal-recessive polycystic kidney disease in neonates, infants, and children with congenital hepatic fibrosis

Suggested Reading:

- Elder JS. Congenital anomalies and dysgenesis of the kidneys. In: Kliegman RM, Stanton BMD, St Geme J, Schor N, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Elsevier Saunders; 2011:1827-1829
- Porter CC, Avner ED. Autosomal recessive polycystic kidney disease In: Kliegman RM, Stanton BMD, St Geme J, Schor N, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Elsevier Saunders; 2011:1796-1798
- Schwartz MZ, Shaul DB. Abdominal masses in the newborn. Pediatr Rev. 1989;11:172-179. doi: 10.1542/pir.11-6-172

Item 179

An 8-year-old boy who was previously well is seen in the emergency department with a 3-day history of neck pain, high-spiking fever, nausea, and vomiting. One day before presentation, the parents noted a "bump" behind his right ear. The patient has a history of recurrent ear infections and has been receiving a "pink antibiotic" for the last several weeks. On physical examination, his temperature is 37°C, pulse rate is 70 beats/min, respiratory rate is 20 breaths/min, and blood pressure is 94/60 mm Hg. Growth parameters are at the 50th percentile for age. The boy is ill-appearing but alert and interactive during the examination. The right tympanic membrane is erythematous and dull-appearing. His right pinna is displaced anteriorly and there is right posterior auricular erythema and swelling that is tender to palpation. Neck motion elicits pain, but meningismus is absent. The remainder of the physical examination findings, including neurologic assessment, is normal. An axial bone targeted computed tomography scan reveals complete tympanomastoid opacification, and contrast enhancement shows opacification of the right sigmoid sinus and jugular bulb (Item Q179).



ITEM Q179: Axial bone targeted computed tomography scan reveals complete tympanomastoid opacification; contrast enhancement shows opacification of the right sigmoid sinus and jugular bulb.

Of the following, the MOST appropriate antimicrobial regimen for this patient is

A. ceftazidime and ciprofloxacin
B. ceftazidime and clindamycin
C. ceftazidime and doxycycline
D. cefotaxime and trimethoprim–sulfamethoxazole
E. ceftazidime and vancomycin

Item 179 Preferred Response: E

The patient described in the vignette has complications of middle ear disease, including mastoiditis and sigmoid sinus thrombosis, and warrants prompt clinical and surgical evaluation and the initiation of broad-spectrum antimicrobial therapy. Ceftazidime and vancomycin cover both gram-negative and gram-positive pathogens likely to be causing disease and are the most appropriate initial antimicrobial agents for this patient. Empirical antimicrobial therapy for patients with central nervous system (CNS) complications of middle ear disease should include bactericidal agents that have adequate CNS penetration and specifically targeting the likely pathogens.

The bacteria most likely to cause mastoiditis in children include Streptococcus pyogenes, Streptococcus pneumoniae (including multidrug-resistant S pneumoniae), and Staphylococcus aureus (including methicillin-resistant S aureus). Vancomycin is a glycopeptide antibiotic that inhibits bacterial cell wall synthesis by blocking glycopeptide polymerization. Vancomycin is bactericidal against gram-positive organisms, such as S aureus, S pyogenes, and S pneumoniae. Neither clindamycin nor doxycycline would be appropriate for the patient described in the vignette because these drugs are bacteriostatic (inhibit bacterial protein synthesis) and lack activity against some of the likely pathogens implicated in CNS complications of middle ear disease. Trimethoprim-sulfamethoxazole is bactericidal against S aureus in sites other than the CNS; however, it is only bacteriostatic against S aureus in the CNS and, therefore, is not used as a first-line agent in this situation. In addition, trimethoprim-sulfamethoxazole does not have activity against S pyogenes.

Pseudomonas aeruginosa should be considered as a possible pathogen in children with a history of recurrent otitis media and recent antibiotic use, such as the patient described in the vignette. P aeruginosa most commonly causes infection in the setting of tympanic membrane perforation and chronic middle ear infection. Ceftazidime is a β-lactam antibiotic, with activity against most Pseudomonas, that inhibits bacterial cell wall synthesis by binding to penicillin-binding proteins, thereby inhibiting cell wall biosynthesis. Ceftazidime is bactericidal against gram-negative pathogens, including Pseudomonas. Ciprofloxacin is a fluoroquinolone antibiotic with activity against Pseudomonas and other gram-negative pathogens that inhibits DNA gyrase and promotes breakage of double-stranded DNA. It is bactericidal but not generally recommended for use in children younger than 18 years. In addition, it is unnecessary to use 2 agents (eg, ceftazidime and ciprofloxacin) with activity against Pseudomonas to treat CNS complications of middle ear disease.

A history of recurrent otitis media is a risk factor for mastoiditis. Extracranial complications of mastoiditis (and otitis media) include cholesteatoma, subperiosteal abscess, facial nerve palsy, hearing loss, labyrinthitis, osteomyelitis, and Bezold abscess (abscess in the sternocleidomastoid muscle). Intracranial complications of mastoiditis (and otitis media) are uncommon and include meningitis, brain (eg, temporal lobe, cerebellum) abscess, epidural or subdural empyema, and carotid artery and venous sinus thromboses.

PREP Pearls

- Empirical antimicrobial therapy for patients with CNS complications of middle ear disease should include bactericidal agents that have adequate CNS penetration and target the likely pathogens (ie, Streptococcus pyogenes, Streptococcus pneumoniae, Staphylococcus aureus, and Pseudomonas aeruginosa).
- Intracranial complications of mastoiditis (and otitis media) are uncommon and include meningitis, brain (eg, temporal lobe, cerebellum) abscess, epidural or subdural empyema, and carotid artery and venous sinus thromboses.

American Board of Pediatrics Content Specification(s):

- Know the central nervous system complications of middle ear disease

Suggested Reading:

- Bradley JS, Sauberan JB. Antimicrobial agents. In: Long SS, Pickering LK, Prober CG, eds. Principles and Practice of Pediatric Infectious Diseases. 4th ed. Philadelphia, PA: Saunders Elsevier; 2012:1453-1483
- Klein JO, Bluestone CD. Otitis media. In: Feigin RD, Cherry JD, DemmlerHarrison GJ, Kaplan SL, eds. Feigin and Cherry's Textbook of Pediatric Infectious Diseases. 6th ed. Philadelphia, PA: Saunders Elsevier; 2009:216-237
- Lewis K, Shapiro NL, Cherry JD. Mastoiditis. In: Feigin RD, Cherry JD, Demmler-Harrison GJ, Kaplan SL, eds. Feigin and Cherry's Textbook of Pediatric Infectious Diseases. 6th ed. Philadelphia, PA: Saunders Elsevier; 2009:238-244
- Pelton SI. Otitis media. In: Long SS, Pickering LK, Prober CG, eds. Principles and Practice of Pediatric Infectious Diseases. 4th ed. Philadelphia, PA: Saunders Elsevier; 2012:213-219
- Wald ER, Conway JH. Mastoiditis. In: Long SS, Pickering LK, Prober CG, eds. Principles and Practice of Pediatric Infectious Diseases. 4th ed. Philadelphia, PA: Saunders Elsevier; 2012:227-230

Item 180

An adolescent girl presents to your office with severe ano-rectal pain, discharge, and tenesmus of 4 days' duration. She has noted some blood in the discharge and occasional loose stools. She has negative past medical and family histories and has not traveled recently. She says she has never had vaginal intercourse and has no history of drug use. On physical examination, she has a low-grade fever and is in pain but is well-appearing. You find a few vesicles and ulcers in the perianal area. The remainder of her physical examination findings are normal. You arrange for a gastrointestinal consultation.

Of the following, the organism MOST likely responsible for the girl's proctitis is

A. cytomegalovirus
B. Entamoeba histolytica
C. herpes simplex virus
D. Shigella dysenteriae
E. Treponema pallidum.

Item 180 I-C Preferred Response: C

The girl in the vignette has symptoms and signs consistent with a diagnosis of herpes simplex virus (HSV) proctitis. She most likely acquired the infection as a result of unprotected receptive anal intercourse. When asking about sexual activity, the physician should ask about anal and oral along with vaginal intercourse because this information may not be shared voluntarily. Many females practicing receptive anal intercourse are only considering pregnancy risks and may not be aware of the risk for sexually transmitted infections (STIs). The most common STIs that cause proctitis are Neisseria gonorrhea, Chlamydia trachomatis, and less commonly HSV, lymphogranuloma venereum, and Treponema pallidum.

Condom use is an effective barrier to the transmission of infections and should be discussed with all adolescents. In addition to anal receptive intercourse, the -anal-oral route may also be responsible for transmission of fecal organisms. Oral-penile contact is termed fellatio, oral-vaginal sex is termed cunnilingus, and oral-anal sex is termed analingus. Adolescent girls need to learn about the need for male partners to wear a condom during acts of fellatio. For cunnilingus and analingus, use of barrier protection such as dental dams (latex sheets) should be encouraged. Protection when using sex toys to prevent the exchange of body fluids should also be discussed.

Organisms that may be transmitted by direct rectal inoculation or the oral route include Shigella, Salmonella, Entamoeba, and Campylobacter. None of these organisms cause vesicles or ulcers, as with the girl in the vignette. Perianal infection with cytomegalovirus would be extremely unlikely in an immunocompetent host, such as the girl in the vignette.

PREP Pearls

- Teenagers who engage in oral and anal intercourse and not vaginal sex may consider themselves virgins and may not disclose this behavior, unless asked directly.
- Teenagers underestimate the risk of STIs from oral and anal sexual contact.
- Barrier methods should be used during all forms of sexual contact to prevent STIs.

American Board of Pediatrics Content Specification(s):

- Understand the importance of counseling adolescents about the necessity of condom use during anal as well as vaginal intercourse

Suggested Reading:

- Cavanaugh RM Jr. Screening adolescent gynecology in the pediatrician's office: have a listen, take a look. Pediatr Rev. 2007;28:332-342. doi:10.1542/pir.28
- Chayavichitsilp P, Buckwalter JV, Krakowski AC, Friedlander SF. Herpes simplex. Pediatr Rev. 2009;30:119-130. doi:10.1542/pir.30-4-119
- Committee on Adolescence. Contraception and adolescents. Pediatrics. 2007;120:1135-1148. doi:10.1542/peds.2007-2535
- Martino SC, Elliott MN, Corona R, Kanouse DE, Schuster MA. Beyond the "big talk": the roles of breadth and repetition in parent-adolescent communication about sexual topics. Pediatrics. 2008;121: e612-e618. doi:10.1542/peds.2007-2156

Item 181

You are seeing a 3-year-old girl who has cognitive and motor impairment. She recently completed a course of antibiotics for the treatment of a right middle lobe pneumonia. Since 6 months of age, she has been treated with lansoprazole for frequent postprandial regurgitation and a presumptive diagnosis of gastroesophageal reflux. Her present dose of lansoprazole is 15 mg twice daily. She is fed pureed foods and a proprietary liquid formula (at an energy concentration of 1 kcal/mL). She also receives occasional sips of water and apple juice. Her parents report a significant reduction in spitting-up episodes since about 12 months of age. Two months ago, however, she began to have frequent bouts of gagging and coughing during feedings.

Of the following, the MOST appropriate next step in this girl's evaluation is to obtain

A. esophageal pH monitoring
B. milk scintigraphy
C. upper gastrointestinal endoscopy
D. upper gastrointestinal tract radiographic series
E. videofluoroscopic swallowing study

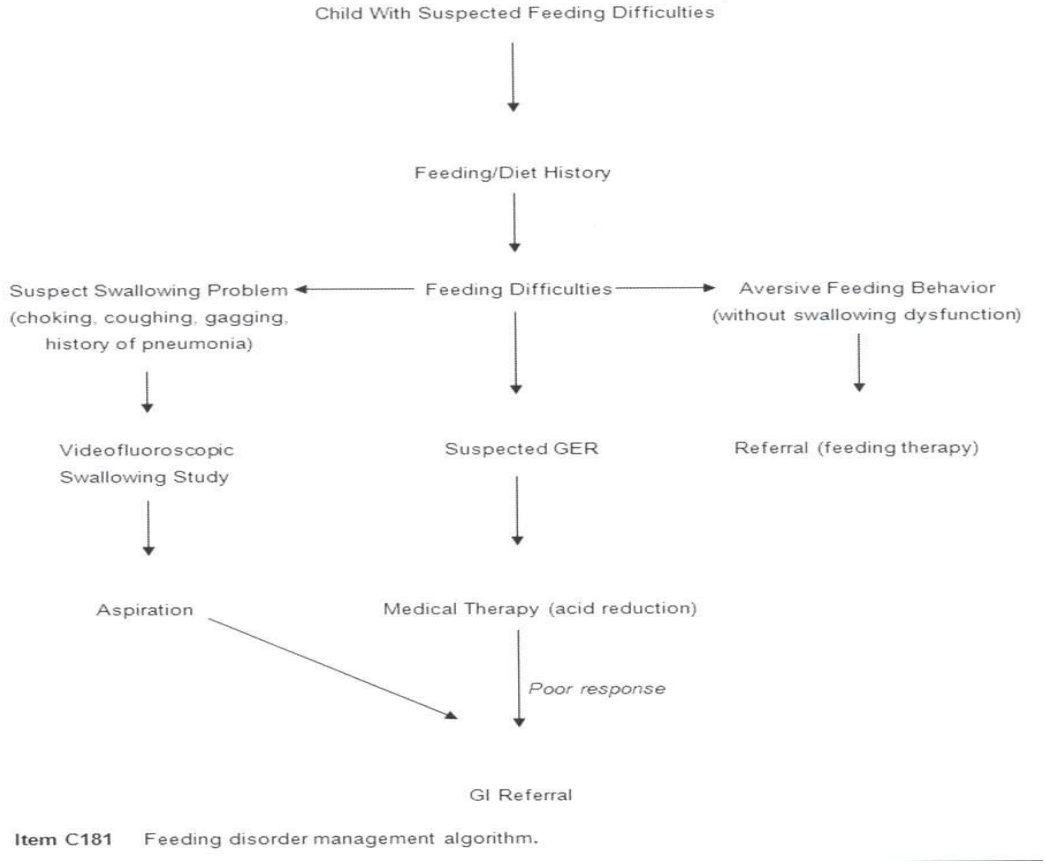
Item 181 TE SBP_S Preferred Response: E

Feeding and swallowing difficulties often complicate the treatment of infants and children with developmental disabilities. A review of published clinical studies reveals that 30% to 90% of individuals (of all age ranges) with major motor or cognitive disorders exhibit feeding difficulties. In nonambulatory children with cerebral palsy, feeding problems are associated with evidence of malnutrition in up to 90% of patients. The most frequently identified feeding difficulties in these neurodevelopmentally challenged individuals include gastroesophageal reflux (GER), oral motor dysfunction (ie, the inability to coordinate movement of an oral bolus retrograde), swallowing disorders, and aversive feeding behaviors. The developmentally disabled girl in the vignette is experiencing "choking and gagging" during feedings. Regurgitation, prominent during infancy and requiring acid reduction therapy, has improved significantly. These historical data point to a significant swallowing problem, which if undiagnosed, may lead to recurrent episodes of aspiration (especially for thin liquids, such as juice or water). The most appropriate method to initially evaluate oral motor and pharyngoesophageal function in this setting is a videofluoroscopic swallowing study (VFSS).

Although GER remains an important cause of aspiration in neurologically impaired individuals, oropharyngeal aspiration, characterized by aspiration of solids or liquids below the level of the vocal cords, may represent a more prevalent cause of secondary pneumonia in this patient group. Early diagnosis and management of swallowing dysfunction in high-risk patients are critically important to prevent chronic lung disease. Aspiration as a result of a swallowing disorder may either be overt (associated with coughing or gagging during feedings) or silent (not associated with acute symptoms). One recent report in 300 children with feeding difficulties evaluated with VFSS found that children with neurodevelopmental delay were significantly more likely to exhibit oropharyngeal aspiration than those with normal neurologic function. Furthermore, neurologically challenged patients were 4 times as likely to manifest silent aspiration as were patients without neurologic dysfunction.

In the child with feeding difficulties, a comprehensive evaluation will involve an assessment by a licensed feeding therapist and diagnostic studies, including a VFSS and determination of GER (either by intraesophageal pH monitoring or a multichannel intraluminal impedance study). However, when choking or gagging accompanies feedings (as for the girl in the vignette) or when aspiration is suspected (particularly when postprandial vomiting is not a prominent symptom), the VFSS is of paramount importance. This fluoroscopic study not only may identify aspiration of both thin and thick liquids but also will detect posterior pharyngeal pooling of liquids, with or without associated laryngeal penetration. Accordingly, the VFSS permits an evaluation of patients who may not exhibit aspiration but who are at risk for this complication of swallowing dysfunction. In some cases, milk scintigraphy may be useful in detecting overt pulmonary aspiration that occurs during feedings. An upper gastrointestinal tract series will identify major anatomical abnormalities (eg, partial thoracic stomach and malrotation), but it is not useful in the routine management of feeding and swallowing disorders. A diagnostic upper endoscopy is indicated in this setting only if major esophageal complications (eg,

esophagitis and esophageal stricture) are suspected. Item C181, page C-143 presents a diagnostic algorithm that may be followed before subspecialist referral.



Item C181 Feeding disorder management algorithm.

PREP Pearls

- Feeding disorders comprise the most common problems leading to malnutrition in developmentally disabled children.
- The most prevalent cause of aspiration in disabled children is oral-motor and swallowing dysfunction.
- Choking or gagging with feedings signals the presence of oropharyngeal dysfunction.

American Board of Pediatrics Content Specification(s):

- Know that recurrent aspiration can recur with swallowing disorders independent of gastroesophageal reflux

Suggested Reading:

- Arvedson A, Rogers B, Buck G, Smart P, Msall M. Silent aspiration prominent in children with dysphagia. Int J Pediatr Otorhinolaryngol.
- 1994;28:173-181. doi:10.1016/0165-5876(94)90009-4
- Arvedson JC. Swallowing and feeding in infants and young children. GI Motility Online. May 16, 2006. doi:10.1038/gimol7
- Mohan P. Aspiration in infants and children. Pediatr Rev. 2002;23:330-331. doi:10.1542/pir.23-9-330

- Newman LA, Keckley C, Petersen MC, Hamner A. Swallowing function and medical diagnoses in infants suspected of dysphagia. Pediatrics. 2001:e108:106. doi:10.1542/peds.108.6.e106
- Schwarz SM. Feeding disorders in children with developmental disabilities. Inf. Young Childr. 2003;16:317-330
- Sullivan PB, Lambert B, Roe M, et al. Prevalence and severity of feeding and nutritional problems in children with neurological impairment: Oxford feeding study. Dev Med Child Neurol. 2000;42:674-680. doi: 10.1111/j.1469-8749.2000.tb00678.x
- Weir KA, McMahon S, Taylor S, Chang AB. Oropharyngeal aspiration and silent aspiration in.children. Chest. 2011;140:589-597

Item 182

An 11-year-old boy who has a history of attention-deficit/ hyperactivity disorder comes to see you because of uncomfortable feelings in his legs and frequent urges to move his legs. The symptoms are worse during rest and decrease when he is moving. They are also worse during the evening and at night. The results of his physical examination, including a thorough neurologic assessment, are unremarkable.
Of the following, the test MOST likely to show an abnormality contributing to this condition is

A. cranial magnetic resonance imaging
B. electroencephalogram
C. serum creative kinase level
D. serum ferritin level
E. serum potassium and calcium levels

Item 182 Preferred Response: D

The child in the vignette has symptoms characteristic of restless leg syndrome (RLS). RLS is defined by 4 criteria: (1) an uncomfortable sensation or unexplainable urge to move, (2) increased symptoms when at rest; (3) decreased symptoms with movement; and (4) worsening of symptoms in the evening or night. Diagnosis is based primarily on clinical findings including the child's own description of the unusual sensations he/she experiences. A sleep study can help confirm the diagnosis, and anterior tibial electromyography may occasionally be indicated. Central nervous system imaging and electroencephalography are not useful to the diagnosis. Because this is a primarily neurologic/dopamine-mediated condition, one would not expect abnormalities in muscle enzymes. Likewise, electrolytes (potassium, calcium) are normal in RLS. Serum ferritin levels less than 50 ng/mL (112 pmol/L) have been found in children with RLS. A recent small study showed that iron supplementation led to both an improvement in ferritin values and RLS symptoms in 90% (27/30) of affected children.

Iron deficiency is one of the most common nutrient deficiencies in both the developed and developing world. Iron deficiency anemia (IDA) has been associated not only with growth impairment, but also with neurocognitive and behavioral delays. Some studies have indicated that even after iron stores are rebuilt the developmental consequences of early iron deficiency may not be reversible. In addition to the direct effects of inadequate iron stores, iron deficiency promotes lead absorption and its neurologic consequences. Iron deficiency without anemia also has significant effects. Iron is found in nearly all cells of the body and, in addition to being crucial for adequate oxygen delivery to the tissues, is a necessary cofactor for many enzymes. It is particularly crucial to the dopaminergic system in the central nervous system. Among the nonhematologic neurologic conditions that have been associated with or have a proposed relationship with iron deficiency are breath-holding spells, attention-deficit/hyperactivity disorder, syncope, stroke, febrile seizures, and RLS.

PREP Pearls

- Iron deficiency, with or without anemia, is associated with growth impairment, neurocognitive delays, and behavioral problems as well as other neurologic abnormalities.
- Some iron deficiency-related conditions such as cognitive delay may not be reversible even when iron stores have been restored.
- Restless leg syndrome has been associated with low ferritin levels, and treatment with iron has been demonstrated in many cases to improve both the ferritin level and the symptoms of the condition.

American Board of Pediatrics Content Specification(s):

- Understand that iron deficiency causes nonhematologic effects such as behavior and learning disturbances

Suggested Reading:

- Baker RD, Greer FR, and the Committee on Nutrition. Clinical report: diagnosis and prevention of iron deficiency and iron deficiency anemia in infants and young children (0-3 years of age). Pediatrics. 2010;126:10401050. doi:10.1542/peds.2010-2576
- Drumer JS, Quraishi GH. Restless legs syndrome, periodic leg movements, and periodic limb movement disorder in children. Pediatr an North Am. 2011;58:591-620. doi:10.1016/j.pc1.2011.03.005
- Mohri I, Kato-Nishimura K, et al. Evaluation of oral iron treatment in pediatric restless legs syndrome (RLS). Sleep. 2012;13(4):429-432. doi:10.1016/j.sleep.2011.12.009

Item 183

You are evaluating a 15-year-old boy in the emergency department who complains of a severe headache of several weeks' duration; the pain does not change throughout the day. He does not report any previous medical problems but, on review of systems, states that he has had some "blurry vision" over the past week. His physical examination is remarkable only for a blood pressure of 195/130 mm Hg and papilledema. You plan the boy's evaluation and elect to begin treatment.

Of the following, the MOST appropriate medication to minister via continuous infusion is

A. diazoxide
B. fenoldopam
C. hydralazine
D. labetalol
E. phentolamine

Item 183 Preferred Response: D

Hypertension, systolic or diastolic blood pressure greater than the 95th percentile on 3 or more occasions, occurs in approximately 5% of the pediatric population. Pediatric hypertensive emergencies, defined as a severe symptomatic elevation in blood pressure with evidence of acute end organ damage, are rare (less than 1% of emergency department visits). Hypertensive emergencies can be seen in newly diagnosed patients as well as those with chronic hypertension who have an acute elevation. No specific systolic or diastolic blood pressure defines an emergency, and therefore it is imperative to recognize the end organ dysfunction to quickly treat the hypertension and prevent morbidity and mortality. The most commonly affected end organs are the brain (altered mental status, seizures, edema, increased intracranial pressure), kidneys (renal insufficiency), heart (heart failure), and eyes (papilledema, retinal hemorrhages). Hypertensive encephalopathy (altered mental status or seizures) is the most common manifestation of a hypertensive emergency and has been reported in up to 50% of cases.

Assessment of hypertensive emergencies should include a thorough history and physical examination to rule out potential causes (head trauma, intracranial masses, abdominal tumors, hyperthyroidism, etc). The physical examination should include a 4-extremity blood pressure measurement to assess for a possible aortic coarctation. Risk factors for hypertensive emergencies include chronic hypertension (especially those who are not compliant with medications), chronic kidney disease, genitourinary abnormalities, renal vascular disease, acute glomerulonephritis, illicit drug use, pheochromocytoma, and pregnancy. Ancillary studies should include electrolyte and renal function measurement to assess for renal disease as well as chest radiography and electrocardiography to evaluate for cardiac hypertrophy. Other studies that might be performed based on results of the history and physical examination include complete blood count, urine toxicology screen, pregnancy test, echocardiography, abdominal ultrasonography, and computed tomography of the head.

Intravenous antihypertensive medications should be started as soon as the emergency is recognized, with the goal of lowering the blood pressure to a level that will stop or at least mitigate the end organ damage. In general, lowering the blood pressure by approximately 20% to 25% over the first 8 hours will achieve this goal. Further lowering of the blood pressure may produce end organ damage because of lack of adequate perfusion secondary to altered autoregulation. The selection of the appropriate agent will depend on the suspected underlying cause but in general labetalol (a β-blocker) or nicardipine (a calcium channel blocker) are generally used as first-line agents because of their proven efficacy and their ability to be given in continuous infusions after an initial bolus.

Hydralazine, a direct arterial smooth muscle dilator, has been used as a first-line agent but has a slower onset of action and longer duration, and cannot be used as a continuous infusion. Phentolamine, an α-adrenergic blocker, would be indicated in the setting of excessive catecholamine levels as might occur with a pheochromocytoma or cocaine overdose. Fenoldopam, a peripheral dopamine receptor agonist, is less potent than either labetalol or nicardipine and, therefore, is not generally recommended for initial treatment.

Because of its ability to improve renal perfusion, it may be a useful drug in patients with renal insufficiency. Sodium nitroprusside is no longer generally recommended as a first-line agent because of potential cyanide toxicity. Diazoxide also is no longer recommended as a first-line agent because of its unpredictable blood pressure lowering effect.

PREP Pearls

- Pediatric hypertensive emergencies, defined as a severe symptomatic elevation in blood pressure with evidence of acute end organ damage, are rare but must be recognized and treated quickly.
- Hypertensive encephalopathy is the most common manifestation of a hypertensive emergency and has been reported in up to 50% of cases.
- The goal in the initial treatment of hypertensive emergencies is to lower the blood pressure by approximately 20% to 25% over the first 8 hours to stop or mitigate end organ damage.

American Board of Pediatrics Content Specification(s):

- Recognize and plan the therapy for a hypertensive emergency

Suggested Reading:

- Lande MB. Systemic Hypertension. In: Kliegman RM, Stanton BMD, St Geme J, Schor N, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Elsevier Saunders; 2011:1639-1647
- Flynn JT. Management of hypertensive emergencies and urgencies in children. UptoDate. Available online only for subscription
- Symons J, Enriquez B. Approach to hypertensive emergencies and urgencies in children. UptoDate. Available online only for subscription

Item 184

A 5-year-old girl with classic 21-hydroxylase deficiency (congenital adrenal hyperplasia) develops gastroenteritis with fever (up to 38.9°C), vomiting, and diarrhea. Her regular medications include hydrocortisone and fludrocortisone. Upon presentation to the emergency department, she is tired-appearing and remains febrile. Her pulse rate is 162 beats/min, blood pressure is 62/40 mm Hg, and capillary refill is poor. Laboratory tests drawn in the emergency department are still pending, but fingerstick glucose level is 42 mg/dL (2.3 mmol/L). The patient is treated with a bolus of normal saline to restore circulatory support.

Of the following, the MOST important therapy to administer to this patient next is

A. cortisone acetate intramuscularly and aldosterone intravenously
B. cortisone acetate intramuscularly and dextrose intravenously
C. dopamine and dextrose intravenously
D. hydrocortisone hemisuccinate and aldosterone intravenously
E. hydrocortisone hemisuccinate and dextrose intravenously

Item 184 Preferred Response: E

The girl described in this vignette has a known diagnosis of adrenal insufficiency due to 21-hydroxylase deficiency and is hemodynamically unstable during a febrile illness. Emergency treatment with high doses of steroids (stress dose steroids) is needed to mimic the high doses of steroids normally made under stress in patients with sufficient adrenal function.

The treatment of choice for adrenal crisis (of any cause) is fluids, hydrocortisone, and dextrose intravenously. Hydrocortisone is quick acting and at an emergency stress dose (100 mg/m2) saturates all steroid receptors, causing a mineralocorticoid and glucocorticoid effect. In addition, hypoglycemia is common during adrenal crisis, so the patient's glucose level should be checked and an intravenous bolus of dextrose given if hypoglycemia is present.

Pediatricians should recognize that some steroids, such as methylprednisolone, commonly used in asthma, have no mineralocorticoid activity at any dose and would not be appropriate for this patient. Hydrocortisone stress dosing guidelines are shown (Item C184).

Item C184. Stress Dosing Guideline for Hydrocortisone	
Physiologic replacement dosing	6-10 mg/m^2 per day
Oral stress dosing (minor febrile illness, taking oral well)	30 mg/m^2 per day three times daily
Intravenous stress dosing (before surgery, major illness but clinically stable)	50 mg/ m^2 one hour before procedure and then continued 50 mg/ m^2 per day divided 4 times daily as needed for continued stress
Intravenous stress dosing (adrenal crisis, sepsis, shock)	100 mg/ m^2 initial dose and then 100 mg/ m^2 per day divided 4 times daily(100 mg maximum per dose)

Because there is no available intravenous form of aldosterone, choices A and D would be incorrect, and instead one should choose a quick-acting steroid with mineralocorticoid activity, in this case, hydrocortisone at high doses. Intramuscular hydrocortisone is commonly given to patients to take at home before coming to the hospital if they are severely ill, but once in the emergency department, intravenous hydrocortisone should be used due to its quick onset of action. Dopamine would not be used in the acute setting of adrenal crisis because intravenous hydrocortisone must be given first.

PREP Pearls

- High-dose hydrocortisone is the steroid of choice for the treatment of adrenal crisis.
- A fingerstick glucose level should be checked on presentation in adrenal crisis, and a bolus of intravenous dextrose should be given if hypoglycemia is present.

American Board of Pediatrics Content Specification(s):

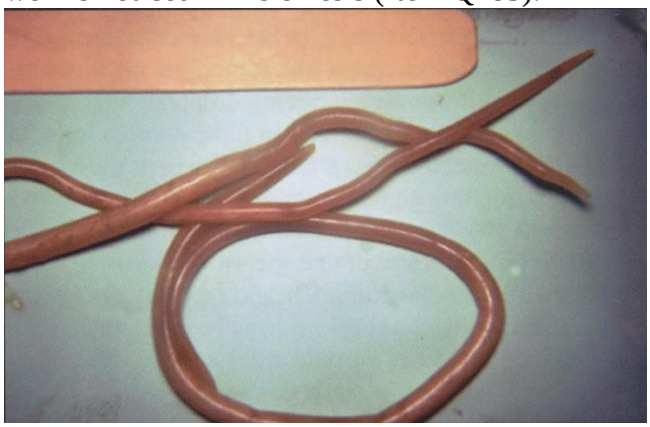
- Plan the treatment for an adrenal crisis in a patient with congenital adrenal hyperplasia

Suggested Reading:

- Shulman DI, Palmert MR, Kemp SF; Lawson Wilkins Drug and Therapeutics Committee. Adrenal insufficiency: still a cause of morbidity and death in childhood. Pediatrics. 2007;119:e484-e494. doi: 10.1542/ peds.2006-1612

Item 185

A 3-year-old boy whose family recently emigrated from Myanmar presents to your office with a 3-month history of increasing crampy abdominal pain, vomiting, and constipation. The mother reports her son has lost 4-pounds over this period and brought in several worms noticed in his emesis (Item Q185).



ITEM Q185: Worms as submitted by the patient.

On physical examination, he is afebrile. The boy weighs 12 kg (5th percentile), length is 93 cm (25th percentile), and head circumference is 49 cm (25th to 50th percentile). Abdominal examination reveals distension without organomegaly. Bowel sounds are normal and mild diffuse tenderness is appreciated.

Of the following, the MOST likely cause of this child's illness is

A. Ascaris lumbricoides
B. Entamoeba histolytica
C. Enterobius vermicularis
D. Giardia intestinalis
E. Trichuris trichiura

Item 185 Preferred Response: A

Ascaris lumbricoides is an intestinal nematode that is distributed worldwide but is most common in tropical regions such as Myanmar. It is the largest nematode (roundworm) infecting people, with adult worms reaching more than 30 cm. Adult worms live in the small intestine and when a male and female coinfect the same person, large numbers of eggs are excreted in the feces and form an embryonate in the soil. When ingested, the eggs mature into larvae which can invade the intestinal mucosa and spread to the lungs. In the lungs, the larvae further mature, penetrate the alveolar wall, ascend the bronchial trees, cross over the epiglottis, and are swallowed. Occasionally a worm may come through the nose during this migration or via regurgitation. Once back in the intestines these larvae then complete their development into adult worms. This life-cycle is depicted in Item C185, page C-146.

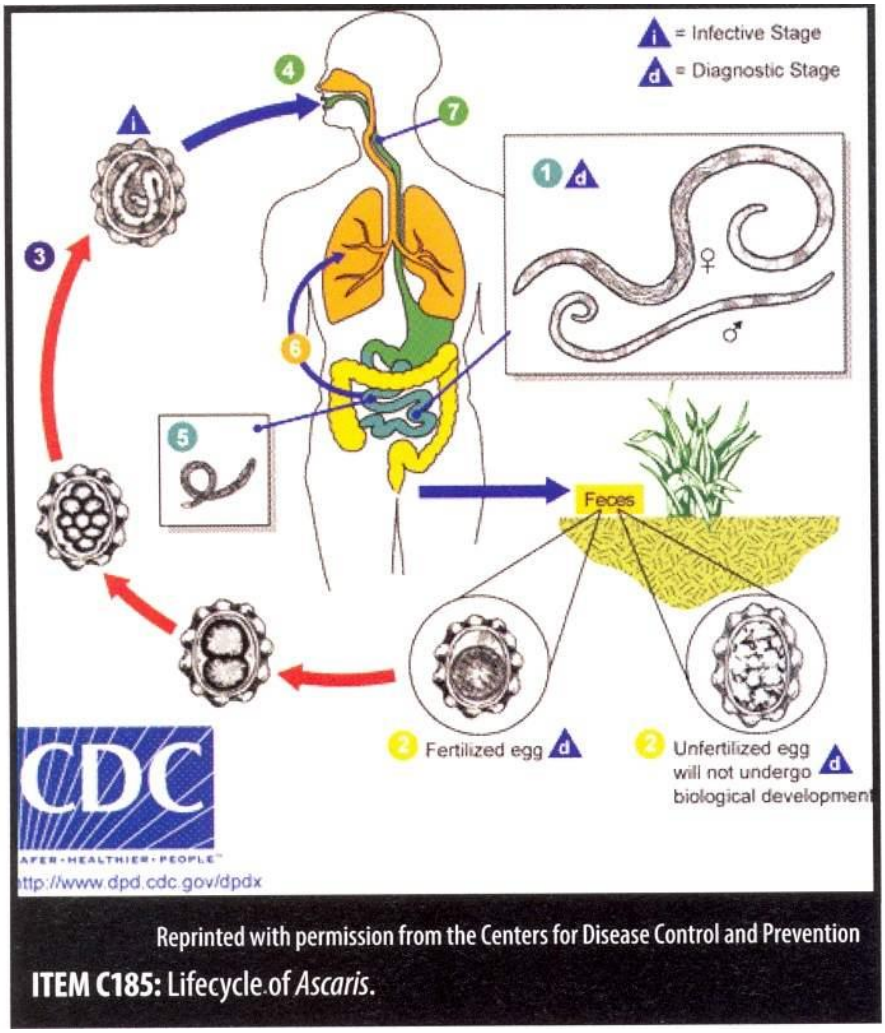
Most Ascaris infections are asymptomatic, however a heavy burden of infection can lead to malabsorption, malnutrition, and growth retardation. A large matted mass of worms can rarely cause intestinal obstruction requiring surgical intervention. In the absence of a visible adult worm, the diagnosis can be made through detection of eggs in stool specimens. The presence of malabsorption and malnutrition, the nation of origin, and the visible worm for the child in the vignette are most consistent with this diagnosis. Entamoeba histolytica is a protozoan and would not produce a visible worm, but such an infection can produce similar indolent gastrointestinal symptoms. More often symptomatic E histolytica infection is associated with dysentery with bloody stools, vomiting, and fever. Symptomatic Enterobius vermicularis or pinworm infection is associated with perirectal itching, not malabsorption, and the worms if seen are much smaller and localized to the perirectal region. Giardia intestinalis (formerly G lamblia) is a protozoan and infection may be associated with bloating and malabsorption, but visible worms are not present. Trichuris trichuria or whipworm when symptomatic produces loose stools that may be bloody or associated with rectal prolapse. Adult whipworms are only up to 4 cm in length.

PREP Pearls

- Ascaris lumbricoides is the largest nematode infecting people with adult worms reaching >30 cm in length.
- Most Ascaris infections are asymptomatic but malabsorption, malnutrition, or obstruction from large worm burden may occur.
- Pulmonary symptoms (eg, wheezing) may be seen during larval migration through the lungs.

American Board of Pediatrics Content Specification(s):

- Recognize the clinical manifestations of ascariasis



Suggested Reading:

- American Academy of Pediatrics. Ascaris lumbricoides infection. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:239-240
- American Academy of Pediatrics. Amebiasis. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:222-225
- American Academy of Pediatrics. Giardia intestinalis infections. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:333-335
- American Academy of Pediatrics. Pinworm infection (Enterobius vermicularis) In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:566-567

- American Academy of Pediatrics. Trichuriasis (whipworm infection. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:731-732
- Leder K, Weller PF. Ascariasis. UptoDate. Available online only for subscription

Item 186

A 15-year-old, African-American girl with systemic lupus erythematosus and lupus nephritis presents to your office for her annual health supervision visit. She has been doing very well and has not had any symptoms suggestive of active disease. Her current medications include prednisone, hydroxy-chloroquine, ranitidine, simvastatin, mycophenolate mofetil, enalapril, depot medroxyprogesterone, vitamin D with calcium, and a multivitamin. She asks you which of these medications are keeping her lupus disease under control.

Of the following, the response you are MOST likely to give is

A. enalapril, hydroxychloroquine, mycophenolate mofetil, prednisone, and simvastatin
B. enalapril, hydroxychloroquine, mycophenolate mofetil, and prednisone
C. hydroxychloroquine, mycophenolate mofetil, and prednisone
D. hydroxychloroquine, mycophenolate mofetil, prednisone, and simvastatin
E. mycophenolate mofetil, ranitidine, and prednisone

Item 186 Preferred Response: C

Systemic lupus erythematosus (SLE) is a complex disease managed with a multisystem approach, as noted in the girl in the vignette. A listing of the various drugs used to treat SLE is shown (Item C186, page C-147) Hydroxychloroquine maintains remission in patients who have SLE and controls skin involvement and disease activity. Mycophenolate mofetil is used as induction and maintenance therapy in lupus nephritis and controls disease activity. Prednisone is also used to control lupus disease activity. Enalapril would be used to control hypertension related to lupus nephritis but would not control SLE disease activity. Simvastatin can reduce the hypercholesterolemia seen in lupus patients and reduces the risk of early atherosclerosis, but it would not control lupus activity. Ranitidine is often used in patients who have lupus for gastrointestinal prophylaxis while on steroids but would not control lupus activity.

Prior to 2011, only 3 drugs-aspirin, hydroxychloroquine, and prednisone-were approved by the US Food and Drug Administration (FDA) for the treatment of SLE in adults. The FDA recently approved belimumab for adult SLE. There are no FDA-approved medications for the treatment of pediatric lupus patients. Live vaccines should be avoided while a lupus patient is on immunosuppressive therapy.

PREP Pearls

- Hydroxychloroquine helps to maintain remission and treats skin involvement in patients who have lupus.

American Board of Pediatrics Content Specification(s):

- Know which drugs are useful in the treatment of systemic lupus erythematosus

Suggested Reading:

- Levy DM, Kamphuis S. Systemic lupus erythematosus in children and adolescents. Pediatr Clin N Am. 2012;59(2):345-364. doi:10.1016/j. pc1.2012.03.007
- Weiss JE. Pediatric systemic lupus erythematosus: more than a positive antinuclear antibody. Pediatr Rev. 2012;33(2):62-73. doi:10.1542/pir.33-2-62

Item 187

A 7-year-old girl is having difficulty establishing relation-ships with other children despite repeated opportunities to do so. The girl prefers to stay near her mother or her teacher and will avoid other children. She sometimes cries and can be difficult to calm down after being dropped off at school, so her mother frequently remains in the classroom for a few minutes before quietly leaving. On days when morning transitions to school are significantly difficult, her mother will allow her to stay home. Her mother reports that, in preschool, things were worse in that she usually "couldn't" leave her daughter in the classroom. The girl typically speaks little when in public, but she speaks normally when home alone with her mother. She is an only child and the parents are divorced. When the girl spends the weekend at her father's house, she often expresses worry that something bad is going to happen to her mother. Her mother frequently allows the girl to sleep with her to avoid temper tantrums or nightmares about sleeping alone.

Of the following, the BEST next step in this child's care is

A. initiate treatment with a selective serotonin reuptake inhibitor
B. reassure her mother that because of the improvement seen since preschool, her daughter's problems should resolve without intervention
C. refer her for a neuropsychological evaluation to determine if she has underlying cognitive impairments
D. refer her to a cognitive behavior therapist to work on skills for managing her distress
E. refer her to a play therapist to assist the child in recognizing the causes of her distress

Item 187 Preferred Response: D

The child described in the vignette has many symptoms of an anxiety disorder, including characteristics of separation anxiety and generalized anxiety. The most appropriate next step would be to refer her to a mental health specialist to initiate cognitive behavior therapy (CBT). Anxiety disorders in particular are very responsive to the psychoeducation, cognitive restructuring, and graded exposure techniques used in CBT.

A well-monitored low-dose trial of a selective serotonin reuptake inhibitor (SSRI) might be appropriate if she had already been engaged in appropriate psychotherapy that was failing to make progress, and she was having significant functional impairment from anxiety such as an inability to attend school. Because there are no controlled trials supporting the use of SSRIs in very young children, it would be prudent to consult a mental health specialist before prescribing an SSRI in this age group.

This child is experiencing significant distress from her current symptoms, so reassurance that this will self-resolve would not be appropriate. Her extensive anxiety symptoms are unlikely to self-resolve but rather are likely to change over time into other ways of expressing anxiety such as generalized anxiety or social phobia. Teaching both the child and her family how to appropriately address anxiety will help to manage her symptoms over the long term.

The only developmental symptom described in the vignette is difficulty making friends, and anxiety symptoms could very easily be responsible for this problem. Therefore a neuropsychological evaluation to look for cognitive impairments would not be warranted. Anxiety disorders can be present in someone with developmental impairments, so a lack of response to anxiety treatment or any other symptoms of developmental impairment could later merit neuropsychiatric testing.

Play therapy is a far less evidence-based treatment for childhood anxiety problems than CBT, and as such would not be the initial preferred approach in this case.

PREP Pearls

- CBT is the preferred treatment for childhood anxiety disorders, particularly for elementary school aged children.
- Watchful waiting with anxiety disorders seldom leads to a full resolution of symptoms.

AAP Mental Health Competency:

- Recognize the types of evidence based psychotherapies for treating childhood anxiety

Suggested Reading:

- Connolly SD, Bernstein GA, and the American Academy of Child and Adolescent Psychiatry. Practice parameter for the assessment and treatment of children and adolescents with anxiety disorders. J Am Acad Child Adolesc Psychiatry. 2007;46(2):267-283

- Ginsburg GS, Kendall PC, Sakolsky D. et al. Remission after acute treatment in children and adolescents with anxiety disorders: findings from the CAMS. J Consult Clin Psychol. 2011;79(6):806-813. doi:10.1037/a0025933

Item C186. Drugs Used in the Treatment of Systemic Lupus Erythematosus

Drug Class	Drugs	Use in treating Systemic Lupus Erythematosus	Major Serious Adverse Effects	Recommended Monitoring
Antimalarial drug	Hydroxychloroquine or chloroquine	Maintenance of remission, mucocutaneous involvement, mild disease	Retinal toxicity	Ophthalmology exam every 6–12 months Reduce dose with renal impairment
Nonsteroidal Anti-inflammatory Drugs	Naproxen, meloxicam, ibuprofen, celecoxib, indomethacin, others	Musculoskeletal complaints, systemic symptoms (arthralgia, myalgia, arthritis, fever)	Aseptic meningitis in SLE patients, GI toxicity	Renal function, monitor closely in renal impairment
Glucocorticoids	Prednisone	Most SLE manifestations	Acute adrenal insufficiency, Cushing syndrome, growth suppression, osteoporosis, avascular necrosis, infection, hypertension, psychosis, mood and behavior changes, cataracts, glaucoma, myopathy, many others	Blood pressure, blood glucose levels, consider bone mineral density monitoring and monitoring for signs and symptoms of adrenal insufficiency
	Methylprednisolone	Severe end-organ involvement, severe exacerbation, life-threatening disease		
Immunosuppressive Agents	Azathioprine	Nephritis, steroid sparing agent	Bone marrow suppression, infection	CBC, hepatic enzymes, renal function
	Cyclophosphamide	Neuropsychiatric disease, nephritis	Bone marrow suppression, infection, infertility, malignancy	CBC, hepatic enzymes, renal function
	Cyclosporine	Rarely used except for specific types of nephritis	Hypertension, renal toxicit	CBC, hepatic enzymes, renal function, magnesium levels
	Methotrexate	Arthritis, mucocutaneous involvement, steroid sparing agent	Hepatitis, bone marrow suppression	CBC, hepatic enzymes
	Mycophenolate mofetil	Nephritis, steroid sparing agent	Bone marrow suppression, infection	CBC, hepatic enzymes
Biologic Therapy	Immune globulin intravenous	Limited role, cytopenia	Anaphylactoid reaction, aseptic meningitis	Infusion reactions
	Anti-CD20 monoclonal antibody (rituximab)	Steroid sparing agent in treatment resistant disease	Infection, progressive multifocal leukoencephalopathy	Check B-cell levels and quantitative immunoglobulins
	Anti–B lymphocyte stimulator (belimumab)	Mucocutaneous involvement, arthritis, mild disease, steroid sparing agent	Infection, anaphylaxis, malignancy	CBC
	Anti-TNF agents (adalimumab, etanercept)	Arthritis, rarely a steroid sparing agent	Infection, malignancy	CBC, hepatic enzymes

Abbreviations: SLE, systemic lupus erythematosus; GI, gastrointestinal; CBC, complete blood cell count; TNF, tumor necrosis factor.

Item 188

You are called to stabilize a neonate born at 30 weeks of gestation before the neonate is transferred to a tertiary center. The mother was delivered by emergency medical services in an ambulance en route to the hospital. The neonate emerged vigorous with a strong cry. You arrive to find a 1,250-gram newborn on the warmer in a 100% oxygen hood. Assessment reveals a temperature of 36.0°C, heart rate of 115 beats/min, respiratory rate of 70 breaths/min, blood pressure of 45/30 mm Hg (mean blood pressure, 35 mm Hg), and oxygen saturation of 100%. The examination is notable for mild grunting, flaring, and retracting with equal breath sounds bilaterally. The bedside glucose level is 50 mg/dL (2.8 mmol/L).

Of the following, the MOST appropriate next step in management is to

A. administer a bolus of 10% dextrose
B. administer intramuscular ceftriaxone
C. decrease hood oxygen concentration
D. initiate dopamine therapy
E. obtain a chest radiograph

Item 188 S Preferred Response: C

The premature infant in the vignette is in a 100% oxygen hood with an oxygen saturation value of 100% and should immediately have the hood oxygen concentration decreased. Hyperoxia has been linked with oxygen toxicity in premature infants. The association between retinopathy of prematurity (ROP) and oxygen exposure was first described in 1951 when high levels of oxygen in incubators were linked to the development of blindness in premature infants. Vasoconstriction of the retinal vessels in response to oxygen exposure leads to the neovascularization seen in ROP. Inflammation, fibrosis, and abnormal pulmonary vascular development caused by exposure to supplemental oxygen also contribute to the development of bronchopulmonary dysplasia in premature infants.

Target oxygen saturation levels for premature infants remain elusive. In one large randomized trial, infants born between 24 weeks 0 days and 27 weeks 6 days of gestation who were exposed to a lower target range of oxygenation (85-89%) had less severe ROP but increased mortality when compared to infants exposed to a higher target range of oxygenation (9195%). This increased mortality has also been found in a second large randomized trial, in which preterm infants born at less than 28 weeks' gestation with a targeted oxygen saturation range of 85-89% had a higher rate of death than those with a target range of 91-95%. At present, the target oxygen saturations of 85-95% at ten minutes after birth outlined by the AAP-AHA Neonatal Resuscitation Program offer a guide for initial supplemental oxygen management in the premature infant. Some experts are suggesting oxygen saturations be maintained in the 90-95% range following the immediate period after birth for premature infants.

The care of a premature infant at the time of birth focuses on the establishment of an airway, initiation and maintenance of adequate ventilation, and ongoing monitoring of cardiovascular status. Premature infants may require respiratory support ranging from hood oxygen, continuous positive airway pressure, or intubation for respiratory distress syndrome (RDS). Oxygenation should be closely monitored starting in the delivery room. After the initial stabilization of the premature infant, attention must be focused on thermoregulation, glucose homeostasis, and infection. The decreased subcutaneous fat and immature epidermis of the preterm infant contribute to increased heat loss after birth, leading to hypothermia. Low glycogen stores and a decreased ability to perform gluconeogenesis leads to a continuous need for parenteral glucose that often begins shortly after birth. Underlying infection may present with preterm delivery, prompting a sepsis evaluation and initiation of broad-spectrum antibiotics (typically ampicillin and gentamicin) after delivery.

The premature infant in the vignette has a stable cardiorespiratory status with mild respiratory distress. If the respiratory status worsens or fails to improve over time a chest radiograph may be obtained to evaluate for RDS, pneumonia or other respiratory disorders. The mean blood pressure of 35 mm Hg is normal for a 30 week gestation and dopamine does not need to be initiated. A bolus of 10% dextrose is not needed for a glucose level of 50 mg/dL, although a continuous infusion of 10% dextrose should be begun. A sepsis evaluation and antibiotics are indicated in the premature infant in the

vignette. Ceftriaxone should not be used due to in vitro studies that demonstrate that it can displace bilirubin from albumin in term and premature infants.

PREP Pearls

- The initial care of a premature infant includes close monitoring of blood glucose and oxygen saturation.

American Board of Pediatrics Content Specification(s):

- Recognize that initial care of a very-low-birth-weight infant includes monitoring of blood glucose and arterial oxygen concentrations
- Recognize that initial care of the very-low-birth-weight infant includes evaluation for sepsis if appropriate

Suggested Reading:

- American Academy of Pediatrics. American Heart Association. Resuscitation of babies born preterm. In: Kattwinkel J, ed. Textbook of Neonatal Resuscitation. 6th ed. Elk Grove, IL: American Academy of Pediatrics; 2011:267-282
- Angert R, Adam HM. Care of the very low-birthweight infant. Pediatr Rev. 2009;30: 32-35
- SUPPORT Study Group of the Eunice Kennedy Shriver NICHD Neonatal Research Network. Target ranges of oxygen saturation in extremely preterm infants. NEJM. 2010;363:1959-1969
- The BOOST II United Kingdom, Australia, and New Zealand Collaborative Groups. Oxygen saturation and outcomes in preterm infants. NEJM. 2013;368:2094-2104
- Weinberger B, Laskin DL, Heck DE, Laskin JD. Oxygen toxicity in premature infants. Toxicol Appl Pharmacol. 2002;181:60-67

Item 189

You are seeing a 15-year-old girl with sickle cell disease who was admitted to the pediatric intensive care unit with fever, shock, and respiratory distress. Upon access of her central venous catheter, bleeding is noted from the puncture site. Examination shows an ill-appearing young woman with petechiae and purpura on her extremities. Additional peripheral intravenous access sites are also oozing blood. Her temperature is 39°C, respiratory rate is 30 breaths/min, pulse rate is 130 beats/min, and blood pressure is 80/40 mm Hg. There are coarse breath sounds bilaterally, a grade 2/6 systolic ejection murmur, and no hepatosplenomegaly or lymphadenopathy. Blood cultures are obtained. She is given an infusion of normal saline and started on cefotaxime and dopamine. Complete blood cell count shows a white blood cell count of 25,000/µL (25×10^9/L), with 75% polymorphonuclear leukocytes, 10% band neutrophils, 10% lymphocytes, and 5% monocytes. Hemoglobin is 7.5 g/dL (75 g/L) and reticulocyte count is 8% (0.08). Platelet count is 15×10^3/µL (15×10^9/L). Prothrombin time is 20 seconds and partial thromboplastin time is 53 seconds.

Of the following, the MOST likely diagnosis in this patient is

A. disseminated intravascular coagulation
B. factor X deficiency
C. hypersplenism
D. immune thrombocytopenic purpura
E. von Willebrand disease

Item 189 Preferred Response: A

The patient in the vignette has a clinical presentation most consistent with disseminated intravascular coagulation (DIC). DIC is characterized by systemic activation of the coagulation system, which leads to release of procoagulants into the circulation, fibrin deposition in end organs, fibrinolysis, formation of fibrin degradation products, impaired platelet aggregation, consumption of clotting factors and platelets, and hemolysis. In neonates, DIC is most often associated with birth asphyxia and sepsis, whereas in children and adults, it is usually associated with sepsis, trauma, or malignant tumors. Less common causes of DIC in children include transfusion reactions, Kasabach-Merritt phenomenon, snake bites, spider bites, and liver disease. Congenital homozygous deficiency of protein C or protein S can present in the newborn period as purpura fulminans and DIC. Laboratory findings in DIC reflect a consumptive coagulopathy and increased fibrinolysis. Prothrombin time (PT) and partial thromboplastin time (PTT) are prolonged and fibrinogen activity is decreased due to the depletion of coagulation factors. Fibrinolysis leads to increased D-dimers and fibrin degradation products. There may be evidence of microangiopathic hemolysis as the red blood cells undergo mechanical shearing by intravascular fibrin strands. Treatment for DIC is aimed at identifying and treating the underlying condition; however, supportive care may be indicated in cases of severe hemorrhage, thrombosis, or end-organ damage until the coagulopathy resolves. Fresh frozen plasma, cryopreciptiate, and platelet transfusions can be given to replace clotting factors, fibrinogen, and platelets, respectively, to control bleeding.

The sickle cell patient described in the vignette most likely has sepsis, given her clinical symptoms and her susceptibility to infection with encapsulated organisms, such as Streptococcus pneumoniae and Neisseria meningitidis. Her laboratory values (prolonged PT and PTT and thrombocytopenia) in this setting suggest DIC. Factor X deficiency is a rare coagulation disorder that is not associated with thrombocytopenia. Hypersplenism would be unusual in an adolescent patient with sickle cell disease because she is likely to have autoinfarction of her spleen by this age. Immune thrombocytopenia purpura is not associated with prolongation of the PT and PTT. Von Willebrand disease (vWD) is the most common inherited bleeding disorder. In severe type 1 vWD or type 3 vWD, very low levels of von Willebrand factor (vWF) lead to increased clearance of factor VIII, which can cause the PTT to be prolonged; however, the PT and platelet count should not be affected. Type 2B vWD can be associated with thrombocytopenia as a result of increased clearance of the platelet-vWF complex; however, the PT and PTT would be normal.

PREP Pearls

- In neonates, DIC is most often associated with birth asphyxia and sepsis, whereas in children and adults, it is usually associated with sepsis, trauma, or malignant tumors.
- Laboratory findings in DIC reflect a consumptive coagulopathy and increased fibrinolysis, prolonged PT and PTT, decreased fibrinogen level, and decreased platelet count which may result in a decreased hemoglobin.
- Treatment for DIC is aimed at identifying and treating the underlying condition; however, supportive care may be indicated in cases of severe hemorrhage,

thrombosis, or end-organ damage until the coagulopathy resolves. Fresh frozen plasma, cryoprecipitate, and platelet transfusions can be given to control bleeding.

American Board of Pediatrics Content Specification(s):

- Identify the need for measuring prothrombin time, partial thromboplastin time, and platelet count as part of the evaluation for disseminated intravascular coagulation in a child with sepsis and purpura

Suggested Reading:

- Kaneko T, Wada H. Diagnostic criteria and laboratory tests for disseminated intravascular coagulation. I Clin Exp Hematopathol. 2011;51:67-76. doi:10.3960/jslrt.51.67
- Hook KM, Abrams CS. The loss of homeostasis in hemostasis: new approaches in treating and understanding acute disseminated intravascular coagulation in critically ill patients. Clin Trans Sci. 2012;5:85-92. doi:10.1111/j.1752-8062.2011.00351.x
- Semeraro N, Ammollo CT, Semeraro F, Colucci M. Sepsis, thrombosis, and organ dysfunction. Thromb Res. 2012;129:290-295. doi:10.1016/j. thromres.2011.10.013
- Wong W, Glader B. Disseminated intravascular coagulation in infants and children. UptoDate. Available online only for subscription

Item 190

A 15-year-old boy was standing in the gym bleachers at a school assembly and started to feel light-headed and dizzy. He heard a ringing noise in his ears and then had tunnel vision. The next thing he remembers is waking up on the gym floor surrounded by teachers. The teachers told the parents that the boy suddenly lost consciousness and started to fall. He was caught by other students and then observed to have symmetric limb jerking that lasted about 30 seconds. He has never had an event like this before and has been healthy. His vital signs and physical examination findings are normal. An electrocardiogram is unremarkable, but an electroencephalogram shows generalized spike-wave discharges interpreted as being consistent with a diagnosis of epilepsy.

Of the following, the MOST likely cause for his symptoms is

A. fugue state
B. seizure
C. somatization
D. syncope
E. vertigo

Item 190 Preferred Response: D

The boy described in the vignette had an episode of syncope. The brief limb jerking that can occur after syncope is termed syncopal convulsion, and should be differentiated from generalized tonic-clonic seizure. In a generalized tonic-clonic seizure, the body stiffens and the limb jerking occurs early, often causing a fall. In syncopal convulsion, the fall occurs first and is followed by the convulsion.

Up to 6% of children have abnormal electroencephalograms and do not have seizures. The child in the vignette has a clearly syncopal event, in which case, obtaining an electroencephalogram is not indicated. If the results are abnormal, they can cause confusion. Electroencephalography results can be helpful if the initial event was not witnessed, or if the details are not clear; abnormal electroencephalography results would suggest that the person could have had a seizure. Electroencephalography can also give a diagnosis of a specific epilepsy syndrome. Fugue state is a psychiatric disorder in which the person has amnesia regarding his or her own identity, lasting for hours to days or longer. Somatization can present with symptoms of syncope or near-syncope; symptoms typically persist or evolve if not diagnosed and treated. Vertigo is characterized by a spinning sensation that can cause imbalance, but not loss of consciousness.

PREP Pearls

- Up to 6% of children without seizures have abnormal electroencephalograms.
- Syncope can be followed by brief tonic stiffening and limb jerking, termed syncopal convulsion, which should not be confused with a generalized tonic-clonic seizure.

American Board of Pediatrics Content Specification(s):

- Understand the value and limitations of neurodiagnostic techniques such as evoked potentials, electromyography, and electroencephalography

Suggested Reading:

- Hirsch LJ, Arif H, Moeller J. Electroencephalography (EEG) in the diagnosis of seizures and epilepsy. UptoDate. Available online only for subscription
- Khoshbin S. Clinical neurophysiology. UptoDate Online. Available online only for subscription

Item 191

A 7-year-old girl presents to your office with her parents for a routine health supervision visit. Her parents report that she has intermittent bilateral knee pain with activity. The girl and her parents have not noticed any joint swelling, limp, recurrent fevers, skin lesions, or change in activity level or appetite. The child does not limit her activities because of pain. On physical examination, you note that the girl is able to hyperextend beyond 10° at both knees and elbows. Knee examination is otherwise unremarkable. Her mother reports that she and several other maternal relatives are "double jointed"

Of the following, the MOST appropriate initial management step is

A. encourage stretching before physical activities
B. order magnetic resonance imaging
C. reassure the family that she does not have a structural knee problem
D. refer to rheumatology
E. restrict participation in sports and physical education

Item 191 Preferred Response: C

The girl described in the vignette has intermittent knee pain as well as hypermobility of the knees and elbows on examination. Joint hypermobility is the term used to describe excess movement of the joints due to ligamentous laxity. Children may have joint hypermobility as a result of a genetic connective tissue disorder, such as Marfan syndrome or osteogenesis imperfecta; additionally, joint hypermobility is common in the general pediatric and adolescent population. Joint hypermobility syndrome describes hypermobility associated with significant arthralgias or other physical findings. Because this girl has mild pain without swelling, limp, or signs of internal derangement of the knee on physical examination, she should be continue with sports and other physical activities as tolerated.

The Beighton score is commonly used to evaluate for joint hypermobility in children and adults; one point is awarded for each knee or elbow that extends beyond 10°, for each thumb that can touch the palmar aspect of the wrist, for each fifth finger that can extend beyond 90° at the metacarpophalangeal joint, and for the ability to touch both palms to the floor with the knees extended. This child can extend both elbows and knees beyond 10° and therefore has a Beighton score of 4 of a possible 9 points. There is no agreed-upon cut-off point for Beighton score that establishes the diagnosis of hypermobility in children. In a study of Dutch schoolchildren aged 6 to 12, more than one-third had a Beighton score of 5 or higher; among 13- and 14-year-old teens in Britain, 28% of girls and 11% of boys had a score of 5 or greater.

There are conflicting data on the association between joint hypermobility and pain. In a cohort of Finnish schoolchildren aged 10 to 12, approximately 70% of subjects reported regional or widespread pain during the preceding 3 months; there was no correlation between pain and Beighton score. However, a study of patients seen in rheumatology clinics for arthralgias demonstrated that this population had high rates of joint hypermobility. Children with joint hypermobility should be counseled to avoid extremes of joint motion and should avoid stretching muscle groups adjacent to hypermobile joints.

Rheumatology consultation to evaluate for inflammatory arthropathies may be appropriate for children with multiple arthralgias, particularly in the presence of joint swelling, stiffness, or systemic symptoms. Although children with hypermobile joints appear to have increased rates of shoulder dislocations and acute knee injuries, the additional risk does not justify restricting sports participation in asymptomatic or minimally symptomatic individuals with clinical evidence of hypermobility. Magnetic resonance imaging is not warranted because she does not have signs of infection or internal derangement on physical examination. Physical therapy for strengthening and proprioception may help individuals with pain that limits activities or with history of injuries related to hypermobility.

PREP Pearls

- Joint hypermobility is very common in the pediatric and adolescent population.
- The term joint hypermobility denotes excessive movement of the joints due to ligamentous laxity, while the term joint hypermobility syndrome describes hypermobility associated with significant arthralgias.

American Board of Pediatrics Content Specifications:

- Know that the treatment of hypermobility syndrome is by explanation (ie, counsel the patient to avoid excessive movement)
- Understand the relationship between hypermobility and joint complaints
- Know the physical findings in hypermobility syndrome

Suggested Reading:

- Adib N, Davies K, Grahame R, Woo P, Murray KJ. Joint hypermobility syndrome in childhood: a not so benign multisystem disorder? Rheumatology (Oxford). 2005;44(6):744-750. doi:10.1093/rheumatology/keh557 Clinch J, Deere K, Sayers A, et al. Epidemiology of generalized joint laxity (hypermobility) in fourteen-year-old children from the UK: a population-based evaluation. Arthritis Rheum. 2011;63(9):2819-2827. doi:10.1002/ art.30435
- Mikkelsson M, El-Metwally A, Kautiainen H, Auvinen A, Macfarlane GJ, Salminen JJ. Onset, prognosis, and risk factors for widespread pain in schoolchildren: a prospective 4-year follow-up study. Pain. 2008;138(3):681-687. doi:10.1016/j.pain.2008.06.005
- Pacey V, Nicholson LL, Adams RD, Munn J, Munns CF. Generalized joint hypermobility and risk of lower limb joint injury during sport: a systematic review with meta-analysis. Am J Sports Med. 2010;38(7):14871497. doi:10.1177/0363546510364838
- Smits-Engelsman B, Klerks M, Kirby A. Beighton score: a valid measure for generalized hypermobility in children. J Pediatr. 2011;158(1):119-123, 123.e1-4. doi:10.1016/j.jpeds.2010.07.021

Item 192

You are following a 4-year-old boy in your practice who was diagnosed with achondroplasia at birth. He has been in generally good health and has been following along the growth curves established for children with this condition. He had no symptoms of apnea in infancy and has mildly delayed motor milestones. On examination, you note rhizomelic limb shortening, a mild thoracolumbar kyphosis that has been present since infancy, normal muscle tone, and nonfocal neurologic findings. The parents have questions about their child's future in terms of his anticipated medical and developmental progress.

Of the following, you are MOST likely to tell the parents at this time that

A. children with achondroplasia should avoid gymnastics, diving, and collision sports
B. FGFR3 gene testing should be performed in order to confirm the diagnosis
C. growth hormone supplementation is recommended for children with achondroplasia
D. mild cognitive delays are common in children with achondroplasia
E. spinal stenosis is common in children with achondroplasia

Item 192 S I-C Preferred Response: A

The child described in the vignette is progressing typically for a 4-year-old with achondroplasia. Children and adults with achondroplasia have narrowing and distortion of the foramen magnum that puts them at greater risk for spinal cord compression with uncontrolled neck movement. Therefore, children and adults with achondroplasia should be cautioned not to participate in sports or activities where collisions are frequent and could result in sudden neck movement. Furthermore, care should be taken during intubation in individuals with achondroplasia to avoid inadvertent hyperextension of the neck.

Hydrocephalus remains a lifelong risk for those with achondroplasia but typically develops before 2 years of age. Therefore, all children with achondroplasia should have frequent head circumference measurements, which should be plotted on the achondroplasia-specific curve. For children with a head circumference that is significantly elevated on that curve or for children whose head circumference is crossing percentiles, consideration should be given to obtaining a baseline or diagnostic computed tomography or magnetic resonance imaging of the brain and craniocervical junction. A consultation with a pediatric neurologist or neurosurgeon may also be indicated under these clinical circumstances.

Most infants with achondroplasia develop a thoracolumbar kyphosis, which can be worsened by unsupported sitting before there is sufficient trunk stability. Parents should there-fore be cautioned not to place an infant who has achondroplasia in soft seats, C-shaped seats, or other devices where the child is unsupported during the first years of life. Although FGFR3 testing is useful in some cases to confirm a suspected diagnosis, achondroplasia is most often a clinical diagnosis, relying on physical examination and radiographs to identify the key findings after birth. The clarification of a specific genetic mutation does not alter medical management but could potentially be used by an affected adult for prenatal or preimplantation genetic diagnosis. Unlike many other genetic conditions, achondroplasia is caused by a mutation in a single base pair in a single gene. This permits easy mutation identification with the ability to perform targeted mutation analysis in those cases where genetic testing is warranted. Although achondroplasia is inherited as an autosomal-dominant condition, about 75% of cases represent de novo events, and there is no family history to suspect this diagnosis before birth. However, for a prospective parent who has achondroplasia, the risk of giving birth to an affected child is 50% (as would be expected with any autosomal dominant condition).

Growth hormone supplementation is ineffective in increasing height in children with achondroplasia or other skeletal dysplasias. Although gross motor delays are quite common in infants and young children with achondroplasia, most individuals with this condition have completely normal intelligence. Whereas spinal stenosis is a relatively common complication seen in adults and presents with numbness, weakness, or altered deep tendon reflexes, it is generally not a complication seen in childhood achondroplasia.

PREP Pearls

- Children and adults with achondroplasia should not participate in collision sports where sudden or exaggerated neck movement might put them at risk for spinal cord compression.
- Infants with achondroplasia require additional spine support to prevent progression of thoracolumbar kyphosis and should not be placed in soft or C-shaped seats for extended periods of time.

American Board of Pediatrics Content Specification(s):

- Recognize the inheritance pattern of achondroplasia
- Recognize the clinical features and complications of achondroplasia

Suggested Reading:

- Pauli RM. Achondroplasia. In: Pagon RA, Bird TD, Dolan CR, eds. GeneReviews. Seattle, Washington: University of Washington; 2013
- Trotter TL, Hall JG; Committee on Genetics. Health supervision for children with achondroplasia. Pediatrics. 2005;116(3):771-783. doi:10.1542/ peds.2005-1440

Item 193

A 17-year-old girl is brought to the emergency department by her parents after they "found her drunk in her room" The girl's parents tell you that she has been spending a lot of time alone in her bedroom since breaking up with her boyfriend last week. Two hours ago, the girl's mother entered her bedroom to check on her and found her lying on the floor and acting confused. The parents state that the girl "smelled strongly of alcohol." The parents' search of the girl's bedroom yielded several empty beer bottles and a half-empty vodka bottle hidden underneath the bed. The girl vomited several times while her parents were driving her to the emergency department.

On physical examination, the girl appears lethargic but responds to verbal stimuli. Her temperature is 37.0°C, heart rate is 70 beats/min, respiratory rate is 16 breaths/min, and blood pressure is 92/68 min Hg. She is able to tell you her name and age, but her speech is slurred and she answers several of your questions inappropriately. You detect the smell of alcohol as you proceed with your examination. There are no signs of traumatic injury. The girl's pupils are midsized and equally reactive. Her skin is pale and clammy. A bedside glucose check yields a normal result, and a urine pregnancy test result is negative. A serum ethanol level is 70 mg/dL. The patient's electrocardiogram reveals a normal sinus rhythm with no abnormalities.

Of the following, the BEST next step in the management of this patient is to

A. administer 1 g/kg of activated charcoal
B. administer a dose of promethazine for nausea
C. close observation in the emergency department over the next 4 to 6 hours for resolution of symptoms
D. obtain emergent child psychiatric consultation
E. obtain serum and urine toxicology screens

Item 193 I-C S Preferred Response: E

The teenage girl described in the vignette is brought to the emergency department with altered mental status, slurred speech, and vomiting after being found in her room with several empty containers of ethanol. Although she appears lethargic, her airway, breathing, and circulation are intact. Although the history provided by her parents strongly suggests a diagnosis of acute ethanol intoxication, obtaining serum and urine toxicology screens to investigate for coingestion of additional (and potentially life-threatening) toxins is the best next step in management at this time.

Although most toxic ingestions in younger children occur unintentionally due to developmentally normal exploratory behavior and involve small quantities of a single agent, toxic exposures in adolescents are much more likely to be intentional and involve several drugs at once (polypharmacy). Drug abuse, experimental risk-taking behaviors, and depression with suicidal ideation place adolescents at risk for poisoning.

The drug-abusing adolescent may present for medical attention after an intentional or unintentional overdose, a suicide attempt, a concerning change in behavior, or sustaining trauma while under the effects of drugs or alcohol. Historical information provided by adolescents presenting with toxic ingestions may not be reliable because of their unwillingness or inability to fully disclose the types and amounts of toxins involved. Clinicians should ask other household members about all medications, vitamin and mineral supplements, herbal and folk remedies, and household chemicals present in the home. Adolescents may also have access to drugs and other toxins in their school, work, or recreational environments. For adolescents presenting after an acute toxic ingestion, family members, friends, or paramedics can provide important information about open containers, empty bottles, spilled contents, drug paraphernalia, or suicide notes at the scene.

Clinicians caring for children presenting with presumed ethanol intoxication should maintain a high index of suspicion for the coingestion of other potentially dangerous toxins. The nonspecific symptoms of lethargy, altered mental status, nausea, vomiting, and ataxia—common findings in patients with ethanol intoxication—can also signal the detrimental effects of other potentially lethal toxins, including acetaminophen and aspirin.

Although toxicology screening is generally unnecessary for children presenting after unintentional ingestions who have clinical findings that are consistent with the history, it is indicated in situations in which the possibility of a potentially harmful coingestion exists. Specifically, screening for acetaminophen and salicylate ingestion is strongly recommended for patients presenting with an uncertain history or intentional poisoning; few early signs may be apparent after lethal doses of these agents, and these signs can be "masked" by the presence of other toxins, such as ethanol. Furthermore, specific treatments for acetaminophen and salicylate poisoning are available and highly effective if implemented early in the patient's clinical course.
Urine screens provide qualitative data about the recent use of substances included in the screen. In most institutions, urine screens test for a limited number of substances,

typically drugs of abuse. These screens are relatively inexpensive and provide rapid results (usually within 1 hour). Although urine toxicology screens provide no information about timing or quantity of ingestion, the information obtained may aid clinicians in "making sense" of patients' clinical findings, anticipating their clinical course and the potential for withdrawal, and determining psychiatric disposition. Serum toxicologic testing provides quantitative data and is important in the diagnosis and management of ingestion of several drugs and medications, including acetaminophen and salicylates. Clinicians should become familiar with the specific drugs tested for on both the urine and serum toxicology screens at their respective institutions.

Although administration of activated charcoal is the decontamination technique of choice for most poisonings, it would not be the best next step in the treatment of the patient presented in this vignette. Although this patient's airway is currently intact, her depressed mental status and nausea with recent vomiting place her at high risk for aspiration. Furthermore, toxic alcohols are very poorly adsorbed by activated charcoal. Activated charcoal should only be administered after securing a definitive airway in this patient.

The administration of promethazine for nausea and vomiting would likely result in increased central nervous system depression in this patient (who is already lethargic) and also carries the potential to produce a dystonic reaction. Therefore, it would not be the next best step in the treatment of this patient. Newer antiemetics, such as ondansetron and granisetron, tend to be less sedating and are much less likely to cause dystonia. Although the girl in the vignette should certainly be monitored closely in the emergency department for worsening of her clinical status or the development of new signs and symptoms, laboratory screening for the possibility of a potentially dangerous, undisclosed coingestion should be completed early in her emergency department course before the clinical decision to simply observe is made.

Although a thorough psychiatric evaluation is warranted in this patient before her ultimate discharge from the hospital, consulting a child psychiatrist urgently is not the best next step in her management. Once she is medically stabilized and no longer intoxicated, psychiatric consultation may be obtained.

PREP Pearls

- Clinicians caring for children presenting with ethanol intoxication should maintain a high index of suspicion for the coingestion of other potentially dangerous toxins.
- Historical information provided by patients presenting with toxic ingestions may not be reliable because they may be unwilling or unable to fully disclose the types and amounts of toxins involved.
- Toxicologic screening, including screening for acetaminophen and salicylate ingestion specifically, is strongly recommended for intoxicated patients presenting with an uncertain history or intentional poisoning.

American Board of Pediatrics Content Specification(s):

- Know that ethanol intoxication may mask toxicities from other drugs

Suggested Reading:

- Velez LI, Shepherd JG, Goto CS. Approach to the child with occult toxic exposure. UptoDate. Available online only for subscription
- Woolf AD. Poisoning in children and adolescents. Pediatr Rev. 1993;14:411. doi:10.1542/pir.14-11-411

Item 194

A 7-year-old girl born with a myelomeningocele presents to your office for her annual health supervision visit. Her mother states that her daughter's teacher is concerned that the girl has limited reading skills and language proficiency for her age. The mother attributes this to the fact that Spanish is the primary language spoken at home, whereas English is the primary language spoken at the school. The child is well-behaved at home but has demonstrated some attention problems at school.

Of the following, your BEST recommendation is

A. the child should participate in an after school English program
B. the child should participate in a summer reading program
C. the child should undergo formal psychoeducational evaluation
D. the mother should join a parenting support group
E. no intervention is needed since this is a normal variation

Item 194 I-C SBP Preferred Response: C

Myelomeningocele is a form of neural tube defect in which the spinal cord (myelon), its covering (meninges), and vertebral arches develop abnormally early in gestation. The cause is uncertain and multifactorial, with both environmental and genetic factors playing a role. The prevalence varies among countries and ethnic groups.

Associated abnormalities of the brain, including hydrocephalus, Chiari type II malformation, agenesis of the corpus callosum, and other structural anomalies, may occur. It is not uncommon for patients with spina bifida to also have intellectual disabilities, including verbal and nonverbal disorders, attention-deficit/hyperactivity disorder, and problems with executive functioning or behavior. Most children with myelomeningocele have overall intelligence in the normal range, so it is imperative that formal psychoeducational evaluation be performed in these individuals when there are concerns raised.

One should not attribute even mild language delay or poor reading skills to the neural tube defect alone without evaluation and treatment. Investigation for intellectual disabilities should be performed. Children who are raised and educated in bilingual environments should not have reading and language delays evident at 7 years of age. Participation in extra tutoring or afterschool programs in English and encouraging summer reading are helpful to assist the child who is lagging academically. This should be encouraged once formal evaluation determines the specific learning issues and the degree of delay, so the educational program can be individualized to meet the needs of the learner. Parenting support groups are a wonderful resource for parents of children with special needs or chronic medical conditions. However, simply referring to a support group or reassuring that a delay is a normal variation is not adequate. Children who have seizure disorders or other central nervous system-based chronic conditions have a significantly increased incidence of learning difficulties and require a full individualized evaluation and treatment plan for any educational or emotional needs. It is the pediatric health care physician's role to advocate for early identification and ongoing services for these children at risk for intellectual disabilities.

PREP Pearls

- Most children with myelomeningocele will have overall intelligence in the normal range.
- it is not uncommon for patients with central nervous system—based chronic conditions, such as epilepsy or myelomeningocele, to have intellectual disabilities.
- Developmental surveillance is key to early identification and referral few formal evaluation

American Board of Pediatrics Content Specification(s):

- Recognize that patients who have CNS-based chronic conditions (eg, epilepsy, myelomeningocele) have an increased incidence of learning disabilities

Suggested Reading:

- Burke R, Liptak GS, and The Council on Children with Disabilities. Providing a primary care medical home for children and youth with spina bifida. Pediatrics. 2011;128:e1645-1657. doi: 10.1542/peds.2011-2219
- Liptak GS, Dosa NP. Myelomeningocele. Pediatr Rev. 2010;31:443-450. doi: 10.1542/pir.31-11-443
- Rimrodt SL, Lipkin PH. Learning disabilities and school failure. Pediatr Rev. 2011;32:315-324. doi: 10.1542/pir.32-8-315

Item 195

Your first patient of the morning is a 7-year-old boy who has been "coughing and wheezing since dinner last night." He required 2 albuterol nebulizer treatments last night and has received 3 treatments since 6:00 AM. You observe that the child is sitting upright and appears breathless. His temperature is 37.1°C, oxygen saturation is 82% on room air, heart rate is 120 beats/min, and respiratory rate is 40 breaths/ min. He is using accessory muscles to breathe, and his nails and tongue are blue-tinged. On auscultation of the lungs, air entry is diminished bilaterally and there is mild expiratory wheezing. He is barely able to speak, and his peak expiratory flow is 35% of predicted.

Of the following, the MOST likely initial diagnosis is

A. acute moderate asthma exacerbation
B. acute severe asthma exacerbation
C. foreign body aspiration
D. pneumonia
E. pneumothorax

Item 195 S SBP Preferred Response: B

The child described in the vignette has signs of severe obstruction in an acute exacerbation of asthma. He is hypoxic and cyanotic and unable to speak whole phrases. He has a markedly increased respiratory rate and uses accessory muscles of respiration. His lung findings are paradoxically and ominously mild in contrast to the other signs of respiratory distress, suggesting severe obstruction and imminent respiratory arrest.
A child with moderate obstruction in contrast will typically be able to speak in phrases; he will have loud expiratory wheeze, peak expiratory flow from 49% to 60% predicted, and pulse oximetry 90% to 95% on room air, in addition to symptoms of obstruction such as increased respiratory rate and chest retractions (Item C195, page C-153). Other features include relief with use of rescue short-acting p agonists and relief in 1 to 2 days after the onset of systemic steroid burst. Note that many of the parameters, and in particular, the correlation with each other, have not been systematically studied. In general, the presence of several parameters helps classify an individual into a particular severity.

There is no history of choking or ingestion of a foreign body. He has bilateral diminished respiratory sounds making pneumothorax (and foreign body aspiration) less likely. He is afebrile and nontoxic, pushing pneumonia lower on the differential diagnosis.

PREP Pearls

- The presence of several symptoms and signs helps indicate severity of obstruction in an acute asthma exacerbation; there is no single best assessment tool or measure.
- Diminution in breath sounds, in a child with other signs of airway obstruction, may indicate imminent respiratory failure.

American Board of Pediatrics Content Specification(s):

- Recognize the signs of severe obstruction in an acute exacerbation of asthma (severe retractions, inability to speak whole phrases, cyanosis, quiet breath sounds in presence of other signs of obstruction, peak expiratory flow rates less than 30% of predicted)

Suggested Reading:

- National Asthma Education and Prevention Program. Expert Panel Report 3 (EPR-3): guidelines for the diagnosis and management of asthma—summary report 2007 [published correction appears in J Allergy Clin Immunol. 2008;121(6):1330]. I Allergy Clin Immunol. 2007;120(5 suppl):S94-S138. doi:10.1016/j.jaci.2007.09.029
- National Heart, Lung, and Blood Institute. Expert Panel Report 3: Guidelines for the Diagnosis and Management of Asthma. National Asthma Education and Prevention Program, Third Expert Panel on the Diagnosis and Management of Asthma. Bethesda, MD: National Heart, Lung, and Blood Institute; 2007

Components of Control		Classification of Asthma Control (Children 0–4 years of age)		
		Well Controlled	Not Well Controlled	Very Poorly Controlled
Impairment	Symptoms	≤2 days/week	>2 days/week	Throughout the day
	Nighttime awakenings	≤1x/month	>1x/month	>1x/week
	Interference with normal activity	None	Some limitation	Extremely limited
	Short-acting $beta_2$-agonist use for symptom control (not prevention of EIB)	≤2 days/week	>2 days/week	Several times per day
Risk	Exacerbations requiring oral systemic corticosteroids	0–1/year	2–3/year	>3/year
	Treatment-related adverse effects	Medication side effects can vary in intensity from none to very troublesome and worrisome. The level of intensity does not correlate to specific levels of control but should be considered in the overall assessment of risk.		

Key: EIB, exercise-induced bronchospasm; ICU, intensive care unit

Notes:

- The level of control is based on the most severe impairment or risk category. Assess impairment domain by caregiver's recall of previous 2–4 weeks. Symptom assessment for longer periods should reflect a global assessment, such as inquiring whether the patient's asthma is better or worse since the last visit.
- At present, there are inadequate data to correspond frequencies of exacerbations with different levels of asthma control. In general, more frequent and intense exacerbations (e.g., requiring urgent, unscheduled care, hospitalization, or ICU admission) indicate poorer disease control. For treatment purposes, patients who had ≥2 exacerbations requiring oral systemic corticosteroids in the past year may be considered the same as patients who have not-well-controlled asthma, even in the absence of impairment levels consistent with persistent asthma.

Courtesy of the National Heart, Lung, and Blood Institute and the National Asthma Education and Prevention Program. *Expert Panel Report 3 (EPR3): Guidelines for the Diagnosis and Management of Asthma*. Bethesda, Md: U. S. Department of Health and Human Services. 2007.

ITEM C195: Classification of asthma control in the urgent or emergency care setting.

Item 196

A 16-year-old girl presents to your office with complaints of dysuria and gross hematuria for the last 2 days. She describes her urine as "red with blood clots in it." She has no complaints of fever, frequency, or flank pain. She has regular monthly periods for the last 2 years and finished her periods 15 days ago. Her urinalysis demonstrates a specific gravity of 1.015, pH of 6.0, 4+ blood, 4+ leukocyte esterase, and no protein or nitrites. Her urine microscopy shows more than 100 red blood cells per high-power field, 10 to 50 white blood cells per high-power field, and no crystals or bacteria.

Of the following, the MOST likely diagnosis in this adolescent is

A. acute pyelonephritis
B. cystitis
C. IgA nephropathy
D. myoglobinuria
E. paroxysmal nocturnal hemoglobinuria

Item 196 Preferred Response: B

The differential diagnosis for red urine is quite extensive. The history and urinalysis findings on urine dipstick and urine microscopy are helpful in differentiating the possible causes. Hematuria is defined as more than 5 red blood cells (RBCs) per high-power field (HPF). Hematuria described as bright red is usually indicative of lower urinary tract bleeding whereas glomerular hematuria (as in nephritis) is usually reported as cola- or tea-colored or brown. Presence of blood clots and dysuria, as in the patient in the vignette, is indicative of cystitis (commonly viral in origin) as the underlying cause of her hematuria. Presence of blood clots also suggests lower urinary tract bleeding as a cause of the hematuria.

Immunoglobulin A nephropathy is associated with glomerular hematuria and is unlikely in this patient presenting with bright red hematuria associated with clots.
Acute pyelonephritis is a clinical diagnosis based on the presenting symptoms of fever, vomiting, and flank pain in a teenage patient. The absence of these symptoms makes acute pyelonephritis unlikely in the patient in the vignette.

Pigmenturia (hemoglobinuria and myoglobinuria) can result in a positive finding on the dipstick test for blood, but both conditions lack the presence of RBCs on microscopy. Myoglobinuria and paroxysmal nocturnal hemoglobinuria are unlikely in this patient because the urine microscopy shows more than 100 RBC/HPF.

PREP Pearls

- Nonglomerular hematuria is usually described as red or pink on a patient's history.
- Glomerular hematuria is usually described as cola-colored, tea-colored, or brown.
- Presenting complaints of bright red hematuria with clots and dysuria are suggestive of underlying cystitis.
- Pigmenturias (hemoglobinuria and myoglobinuria) can result in a positive dipstick test result for blood, but urine microscopy in such patients does not show red blood cells.

American Board of Pediatrics Content Specification(s):

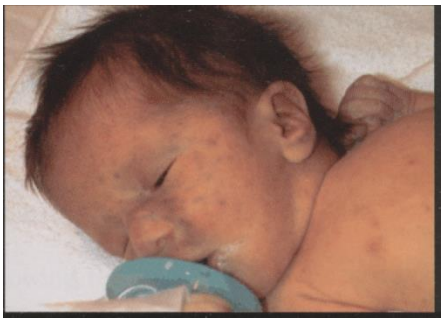
- Determine by history and laboratory evaluation the etiology of red urine

Suggested Reading:

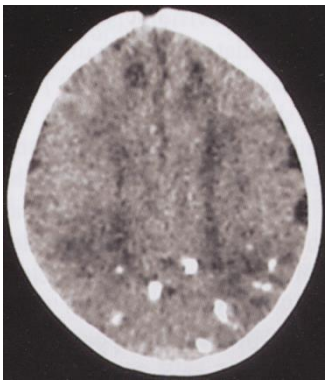
- Massengill SF. Hematuria. Pediatr Rev. 2008;29:342-348. doi: 10.1542/ pir.29-10-342
- Reidy KJ, Rio MD. Hematuria In: McInerny TK, Adam HM, Campbell DE, Kamat DM, Kelleher KJ, eds. American Academy of Pediatrics Textbook of Pediatric Care. 19th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2009;1566-1570

Item 197

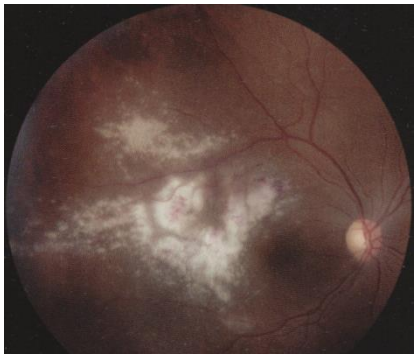
A 12-day-old term neonate is being evaluated for congenital infection. On physical examination, the vital signs are normal, the head circumference is at the 5th percentile, and length and weight are at the 20th percentile for age. The facies are normal, and the palate is intact. The liver edge and spleen are palpated at 5 cm and 4 cm below the costal mar-gins, respectively. There is a rash that involves the face, trunk, and extremities (Item Q 197A). The remainder of the physical examination findings is normal.



ITEM Q197A: Rash as described for the infant in the vignette.



ITEM Q197B: Computed tomography of the head for the infant in the vignette.



ITEM Q197C: Fundoscopic findings exhibited by the infant in the vignette

Laboratory findings include the following:

- White blood cell count, 15,600/µL (15.6 x 10^9/L), with 20% neutrophils, 68% lymphocytes, and 12% monocytes
- Hemoglobin, 10 g/dL (100 g/L)
- Platelet count, 55 x 103/µL (55 x 10^9/L)
- Aspartate aminotransferase, 55 U/L
- Alanine aminotransferase, 38 U/L

Cerebrospinal fluid examination reveals the following:

- White blood cells, 15/µL, with 80% lymphocytes and 20% monocytes
- Red blood cells, 100/µL
- Protein, 60 mg/dL
- Glucose, 80 mg/dL

A chest radiograph is unremarkable. Computed tomography scan of the brain (Item Q197B) and an ophthalmologic examination (Item Q197C) are performed.

Of the following, the MOST likely cause of the infant's dis-order is infection with

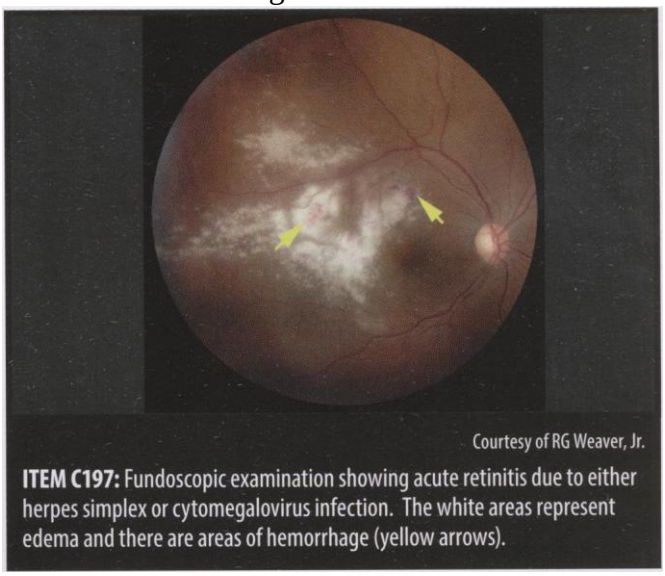
A. cytomegalovirus
B. herpes simplex virus
C. human immunodeficiency virus
D. rubella virus
E. Treponema pallidum

Item 197 Preferred Response: A

The infant described in the vignette has microcephaly, periventricular calcifications on computed tomography, chorioretinitis on ophthalmologic examination, hepatosplenomegaly, a purpuric rash, anemia, and thrombocytopenia. This constellation of signs and symptoms strongly suggests congenital infection due to cytomegalovirus (CMV).

Approximately 1% of newborns have congenital CMV infection, 90% of whom are asymptomatic. The 10% of infants with symptomatic congenital infection may have manifestations, including intrauterine growth retardation, microcephaly, intracranial calcifications (often periventricular), chorioretinitis, petechial or purpuric rash, hepatosplenomegaly, and jaundice. Developmental delay and sensorineural hearing loss are common. Laboratory abnormalities can include thrombocytopenia, hemolytic anemia, transaminitis, and hyperbilirubinemia (direct and indirect). Isolation of CMV from urine, cerebrospinal fluid (CSF), stool, or respiratory tract secretions collected within the first 2 to 4 weeks of life can establish the diagnosis of congenital CMV infection. Detection of CMV DNA in urine, serum, or newborn dried blood spots using the polymerase chain reaction (PCR) assay also may be used to establish the diagnosis, but these methods are costly and less widely available.

Congenital herpes simplex virus (HSV) infection is uncommon and is most often encountered after maternal primary HSV infection during pregnancy. Intrauterine demise is common. Survivors with congenital HSV infection may have skin vesicles or scarring. Severe central nervous system manifestations can include microcephaly or hydranencephaly. Eye abnormalities, including retinitis (C197), can occur. Rarely, neonates with intrauterine infection present with symptoms of disseminated HSV. The diagnosis is established by isolation of virus from blood, skin lesions, CSF, eye or conjunctivae, mouth or oropharyngeal secretions, and stool or rectum. The detection of HSV DNA, especially in the first 48 to 72 hours of life, in the blood or CSF using PCR also confirms the diagnosis.



Courtesy of RG Weaver, Jr.

ITEM C197: Fundoscopic examination showing acute retinitis due to either herpes simplex or cytomegalovirus infection. The white areas represent edema and there are areas of hemorrhage (yellow arrows).

The most common manifestations of congenital rubella syndrome in neonates include ophthalmologic abnormalities (cataracts, congenital glaucoma, pigmentary retinopathy, or microphthalmos), congenital heart disease (patent ductus arteriosus or peripheral pulmonary artery stenosis), hearing impairment, and neurologic abnormalities (developmental delay, meningoencephalitis, or microcephaly). Other manifestations can include intrauterine growth retardation, interstitial pneumonitis, hepatosplenomegaly, dermal erythropoiesis ("blueberry muffin" rash), jaundice, thrombocytopenia, and radiolucent bone disease. The diagnosis is established by detecting rubella IgM antibody at birth or stable or increasing rubella IgG titers in the first several months of life. Rubella virus can be isolated from blood, urine, cataract specimens, and throat or nasal secretions. Reverse-transcriptase RNA PCR testing for the detection of rubella virus in throat secretions or urine may be available in some laboratories.

Most infants born with congenital infection due to Treponema pallidum (syphilis) are asymptomatic. Intrauterine infection can result in stillbirth, fetal hydrops, or preterm birth. Manifestations in symptomatic infants can include fever, rash, mucocutaneous lesions, rhinitis ("snuffles"), lymphadenopathy, hepatosplenomegaly, pneumonia, myocarditis, edema, osteochondritis, extremity pain (pseudoparalysis), hemolytic anemia, and thrombocytopenia. Definitive diagnosis is established by identifying spirochetes by darkfield microscopy examination of tissues, lesions, or exudates (rarely performed). A presumptive diagnosis is made using nontreponemal (rapid plasma reagin or VDRL tests) and treponemal (fluorescent treponemal antibody absorption, T pallidum particle agglutination, or microhemagglutination for T pallidum) tests.

Neonates with congenitally acquired human immunodeficiency virus (HIV) infection are asymptomatic. An HIV DNA PCR assay is the preferred test for diagnosing HIV infection in infants and children younger than 18 months, who may still have circulating maternal antibodies against the virus.

PREP Pearls

- Approximately 1% of newborns have congenital CMV infection; 90% of infected infants are asymptomatic.
- Microcephaly, periventricular calcifications, chorioretinitis, hepatosplenomegaly, a purpuric rash, anemia, and thrombocytopenia suggest congenital CMV infection.
- Isolation of CMV from urine, CSF, stool, or respiratory tract secretions collected within the first 2 to 4 weeks of life can establish the diagnosis of congenital infection.
- Detection of CMV DNA in urine, serum, or newborn dried blood spots using PCR assay also may be used to establish the diagnosis, but these methods are less widely available and costly.

American Board of Pediatrics Content Specification(s):

- Know the diagnostic tests for congenital or acquired cytomegalovirus infection

Suggested Reading:

- American Academy of Pediatrics. Cytomegalovirus infection. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:300-305
- American Academy of Pediatrics. Herpes simplex virus. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:398-408
- American Academy of Pediatrics. Human immunodeficiency virus infection. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:418-439
- American Academy of Pediatrics. Rubella. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:629-634
- American Academy of Pediatrics. Syphilis. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:690-703

Item 198

You receive a message from the mother of a 14-year-old girl who you will be seeing in the office today. The mother requests that you test her daughter for drugs and that you not tell her daughter that she made such a request. When they arrive, both mother and daughter enter the examination room together.

Of the following, the MOST appropriate way to manage this situation is to

A. ignore the request as the subject is not brought up during the encounter
B. inform the patient of her mother's request and ask for a urine sample
C. obtain a urine sample and then inform the patient you will be testing her
D. request to speak with the patient alone before proceeding with testing
E. send a urine drug screen without the patient's knowledge

Item 198 I-C P Preferred Response: D

In addition to speaking with parents and their adolescents together, speaking with a parent and an adolescent separately to discuss confidentiality and its limits is important to developing a trusting relationship. This will in turn allow for open communication. The history is the most important part of the visit with an adolescent and will aid in the diagnosis in most cases. This should occur irrespective of the age of the adolescent. Knowing that the adolescent has access to a trusted physician will allow the adolescent to open up, if not at a current visit then in the future, when ready to reveal details.

Requests made by parents should never be ignored. In the vignette, the mother's concerns should be addressed when speaking with her alone. She needs to understand that obtaining a drug screen without her daughter's knowledge will make it more difficult to help her if there is a substance use problem because the physician-patient relationship will be compromised. After speaking with the mother alone, speaking with the adolescent alone will allow for developing a bigger picture of her functioning at home, at school, and with her peers, along with assessing her mental health status. This information will be more useful in deciding next steps. Asking for a urine sample in the mother's presence without previous discussion will not give the adolescent a chance to discuss the situation, and she may feel pressured to comply, with resultant loss of trust.

PREP Pearls

- The role of confidentiality in being able to obtain a complete psychosocial history cannot be underestimated.
- Confidentiality should be discussed with families and provided to all adolescents of all ages.
- Drug screens should not be obtained without an adolescent's knowledge.

American Board of Pediatrics Content Specification(s):

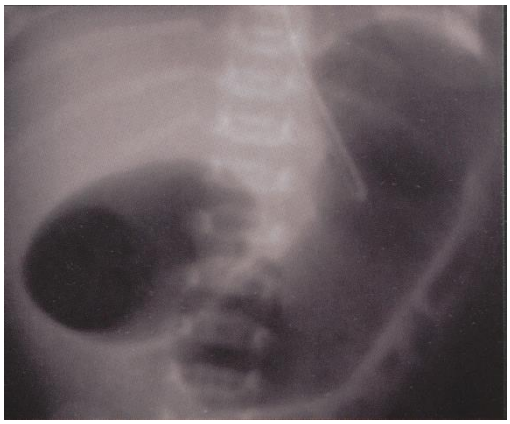
- Know that age alone does not dictate whether parents accompany adolescents during the physical examination and history
- Understand the importance of interviewing an adolescent without the parents present

Suggested Reading:

- American Academy of Pediatrics: Policy Statement: Substance Use Screening, Brief Intervention, and Referral to Treatment for Pediatricians Committee on Substance Abuse. Pediatrics. 2011;128:e1330-e1340. DOI: 10.1542/peds.2011-1754
- Britto MT, Tivorsak TL, Slap GB. Adolescents' needs for health care privacy. Pediatrics. 2010;126: e1469-e1476. doi:10.1542/peds.2010-0389
- Committee on Substance Abuse and Council on School Health. Testing for drugs of abuse in children and adolescents: addendum—testing in schools and at home. Pediatrics. 2007;119: 627-630. DOI: 10.1542/peds.2006-3688
- Henry-Reid LM, O'Connor KB, Klein JD, Cooper E, Flynn P, Futterman DC. Current pediatrician practices in identifying high-risk behaviors of adolescents. Pediatrics. 2010;125: e741-e747. doi:10.1542/peds.2009-0271
- Levine SB. Adolescent consent and confidentiality. Pediatr Rev. 2009;30:457-459. DOI: 10.1542/pir.30-11-457

Item 199

You are called to the nursery to see a 6-hour-old, full-term neonate who has just vomited a large amount of clear, yellow fluid. The baby was the 2,500-g product of a full-term, uncomplicated pregnancy and was born to a gravida 2, para 2 mother. You ask the resident on call to pass a nasogastric tube and obtain an abdominal radiograph (Item Q199).



ITEM Q199: Abdominal radiograph for the infant in the vignette.

Of the following, this radiographic finding is MOST likely to be associated with

A. annular pancreas
B. congenital Ladd bands
C. Peutz–Jegher syndrome
D. trisomy 18
E. trisomy 21

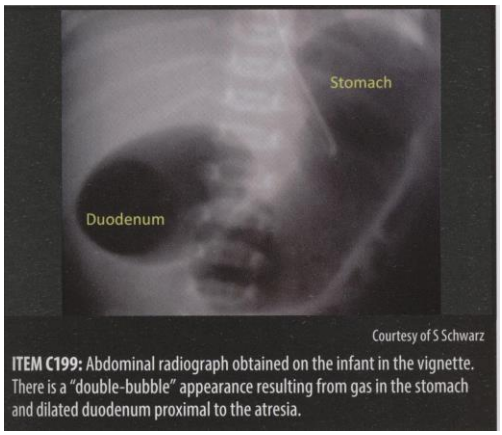
Item 199 Preferred Response: E

When bilious emesis is suspected in any patient, especially when bile-tinged fluid appears with the initial episode of vomiting (in contrast to protracted vomiting and retching), management must include assessment for functional or mechanical bowel obstruction. In the vignette, a 6-hour-old infant has vomited a large amount of bilious fluid. The emergency response should be to temporarily relieve obstructive symptoms by passing a nasogastric tube, followed by a diagnostic evaluation. In the newborn, this scenario suggests mechanical obstruction as the consequence of one of the following gut malformations: intestinal malrotation with midgut volvulus, intestinal atresia, or intestinal stenosis. The abdominal radiograph for the infant in the vignette demonstrates the presence of a typical "double bubble" sign characteristic of duodenal atresia (Item C199). This anomaly represents the most common form of fetal intestinal atresia, occurring in 1 in 10,000 births. Importantly, duodenal atresia is most commonly encountered in patients who have trisomy 21; such infants account for 25% to 40% of cases. In trisomy 21, the incidence of duodenal atresia has been reported to be as high as 1 in 12 fetuses. Duodenal atresia is not only the most common intestinal atresia but is also the most important cause of complete duodenal obstruction. Although the causative factors are not fully understood, duodenal atresia is thought to arise from a failure of duodenal recanalization between the 9th and 11th weeks of gestation. This is in contrast to jejunal and ileal atresias, which are thought to be related to intrauterine vascular accidents. The postpartum, double bubble radiographic appearance corresponds, respectively, to a gas- and fluid-filled stomach and duodenum proximal to the atresia. Polyhydramnios occurs as the result of impaired absorption of amniotic fluid by the fetal intestine in approximately half of the cases.

Fifty percent of cases of duodenal obstruction (stenosis or atresia) occur in the presence of other anomalies, including the following:

- Intestinal malrotation
- Esophageal atresia
- Ectopic or imperforate anus
- Annular pancreas
- Biliary atresia
- Renal anomalies
- Vertebral anomalies

Other, less frequent diagnoses include partial or complete obstruction caused by a duodenal web, Ladd bands, or a preduodenal portal vein. Another chromosomal disorder, trisomy 18 (Edwards syndrome), may be associated with various gastrointestinal anomalies, including malrotation. In any newborn with suspected duodenal obstruction, midgut volvulus remains the most important differential diagnosis to consider. When present at birth, midgut volvulus typically appears as partial duodenal obstruction on a small bowel radiographic series. However, complete obstruction may also be present, making the distinction between midgut volvulus and duodenal atresia difficult. When duodenal obstruction occurs after the immediate newborn period, a diagnosis of midgut volvulus must be assumed until ruled out.



ITEM C199: Abdominal radiograph obtained on the infant in the vignette. There is a "double-bubble" appearance resulting from gas in the stomach and dilated duodenum proximal to the atresia.

PREP Pearls

- Bilious vomiting in the newborn is always a sign of intestinal obstruction
- Trisomy 21 represents the most common genetic abnormality associated with intestinal atresias
- In the newborn with Bilious vomiting, the upper GI series to the ligament of Treitz is indicated to rule out malrotation

American Board of Pediatrics Content Specification(s):

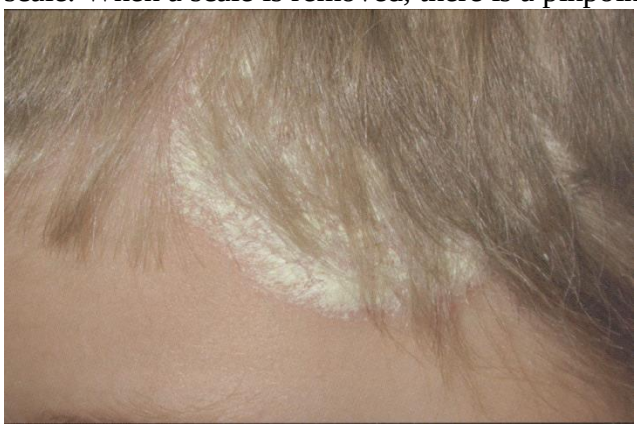
- Know the clinical situations in which duodenal atresia may occur

Suggested Reading:

- Applebaum H, Lee SL, Puapong DP. Duodenal atresia and stenosis: annular pancreas. In: Grosfeld JL, O'Neill JA, Fonkalsrud E, Coran AG. Pediatric Surgery. Philadelphia, PA: Mosby Elsevier; 2006:1260-1268
- Choudhry MS, Rahman N, Boyd P, Lakhoo K. Duodenal atresia: associated anomalies, prenatal diagnosis and outcome. Pediatr Surg Int. 2009;25:727-730. doi:10.1007/s00383-009-2406-y
- Escobar MA, Ladd AP, Grosfeld JL, et al. Duodenal atresia and stenosis: long-term follow-up over 30 years. I Pediatr Surg. 2004;39:867-871. doi:10.1016/j.jpedsurg.2004.02.025
- Freeman SB, Torfs CP, Romitti PA, et al. Congenital gastrointestinal defects in Down syndrome: a report from the Atlanta and National Down Syndrome Projects. Clin Genet. 2009;75:180-184. doi:10.1111/j.1399- 0004.2008.01110.x
- Piper HG, Alesbury J, Waterford SD, Zurakowski D, Jaksic T. Intestinal atresias: factors affecting clinical outcomes. J Pediatr Surg. 2008;43:12441248. doi:10.1016/j.jpedsurg.2007.09.053

Item 200

A 9-year-old boy has had a rash for several months with-out associated systemic symptoms. Examination shows red papules and plaques with sharply demarcated but irregular borders that mainly involve the scalp (Item Q200), knees, elbows, umbilicus, and gluteal cleft. On the larger plaques, there is silvery to yellow-white, thick, adherent scale. When a scale is removed, there is a pinpoint area of bleeding.



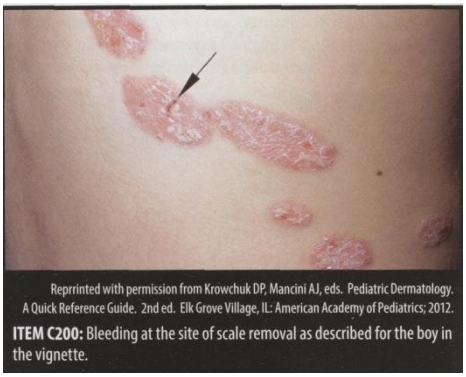
ITEM Q200: Lesions on the scalp as described for the boy in the vignette.

Of the following, the MOST appropriate initial treatment for this child is

A. oral corticosteroid
B. oral erythromycin
C. topical calcipotriene
D. topical 1% hydrocortisone
E. topical mupirocin

Item 200 Preferred Response: C

Psoriasis is a common inflammatory skin condition accounting for approximately 4% of all pediatric dermatoses. It is characterized by inflammation and hyperproliferation of the epidermis. Approximately 35% of all cases have their onset before age 20 years. Genetics play a role in the development of psoriasis, with many pediatric patients having an affected family member. A strong association has been found between the HLA-Cw6 allele and pediatric onset of the condition. Trauma, infections (group B Streptococcus, human immunodeficiency virus), drugs (lithium,β-blockers, steroid withdrawal), and stress have all been implicated as triggers. The Koebner phenomenon occurs when psoriatic lesions are seen at the site of preceding trauma. A recent study also indicated that exposure to tobacco smoke in the home, a stressful life event in the preceding year, and body mass index greater than 76 are all more common in pediatric psoriasis sufferers compared with control subjects.



Reprinted with permission from Krowchuk DP, Mancini AJ, eds. Pediatric Dermatology. A Quick Reference Guide. 2nd ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012.

ITEM C200: Bleeding at the site of scale removal as described for the boy in the vignette.

Psoriasis has several subcategories.

1. The most common presentation in children is the plaque form. In this variety, one sees well-defined red papules and plaques with silvery scales. The Auspitz sign is nearly diagnostic: pinpoint areas of bleeding occur in areas where scales are removed (Item C200). The lesions are often pruritic and are distributed widely, including the trunk, extensor surfaces of the extremities, and scalp. A rarer form called inverse psoriasis affects the face and flexor surface.
2. Linear psoriasis involves psoriatic lesions that follow the lines of Blaschko (linear skin lines, usually invisible. thought to represent pathways of epidermal cell movement during embryonic development). Sonic experts feel this is not a distinct category of psoriasis.
3. psoriatic diaper rash affects children younger than 2 years of age with sharply demarcated, bright red papules in the diaper area including the inguinal folds.
4. Guttate psoriasis follows a group A streptococcal infection either of the throat or perianal area. Onset is rapid with 1-cm papules symmetrically distributed on the trunk, proximal extremities, face, and scalp. It typically resolves in 3 to 4 months but frequently recurs.
5. Pustular psoriasis is rare in children but can be seen in either generalized or localized distributions. The localized form is especially common on the plantar or palmar areas and consists of sterile pustules on an erythematous base. One generalized form presents with annular lesions.

Although nail changes can occur with several forms of psoriasis, they are most common with digital rash or with psoriatic arthritis, which are more frequent in adults. Nail changes include pits, discoloration, splitting, hyperkeratosis, onychodystrophy, and splinter hemorrhages.

Treatment for psoriasis can be either topical or systemic. Topical therapies may include mid- to high-potency steroids, phototherapy, vitamin D3 analogs, calcineurin inhibitors,

and tar derivatives such as anthralin. Vitamin D3 analogs, including calcipotriene, would be an appropriate treatment for the patient in the vignette who has mild to moderate plaque psoriasis. Low-potency steroids such as 1% hydrocortisone would be unlikely to benefit the patient. Oral corticosteroids and oral or topical antibiotics would not be the most appropriate initial therapy for this patient.

If the patient does not respond adequately to topical treatment, systemic therapy may be required; however, data about the effectiveness of systemic therapy for children are limited. Acitretin, a retinoid, is often used in combination with topical therapies. The immunosuppressive agents methotrexate and cyclosporine have been used successfully in children with severe disease. Tumor necrosis factor a inhibitors are being used increasingly for adults, particularly those with psoriatic arthritis. Case reports have shown successful use in children, but no systematic evaluations of these agents have been made at this point.

PREP Pearls

- Psoriasis is a relatively common dermatosis even in childhood, with 20% of all sufferers reporting onset of disease before age 20 years.
- Genetics play an important role in the causation of this condition with a strong association between the HLA Cw6 allele and childhood onset of disease.
- The primary forms seen in childhood include plaque, diaper rash, and guttate psoriasis.
- Topical treatments for psoriasis include mid- to high-potency corticosteroids, vitamin D3 analogs (calcipotriene), calcineurin inhibitors, tar derivatives, and phototherapy.
- Immunosuppressive agents and biologics may prove very useful in the treatment of psoriasis.

American Board of Pediatrics Content Specification(s):

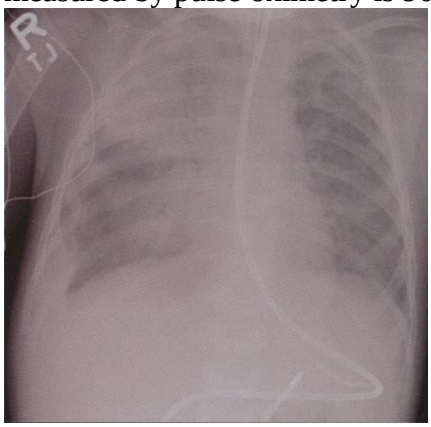
- Identify psoriasis

Suggested Reading:

- Bard S, Torchia D, Schachner LA. Managing pediatric patients with psoriasis. Am I Clin Dermatol. 2010;11(suppl 1):15-17. doi: 10.2165/1153415 SO-000000000-00000
- Benoit S, Hamm H. Childhood psoriasis. Clin Dermatol. 2007;25:555-562. doi: 10.1016/j.clindermato1.2007.08.009
- Psoriasis. In: Krowchuk DP, Mancini AL eds. Pediatric Dermatology:
- A Quick Reference Guide. Elk Grove Village, IL: American Academy of Pediatrics; 2012:301-306
- Ozden MG, Tekin NS, Gurer MA, et al. Environmental risk factors in pediatric psoriasis: a multicenter case-control study. Pediatr Dermatol.
- 2011;28:306-312. doi:10.1111/j.1525-1470.2011.01408.x

Item 201

A 2-year-old boy is admitted to the pediatric inpatient service 4 hours after swallowing a small amount of kerosene. On physical examination, the boy has a temperature of 38.0°C, heart rate of 130 beats/min, blood pressure of 85/50 min Hg, and respiratory rate of 40 breaths/min. He is awake and alert with mild nasal flaring; he has bilateral breath sounds with mild wheezing and mild intercostal retractions. His oxygen saturation on room air as measured by pulse oximetry is 90%. A chest radiograph is obtained (Item Q201).



ITEM Q201: Radiographic findings for the boy in the vignette.

Of the following, the MOST appropriate next step in the management of this boy is administration of

A. activated charcoal
B. albuterol
C. antibiotics
D. corticosteroids
E. isoproterenol

Item 201 S Preferred Response: B

Hydrocarbons represent about 2% of all pediatric non-pharmaceutical ingestions in the United States. Hydrocarbons generally are ingested in small amounts because of the foul taste, but can produce significant morbidity and mortality as evidenced by the fact that hydrocarbons comprise 10% of pediatric ingestion fatalities in the United States. Symptoms of hydrocarbon ingestion can be limited to the respiratory system (direct effect of the hydrocarbon on pulmonary tissue) or they can be systemic (due to gastrointestinal and respiratory absorption). Respiratory symptoms include coughing, choking, tachypnea, and cyanosis. Systemic symptoms and clinical findings include central nervous system depression, cardiac dysrhythmias, fever, leukocytosis, hemolysis, and hemoglobinuria. Hydrocarbons produce both destruction of the respiratory components (airway epithelium, alveolar septae, and pulmonary capillaries) and dissolution of the lipid surfactant layer. These processes can result in chemical pneumonitis, pneumothorax, necrotizing pneumonia, respiratory failure, and death.

Management is primarily supportive. Patients often develop bronchospasm and should be treated with a 82 selective agent such as albuterol. Isoproterenol should be avoided because of myocardial sensitization and subsequent risk of fatal dysrhythmias. The pneumonitis associated with hydrocarbon ingestion is chemical and, therefore, antibiotics are indicated only if there are signs of secondary infection (fever or progression of infiltrates on chest radiography after 48 hours). Corticosteroids have not been shown to reduce respiratory symptoms or damage. External decontamination (removal of contaminated clothes and cleansing of skin) should be performed. Gastric decontamination (administration of syrup of ipecac or nasogastric lavage) should be avoided because of the risk of aspiration. The use of activated charcoal is contraindicated in single-agent hydrocarbon ingestion and when the patient presents with pulmonary symptoms, as seen in the patient in the vignette. Activated charcoal might be recommended by the poison control center in a multiple substance ingestion with systemic toxicity.

PREP Pearls

- Ingestion of hydrocarbons represents about 2% of all pediatric nonpharmaceutical ingestions in the United States but cause 10% of pediatric ingestion fatalities.
- Hydrocarbon ingestion can manifest in respiratory symptoms or systemic symptoms if gastrointestinal or respiratory absorption occurs.
- Gastric decontamination (administration of syrup of ipecac or nasogastric lavage) should be avoided in cases of hydrocarbon ingestion because of the risk of aspiration.

American Board of Pediatrics Content Specification(s):

- Understand that hydrocarbon pneumonitis may cause acute and chronic lung disease

Suggested Reading:

- Colombo JL. Aspiration syndromes. In: Kliegman RM, Stanton BMD, St Geme J, Schor N, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Elsevier Saunders; 2011:1469-1471
- Lewander WJ, Aleguas A. Hydrocarbon poisoning. UptoDate. Available online only for subscription

Item 202

A 13-year-old girl presented to your office 6 months ago for evaluation of weight gain and was diagnosed with subclinical hypothyroidism (thyroid-stimulating hormone level of 7.5 mIU/L, with a normal free thyroxine [FT4] of 1.2 ng/ dL [15 pmol/L]). She was not treated with any medication at that time. She returns now for follow-up and you notice a small goiter on physical examination. She is still overweight. Her linear growth is normal, and she still has regular menses. She denies constipation or dry skin. Repeat thyroid function testing shows her thyroid-stimulating hormone and FT4 are unchanged from 6 months ago.

Of the following, the test that is MOST likely to predict progression of this patient's thyroid disease is

A. iodine-123 uptake scan
B. thyroid-stimulating immunoglobulin
C. thyroid ultrasonography
D. thyroid peroxidase antibody titer
E. triiodothyronine (T3) level

Item 202 Preferred Response: D

Subclinical hypothyroidism (SCH), defined as a normal thyroxine (T4) level with a mildly elevated thyrotropin (TSH) level (typically 5-10 mIU/L), is common in children. Several pediatric studies indicate that progression of SCH to overt hypothyroidism is uncommon and that in a period of several years elevated TSH levels can normalize or, in many cases, not increase.

The most common cause of acquired overt hypothyroidism in both children and adults is autoimmune thyroiditis (Hashimoto disease). An elevated thyroid peroxidase (TPO) antibody concentration would be most consistent with this diagnosis. In general, larger thyroid gland volume (goiter) and higher TSH levels predict further elevation of TSH in the future, indicating progression to overt disease. Hashimoto thyroiditis can be present for years without progression to overt hypothyroidism, and it is not clear whether treatment is indicated with only a mild elevation of TSH.

Recent studies have reported that obesity is linked to mild elevations of TSH and that elevated leptin levels may result in increased TSH levels, suggesting obesity as a possible cause of SCH in some patients. Thus, in obese patients who have a mild elevation of TSH and no evidence of goiter and are TPO antibody negative, treatment with T4 may not be needed. The patient in this vignette ultimately developed a goiter and ended up having positive TPO antibodies, suggesting her disease was likely to progress; therefore, treatment would be indicated.

Thyroid imaging studies can reveal changes in uptake on iodine-123 scan, or thyroid ultrasonography may reveal a diffusely enlarged heterogeneous gland, but neither of these is diagnostic of Hashimoto thyroiditis.

Thyroid-stimulating immunoglobulins are generally elevated in patients with Graves disease, in whom elevated T4 and triiodothyronine (T3) levels can be seen.

PREP Pearls

- TPO antibodies, goiter, and higher TSH levels increase the likelihood of future development of overt hypothyroidism.
- Progression of SCH to overt disease is uncommon in patients who do not have TPO antibodies, goiter, or a higher baseline TSH level in the absence of associated conditions (eg, trisomy 21 or type 1 diabetes mellitus and other autoimmune diseases).

American Board of Pediatrics Content Specification(s):

- Know the natural history of Hashimoto thyroiditis

Suggested Reading:

- Kaplowitz PB. Subclinical hypothyroidism in children: normal variation or sign of a failing thyroid gland? Int 1 Pediatr Endocrinol. 2010:281453. doi:10.1155/2010/281453
- Reinehr T. Thyroid function in the nutritionally obese child and adolescent. Curr Opin Pediatr. 2011;23:415-420. doi:10.1097/ MOP.0b013e328344c393

Item 203

A 5-year-old girl presents to your office for evaluation of a 3-week history of cervical lymph node swelling and temperature up to 39.2°C. She was evaluated 7 days ago in an urgent care center and treated with clindamycin without improvement. The family reports that they have a 6-month-old kitten at home and that the girl plays with her frequently. Physical examination reveals a temperature of 38.8°C and a 2.5-cm erythematous, swollen, and tender cervical lymph node. Complete blood cell count reveals a white blood cell count of 14,600/pL (14.6 x 10A), with 72% neutrophils, 19% lymphocytes, 7% monocytes, and 2% eosinophils.

Of the following, the BEST test for confirming the diagnosis is

A. Bartonella henselae antibody titer
B. culture obtained from incision and drainage of the lymph node
C. intradermal skin test with cat scratch disease–antigen
D. Toxocara cati antibody titer
E. Toxoplasma gondii antibody titer

Item 203 Preferred Response: A

Regional erythematous cervical lymphadenitis persisting over several weeks, the failure to respond to clindamycin, and the exposure of the child in the vignette to a kitten suggest a diagnosis of cat scratch disease (CSD). Bartonella henselae is the most common cause of CSD and a specific serologic test for this organism is the best method for confirming this diagnosis.

If a diagnosis of CSD is confirmed, incision and drainage of the involved lymph node is not indicated unless there is marked fluctuantes. If a lymph node biopsy is performed, the diagnosis of CSD can be confirmed by observing the organism on a Warthin-Starry silver stain of the tissue. Polymerase chain reaction (PCR) assays for detecting B henselae in tissue specimens are available through the Centers for Disease Control and Prevention. An intradermal skin test for CSD prepared from antigenic material obtained from infected lymph nodes was an early diagnostic test for this condition but because of lack of standardization and concerns about using human tissue, this test is no longer available. Toxocara infection is not associated with lymphadenitis. Symptomatic toxocariasis is associated with symptoms that may include wheezing, asymptomatic eosinophilia, and visceral or ocular involvement. Toxoplasma gondii is a parasite found in contaminated soil, undercooked meat, and cat intestines. Symptomatic infection is associated with a mononucleosis-like illness or generalized lymphadenopathy.

Regional lymphadenitis in the region draining from the site of the scratch is the most common clinical manifestation of CSD. A papule at the site of inoculation may be seen and systemic manifestations with low-grade fever also may occur. Inoculation of the conjunctiva may result in oculoglandular syndrome with conjunctivitis and ipsilateral preauricular lymphadenopathy. Less common manifestations of CSD include fever of unknown origin, aseptic meningitis, encephalopathy, neuroretinitis, osteolytic lesions, granulomatous hepatitis, erythema nodosum, and endocarditis.
Bartonella henselae is sensitive to a number of antibiotics including azithromycin, ciprofloxacin, trimethoprim-sulfamethoxazole, rifampin, and gentamicin; however, the benefit of treating localized disease in a normal host is uncertain.

PREP Pearls

- Bartonella henselae is the causative agent of cat scratch disease (CSD); diagnosis is confirmed by detecting antibodies against the organism.
- Regional lymphadenitis is the leading clinical manifestation of CSD.

American Board of Pediatrics Content Specification(s):

- Know how to diagnose cat-scratch disease

Suggested Reading:

- American Academy of Pediatrics. Cat Scratch Disease (Bartonella henselae). In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012: 269-271

- Florin TA, Zaoutis TE, Zaoutis LB. Beyond cat scratch disease: widening spectrum of Bartonella henselae infection. Pediatrics.

Item 204

A 3-year-old girl is seen for complaints of persistent left-knee swelling and limping that began 6 weeks ago. She was taken to the emergency department for evaluation shortly after the symptoms began. A plain radiograph of the knee showed soft tissue swelling, synovial fluid evaluation was unremarkable, and results of her complete blood cell count and basic metabolic panel were within reference range. The family has treated her with acetaminophen with mild improvement of her limp but no improvement in the swelling. There is no history of trauma, fever, or recent illness. Findings on physical examination reveal a warm, erythematous, swollen left knee that is tender to touch and has decreased range of motion. A repeat complete blood cell count and complete metabolic panel are within reference range, Lyme titer is negative, and erythrocyte sedimentation rate is 40 mm/h. A diagnosis of juvenile idiopathic arthritis is made.

Of the following, the BEST initial treatment is

A. codeine
B. hydroxychloroquine
C. prednisolone
D. naproxen
E. sulfasalazine

Item 204 Preferred Response: D

The patient described in the vignette has been diagnosed with juvenile idiopathic arthritis (JIA) by ruling out infection and trauma and by the presence of arthritis for at least 6 weeks.

Nonsteroidal anti-inflammatory drugs (NSAIDs) are the recommended initial treatment for JIA (Item C204, page C-161). NSAIDs are good analgesics and mild antiinflammatory medications that work by blocking the cyclooxygenase enzymes. Naproxen is an NSAID that is recommended as the first-line treatment for both JIA and reactive arthritis. Common adverse effects include gastrointestinal irritation manifested by abdominal pain, nausea, or heartburn. The most commonly used NSAIDs are ibuprofen, naproxen, and meloxicam. These come in a liquid or pill form and are inexpensive and generally well tolerated. Celecoxib and other NSAIDs are also used for patients who have JIA. Codeine would treat the pain but has no antiinflammatory effect that would benefit the patient in the vignette.

Prednisone can be used to treat arthritis but would not be the best initial treatment. In addition to having a greater profile of adverse effects than NSAIDs, steroids can mask symptoms of a chronic arthritis and make it difficult for ongoing assessment by a rheumatologist.

Disease-modifying antirheumatic drugs (DMARDs) are also used in JIA to inhibit the immune system and slow progression of the disease; they include methotrexate, leflunomide, hydroxychloroquine, and sulfasalazine. Disease-modifying antirheumatic drugs are used off-label in JIA and are best monitored by a rheumatologist. Methotrexate is a folic acid analogue that competitively inhibits dihydrofolate reductase. Oral ulcers, nausea, bone marrow suppression, and hepatic transaminitis are the most commonly reported adverse effects. Leflunomide blocks activated lymphocyte proliferation by blocking de novo pyrimidine synthesis. Hydroxychloroquine, which inhibits the synthesis of DNA and RNA, is sometimes used to treat arthritis, but it does not have strong evidence for effectiveness and should not be used alone. Sulfasalazine, which has several proposed mechanisms of action in arthritis, should not be used as a first-line agent Newer biologic therapies used in treating JIA target and block the function of cytokines and specific immune cell receptors. These include tumor necrosis factor-alpha (TNF-α) inhibitors (adalimumab and etanercept, US Food and Drug Administration [FDA] approved) and interleukin (IL) 1-receptor antagonists (anakinra, off label), T-cell activation inhibitors (abatacept, FDA approved), and IL6-receptor antagonists (tocilizumab, FDA approved). There has been an increased risk of infections (ie, fungal and tuberculosis) and rare cancers reported with some of these agents. Liver enzymes and blood cell counts should be monitored while on biologic medications.

Current treatment recommendations for JIA include an initial NSAID trial for low disease activity. The NSAID trial should not last longer than 2 months in a patient with active arthritis. In patients with 4 or fewer joints involved, intra-articular steroid injection may be used alone to control the arthritis. If the arthritis is not responsive or if more than 4 joints are involved, methotrexate with or without systemic or intra-articular

glucocorticoids is recommended. If the patient fails methotrexate, a TNF-α inhibitor, usually adalimumab or etanercept, is added to therapy. If the patient fails these regimens, abatacept, another biologic therapy, is recommended. In patients with 5 or more active joints, NSAIDs are not used as monotherapy. The recommendations are different for patients with systemic JIA (fevers, rash, and arthritis). The first-line drugs are still NSAIDs, but systemic corticosteroids are used if there is inadequate response. If there is failure to control the arthritis, methotrexate is used in patients without fever and rash. If fever and rash are present, anakinra is recommended.

PREP Pearls

- Nonsteroidal anti-inflammatory medications are recommended as first-line agents in JIA.
- Prednisone can mask symptoms of joint swelling and make the diagnosis of chronic arthritis difficult.
- Appropriate referral to rheumatology should be made for patients with 6 weeks of joint symptoms after infection, reactive arthritis, and trauma have been ruled out.

Item C204. Commonly Used Nonsteroidal Anti-inflammatory Drugs (NSAID)

Nonsteroidal Anti-inflammatory Drugs	Anti-inflamatory Dose*	Dosage Forms
Ibuprofen	10 mg/kg per dose four times a day OR 13mg/kg per dose three times a day (Maximum dose 2400mg/day)	Pill or liquid forms
Naproxen	10 mg/kg per dose twice daily (Maximum dose, 1,000 mg/d)	Pill or liquid forms
Meloxicam	0.25 mg/kg per dose once daily (Maximum dose, 15mg/day)	Pill or liquid forms
Celecoxib	50 mg twice daily for 2- to 17-year olds and those weighing 10–25kg. 100 mg twice daily for patients >25kg (Maximum dose, 200mg/day)	Caps may be opened, sprinkled onto apple sauce, and taken immediately with water

*The doses listed are anti-inflammatory doses for Nonsteroidal Anti-inflammatory Drugs. Smaller doses are used for fever and pain relief.

American Board of Pediatrics Content Specification(s):

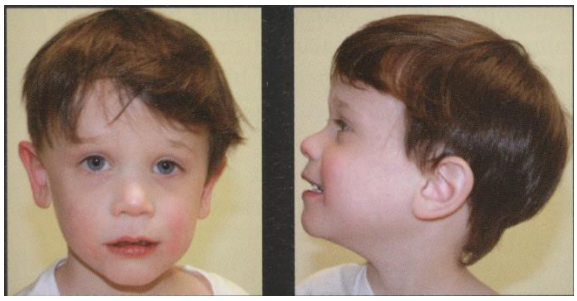
- Understand the pharmacologic treatment of juvenile rheumatoid (idiopathic) arthritis

Suggested Reading:

- Berard R. Approach to the child with joint inflammation. Pediatr Clin N Am. 2012;59(2):245-262. doi:10.1016/j.pc1.2012.03.003
- Espinosa M, Gottlieb B. Juvenile idiopathic arthritis. Pediatr Rev. 2012; 33:303-313; doi:10.1542/pir.33-7-303

Item 205

A 3-year-old boy is seen for a health supervision visit. He attends a special education preschool program designed for children who have developmental impairments. His mother notes that he has not yet begun to talk, but he appears to understand some of her requests. He is not interested in playing or interacting with other children. He tends to play with objects in a stereotyped fashion, such as pushing the same toy car back and forth repetitively. He insists on only wearing sweatpants and cotton T-shirts, regardless of the weather. The mother's pregnancy and delivery were uneventful. The child has not had any major medical illnesses beyond frequent ear infections. On physical examination, he makes no eye contact and has no spontaneous speech. His height and weight are both at the 50th percentile. He has large ears, prominent forehead and a slightly elongated face (Item Q205). His skin is dry but otherwise unremarkable. He has no syndactyly. His gait is normal and he is active, but he cannot jump or hop. Results of his cardiac examination are within normal limits



ITEM Q205: Boy described in the vignette.

Of the following, the MOST likely diagnosis is

A. fetal alcohol syndrome
B. fragile X syndrome
C. Rett syndrome
D. Smith–Lemli–Opitz syndrome
E. tuberous sclerosis

Item 205 Preferred Response: B

The boy described in the vignette most likely has an autism spectrum disorder evidenced by developmental delay, stereotyped interests, repetitive play with the same toy, lack of social interest in other children, and lack of eye contact during the examination. Fragile X syndrome is the most common genetic or developmental comorbidity with an autism spectrum disorder (present in about 5% of cases), and as such should be considered the most likely diagnosis from the choices provided. Furthermore, this patient has a few physical features that taken together should make one suspect fragile X syndrome: large ears, elongated face, and prominent forehead (though these are nonspecific). Other signs of fragile X syndrome could include a history of mild intellectual impairment in the mother, a high-arched palate, hyper-extensible joints, flat feet, hypotonia, and enlarged testes in older boys. Genetic testing for fragile X syndrome should be considered for patients with autism who have physical features or a family history suggestive of fragile X.

Fetal alcohol syndrome is a possible cause of retardation, but would be associated with different physical features including a smooth philtrum, thin upper lip, small palpebral fissures, and growth impairment. Fetal alcohol syndrome is typically associated with a socially outgoing personality and moderate-to-severe attention-deficit/hyperactivity disorder-like behavioral symptoms, which is different from the symptoms described for the patient in the vignette.

Children with Rett syndrome often demonstrate symptoms of autism, but this syndrome is a far rarer disorder than fragile X. Rett syndrome is also typically associated with other unique characteristics such as hand wringing, seizures, deceleration of head growth, scoliosis, and constipation. Rett syndrome is also rarely found in boys because of the relative lethality of the unique MECP2 X-linked genetic mutation; male patients with this condition who survive typically only live for a few years.

Smith-Lemli-Opitz syndrome is an autosomal recessive defect in cholesterol metabolism characterized by microcephaly, syndactyly or polydactyly, growth retardation, intellectual disability, cleft palate, and hypospadias.

Tuberous sclerosis is sometimes associated with autism spectrum disorder, but it is distinguished by different and unique physical signs such as hamartomas and angiofibromas on the skin, ash leaf spots, shagreen patches, and epilepsy.

PREP Pearls

- The most common genetic syndrome associated with autism is fragile X.
- Many syndromes manifested by cognitive disability are associated with autism.

AAP Mental Health Competency:

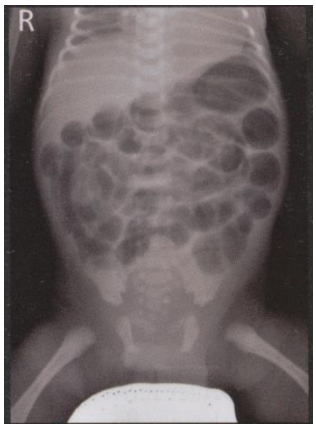
- Identify the key differential diagnoses to consider in the presence of an autistic spectrum disorder

Suggested Reading:

- American Academy of Pediatrics Committee on Genetics. Fragile X syndrome. In: Saul RA, ed. Medical Genetics in Pediatric Practice. Elk Grove Village, IL: American Academy of Pediatrics 2013:197-200
- Bacino CA, Brenda L. Fragile chromosome sites. In: Kliegman RM, Stanton BMD, St Geme J, Schor N, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Elsevier Saunders; 2011:411

Item 206

An infant develops abdominal distention and bilious vomiting at 36 hours after birth. The 3.9-kg infant was born at 37 weeks of gestation to a mother with type 1 diabetes mellitus. The prenatal course was unremarkable, with negative carrier testing for cystic fibrosis. According to the mother, the infant has been breastfeeding and has had 4 wet diapers and 1 small smear of meconium. Examination reveals an uncomfortable infant with a distended, firm abdomen that is slightly tender to deep palpation. The rectum appears to be externally patent with an anal wink. A radiograph is obtained (Item Q206).



ITEM Q206: Radiograph for the infant described in the vignette.

Of the following, the MOST likely cause of the infant's findings is

A. anorectal malformation
B. Hirschsprung disease
C. meconium ileus
D. neonatal small left colon syndrome
E. pneumatosis coli

Item 206 Preferred Response: D

An infant of a diabetic mother (IDM) who has evidence of an intestinal obstruction is likely to have neonatal small left colon syndrome (NSLCS). Neonatal small left colon syndrome presents with features of intestinal obstruction including abdominal distention, vomiting (bilious or non-bilious), and delayed passage of meconium. Contrast enema in NSLCS reveals a narrowed left colon with a transition zone at the splenic flexure similar to Hirschsprung disease. Rectal biopsy demonstrates normal bowel innervation with the presence of ganglion cells. The enema in NSLCS is curative, with bowel function returning without further surgical intervention. NSLCS may be found in up to 5% of IDMs and its cause remains unknown

Diabetes during pregnancy puts the exposed infant at increased risk of multiple transitory issues in the neonatal period, including hypoglycemia, hypocalcemia, polycythemia, hypertrophic cardiomyopathy, and NSLCS. Hypoglycemia may be seen in up to 27% of IDMs. In infants with macrosomia, the hypoglycemia is caused by hyperinsulinemia and will persist for 2 to 4 days. Infants of a diabetic mother that are premature or small for gestational age may have both decreased glycogen stores and hyperinsulinemia, exaggerating the degree and duration of the hypoglycemia. Hypocalcemia is seen in IDMs between 24 and 72 hours after birth and may be related to reduction of parathyroid hormone secretion. Screening is recommended only in symptomatic infants or those with additional risks including prematurity, respiratory distress syndrome, or asphyxia. Infants of diabetic mothers have an increased incidence of polycythemia that is attributed to increased erythropoietin secondary to chronic fetal hypoxemia. Infants of diabetic mothers may also have transient hypertrophic cardiomyopathy with thickening of the intraventricular septum and outflow obstruction. Some affected infants have poor cardiac output or heart failure that requires supportive care. The cardiomyopathy resolves within 6 to 12 months after birth.

The infant described in the vignette is an IDM, therefore the most likely cause of the clinical findings is NSLCS. Radiography suggests an intestinal obstruction and could also be seen with Hirschsprung disease or meconium ileus. It is estimated that more than 10 cases of NLSCS are seen for every case of Hirschsprung disease in IDMs. Meconium ileus is associated with cystic fibrosis, which is unlikely in the infant in the vignette because the mother is not a carrier. The passage of a meconium smear coupled with the normal external examination findings seen in the infant in the vignette make the diagnosis of anal atresia unlikely. Radiography does not demonstrate any pneumatosis intestinalis and does not support the diagnosis of pneumatosis coli.

PREP Pearls

- An infant of a diabetic mother is at risk for hypoglycemia, hypocalcemia, polycythemia, and neonatal small left colon syndrome.

American Board of Pediatrics Content Specification(s):

- Recognize that an infant of a diabetic mother is at risk for hypoglycemia, hypocalcemia, polycythemia, and neonatal small left colon syndrome.

Suggested Reading:

- Ellis H, Kumar R, Kostyrka B. Neonatal small left colon syndrome in the offspring of diabetic mothers: an analysis of 105 children. J Pediatr Surg. 2009;44:2343-2346. doi: 10.1016/j.jpedsurg.2009.07.054.
- Loening-Baucke V, Kimura K. Failure to pass meconium: diagnosing neonatal intestinal obstruction. Am Fam Physician. 1999;60:2043-2050
- Ogata ES. Problems of the infant of the diabetic mother. NeoReviews. 2010;11:e627-e631. doi: 10.1542/neo.11-11-e627
- Riskin A, Garcia-Prats JA. Infant of a diabetic mother. UptoDate. Available online only for subscription

Item 207

A 12-month-old boy who is visiting from Peru is seen in the emergency department with pallor. His mother states that he recovered 1 week ago from a febrile illness, for which he was treated with trimethoprim-sulfamethoxazole. His oral temperature is 36.2°C, pulse rate is 120 beats/min, respiratory rate is 24 breaths/min, and blood pressure is 85/55 mm Hg. On physical examination, he is jaundiced and has a grade 2/6 systolic ejection murmur. The remainder of the physical examination is normal. The following are the results of the child's laboratory tests:

- White blood cell count, 11,500/µL (11.5 x 10^9/L), with 30% polymorphonuclear leukocytes, 61% lymphocytes, 6% monocytes, and 3% eosinophils
- Hemoglobin, 5.2 g/dL (52 g/L)
- Mean corpuscular volume, 78/μm^3 (78 fL)
- Platelet count, 338 x 10^3/µL (338 x 10^9/L)
- Reticulocyte count, 8% (0.08)
- Conjugated bilirubin, 0.4 mg/dL (6.8 µmol/L)
- Unconjugated bilirubin, 3.5 mg/dL (59.9 µmol/L)
- Direct antiglobulin (Coombs) test, negative

Of the following, the test MOST likely to diagnose the cause of anemia in this child is a

A. bone marrow biopsy
B. glucose-6-phosphate dehydrogenase level
C. hemoglobin electrophoresis
D. liver biopsy
E. gene-mutation testing for Gilbert syndrome

Item 207 S Preferred Response: B

The child described in the vignette has a clinical presentation consistent with a hemolytic crisis secondary to glucose-6-phosphate dehydrogenase (G6PD) deficiency.
The main role of G6PD in the pentose-phosphate pathway is to produce the reduced form of nicotinamide adenine dinucleotide phosphate (NADPH), which has a crucial role in preventing oxidative damage in all cells. NADPH is particularly important in red blood cells, which are constantly exposed to oxygen radicals during methemoglobin production. The gene responsible for G6PD deficiency is located on the long arm of the X chromosome (band Xq28), which accounts for the X-linked recessive inheritance pattern of G6PD deficiency. G6PD deficiency has a high prevalence, occurring in more than 500 million people worldwide, many of whom are in, or descended from, the tropical and subtropical parts of the world with an increased exposure to malaria. The most common form of G6PD deficiency seen in the United States is the A-minus variant, an X-linked recessive disorder commonly seen in African American males. Because of the instability of the A-minus variant enzyme, its levels diminish more rapidly than normal as the erythrocyte ages and causes few chronic symptoms in affected individuals until they are exposed to oxidant drugs or have a serious infection. The form of G6PD deficiency that is seen predominantly in Mediterranean patients (G6PD Mediterranean) is characterized by an enzyme deficiency in erythrocytes of all ages, including reticulocytes, which leads to a more severe hemolysis in response to oxidant drugs.

Although children affected with G6PD deficiency may remain asymptomatic most of the time, on exposure to oxidative agents or fava beans or during the course of an infection, an acute hemolytic crisis can occur. Fava beans are popular in various ethnic cuisines, including that of Peru, Mexico, China, Pakistan, and the Mediterranean countries. Patients may present with jaundice, pallor, dark urine, and lethargy. The dark urine, due to hemoglobinuria, will show few or no red blood cells despite the initial screening urinalysis result being positive for heme. There may be a moderate to severe normocytic, normochromic anemia and a brisk reticulocytosis.

The boy described in the vignette has recently taken trimethoprim-sulfamethoxazole, a sulfa drug. Sulfa drugs are oxidative agents that can precipitate hemolytic crises in G6PD-deficient individuals. This antibiotic or the infection itself could have led to the hemolytic crisis in this patient.

A bone marrow biopsy would be useful if there was concern for decreased erythrocyte production or an infiltrative process (eg, leukemia) as the cause for the anemia; however, the elevated reticulocyte count and lack of other cytopenias, respectively, make these diagnoses less likely. A hemoglobin electrophoresis would help in the diagnosis of hemoglobin disorders, such as thalassemia or sickle cell anemia. Thalassemia, which produces a microcytic anemia, would be unlikely in this patient whose anemia is normocytic. Sickle cell anemia is a possibility, but given the ethnic background of this patient, G6PD deficiency would be more likely. A liver biopsy would be important in a patient with direct hyperbilirubinemia, which is more suggestive of a primary hepatic cause of jaundice. Isolated indirect hyperbilirubinemia with anemia suggests hemolysis. Uridine-diphosphate-glucuronosyltransferase isoform 1A1 (UGT1A1) is an enzyme

involved in converting bilirubin from its unconjugated (indirect) to its conjugated (direct) form. Gilbert syndrome is caused by a mutation in UGT1A1, leading to an enzyme decrease and consequently an elevated serum unconjugated bilirubin level. This is a benign condition that affects 5% to 10% of the population and is not associated with hemolysis or anemia.

PREP Pearls

- G6PD deficiency is an X-lined recessive disorder that often presents with anemia, pallor, jaundice, dark urine, and lethargy.
- Common causes for hemolytic crisis secondary to G6PD deficiency include infection, medications, and ingestion of fava beans.

American Board of Pediatrics Content Specification(s):

- Know that G6PD deficiency is a common X-linked disorder

Suggested Reading:

- Cappellini MD, Fiorelli G. Glucose-6-phosphate dehydrogenase deficiency. Lancet. 2008;371:64-74. doi: 10.1016/S0140-6736(08)60073-2. http://www.ncbi.nlm.nih.gov/pubmed/18177777
- Luzzato L, Poggi V. Glucose-6-phosphate dehydrogenase deficiency. In: Nathan and Oski's Hematology of Infancy and Childhood. 7th ed. Philadelphia, PA: Saunders Elsevier; 2009:883-907
- Schuurman M, van Waardenburg D, Da Costa J, Niemarkt H, Leroy P. Severe hemolysis and methemoglobinemia following fava beans ingestion in glucose-6-phosphatase dehydrogenase deficiency: case report and literature review. Eur J Pediatr. 2009;168:779-782. doi:10.1007/s00431-009-0952-x

Item 208

An 8-year-old girl has had frequent, severe headaches for the past 8 months. They are bifrontal and associated with nausea, photophobia, and blurry vision. They last 2 to 3 hours and occur 1 to 2 times per week, mostly at the end of a school day. She also has asthma and attention-deficit/hyperactivity disorder. Her father and paternal aunt have migraine headaches. She is typically a good student, but lately her grades have fallen due to absenteeism caused by the headaches. On physical examination, she is a thin, slightly nervous-appearing girl. Her funduscopic examination shows crisp optic disk margins, and extraocular movements are conjugate and intact in all directions. There is no nystagmus. The remainder of her physical examination findings are normal.

Of the following, the BEST prophylactic medication for her headaches is

A. cyproheptadine
B. ergotamine
C. fluoxetine
D. propranolol
E. topiramate

Item 208 Preferred Response: A

Of the choices listed, cyproheptadine is the best migraine prophylaxis medication for this girl. Potential side effects such as sedation and weight gain are dose dependent, and it is often possible, in a young child, to achieve an adequate dose for migraine prevention without side effects.

In addition to medications, a treatment plan for migraine prophylaxis should involve regular, restful sleep, exercise, and a healthy diet. A headache diary can help patients and families identify food triggers and other patterns that may provoke headaches.

Ergotamine is an acute treatment for migraine. It is not used for prophylaxis. Selective serotonin reuptake inhibitors like fluoxetine are also not used for migraine prophylaxis. Propranolol is a β-blocker used for migraine prophylaxis but should be avoided in a patient with asthma. Topiramate is an anticonvulsant that is also used for migraine prophylaxis, but can cause weight loss. It is not the first treatment choice for the girl in the vignette.

PREP Pearls

- Select a migraine prophylactic medication based on likelihood of efficacy, side effect profile, and other comorbid conditions.

American Board of Pediatrics Content Specification(s):

- Plan prophylactic treatment for recurrent migraine

Suggested Reading:

- Blume HK. Pediatric Headache: A Review. Pediatr Rev 2012; 33:562-576; doi:10.1542/pir.33-12-562
- Hershey A. Migrane. In: In: Kliegman RM, Stanton BMD, St Geme J, Schor N, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Elsevier Saunders; 2011: 2040-2045

Item 209

A 16-year-old girl presents to your office following an injury she sustained playing soccer. One week ago, another player hit her in the chest, causing her head to snap back. She had near-immediate onset of a diffuse frontal headache and mild nausea that lasted several hours after the injury. She did not lose consciousness. Her mother reports that the girl has seemed more forgetful and has been having trouble sleeping since the injury. She has not played soccer since the injury.

Of the following, you are MOST likely to tell the girl and her mother that this injury was

A. a concussion and she can return to soccer since it has been 1 week since the injury
B. a concussion and she can return to soccer since she no longer has headache
C. a concussion and she should continue to rest from all physical activities
D. not a concussion because she did not get hit in the head
E. not a concussion because she did not have loss of consciousness

Item 209 S Preferred Response: C

The girl described in the vignette sustained a concussion; she should not return to physical activities because she is still symptomatic. Among commonly played high school sports, the highest rates of sports concussion occur in American foot-ball, boys' ice hockey, boys' lacrosse, and girls' soccer. A concussion is a type of traumatic brain injury that occurs from a direct blow to the head or from a transmitted force causing linear or rotational acceleration. Individuals with concussion generally experience onset of symptoms within several hours following injury. Standard brain imaging, such as computed tomography and magnetic resonance imaging, yields normal results in concussed individuals. Symptoms can include somatic symptoms (eg, headache, nausea, or visual disturbance), cognitive symptoms (eg, difficulty concentrating or feeling confused), mood symptoms (eg, anxiety or irritability), and sleep disturbance. Over the past decade, the definition of concussion has evolved, with loss of consciousness no longer a criterion for establishing the diagnosis.

Children and adolescents who have concussion experience a 10- to 14-day average duration of symptoms. Younger individuals experience a more prolonged recovery. Treatment for concussion includes "brain rest:' such as deferring tests at school, avoiding "screen time" (ie, computers, television, texting, and video games), as well as complete rest from physical activity. The current consensus guidelines for the management of sports concussion stipulate that concussed individuals can begin a progression back to physical activity if neurologic examination results are normal and athletes are asymptomatic or at symptom baseline. Use of additional evaluation, such as computerized neuropsychological testing, may be used to determine whether return to sports is appropriate.

PREP Pearls

- Loss of consciousness is not required for the diagnosis of concussion.
- A concussion can occur with a transmitted force to the head.
- In individuals with concussion, standard brain imaging yields normal results.
- Generally, individuals with concussions can begin a progression back to physical activity if neurologic examination results are normal and athletes are asymptomatic or at symptom baseline.
- Treatment for concussion includes "brain rest" as well as complete rest from physical activity.

American Board of Pediatrics Content Specification(s):

- Know the sports most commonly associated with a head injury.

Suggested Reading:

- Guskiewicz KM, Valovich McLeod TC. Pediatric sports-related concussion. PM R. 2011;3(4):353-364. doi:10.1016/j.pmrj.2010.12.006
- Marar M, Mcllvain NM, Fields SK, Comstock RD. Epidemiology of concussions among United States high school athletes in 20 sports. Am Sports Med. 2012;40(4):747-755. doi:10.1177/0363546511435626
- McCrory P, Meeuwisse W, Johnston K, et al. Consensus Statement on Concussion in Sport: the 4th International Conference on Concussion in Sport held in Zurich, November 2012. Br I Sports Med. 2013;47:250-258. doi:10.1136/ bjsports-2013-092313

Item 210

A 15-year-old boy is followed in your practice and is seen for a routine health supervision visit and sports participation physical. He is planning to start as the center on his high school's junior varsity basketball team and denies any health problems. You note that he has a tall (>95th percentile for age) thin habitus with long arms and legs, arachnodactyly, flat feet, and a mild pectus excavatum. He wears contacts for significant myopia but has no other visual complaints. The family history is negative for anyone with similar physical features.

Of the following, you are MOST likely to recommend at this time

A. a chest radiograph to rule out apical blebs
B. an echocardiogram to rule out aortic root dilatation
C. magnetic resonance imaging of the brain to rule out cerebral aneurysm
D. a slit-lamp examination to rule out corneal thinning
E. spinal radiography to rule out dural ectasias

Item 210 S Preferred Response: B

The young man described in the vignette has clinical finding suggestive of a diagnosis of Marfan syndrome (MFS). However, in the absence of specific clinical findings such as aortic root dilatation or ocular lens subluxation, the diagnosis cannot be confirmed. Because aortic root dilatation is one of the cardinal features of MFS and could have serious clinical consequences if not identified and addressed, an echocardiogram is the best first test to obtain. In addition, mitral valve prolapse is seen in a large percentage of affected individuals, although it carries less weight in making the diagnosis because of its relative frequency in the general population.

The only cardinal feature in the ocular system is ectopia lentis (or lens subluxation) as seen on dilated eye examination, but high myopia, corneal flattening, iris hypoplasia, or ciliary muscle hypoplasia may also be identified on eye examination in affected individuals. Although spontaneous pneumothorax occurs in some individuals with MFS, presumably due to apical blebs in the lungs, the finding of blebs on chest radiography is no longer considered a diagnostic criterion. In 2010, diagnostic criteria for MFS were revised, eliminating the previous major and minor criteria. Instead, this new system relies on a combination of the cardinal features to classify patients: aortic root dilatation and ectopia lentis, other systemic clinical findings, results of molecular analysis of the fibrillin gene (FBN1), and the family history. Because there is overlap between the clinical features of MFS and other connective tissue disorders (Item 210A, page C-165), it is important to assess patients for features of these other conditions as well. The revised criteria as outlined (Item 210B, page C-165), help to better define subsets of patients and incorporate newer molecular technologies into practice. Points are assigned for systemic findings as well (Item C210C, page C-165), with elimination of some of the less helpful clinical features. The Z score, or degree of aortic root dilatation, that is used in this diagnostic paradigm is slightly different for children compared to adults, with a Z score greater than 2 for adults and greater than 3 for children. It is anticipated that application of these revised criteria will improve the accuracy of diagnosis and subsequently reduce the number of individuals who are given an incorrect diagnosis of MFS, which may be life-changing and stigmatizing.

Approximately 65% to 75% of individuals with MFS inherit the autosomal dominant gene mutation in FBNI from a parent, whereas 25% to 35% of the time the condition arises from a de novo mutation. Sequencing of the FBNI gene is being used more often for diagnostic clarification, but this testing is still quite expensive, and identifiable mutations are seen in only 70% to 93% of individuals with MFS. However, over time as the testing becomes less costly and the methodology improves, it is very likely that this testing will become standard of care in the diagnosis of MFS.

Routine radiographs looking for spinal dural ectasias or protrusio acetabuli (medial displacement of the femoral head) are not routinely recommended for individuals being evaluated for MFS, but if present can provide diagnostic support. Individuals with MFS are not at increased risk for cerebral aneurysms, so magnetic resonance imaging of the brain is not indicated. Corneal flattening but not thinning is seen in some affected individuals.

PREP Pearls

- Individuals suspected to have Marfan syndrome may benefit from an echocardiogram to rule out mitral valve prolapse and aortic root dilatation, as well as a slit-lamp and dilated eye examination assessing for lens subluxation (ectopia lentis), high myopia, and a few other minor anatomic changes.
- Sequencing of the fibrillin (FBNJ) gene associated with Marfan syndrome has some clinical diagnostic utility and may become standard of care in diagnosis of Marfan syndrome in the near future.

American Board of Pediatrics Content Specification(s):

- Know the clinical and laboratory findings of Marfan syndrome

Suggested Reading:

- Committee on Genetics. Health supervision for children with Marfan syndrome. Pediatrics. 1996;98(5):978-982
- Dietz HC. Marfan syndrome. In: Pagon RA, Bird TD, Dolan CR, eds. GeneReviews. Seattle, Washington: University of Washington; 2013
- Loeys BL, Dietz HC, Braverman AC, et al. The revised Ghent nosology for Marfan syndrome. J Med Genet. 2010;47(7):476-485. doi:10.1136/jmg.2009.072785

Item C210A. Discriminating features in the differential diagnosis of Marfan syndrome

Differential diagnosis	Gene	Discriminating features
Loeys-Dietz syndrome (LDS)	*TGFBR1/2*	Bifid uvula/cleft palate, arterial tortuosity, hypertelorism, diffuse aortic and arterial aneurysms, craniosynostosis, clubfoot, cervical spine instability, thin and velvety skin, easy bruising
Shprintzen-Goldberg syndrome (SGS)	*FBN1* and other	Craniosynostosis, mental retardation
Congenital contractural arachnodactyly (CCA)	*FBN2*	Crumpled ears, contractures
Weill-Marchesani syndrome (WMS)	*FBN1* and *ADAMTS10*	Microspherophakia, brachydactyly, joint stiffness
Ectopia lentis syndrome (ELS)	*FBN1* *LTBP2* *ADAMTSL4*	Lack of aortic root dilatation
Homocystinuria	*CBS*	Thrombosis, mental retardation
Familial thoracic aortic aneurysm syndrome (FTAA)	*TGFBR1/2, ACTA2*	Lack of Marfanoid skeletal features, levido reticularis, iris flocculi
FTAA with bicupid aortic valve (BAV)		
FTAA with patent ductus arteriosus (PDA)	*MYH11*	
Arterial tortuosity syndrome (ATS)	*SLC2A10*	Generalised arterial tortuosity, arterial stenosis, facial dysmorphism
Ehlers-Danlos syndromes (vascular, valvular, kyphoscoliotic type)	*COL3A1, COL1A2, PLOD1*	Middle sized artery aneurysm, severe valvular insufficiency, translucent skin, dystrophobic scars, facial characteristics

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Item C210B. Revised Ghent criteria for diagnosis of Marfan syndrome and related conditions

In the absence of family history:
(1) Ao (Z ≥2) AND EL=MFS*
(2) Ao (Z ≥2) AND *FBN1*=MFS
(3) Ao (Z ≥2) AND Syst (≥7pts)=MFS*
(4) EL AND *FBN1* with known Ao=MFS

EL with or without Syst AND with an *FBN1* not known with Ao or no *FBN1*=ELS
Ao (Z < 2) AND Syst (≥5 with at least one skeletal feature) without EL=MASS
MVP AND Ao (Z <2) AND Syst (<5) without EL=MVPS

In the presence of family history:
(5) EL AND FH of MFS (as defined above)=MFS
(6) Syst (≥7 pts) AND FH of MFS (as defined above)=MFS*
(7) Ao (Z≥2 above20 years old, ≥3 below 20 years) +FH of MFS (as defined above)=MFS*

*Caveat without discriminating features of SGS, LDS or vEDS (as defined in table 210A) AND after *TGFBR1/2*, collagen biochemistry, *COL3A1* testing if indicated. Other conditions/genes will emerge with time. Ao, aortic diameter at the sinuses of Valsalva above indicated Z-score or aortic root dissection; EL, ectopia lentis, ELS, ectopia lentis syndrome; *FBN1*, fibrillin-1 mutation; *FBN1* not known with Ao, *FBN1* mutation that has not previously been associated aortic root aneurysm/dissection; *FBN1* with known Ao, *FBN1* mutation that has been identified in an individual with aortic aneurysm, MASS, myopia, mitral valve prolapse, borderline (Z<2) aortic root dilatation, striae, skeletal findings phenotype; MFS, Marfan syndrome; MVPS, mitral valve prolapse syndrome, Syst, systematic score (see C210C); and Z, Z-score.

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Item C210C. Scoring of systemic features seen in Marfan syndrome

- Wrist AND thumb sign – 3 (wrist OR thumb sign – 1)
- Pectus carinatum deformity – 2 (pectus excavatum or chest asymmetry – 1)
- Hindfoot deformity – 2 (plain pes planus – 1)
- Pneumothorax – 2
- Dural ectasia – 2
- Protrusio acetabuli – 2
- Reduced US/LS AND increased arm/height AND no severe scoliosis – 1
- Scoliosis or thoracolumbar kyphosis – 1
- Reduced elbow extension – 1
- Facial features (3/5) – 1 (dolichocephaly, enophthalmos, downslanting palpebral fissures, malar hypoplasia, retrognathia)
- Skin striae – 1
- Myopia > 3 diopters – 1
- Mitral valve prolapse (all types) – 1

Maximum total: 20 points; score ≥7 indicates systemic involvement; US/LS, upper segment/lower segment ratio.

Reprinted with permission from Loeys BL, Dietz HC, Braverman AC, et al. The revised Ghent nosology for Marfan syndrome. *J Med Genet*. 2010;47(7):476-485

Item 211

You are seeing a 14-year-old boy in your office for his annual health supervision visit. Over the past 6 months, he has had episodes of epistaxis that are increasing in frequency and duration. Over the past month, the episodes have occurred 3 to 4 times per week. Some have lasted as long as 30 minutes. The boy tells you he has also had increasing difficulty breathing through the left side of his nose over the past 2 months. At today's visit, the boy's height and weight are found to be at the 50th percentile for age. On physical examination, he is in no acute distress. You find no abnormalities of the anterior aspect of his nose. On inspection of the nares using a nasal speculum, you note a mass in the left posterior nasal passage.

Of the following, the diagnostic study that is MOST useful in identifying the underlying cause of the boy's epistaxis is

A. coagulation studies
B. complete blood cell count with differential
C. computed tomography of the sinuses
D. serum immunoglobulin E
E. sweat chloride test

Item 211 Preferred Response: C

The symptoms and examination findings for the male teenager in the vignette highly suggest juvenile nasopharyngeal angiofibroma (JNA) as the etiology of his epistaxis. Computed tomography of the sinuses would be the most useful diagnostic study for this patient.

Juvenile nasopharyngeal angiofibroma is a benign tumor and a rare but important cause of epistaxis in prepubertal and adolescent males, accounting for 0.05% of all head and neck tumors. It occurs almost exclusively in males in the first or second decade of life, with a mean age of onset of 15 years, and must be considered in boys with recurrent epistaxis with associated nasal obstruction. A hormonally mediated origin has been suggested for development of JNA.

Juvenile nasopharyngeal angiofibroma lesions originate close to the posterior attachment of the middle turbinate, near the superior border of the sphenopalatine foramen. These lesions derive their blood supply from the internal maxillary artery. Although a histologically benign tumor, JNA is highly vascular and can result in severe epistaxis. Furthermore, the tumor can cause severe problems through local invasion of adjacent structures.

The most common presenting symptoms found in boys with JNA are nasal obstruction, recurrent unilateral epistaxis, nasal drainage, headache, and facial swelling. The most common signs identified on physical examination include unilateral nasal mass, orbital mass, and proptosis.

When suspected, JNA can be confirmed by contrast-enhanced computed tomography or magnetic resonance imaging. These studies are indicated in children with epistaxis if a nasopharyngeal mass is visualized or suspected; in addition to confirming JNA, these diagnostic studies can identify and delineate other types of neoplasms of the nasal cavity, including rhabdomyosarcomas and nasopharyngeal carcinomas.

The differential diagnosis for epistaxis in children includes causes ranging from self-limited mucosal irritation to life-threatening neoplasms. The most common causes of epistaxis in children include mucosal dryness, local trauma, nasal foreign body, and rhinitis (allergic or infectious). Less common but extremely important underlying causes of epistaxis include bleeding disorders, medications, illicit drugs use, neoplasms, inflammatory disorders, and hypertension. Children with systemic causes of epistaxis rarely present with epistaxis as the only clinical manifestation.

Although obtaining coagulation studies and a complete blood cell count with differential would not be unreasonable in this adolescent with recurrent epistaxis, these studies would not identify the underlying problem for this boy. The finding of a unilateral nasal mass and the patient's report of progressive nasal obstruction should raise clinical suspicion for JNA. The history of unilateral, rather than bilateral, epistaxis further indicates an isolated nasopharyngeal lesion rather than coagulopathy, thrombocytopenia, or other systemic disease. Nasal cavity neoplasms in children may include benign lesions (such as JNA,

hemangioma, pyogenic granuloma, and inverting papilloma) and rare but extremely important malignant tumors, such as rhabdomyosarcomas and nasopharyngeal carcinomas.

Testing for elevated serum immunoglobulin E level, which can be an indicator of allergy-related disease, is typically not included in the initial evaluation of a child with recurrent or severe nosebleeds.

A sweat chloride test to confirm the diagnosis of cystic fibrosis would not be indicated in this patient. The boy's normal height and weight and absence of significant prior illnesses are inconsistent with the diagnosis of underlying cystic fibrosis.

PREP Pearls

- The diagnosis of JNA should be considered in prepubescent and adolescent males with recurrent unilateral epistaxis and nasal obstruction.
- Systemic conditions that may result in epistaxis include bleeding disorders, medication effects, hypertension, infectious and inflammatory disorders, and malignant tumors.
- Radiologic imagings are indicated in children presenting with epistaxis and a nasopharyngeal mass lesion.

American Board of Pediatrics Content Specification(s):

- Understand the evaluation of a child with severe recurrent epistaxis
- Know the differential diagnosis of epistaxis

Suggested Reading:

- Messner AH. Epidemiology and etiology of epistaxis in children. UPtoDate. Available online only for subscription.
- Messner AH. Evaluation of epistaxis in children. UPtoDate. Available online only for subscription.

Item 212

An 18-month-old boy is seen for his health supervision visit. Although he is clinically well today, his mother reports that he was treated with amoxicillin 2 months ago for acute otitis media. He had 1 previous episode of otitis media at 12 months of age. His growth and development are normal. He is up to date on his immunizations. On physical examination, you note that both tympanic membranes are cloudy and grayish white and have limited mobility on insufflation.

Of the following, your BEST recommendation today is

A. amoxicillin–clavulanate
B. ceftriaxone
C. reassessment in 4 to 6 weeks
D. referral to otolaryngology
E. repeat the pneumococcal vaccine

Item 212 C Preferred Response: C

The boy described in the vignette exhibits the characteristic physical findings of persistent middle ear effusion after acute otitis media (AOM). On physical examination the normal tympanic membrane should be translucent gray with important landmarks visible (Item C212) plus the tympanic membrane should be in its natural position and mobile. The diagnosis of AOM or otitis media with effusion (OME) is established by documenting the presence of fluid behind the tympanic membrane. The presence of inflammation differentiates AOM from OME. With OME, the tympanic membrane may appear opaque or cloudy, an air-fluid level or air bubbles may be evident on examination, and mobility on pneumatoscopy is limited. Pneumatic otoscopy remains the preferred method to diagnose middle ear effusion (MEE). Tympanometry or acoustic reflectometry are diagnostic adjuncts that may be used.

Otitis media with effusion or persistent MEE after an episode of AOM is common and resolves within 3 months in more than 75% of cases. The American Academy of Pediatrics policy on OME recommends watchful waiting for 3 months after diagnosis. Therefore, the correct response is reassurance and reassessment 3 months after the initial diagnosis of AOM.

In this case, there are no signs or symptoms of acute infection, so treating with amoxicillin—clavulanate or ceftriaxone is not indicated. Most AOM treatment failures manifest within 48 to 72 hours after initiation of therapy, and in these cases changing the antibiotic coverage would be appropriate.

Patients may experience conductive hearing loss that is temporary and due to the effusion, so physicians should make parents aware that ongoing developmental surveillance is important. Medical therapies for management of persistent MEE, such as the use of antihistamines, decongestants, or intranasal corticosteroids, have not proven effective. When OME persists beyond 3 months or if language delay is a concern, hearing should be evaluated. Children who are found to have normal hearing despite persistent MEE should be reevaluated every 3 months. Those who have hearing loss, other symptoms attributable to the effusion (otalgia or vestibular disturbance), or abnormal findings on physical examination beyond simple effusion should be referred to an otolaryngologist.

Although the introduction of the pneumococcal vaccine has decreased the prevalence of AOM due to Streptococcus pneumoniae, there is no indication for repeating the vaccine in a fully immunized normal host.

PREP Pearls

- Presence of inflammation, such as erythema and bulging of the tympanic membrane, helps to differentiate AOM from MEE.
- MEE may persist for 2 to 3 months or longer after AOM, so watchful waiting is recommended during this time in the asymptomatic child. Antibiotics do not need to be given.
- Reassessment for resolution of MEE is best performed 3 months after the initial diagnosis of AOM in the asymptomatic child.

American Board of Pediatrics Content Specification(s):

- Know that effusion may persist for 2 to 3 months or longer following acute otitis media

Suggested Reading:

- American Academy of Pediatrics Clinical Practice Guideline: The diagnosis and management of acute otitis media. Pediatrics. 2013;131:e964-e999. doi:10.1542/peds.2012-3488
- Daly KA, Hunter LL, Giebink GS. Chronic otitis media with effusion. Pediatr Rev. 1999;20:85-94. doi: 10.1542/pir.20-3-85
- Gould JM, Matz PS. Otitis media. Pediatr Rev. 2010;31:102-116. DOI: 10.1542/pir.31-3-102
- Kerschner JE. Otitis media. In: Kliegman RM, Stanton BMD, St Geme J, Schor N, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Elsevier Saunders; 2011:2199-2213

Item 213

You are seeing a 3-year-old girl after hospital discharge. The girl was seen in the emergency department yesterday after she developed an erythematous, itchy, raised, blotchy rash, marked swelling of the eyes and lips, and raspy breathing. According to the mother, the child was given injectable epinephrine, diphenhydramine, and systemic corticosteroids. Although the symptoms improved after a couple of hours, the child was admitted overnight for observation. The girl's mother is shaken by this event and would like you to determine what triggered this reaction.

Of the following, the MOST common triggers for events such as those experienced by the girl are

A. aeroallergens, contact allergens, latex, cleaning agents
B. foods, medications, hereditary angioedema, idiopathic triggers
C. infections (viral or bacterial), foods, medications, insect stings
D. mast cell disorders, hereditary angioedema, urticarial vasculitis
E. nonsteroidal anti-inflammatory drugs, antibiotics, narcotics, and radiocontrast media

Item 213 S Preferred Response: C

The child described in this vignette had an episode of acute urticaria/angioedema. The most common causes of acute urticaria are infections (viral or bacterial), foods, medications, and insect sting reactions. The child had symptoms and signs of urticaria (erythematous, itchy, raised, blotchy rash) as well as angioedema (marked swelling of the eyes and lips). Her raspy breathing could have been due to pharyngolaryngeal angioedema or a sign of bronchospasm suggesting impending anaphylaxis. The epinephrine followed by diphenhydramine and corticosteroids treats both conditions simultaneously. The absence of cardiovascular (eg, hypotension) and respiratory (wheezing) decompensation and additional systemic signs helps provide reassurance against the diagnosis of anaphylaxis.

Although no specific cause can be identified in many patients, acute urticaria is more likely to have an identifiable etiology then chronic urticaria (>6 weeks' duration). Viral or bacterial infections account for greater than 80% of cases, particularly in children. IgE-mediated, allergic reactions causing acute urticaria can be triggered by medications, stinging insects, foods and food additives, aeroallergens, contact allergens, latex, or blood products. Allergic reactions may be limited to the skin or be a part of a systemic allergic reaction (ie, anaphylaxis). Generalized urticaria or angioedema following exposure to a potential allergen should be interpreted as a systemic reaction with an increased risk of anaphylaxis with subsequent exposure. The antibiotics most frequently implicated in causing IgE-mediated urticaria include beta-lactams (penicillins and cephalosporins), although antibiotics from virtually all classes have been reported. Certain drugs cause urticaria owing to mast cell degranulation through a nonIgE-mediated mechanism. The most frequently implicated are narcotics, muscle relaxants, vancomycin, and radiocontrast medium. Nonsteroidal anti-inflammatory drugs, such as aspirin, ibuprofen, naproxen sodium, and others can trigger urticaria and/or angioedema owing to both allergic and nonallergic mechanisms.

Across all age groups, the most common triggers for anaphylaxis are ingested foods (33%), insect stings (19%), and medications (14%). Less common triggers include cats, latex, cleaning agents, environmental allergens, and exercise. For approximately one-quarter of cases, the trigger is unknown. In children, food-induced anaphylaxis is the most common trigger and accounts for 37% to 85% of cases, whereas insect bites/stings account for 5% to 13% and medications account for 5% to 12%.

Acute angioedema that occurs in association with urticaria is similar in pathology to urticaria, although it takes place in the deeper levels of the dermis and subcutaneous tissues. Angioedema that is mast cell-mediated is associated with urticaria and/or pruritus in most cases. Urticarial vasculitis, on the other hand, should be suspected when the urticarial lesions are painful, last for more than 72 hours at a location, are purplish or discolored, and cause scarring. Hereditary angioedema should be considered when angioedema occurs in the absence of urticaria; occurs at sites of trauma or around the mouth and extremities; is preceded by the appearance of a transient, serpiginous rash; occurs with abdominal cramping suggesting bowel edema; requires prolonged treatment; or occurs with a family history of similar symptoms. The presence of urticaria

pigmentosa, solitary reddish-brown lesions that urticate on stroking (Darier sign), or of persistently elevated tryptase should raise concern regarding underlying mast cell disorders.

PREP Pearls

- Viral or bacterial infections are commonly associated with acute urticaria in children.
- Acute urticaria can be triggered by medications, stinging insects, foods and food additives, aeroallergens, contact allergens, latex, or blood products.
- In children, anaphylaxis is most commonly triggered by foods, insect stings, and medications.

American Board of Pediatrics Content Specification(s):

- Know the etiologic agents that commonly cause urticaria/angioedema/ anaphylaxis

Suggested Reading:

- Lieberman P, Nicklas RA, Oppenheimer J, et al. The diagnosis and management of anaphylaxis practice parameter: 2010 update [published correction appears in JAllergy Clin Immunol. 2010;126(6):1104]. JAllergy Clin Immunol. 2010;126(3):477-480.e1-42. doi:10.1016/j.jaci.2010.06.022

Item 214

A 4-year-old girl presents to your office for evaluation 1 month after an episode of pyelonephritis, after which she was diagnosed with grade III vesicoureteral reflux. The patient is healthy with normal growth parameters and development. She has no significant past medical history or past surgical history. According to her parents, she has been toilet trained since 18 months of age. Findings on physical examination are unremarkable; vital signs are normal. Her urine analysis in the office shows specific gravity of 1.010, pH of 6.0, and no protein, blood, leukocyte esterase, or nitrites. There is no history of urinary tract infections in the parents or the 2-year-old sister.

Of the following, the MOST appropriate next step in the management of this patient is to

A. evaluate the patient for voiding dysfunction
B. order urine culture for evaluating resolution of the urinary tract infection
C. order serum electrolytes and serum creatinine for evaluating renal function
D. refer the patient for surgical correction of her reflux
E. screen the 2-year-old sibling with voiding cystourethrography

Item 214 Preferred Response: A

Vesicoureteral reflux (VUR) is the retrograde passage of urine from the bladder to the kidneys. Normally reflux of urine is prevented by compression of the intravesical ureter by the contracting bladder muscles. A shorter intravesical ureter (which may be genetically linked) has been implicated in the failure of the antireflux mechanism, thus leading to primary VUR. Primary VUR may resolve spontaneously with patient growth.

Secondary VUR occurs because of abnormally high pressures in the bladder leading to incompetence of the ureterovesical junction and associated reflux. The high pressure could be the result of anatomic obstruction as seen in posterior urethral valve (in boys only) or bladder contraction against a closed urethral sphincter as seen in dysfunctional voiding. Dysfunctional voiding may be associated with neurogenic (cerebral palsy, myelomeningocele) or non-neurogenic causes. Bladder or bowel dysfunction leading to dysfunctional elimination has been associated with recurrent infections, increased time for spontaneous resolution of reflux, and reduced success of endoscopic surgery. Evaluation for voiding dysfunction include a history of constipation, urgency, urge incontinence, infrequent voiding, or recurrent infections. Appropriate management of voiding dysfunction as recommended for this patient may help in spontaneous resolution of reflux and decrease the risk for urinary tract infection (UTI), thereby improving overall patient prognosis. Treatment for voiding dysfunction focuses on prevention and treatment of constipation, behavioral modification, such as timed and frequent voiding prior to urgency symptoms and positive reinforcement for normal voiding habits. Use of a step stool helps children sit comfortably on the toilet seat and promotes normal and comfortable voiding habits.

Testing serum chemistry or repeating urine culture on follow-up is not indicated for an asymptomatic patient with a history of appropriately treated UTI.

In more than 50% of patients, the reflux resolves spontaneously. Lower grade of reflux, unilateral reflux, prenatal hydronephrosis, and diagnosis before age 1 year have been favorably associated with spontaneous resolution of VUR. Spontaneous resolution has been reported in up to 60% to 80% of the patients with unilateral grade Ito grade IV VUR. Although patients with grade Ito grade II VUR continue to have high rates of bilateral reflux resolution, only 10% to 20% of patients with grade III to grade IV bilateral reflux experience spontaneous resolution. Grade V reflux rarely resolves spontaneously, therefore these patients usually need surgical intervention.

Studies have reported that nearly 30% of the siblings of patients with VUR and 35% of children of parents with VUR have VUR. Despite this genetic predisposition, current evidence does not support routine screening with voiding cystourethrogram (VCUG) for asymptomatic siblings of patients with reflux. According to the American Urological Association, siblings of patients with reflux may be screened with renal ultrasonography. A VCUG is indicated in siblings in the presence of abnormal renal ultrasound findings or history of febrile UTI.

PREP Pearls

- More than 50% of patients experience spontaneous resolution of vesicoureteral reflux (VUR).
- Lower grade of reflux, unilateral reflux, prenatal hydronephrosis, and diagnosis before age 1 year have been favorably associated with spontaneous resolution of VUR.
- Current evidence does not support routine screening with voiding cystourethrogram for asymptomatic siblings of patients with reflux.
- Bladder bowel dysfunction/dysfunctional elimination has been associated with recurrent infections, increased time for spontaneous resolution of reflux, and reduced success of endoscopic surgery
- Appropriate management of voiding dysfunction may help in spontaneous resolution of reflux and decrease risk for urinary tract infection.

American Board of Pediatrics Content Specification(s):

- Know the natural history (eg, etiology, familial association, outcome) of vesicoureteral reflux

Suggested Reading:

- American Academic of Pediatrics, Subcommittee on Urinary Tract Infection, Steering Committee on Quality Improvement and Management. Urinary tract infection: clinical practice guideline for the diagnosis and management of the initial UTI in febrile infants and children 2 to 24 months. Pediatrics. 2011;128(3);595-610. doi:10.1542/peds.2011-1330
- Elder JS. Vesicouretral reflux. In: Kliegman RM, Stanton BMD, St Geme Schor N, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Elsevier Saunders; 2011:1834-1838
- Estrada CR Jr, Passerotti CC, Graham DA, et al. Nomograms for predicting annual resolution rate of primary vesicoureteral reflux: results from 2,462 children [published online ahead of print August 15].1 Urol. 2009;182(4):1535-1541. doi:10.1016/j.juro.2009.06.053
- Feld LG, Mattoo TK. Urinary tract infections and vesicoureteral reflux in infants and children. Pediatr Rev. 2010;31(11):451-463. doi:10.1542/pir. 31-11-451

Item 215

A 7-year-old girl who has moderate asthma presents in December with a 2-day history of fever, chills, diffuse myalgias, and a nonproductive cough. Today she developed nasal congestion and rhinorrhea. On physical examination, her temperature is 38.5°C, heart rate is 120 beats/min, and respiratory rate is 25 breaths/min; her blood pressure is normal for age. The girl appears ill but not toxic. She has clear nasal secretions and mild oropharyngeal hyperemia without discharge. Auscultation of the lungs reveals good air entry bilaterally without wheezing, rhonchi, or rales. The remainder of her physical examination findings are normal. A rapid diagnostic test result for influenza is positive.

Of the following, the BEST agent to prescribe for the treatment of this patient is

A. albuterol
B. oseltamivir
C. rimantadine
D. saline drops
E. zanamivir

Item 215 Preferred Response: B

The patient described in the vignette presents within the first 48 hours of illness due to influenza virus and should be treated with oseltamivir (approved for patients 6 months and older). Oseltamivir and zanamivir are neuraminidase inhibitors with activity against both influenza A and B viruses. However, zanamivir is not recommended for use in patients with underlying respiratory disease, such as asthma. Antiviral resistance to oseltamivir and zanamivir is low. Because resistance to amantadine and rimantadine (ie, adamantanes) among influenza A viruses has increased rapidly worldwide, these agents are not recommended for influenza treatment or prophylaxis. Albuterol is appropriate for treating wheezing due to an asthma exacerbation, but the patient described in the vignette has reassuring pulmonary examination findings without wheezing. Nasal saline drops are helpful for symptomatic relief of illness due to upper respiratory tract infection but would not be the most appropriate therapy for this patient who is at high risk for influenza complications.

Early antiviral treatment decreases the duration of influenza illness symptoms (including fever) and may reduce influenza-related complications (eg, pneumonia and otitis media) and death and shorten the length of hospitalization. Antiviral treatment as soon as possible, but ideally within 48 hours of the onset of illness, is recommended for known or suspected influenza illness that is severe or leads to hospitalization or for any individuals at high risk for influenza-related complications. Persons at high risk for influenza-related complications include the very young (<2 years), the elderly (>64 years), those with chronic conditions (eg, cardiovascular, hematologic, hepatic, metabolic, neurologic or neuromuscular, pulmonary, or renal conditions), those in chronic care facilities, those with immunosuppression, morbidly obese patients, pregnant women, American Indians and Alaskan natives, and persons younger than 19 years undergoing long-term aspirin therapy.

PREP Pearls

- Oseltamivir and zanamivir are recommended for the treatment of influenza A and B viruses; zanamivir should not be used in patients who have underlying respiratory conditions.
- Because resistance to amantadine and rimantadine among influenza A viruses has increased rapidly worldwide, these agents are not recommended for influenza treatment or prophylaxis.
- Antiviral treatment as soon as possible, but ideally within 48 hours of the onset of illness, is recommended for known or suspected influenza illness that is severe or leads to hospitalization or for any individuals at high risk for influenza-related complications.

American Board of Pediatrics Content Specification(s):

- Know the indications for the use of antiviral medications for the treatment of influenza, while recognizing that some strains of the virus are resistant to antiviral medications

Suggested Reading:

- American Academy of Pediatrics. Influenza. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:439-453
- Centers for Disease Control and Prevention. Seasonal influenza (flu): Antiviral drugs. www.cdc.gov/flu

Item 216

A 14-year-old girl is being evaluated for vaginal bleeding. Her menarche was about 18 months ago, and her periods have occurred at 5- to 6-week intervals; the flow is variable and she has had no dysmenorrhea. The current menstrual period has been heavier and she had cramps at the start. She remembers passing a few clots about 2 cm in size. She denies sexual activity. On physical examination, she is not pale, her heart rate is 68 beats/min, and her body mass index is 24. She is at sexual maturity rating 5 for pubertal development. She has vague tenderness diffusely in her lower abdomen with no guarding or rebound. A rapid pregnancy test result is negative, and her hemoglobin level is 12.6 g/dL (126 g/L).

Of the following, the MOST likely explanation for the girl's symptoms is

A. a bleeding disorder
B. dysfunctional uterine bleeding
C. ectopic pregnancy
D. hypothyroidism
E. pelvic inflammatory disease

Item 216 Preferred Response: B

Painless, irregular, or prolonged bleeding of endometrial origin without any accompanying structural disease is referred to as dysfunctional uterine bleeding (DUB) and is the result of physiologic anovulation seen in early puberty with immaturity of the hypothalamic-pituitary axis. It is important to know the normal parameters for menstrual bleeding to exclude other causes of bleeding when the pattern is abnormal. The girl in the vignette is within 2 years of menarche, when anovulatory bleeding is common. Her menstrual interval is at the upper limit of normal, and the fact that she has had no prior pain is suggestive of DUB. On average, ovulatory cycles start at 20 months after menarche, and her history of new onset of cramping suggests that her periods are now becoming ovulatory.

The origin of abnormal bleeding can be divided into local and systemic causes. Local causes of bleeding may involve the vulva, vagina, cervix, uterus, or ovaries and may be the result of trauma, foreign bodies, infections, pregnancy-related problems, and rarely, estrogen-producing ovarian tumors. The girl in the vignette denies sexual activity, and her pregnancy test result is negative. This would make pelvic inflammatory disease and ectopic pregnancy very unlikely. Common systemic issues include bleeding disorders and thyroid disease. The lack of other bleeding issues and a normal hemoglobin level make the presence of a bleeding disorder unlikely. Hypothyroidism may be associated with irregular or prolonged bleeding, whereas hyperthyroidism is more likely to be associated with amenorrhea. Chronic lymphocytic thyroiditis is the most common cause of hypothyroidism in adolescents and usually presents with an asymptomatic goiter. Growth issues and exacerbation of obesity during the adolescent years may also occur. This girl's history and physical examination findings are not suggestive of hypothyroidism.

PREP Pearls

- Irregular, painless menstrual bleeding in the first 2 years after menarche is most likely the result of physiologic anovulation.
- Pregnancy and sexually transmitted infections need to be considered in adolescents with vaginal bleeding.
- Heavy vaginal bleeding with anemia suggests a bleeding disorder, such as von Willebrand disease.

American Board of Pediatrics Content Specification(s):

- Know that a working differential diagnosis for dysfunctional uterine bleeding includes threatened abortion, tubal pregnancy, pelvic inflammatory disease, endocrinopathies such as hyperthyroidism, and coagulopathies

Suggested Reading:

- Bordini B, Rosenfield RL. Normal pubertal development: part II: clinical aspects of puberty. Pediatr Rev. 2011;32:281-292. doi:10.1542/pir.32-7-281
- Emans SJ. Dysfunctional uterine bleeding. In: Emans SJ, Laufer MR, Goldstein DP, eds. Pediatric and Adolescent Gynecology. Philadelphia, PA: Lippincott Williams & Wilkins; 2005:270-286
- Gray SH, Emans SJ. Abnormal vaginal bleeding in adolescents. Pediatr Rev. 2007;28:175-182. doi:10.1542/pir.28-5-175
- Mitan LAP, Slap GB. Dysfunctional uterine bleeding. In: Neinstein LS, Gordon CM, Katzman DK, Rosen DS, Woods ER, eds. Adolescent Health Care: A Practical Guide. 5th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2008:687-690

Item 217

A 17-year-old girl is seen for her initial health supervision visit with you. She recently moved into the area from another state, where she was being treated with levothyroxine because of a thyroid disorder. Although her thyroid hormone levels have been under good control, she has recently become increasingly tired during the day. She is on her high school soccer team but has had difficulty finishing practices because of fatigue. She denies jaundice, vomiting, abdominal pain, easy bruisability, sleep disturbance, or any musculoskeletal symptoms. Physical examination demonstrates a well-developed, pleasant, and cooperative young woman. The remainder of the physical examination findings are unremarkable. Screening laboratory tests demonstrate the following:

- Hemoglobin, 12.1 g/dL (121 g/L)
- White blood cell count, 8,000/µL (8×10^9/L)
- Aspartate aminotransferase, 125 U/L; reference range, <_40 U/L
- Alanine aminotransferase, 180 U/L; reference range, <30 U/L
- Total thyroxine (T4), 7.0 µg/mL (120 nmol/L); reference range, 4.5 to 12.5 µg/mL (77-214 nmol/L)

Urinalysis results are normal.

Of the following, the laboratory test that is MOST likely to lead to this patient's diagnosis is

A. α1-antitrypsin
B. anti-smooth muscle antibody
C. ceruloplasmin
D. free thyroxine (T4) and triiodothyronine (T3)
E. thyroid-stimulating hormone

Item 217 Preferred Response: B

Autoimmune hepatitis (AIH) is a chronic disorder of unknown origin characterized by hepatocellular inflammation, leading to cell necrosis, fibrosis with portal tract to portal tract bridging, and, ultimately, progression to cirrhosis. Patients may present with a picture of acute hepatitis, characterized by jaundice and a tender enlarged liver, or with coagulopathy, reflecting fulminant disease. Accordingly, any patient with acute liver failure should be evaluated for possible AIH. In many patients, the mode of presentation reflects chronic liver involvement, with only vague, nonspecific symptoms (eg, fatigue, headache, myalgias, exercise intolerance, and weight loss). However, AIH may be diagnosed in asymptomatic patients found to have elevated levels of aspartate aminotransferase (AST) and alanine aminotransferase (ALT) on routine screening tests. Importantly, AIH is frequently associated with other autoimmune conditions. In one report, 111 of 278 adult patients with AIH had additional autoimmune diseases. Excluding overlap syndromes (primary biliary cirrhosis and primary sclerosing cholangitis), autoimmune thyroiditis was the most common concurrent disease, occurring in 28 patients (10%). The girl described in the vignette presents with a history of thyroid disease and laboratory study results that indicate hepatocellular inflammation. A diagnosis of AIH must be strongly considered. Virtually all pediatric cases of AIH demonstrate immune serum markers, including autoantibodies against liver-specific and non-liver-specific antigens, as well as increased IgG levels. In the evaluation of suspected AIH, laboratory studies should assess these autoantibodies, including antinuclear antibodies (ANAs), anti-liver-kidney microsomal 1 (anti-LKM-1) antibodies, and anti-smooth muscle antibodies (ASMAs).

In pediatric patients, 2 types of AIH have been described. Type 1 (two-thirds of cases) is characterized by the presence of ASMAs or ANAs. Type 2 is identified by the presence of anti-LKM-1 antibodies. Both types are seen predominantly in girls (up to 80%). Anti-LKM-1-positive patients tend to be younger and are more likely to present with acute liver failure. Both adult and childhood AIH are frequently associated with other autoimmune disorders (20%) and a family history of autoimmune disease (40%).

The clinical presentation of AIH is highly variable. In 40%, signs and symptoms suggest acute viral hepatitis, marked by nonspecific symptoms, followed by liver-specific signs, including jaundice, dark urine, and light-colored stools. In this AIH subgroup, patients positive for anti-LKM-1 anti-bodies are at particular risk because some will develop acute hepatic failure 2 to 8 weeks from the onset of symptoms. Up to 40% of patients present with nonspecific symptoms that may persist for months to years before diagnosis. In approximately 10% of patients, complications of portal hypertension, including splenomegaly and bleeding from esophageal varices, may be the first signs of illness. Considering this wide spectrum of disease, associated with a variable clinical course, AIH should be considered in all children presenting with symptoms and clinical and/or laboratory signs of prolonged or severe liver disease. In addition to the laboratory studies described previously, the evaluation should include a careful physical examination for any stigmata of underlying chronic liver disease, including cutaneous abnormalities (eg, spider nevi, palmar erythema, and striae), firm liver, and splenomegaly. These signs should also be assessed in patients with acute hepatitis.

In any child suspected of having AIH, other chronic liver disorders must be considered. Although autoantibodies are important AIH diagnostic markers, up to 50% of children with hepatitis B and C are positive for ANAs or ASMAs, and 5% of patients with chronic hepatitis C have anti-LKM-1 antibodies. In these viral liver diseases, however, the auto-antibody titers are typically much lower than found in AIH, and identification of the appropriate viral markers will allow for a correct diagnosis.

Wilson disease is an important consideration in children who have chronic liver disease. It can be difficult to differentiate Wilson disease from AIH. Patients may have autoantibodies and elevated IgG levels. More importantly, the liver histopathologic findings in Wilson disease may be strikingly similar to that of AIH. Therefore, a serum ceruloplasmin, ophthalmologic examination for Kayser-Fleischer rings, and urine and (if necessary) liver copper studies should be performed in all cases. However, by 17 years of age, the age of the girl in the vignette, Wilson disease commonly produces neuropsychiatric findings. Furthermore, in a clinical setting that strongly suggests autoimmune thyroid disease, a diagnosis of AIH is far more likely.

Other causes of chronic liver disease may be considered but are much less common. Liver involvement associated with α1 -antitrypsin (A1AT) deficiency, identified by the ZZ A1AT phenotype, is characterized by neonatal cholestasis in 10% of affected individuals. Progression to cirrhosis may occur, and A1AT assessment is part of the evaluation of chronic liver disease. However, the clinical findings for the girl in the vignette are most consistent with AIH. Finally, evaluation of thyroid function should be part of any evaluation in patients with a history of thyroid disease. However, these studies would not assist in diagnosing this patient's liver disorder.

PREP Pearls

- Autoimmune hepatitis (AIH), is frequently associated with other autoimmune disorders, especially thyroiditis.
- Virtually all cases of AIH in children and adolescents are associated with the presence of autoimmune markers.
- Any child with unexplained, persistent elevations in AST and ALT should be screened for AIH.

American Board of Pediatrics Content Specification(s):

- Recognize the signs and symptoms of chronic hepatitis

Suggested Reading:

- Gregorio GV, Portmann B, Reid F, et al. Autoimmune hepatitis in childhood: a 20-year experience. Hepatology. 1997;25:541-547. doi:10.1002/hep.510250308.
- Mieli-Vergani G, Vergani D. Autoimmune hepatitis in children: what is different from adult AIH? Semin Liver Dis. 2009;29:297-306. doi:10.1055/s-0029-1233529
- Mieli-Vergani G, Vergani D. Autoimmune paediatric liver disease. World Gastroenterol. 2008;14:3360-3367. doi:10.3748/wjg.14.3360
- Strassburg CP, Manns MP. Treatment of autoimmune hepatitis. Semin Liver Dis. 2009;29:273-285. doi:10.1055/s-0029-1233534
- Teufel A, Weinman A, Kahaly GJ, et al. Concurrent autoimmune diseases in patients with autoimmune hepatitis. I Clin Gastroenterol. 2010;44:208213. doi:10.1097/MCG.0b013e3181c74e0d

Item 218

During the health supervision visit for a 5-year-old girl, the mother raises concerns about her daughter's aggressive behavior toward her and the child's frequent headaches, for which she is often sent home from school. The headaches do not occur on weekends or during school vacations. While the teachers feel she is a bright child, they report she is not achieving well at school. The mother is very concerned that there is a serious neurologic condition causing these symptoms.
Reviewing the child's medical record, you see that she has been in your office many times over the past year for minor illnesses. You also note that she was hospitalized at 2 months of age for viral meningitis. The results of a thorough physical examination, including detailed neurologic evaluation, are normal, and the child is growing and developing appropriately.

Of the following, the MOST appropriate management of this patient is to

A. discuss with the mother that a previous frightening illness can affect the parent-child interaction and parental perception of the child's vulnerability to illness
B. perform screening laboratory studies, including complete blood cell count and immunoglobulins, to rule out an underlying immune system disorder
C. reassure the mother that you do not find anything wrong today and plan for follow-up at the next well-child check in 1 year
D. refer the child to neurology to evaluate the headaches and to assess for sequelae of meningitis
E. refer the child to a neuropsychologist for educational testing to guide appropriate school placement

Item 218 I-C P Preferred Response: A

The vulnerable child syndrome (VCS) was first described in 1964 as a condition in which the parent perceives their child as vulnerable to illness or harm after a real or perceived life-threatening event to the child, and that apparent vulnerability causes long-term psychological effects for both the parent and the child. It is a dysfunction involving the entire family environment as well as the individual parent and child. Family risk factors include environmental or family stress, low levels of social support, and low socioeconomic status. Parents often rate their own health as poor and/or have underlying psychological conditions. Parental psychological conditions associated with VCS include postpartum depression, obsessive-compulsive behavior, anxiety, unresolved grief reactions, and parentally perceived unacceptable feelings toward the child (eg, anger, ambivalence). Events that lead to VCS can occur at any stage of the child's life including before conception. Associated factors occurring before birth include fertility problems, previous miscarriage, inability to have more children, and risks—real or potential—to the fetus during pregnancy. In the newborn period, risk factors include being firstborn, preterm birth, neonatal hyperbilirubinemia, congenital anomalies, possibility of a hereditary disease, and heart conditions even innocent murmurs. Infants who cry excessively or have feeding problems also may be perceived as vulnerable. After the newborn period, serious illnesses or injuries that threaten the child's life can lead to VCS.

Vulnerable child syndrome entails not only the perceived risk to the child, but also the behavioral consequences. Common symptoms include difficulty with separation, sleep disturbance, frequent somatic complaints, and school under-achievement. These patients often use health services excessively. Parents frequently are unable to set appropriate limits for the child, and may tolerate physical aggression by the child.

Although management of VCS is not well-studied, expert opinion recommends clear and direct communication that past illness can affect current perception of the child's health. The physician should neither exaggerate nor underplay the importance of a preceding illness or injury, but s/he should clearly state (if appropriate) that it is now resolved and would not be expected to occur again. The family may need to have periodic, regularly scheduled visits to control the use of health services; during these visits the physician and family can address health concerns as well as behavior issues. Evaluation of medical problems (such as headaches for the child in the vignette) is most appropriately confined to detailed history and physical examination, and referrals and laboratory studies should be reserved for specific indications. Likewise, educational testing would only be indicated if there were concerns about learning problems once the behavioral components of the VCS had been addressed.

PREP Pearls

- The vulnerable child syndrome occurs when a parent perceives a child as being at risk for illness or harm because of a previous threat to their health.
- The syndrome involves not only the perception of risk but also the behavioral consequences of that perception.

- These children overuse health care resources, have frequent somatic complaints, may experience school underachievement, and may act aggressively toward caregivers.
- Most experts recommend clear and direct communication to the parent about the effect of prior illness on current perception of the child's health.
- Evaluation for medical complaints by these children should primarily rely on thorough history and physical examination rather than extensive laboratory or radiographic studies.

American Board of Pediatrics Content Specification(s):

- Understand the factors that predispose a child to the vulnerable child syndrome
- Recognize how a health crisis, real or perceived, of a child may affect parenting of the child
- Know how to provide anticipatory guidance to prevent vulnerable child syndrome

Suggested Reading:

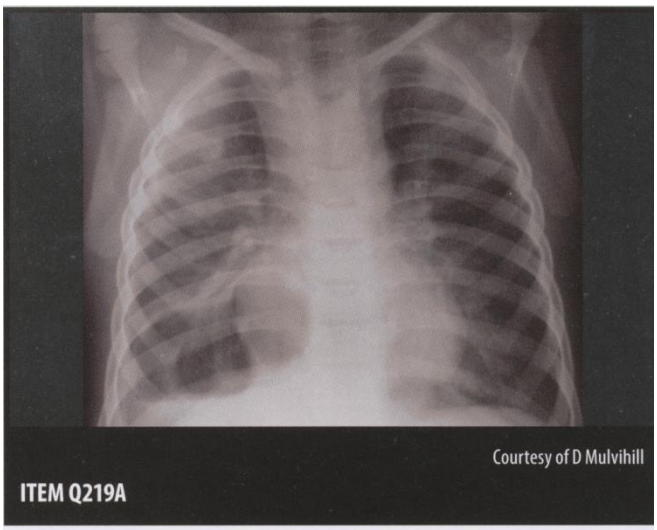
- Chambers PL, Mahabee-Gittens M, Leonard AC. Vulnerable child syndrome, parental perception of child vulnerability, and emergency department usage. Pediatr Emerg Care. 2011;27:1009-1013. doi:10.1097/ PEC.0b013e318235bb4f
- Duncan AF, Caughy MO. Parenting style and the vulnerable child syndrome. J Child Adolesc Psychiatr Nurs. 2009; 22:228-234. doi:
- 10.1111/j.1744-6171.2009.00203.x
- Forsyth BWC, Horwitz SM, Leventhal JM, Burger J. The child vunerability scale; an instrument to measure parental percetions of child vulnerability. J Pediatr Psychol. 1996;21 :89— 101 doi:10.1093/jpepsy/21.1.89
- Kokotos F. The vulnerable child syndrome. Pediatr Rev. 2009;30:193-194. doi:10.1542/pir.30-5-193
- Pearson SR, Boyce T. Consultation with the specialist: the vulnerable child syndrome. Pediatr Rev. 2004;25:345-349. doi:10.1542/pir.25-10-345

Image 219

You are called to assist in the resuscitation of a 3-month old infant in the emergency department. On your arrival, the patient is being managed with bag-valve-mask ventilation and is noted to have a heart rate of 120 beats per minute, palpable peripheral pulses, and an oxygen saturation of 94% as measured by pulse oximetry. Breath sounds are equal bilaterally. The patient has no spontaneous respiratory effort. Chest rise during ventilation is marked, and, as you prepare for intubation, you notice that the heart rate is falling and the oxygen saturation has dropped to 75%. Chest rise is now asymmetric with decreased breath sounds noted on the left side.

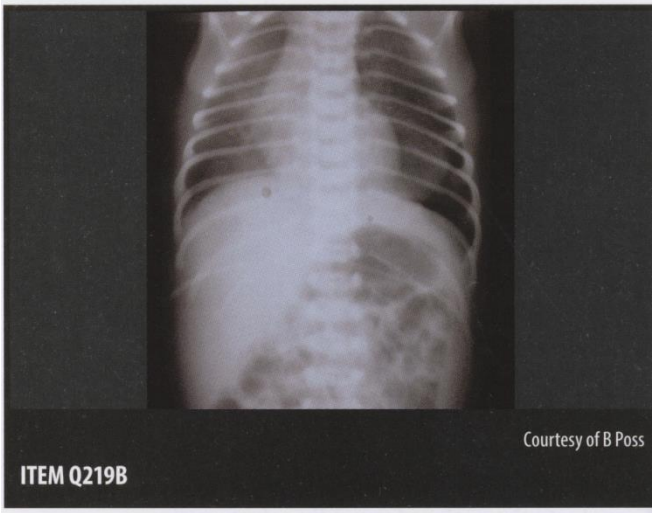
Of the following, the chest radiograph that is MOST consistent with the sudden deterioration in the patient's condition is

A. Item Q219A
B. Item Q219B
C. Item Q219C
D. Item Q219D, page Q-57
E. Item Q219E, page Q-57



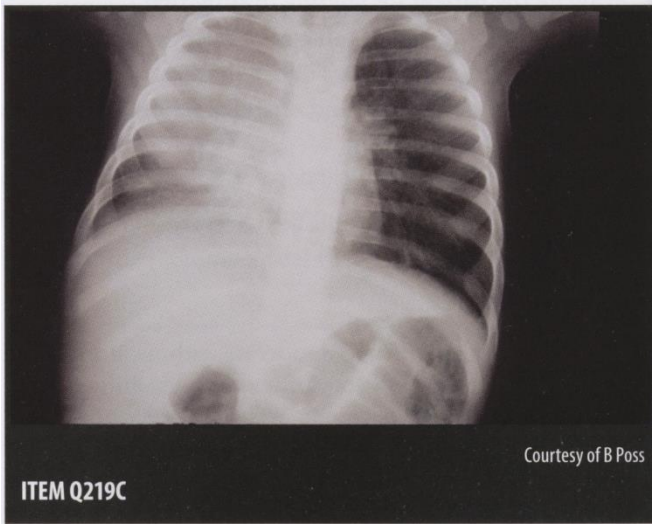
Courtesy of D Mulvihill

ITEM Q219A



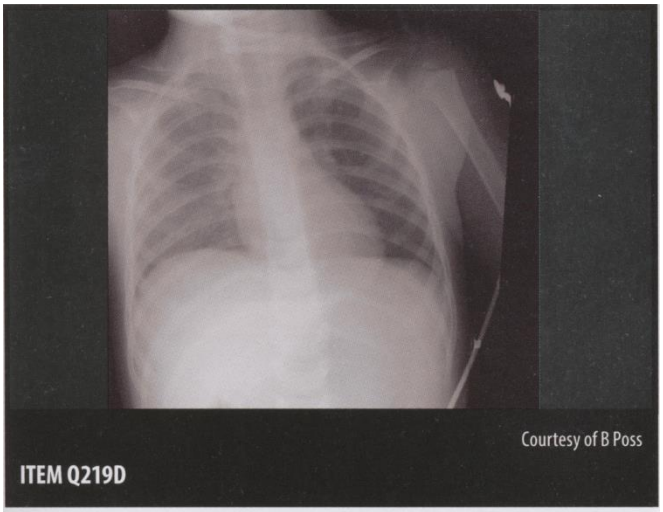
Courtesy of B Poss

ITEM Q219B



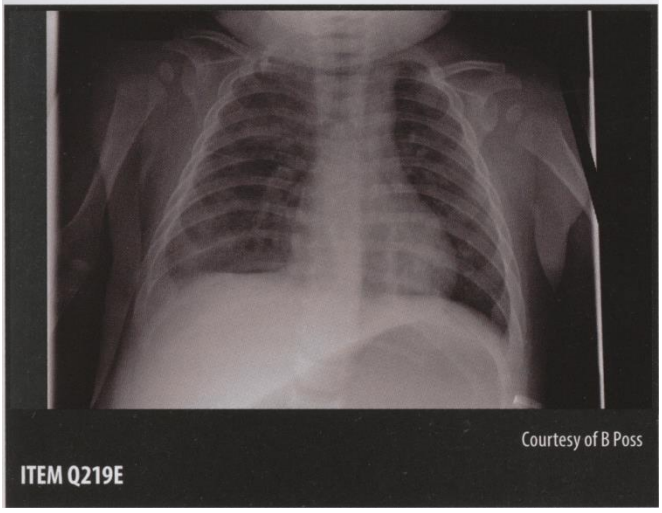
Courtesy of B Poss

ITEM Q219C



Courtesy of B Poss

ITEM Q219D



Courtesy of B Poss

ITEM Q219E

Item 219 Preferred Response: B

Positive pressure ventilation (bag-valve-mask or with an endotracheal tube) can result in a pneumothorax secondary to high pressures or excessive volumes being delivered to lungs that are often already injured by an underlying disease process. For the infant in the vignette, asymmetric chest rise, decreased aeration of the left chest, and deteriorating clinical condition are consistent with a pneumothorax as shown in option B. A useful mnemonic for recalling the causes of acute deterioration in a patient receiving positive pressure ventilation is DOPE-displacement of the tube, obstruction of the tube, pneumothorax, equipment failure.

Pneumothoraces may be classified as simple, communicating, or tension. Simple pneumothoraces do not communicate with the atmosphere, are the most common, and usually arise spontaneously with or without underlying lung pathology. A communicating pneumothorax is normally caused by open chest trauma with rapid accumulation of pleural air possible in the spontaneously breathing patient because of negative intrathoracic pressure. Tension pneumothoraces represent a medical emergency and develop when continued accumulation of air produces a mediastinal shift to the contralateral side and subsequent compression and decreased function of the lung and vascular structures.

Chest radiography remains the gold standard for diagnosis but clinical conditions may necessitate rapid treatment before imaging. Small asymptomatic pneumothoraces may resolve without treatment. Oxygen therapy is often administered but current evidence does not support that it will increase the speed of resolution. Analgesics are often required secondary to pain. Larger (>5% collapse) or symptomatic pneumothoraces require definitive treatment with thoracentesis or thoracic tube placement.

Option A is a radiograph of a patient who has pulmonary sequestration and cysts in the right lower lobe whereas option C is consistent with atelectasis involving the right upper and middle lobes. Option D illustrates a pulmonary contusion with diffuse haziness of the right lung, and option E demonstrates diffuse bilateral interstitial infiltrates and hyperinflation characteristic of viral bronchiolitis.

PREP Pearls

- Positive pressure ventilation (bag-valve-mask or with an endotracheal tube) can result in a pneumothorax secondary to high pressures or excessive volumes.
- Common causes of acute deterioration in a patient receiving positive pressure ventilation include displacement of the tube, obstruction of the tube, pneumothorax, equipment failure (DOPE).
- Chest radiography remains the gold standard for diagnosing a pneumothorax but clinical conditions may necessitate rapid treatment before imaging.

American Board of Pediatrics Content Specification(s):

- Know the appropriate therapy for a child with pneumothorax

Suggested Reading:

- Kleinman ME, Chameides L, Schexnayder SM, et al. Pediatric advanced life support: 2010 American Heart Association guidelines for cardiopulmonary resuscitation and emergency cardiovascular care. Pediatrics. 2010;126:e1361-e1399. doi: 10.1542/peds.2010-2972D
- Winnie GB, Lossef SV. Pneumothorax. In: Kliegman RM, Stanton BMD, St Geme J, Schor N, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Elsevier Saunders; 2011:1509-1512

Item 220

A 4-year-old boy presents with new-onset type 1 diabetes mellitus and diabetic ketoacidosis. He appears tired but, on physical examination, shows only mild signs of dehydration. Initial laboratory tests reveal the following results:

- Serum glucose, 884 mg/dL (49.1 mmol/L)
- Serum sodium, 131 mEq/L (131 mmol/L)
- Serum potassium, 4.5 mEq/L (4.5 mmol/L)
- pH, 6.92 on arterial blood gas

Of the following, the MOST important first step in managing this patient is to administer

A. an insulin drip, beginning at 0.1 units/kg/h
B. an intravenous insulin bolus of 0.1 units/kg
C. lactated Ringer solution, 20 mL/kg over 15 minutes
D. normal saline, 10 to 20 mL/kg over 1 to 2 hours
E. sodium bicarbonate, 1 to 2 mmol/kg over 60 minutes

Item 220 Preferred Response: D

The patient described in the vignette is in diabetic ketoacidosis (DKA) with a significant acidosis. Current guidelines from the American Diabetes Association (ADA), the International Society for Pediatric and Adolescent Diabetes, and the Pediatric Endocrine Society recommend initial management with a bolus of isotonic fluids, typically 10 to 20 mL/kg over 1 to 2 hours.

The reason to give a bolus slowly is to prevent a rapid decrease in glucose concentration while restoring peripheral circulation because a rapid decrease in blood glucose may be one of many risk factors for the development of cerebral edema. A rapid bolus that could lead to osmotically mediated fluid shifts is not appropriate, although recent data suggest that vasogenic, rather than cytotoxic, cerebral edema may be the predominant finding in DKA.

All 3 guidelines recommend starting insulin therapy 1 to 2 hours after fluid resuscitation has begun because most children with DKA are 7.5% to 10% volume depleted at presentation and need volume resuscitation first. In some studies, early initiation of insulin therapy within the first hour of fluid therapy has been associated with the development of cerebral edema, although the reasons for this are not clear. Intravenous fluids and treatment with an insulin drip will serve to correct the high anion gap acidosis present in DKA. Insulin stops further ketoacid production and allows ketoacids to be metabolized. Treatment of hypovolemia improves tissue perfusion and renal function, increasing the excretion of organic acids. Alternative treatments to correct acidosis, such as treatment with intravenous sodium bicarbonate, are not routinely recommended. Controlled trials have reported no clinical benefit from sodium bicarbonate administration, and such use is also linked to an increased risk of cerebral edema. However, as noted in the ADA guidelines, for patients with an arterial pH less than 6.9, in whom decreased cardiac contractility and peripheral vasodilatation can further impair tissue perfusion, and in patients with life-threatening hyperkalemia, treatment with intravenous sodium bicarbonate can be used. Pediatricians must recognize, however, that there are well-recognized adverse effects of sodium bicarbonate therapy, including paradoxical central nervous system acidosis, hypokalemia, and an increased risk of cerebral edema.

An intravenous bolus of insulin is unnecessary, may increase the risk of cerebral edema, and should not be routinely used at the start of therapy.

PREP Pearls

- In diabetic ketoacidosis (DKA), the first therapeutic intervention should be a slow bolus infusion of normal saline.
- Intravenous sodium bicarbonate should not be used in the routine treatment of DKA in children.
- In children who have DKA with an arterial pH less than 6.9 and evidence of hemodynamic instability, or life-threatening hyperkalemia, treatment with intravenous sodium bicarbonate can be used, although there are well-recognized risks with this treatment.

American Board of Pediatrics Content Specification(s):

- Understand the risks of using bicarbonate in diabetic ketoacidosis

Suggested Reading:

- Dunger DB, Sperling MA, Acerini CL, et al; European Society for Paediatric Endocrinology; Lawson Wilkins Pediatric Endocrine Society. European Society for Paediatric Endocrinology/Lawson Wilkins Pediatric Endocrine Society consensus statement on diabetic ketoacidosis in children and adolescents. Pediatrics. 2004;113:e133-e140. doi:10.1542/ peds.113.2.e133
- Wolfsdorf J, Glaser N, Sperling MA; American Diabetes Association. Diabetic ketoacidosis in infants, children, and adolescents: a consensus statement from the American Diabetes Association. Diabetes Care. 2006;29:1150-1159. doi:10.2337/dc06-9909
- Wolfsdorf J, Craig ME, Daneman D, et al. Diabetic ketoacidosis in children and adolescents with diabetes. Pediatr Diabetes. 2009;10(suppl 12):118-133. doi:10.1111/j.1399-5448.2009.00569.x

Item 221

A 4-month-old, previously well infant is seen in the emergency department for evaluation of a 5-day history of bloody diarrhea, an increasing number (up to 10-12) of loose bowel movements per day, and high fever. His oral intake has decreased, and his mother is uncertain about urine output since his diapers have all had loose stool in them.
He was seen by his pediatrician 3 days earlier, and stool culture from that visit is reported as growing colonies suspicious for Salmonella. The infant has a temperature of 40.1°C, heart rate of 150 beats/min, blood pressure of 82/58 mm Hg, and respiratory rate of 28 breaths/min. Capillary refill time is 3 seconds. The anterior fontanelle is slightly sunken. Abdominal examination reveals increased bowel sounds without hepatosplenomegaly or focal tenderness. He is assessed as 3% to 6% dehydrated and is being admitted to the hospital for further management. Stool, blood, urine, and cerebrospinal fluid specimens for culture are obtained and 20 mL/kg of normal saline is administered.

Of the following, the BEST next step would be administration of

A. ampicillin
B. ceftriaxone
C. cefazolin
D. metronidazole
E. vancomycin

Item 221 Preferred Response: B

Salmonella infection is the most common foodborne illness in the United States. Infection also can be acquired through contact with contaminated pets such as turtles, lizards, and hedgehogs. Nontyphoidal Salmonella infection in the normal host is generally limited to the gastrointestinal tract with manifestations ranging from asymptomatic to dysentery with bloody diarrhea and severe cramping. Invasive disease including bacteremia may occur especially in the very young, elderly, or hosts with underlying immunocompromised conditions, including malignancy, human immunodeficiency virus infection, recipients of tumor necrosis factor α-blocking medications, and sickle cell disease. In an infant younger than 1 year, as in this patient, especially in the face of high fever, empiric antibiotic therapy is advisable pending the results of blood culture. Salmonella typhi infection may cause invasive bacteremia at any age. In view of current resistance patterns among Salmonella isolates, a third-generation cephalosporin such as ceftriaxone would be the best choice for treatment pending sensitivity testing results.

Ampicillin would be an appropriate agent for treating invasive Salmonella infection if the isolate is found to be sensitive in vitro, but given high levels of resistant strains it would not be the best choice for initial empiric therapy. First-generation cephalosporins, metronidazole, and vancomycin are not active against Salmonella. Additional agents that may be effective against Salmonella include fluoroquinolones, trimethoprim-sulfamethoxazole, carbapenems, and azithromycin.

PREP Pearls

- Nontyphoidal strains of Salmonella may cause invasive disease in young children and individuals with underlying immunocompromised states.
- A third generation cephalosporin is the drug of choice for treating invasive Salmonella infections pending results of sensitivity testing.

American Board of Pediatrics Content Specification(s):

- Plan the treatment of an invasive Salmonella infection

Suggested Reading:

- American Academy of Pediatrics. Salmonella infections. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:635-640
- Centers for Disease Control and Prevention. Vital signs: Incidence and trends of infection with pathogens transmitted commonly through food—foodborne diseases active surveillance network, 10 sites, 1996-2012. MMWR Recomm Rep. 2011;60(22):749-755

Item 222

A 10-year-old girl presents to your office with 3 days of knee and ankle pain with swelling. There is no history of trauma. Review of systems is negative for fever, rash, or other signs of systemic illness. She was seen in your office 5 weeks ago for a diarrheal illness that resolved after 5 days. Physical examination reveals warmth, swelling, and slightly decreased range of motion of knees and ankles bilaterally. Vital signs are normal. The girl's complete blood cell count and erythrocyte sedimentation rate are within normal limits.

Of the following, the BEST initial treatment is

A. acetaminophen
B. amoxicillin
C. ceftriaxone
D. naproxen
E. prednisone

Item 222 Preferred Response: D

This patient described in the vignette is well appearing, afebrile, with normal blood cell counts and inflammatory markers, presenting a few weeks after a diarrheal illness. She most likely has reactive arthritis. Naproxen is a nonsteroidal anti-inflammatory drug (NSAID) that is recommended as the first line treatment for reactive arthritis. The drug has both pain relieving and mild anti-inflammatory properties and would be the best initial choice for this patient. With no history of sexual activity, and a physical examination and laboratory studies not consistent with a pyogenic arthritis, no antibiotic therapy is needed. Acetaminophen has pain relieving properties, but no anti-inflammatory properties that would benefit this patient. Prednisone is not recommended as this can mask symptoms of a chronic arthritis which would require referral to a rheumatologist.

Reactive arthritis is associated with an infection outside the affected joint. The diagnosis of reactive arthritis is a clinical diagnosis based on mono or oligoarthritis usually of the lower extremities and exclusion of other types of arthritis, such as septic arthritis, Lyme arthritis, acute rheumatic fever, as well as trauma, neoplasm and osteomyelitis. The arthritis is usually asymmetric and affects large joints such as the knee, hip, and ankle. Sacroiliac joints and the joints of the upper extremities can be affected. Reactive arthritis is commonly associated with sexually transmitted infections such as chlamydia and gonorrhea. All patients with a history of sexually transmitted infections and arthritis should be screened for these infections. Reactive arthritis is also associated with other genitourinary, gastrointestinal and upper respiratory infections. The arthritis can appear within days or up to six weeks after the infection. After 6 weeks of joint swelling, the arthritis is considered chronic and a rheumatology referral for possible autoimmune disease should be considered. Treatment of reactive arthritis is supportive with nonsteroidal anti-inflammatory drugs (Item C222), and the use of conservative treatment such as rest and cold therapy. Activity should be limited secondary to pain and can be resumed as pain improves.

PREP Pearls

- Nonsteroidal anti-inflammatory medications are recommended as first line agents in reactive arthritis.
- Prednisone can mask symptoms of joint swelling and make the diagnosis of chronic arthritis difficult.
- All sexually active patients with arthritis should be screened for sexually transmitted infections.

American Board of Pediatrics Content Specification(s):

- Plan the management of a patient with postinfectious arthritis

Suggested Reading:

- Berard R. Approach to the child with joint inflammation. Pediatr Clin N Am. 2012;59(2):245-262. doi:10.1016/j.pc1.2012.03.003
- John J Chandran L. Arthritis in children and adolescents. Pediatr Rev. 2011;32(11):470-480. doi:10.1542/pir.32-11-470

Item C222. Treatment of reactive arthritis

Nonsteroidal Anti-inflammatory Drugs	Anti-inflamatory Dose*	Dosage Forms
Ibuprofen	10 mg/kg per dose four times a day OR 13mg/kg per dose three times a day (Maximum dose 2400mg/day)	Pill or liquid forms
Naproxen	10 mg/kg/dose twice a day (Maximum dose, 1,000 mg/d)	Pill or liquid forms
Meloxicam	0.25 mg/kg/dose once daily (Maximum dose, 15mg/day)	Pill or liquid forms (very small pill)
Celecoxib	50 mg twice a day for 2 to 17 years old 10-25kg. 100mg twice a day for patients >25kg (Maximum dose 200 mg/day)	Caps may be opened, sprinkled onto apple sauce, and taken immediately with water

*The doses listed are antiinflammatory doses for the nonsteroidal anti-inflammatory drugs. Smaller doses are used for fever and pain relief.

Item 223

The parents of a 6-year-old boy report that he frequently resists their requests to do things, such as pick up his toys, and will have temper tantrums, often leading them to give in to his demands. He is pleasant when his parents permit him to do whatever he wants. He frequently disrupts his classroom both by not following the rules and by defying his teacher. Last year's kindergarten teacher reported that he was a "difficult child" who often resisted sharing and did not follow the rules. His current teacher has reported that when the boy is interested and engaged in his schoolwork, he performs at or above grade level academically and does not appear to have problems focusing on tasks. The parents report that the boy's behavior problems began 2 years ago, after the birth of their second child.

Of the following, the MOST appropriate recommendation is to

A. ask the parents to record what happens before, during, and after his defiant episodes for 3 days and return for follow-up
B. prescribe methylphenidate and have the family return in 2 weeks
C. refer the child to a pediatric neurologist to determine if the angry outbursts have a neurologic basis
D. refer the child to a play therapy specialist
E. request a comprehensive learning disability evaluation at the school

Item 223 Preferred Response: A

The boy described in the vignette appears to have oppositional defiant disorder (ODD) which would be diagnosed by the presence of pervasive oppositional and defiant symptoms present for more than 6 months, as well as a lack of other contributing problems. The dynamics within the household likely changed upon the birth of their second child 2 years ago, and a loss of parental focus on the patient could have contributed to the development of a pattern of defiant behaviors. ODD is best treated with behavior management training (also termed parent management training), in which caregivers are coached to use skilled techniques to deal with oppositional and defiant behavior, and to ensure that the child is also receiving positive parental attention. Many specific models of behavior management training have common elements, which include the reincorporation of praise and positive attention, setting up consistent limits, and analyzing the patterns of negative behaviors. To analyze a specific negative behavior pattern, one needs to look at the events leading up to, during, and following the negative behavior. By identifying the reinforcing and perpetuating factors, the way to break the cycle can become obvious. This is also known as functional analysis of behavior. Asking the parents or a school representative to keep track of what happens before, during, and after episodes of defiance for 2 to 3 days will help to reveal the patterns that perpetuate the problem.

An example of functional behavior analysis in a child who throws tantrums could be as follows: for a child who, by throwing a tantrum, ultimately gets what he originally wanted (like candy or a new toy), the way to make the tantrum behavior go away is to coach parents to help the child learn that having a tantrum no longer "works." Yelling at the child and aversively demanding compliance may reinforce the behavior if the child receives minimal one-on-one parental attention at other times. Therefore, both minimizing parental aversive reactions and ensuring that the child receives one-on-one attention routinely, rather than during a tantrum, should improve the behavior.

Prescribing methylphenidate would not be recommended because this child does not consistently demonstrate the problems with attention and hyperactivity in multiple settings that are required for a diagnosis of attention-deficit/ hyperactivity disorder. Referral to a pediatric neurologist would not be helpful because there is no focal neurologic symptom in his history. A neurologic evaluation would have been necessary if the child demonstrated neurologic signs such as outbursts that were completely unprovoked followed by sedation or sleep that might indicate a postictal state.

A play therapist may be able to help this child express any worries or concerns that he might otherwise be reluctant to communicate. However, this is far less likely to yield positive behavioral change than would behavioral management techniques. Knowing that at least some of his behaviors are perpetuated because his parents give in to his demands indicates that there is likely far more to be gained by working with the parents than working solely with the patient.

His ability to perform at or above grade level in school argues against a learning disability being responsible for his disruptive behaviors.

PREP Pearls

- Defining the sequence of events before, during and after child behavioral outbursts will frequently reveal the best means of intervention for ODD problems.
- One-on-one individual counseling or play therapy with a young child with ODD is less likely to produce behavior change than working directly with the parents on modifying their responses to their child's behaviors.

AAP Mental Health Competency:

- Know the key role of doing a functional analysis of behavior in order to know how to treat what is seen as maladaptive behavior

Suggested Reading:

- Chorpita BF, Daleiden EL; PracticeWise LLC. CAMHD biennial report: effective psychosocial interventions for youth with behavioral and emotional needs. Hawaii Department of Health. http://hawaii.gov/health/ mental-health/camhd/library/pdf/ebs013.pdf
- Steiner H, Remsing L, and the American Academy of Child and Adolescent Psychiatry. Practice parameter for the assessment and treatment of children and adolescents with oppositional defiant disorder. JAm Acad Child Adolesc Psychiatry. 2007;46(1):126-141. doi:10.1097/01. chi.0000246060.62706.af

Item 224

A 24-week-premature infant who is now 46 weeks of corrected gestational age is being seen in your office for concerns of decreasing oral intake and poor weight gain. The infant was discharged to home from the neonatal intensive care unit 2 weeks ago weighing 2,400 grams. His medical problems include bronchopulmonary dysplasia that requires diuretics and gastroesophageal reflux that has been treated with omeprazole. The infant was sent home feeding a minimum of 45 mL of premature formula concentrated to 27 calories per ounce orally every 3 hours using a slow flow nipple. At the first office visit 1 week ago, the infant weighed 2,505 grams and was described as feeding slowly but taking the minimum volume of formula daily. Currently, the infant weighs 2,610 g. The mother reports that the infant is feeding 35 to 45 mL of formula every 3 hours. The mother describes her son as waking and hungry every 3 hours, although he often "shuts down" after 10 minutes of feeding and refuses to take more. The infant does not spit, arch, or turn red with feedings. He passes 2 soft, brown stools daily. The physical examination is otherwise unremarkable. When you observe a feeding, the infant has unlabored respiratory effort and maintains an oxygen saturation of 95%.

Of the following, the MOST appropriate approach to help with the poor oral intake is to

A. advance the caloric content of the formula
B. change to a regular-flow nipple
C. consult occupational therapy
D. increase the diuretic dosage
E. initiate oxygen via nasal cannula

Item 224 TE I-C Preferred Response: C

The premature infant described in the vignette is taking limited feeding volumes, resulting in inadequate weight gain, and therefore would benefit from a consultation with occupational therapy. Premature infants, especially those born at less than 1,000 g, are at increased risk of having oral feeding issues. These issues may evolve into long-term feeding disorders including oral aversion. Studies suggest that feeding problems are 7 times more common in infants born prematurely, with 33% of infants born at less than 26 weeks' gestation having continued feeding issues at 30 months' corrected gestational age. The presence of feeding problems in some premature infants may suggest an underlying neurologic impairment and in others may contribute to poor growth. Early identification and management by a multidisciplinary team experienced with premature infants, including an occupational therapist, will lead to improved outcome for these patients.

Coordination of sucking, swallowing, and breathing is needed for effective oral feeding. This skill set begins to emerge between 32 and 34 weeks' postmenstrual age and matures with time. State regulation and behavioral organization are also required for successful feeding and develop with advancing gestational age. An infant who is born extremely prematurely often cannot adequately feed orally until at or beyond the actual due date, which plays a critical role in timing the discharge. Premature infants sometimes are discharged from the hospital when the feeding volumes appear to be sufficient for growth. However, their oral skills and coordination may not have adequately matured and these infants may develop compensatory strategies. These strategies may be dysfunctional and present as feeding problems when required feeding volumes increase or solids are introduced. Oral aversion is a sensory-based feeding disorder that may be seen in premature infants after discharge. Often it does not present until sucking transitions from reflexive to volitional at 3 to 4 months' postmenstrual age. Oral experiences associated with pain such as intubation, suctioning, feeding tubes, severe reflux, and aggressive oral feedings all contribute to the heightened risk for oral aversion in premature infants.

The complication of bronchopulmonary dysplasia (BPD) has been closely linked to delayed oral feedings and later feeding problems. Affected infants have more issues with suck-swallow-breathe coordination with feeding during their initial neonatal care. These issues persist after discharge, with infants with BPD having more desaturation events with feedings, problems with oral motor coordination, less oral intake, and poor growth. Other complications of prematurity including necrotizing enterocolitis and gastroesophageal reflux are also associated with feeding problems.

The infant described in the vignette has BPD that is well managed with diuretic treatment and shows no evidence of respiratory distress or desaturation with feeding. He does not require an increase of the diuretic dosage or initiation of oxygen via nasal cannula. The use of a regular flow nipple may decrease the work required for oral feeding by allowing formula to flow faster, but the additional volume may lead to the development of dysfunctional compensatory feeding strategies. The change to a regular flow nipple is not recommended because of the risk of worsening feeding issues. Increasing the caloric content would enhance weight gain, but not address the underlying cause of the

inadequate feeding volumes. Consultation with an occupational therapist experienced with premature infants is recommended for an effective management plan to be developed which both sup-ports oral skill maturation and adequate growth.

PREP Pearls

- Feeding problems are 7 times more common in infants born prematurely, with 33% of infants born at less than 26 weeks' gestation having continued feeding issues at 30 months' corrected gestational age.
- Consultation with an occupational therapist experienced with premature infants is recommended when feeding problems are identified in discharged premature infants.

American Board of Pediatrics Content Specification(s):

- Recognize that aversive oral motor behavior is associated with bronchopulmonary dysplasia, thereby limiting ways to feed such infants

Suggested Reading:

- Bakewell-Sachs S, Medoff-Cooper B, Escobar GJ, Silber JH, Lorch SA. Infant functional status; the timing of maturation of premature infants. Pediatrics. 2009;123:e878-e886. doi: 10.1542/peds.2008-2568
- Lau C. Oral feeding in the preterm infant. NeoReviews. 2006;7:e19-e27. doi: 10.1542/neo.7-1-e19
- Oral aversion. In: Brodsky D, Ouellette MA, eds. Primary Care of the Premature Infant. Philadelphia, PA: Elsevier Saunders; 2008:101-104
- Thoyre SM. Feeding outcomes of extremely premature infants after neonatal care.l Obstet Gynecol Neonatal Nurs. 2007;36:366-376. doi: 10.1111/j.1552-6909.2007.00158.x

Item 225

A 6-month-old male infant is brought to your office for poor feeding. The mother reports that he refuses to drink from a bottle and cries when she tries to feed him. He has been seen in your office in the past for various infections, including a perirectal abscess, otitis media, and pneumonia. His axillary temperature is 38.3°C, pulse rate is 110 beats/ min, respiratory rate is 24 breaths/min, and blood pressure is 100/60 mm Hg. On examination, the child is fussy but consolable. He has erythematous ulcerations in his oral mucosa, but no hepatosplenomegaly or lymphadenopathy. The remainder of the physical examination is normal. The following are the results of the infant's complete blood cell count:

- White blood cell count, 6,000/µL (6.0 x 10'/L), with 5% polymorphonuclear leukocytes, 85% lymphocytes, 7% monocytes, and 3% eosinophils
- Hemoglobin, 12.5 g/dL (125 g/L)
- Mean corpuscular volume, 85/μm^3 (85 fL)
- Platelet count, 300 x 10^3/µL (300 x 10A)

Of the following, the MOST appropriate next step in management for this patient is to

A. admit for bone marrow studies to rule out leukemia
B. draw a blood culture and give parenteral antibiotics
C. give intravenous hydration and a prescription for pain medication
D. reassure the parents that this is most likely a self-limited viral infection
E. repeat complete blood cell count with differential the following day

Item 225 Preferred Response: B

The infant described in the vignette with fever and severe neutropenia (absolute neutrophil count [ANC], 300/µL (0.3 x 10^9/L) should have a blood culture drawn and receive parenteral antibiotics including coverage for Pseudomonas. Normal values for ANC vary with age (Item C225A, page C-176). From 2 months to 1 year of age, neutropenia is defined as ANC less than 1000/4 (1.0 x 10^9/L), whereas in children older than 1 year and adults, the lower limit of normal for ANC is 1500/ µL. (1.5 x 10^9/L). In addition, there is variation in normal neutrophil counts among different racial groups, with 3% to 5% of patients of African descent having ANC values less than 1500/ µL. (1.5 x 10^9/L). The risk of infection is related to the severity (Item C225B, page C-176), duration, and mechanism of neutropenia. Long periods of neutropenia without recovery lead to an increased risk of infection. Conditions in which there is adequate bone marrow reserve to mobilize neutrophils, as in immune neutropenia, are less likely to be associated with severe infection than in disorders of decreased marrow production, such as bone marrow failure syndromes or myelosuppression secondary to chemotherapy.

In evaluating an infant or child with neutropenia, it is important to ask whether there is a history of recurrent infections in the patient or the family. A thorough physical examination for signs of neutropenia (eg, skin infections, mouth ulcers, and gingivitis) and congenital anomalies is necessary to evaluate for inherited syndromes. The presence of other cytopenias would suggest a generalized bone marrow disorder, such as aplastic anemia or leukemia.

In the pediatric population, congenital forms of neutropenia are much less common than acquired disorders. Severe congenital neutropenia (SCN) can lead to life-threatening infections and increased risk of malignant tumors. Patients with SCN often have very severe neutropenia (ANC <200/mL [0.2 x 10^9/L]) and present in infancy with multiple umbilical, mucosal, or skin infections. The inheritance pattern can be autosomal dominant or autosomal recessive, or it may be sporadic. It has been associated with ELANE mutations. The risk of leukemia is 5% to 10% in SCN. Kostmann syndrome is an autosomal recessive form of SCN that is associated with mutations in the HAX1 gene and has a 15% to 20% risk of leukemia. Patients at highest risk for malignant transformation are those with poor response to high doses of granulocyte colony-stimulating factor (G-CSF) and those with certain mutations in G-CSF receptor gene or ELANE gene.

Cyclic neutropenia is associated with different mutations within the same ELANE gene than those seen in SCN. Serial blood cell counts performed twice a week reveal a 21-day cycle of the neutropenia during which fever and oral ulcers can erupt when the ANC is at its nadir.

Granulocyte-colony-stimulating factor may be required to prevent infections in patients with SCN or cyclic neutropenia and to treat infection or symptoms related to neutropenia (eg, mouth sores). Treatment with G-CSF is not generally indicated in asymptomatic patients with benign forms of neutropenia.

The child described in this vignette has a history and clinical findings concerning for SCN. A bone marrow examination would not be the first step in management because there are no other cytopenias to suggest bone marrow failure or malignant tumors. Hydration and pain medication may be useful if the oral ulcerations are prohibiting adequate oral fluid intake, but empiric treatment for an infection should be started first. It cannot be assumed that the severe neutropenia is caused by a self-limited viral infection, especially in an infant who has had multiple prior infections. Reassurance and repeating the complete blood cell count the following day is not adequate management for this infant with fever and severe neutropenia. Furthermore, if the neutropenia was thought to be transient (eg, viral mediated), the ANC is not likely to reveal a significant change in 1 day. Drawing a blood culture and giving empiric parenteral antibiotics for presumed bacteremia would be the most appropriate next step in managing this patient with suspected SCN.

Item C225B. Classification of Severity of Neutropenia in Patients 1 Year and Older

Degree of Severity	Absolute Neutrophil Count	Risk of Infection
Mild neutropenia	1000-1500/μL (1.0 to 1.5×10^9/L)	Little or none
Moderate neutropenia	500-1000/μL (0.5 to 1.0×10^9/L)	Less frequent, less severe
Severe neutropenia	<500/μL (0.5×10^9/L)	Highly susceptible

PREP Pearls

- An infant with fever and severe neutropenia (ANC <500/μL [0.5 x10^9/L) should have a blood culture drawn and receive parenteral antibiotics including coverage for Pseudomonas.
- Severe congenital neutropenia is associated with an increased risk of malignant tumors.
- The risk of infection is related to the severity, duration, and mechanism of neutropenia.

American Board of Pediatrics Content Specification(s):

- Recognize that congenital neutropenia may be persistent or cyclical, and manage appropriately

Suggested Reading:

- Bouma G, Ancliff PJ, Thrasher A, Burns SO. Recent advances in the understanding of genetic defects of neutrophil number and function. Br I Hernatol. 2010;151:312-326. doi:10.1111/j.1365-2141.2010.08361.x

- Bortug K, Klein C. Genetic etiologies of severe congenital neutropenia. Curr Opin Pediatr. 2011;23:2321-2326. doi:10.1097/M0P.0b013e32834262f8
- Segel GB, Halterman JS. Neutropenia in pediatric practice. Pediatr Rev. 2008;29:12-24. doi:10.1542/pir.29-1-12

Item C225A: Normal Blood Leukocyte Counts*

	Total Leukocytes		Neutrophils			Lymphocytes			Monocytes		Eosinophilis	
Age	Mean	(Range)	Mean	(Range)	%	Mean	(Range)	%	Mean	%	Mean	%
Birth	18.1	(9.0 to 30.0)	11.0	(6.0 to 26.0)	61	5.5	(2.0 to 11.0)	31	1.1	6	0.4	2
12 h	22.8	(13.0 to 38.0)	15.5	(6.0 to 28.0)	68	5.5	(2.0 to 11.0)	24	1.2	5	0.5	2
24 h	18.9	(9.4 to 34.0)	11.5	(5.0 to 21.0)	61	5.8	(2.0 to 11.5)	31	1.1	6	0.5	2
1 wk	12.2	(5.0 to 21.0)	5.5	(1.5 to 10.0)	45	5.0	(2.0 to 17.0)	41	1.1	9	0.5	4
2 wk	11.4	(5.0 to 20.0)	4.5	(1.0 to 9.5)	40	5.5	(2.0 to 17.0)	48	1.0	9	0.4	3
1 mo	10.8	(5.0 to 19.5)	3.8	(1.0 to 9.0)	35	6.0	(2.5 to 16.5)	56	0.7	7	0.3	3
6 mo	11.9	(6.0 to 17.5)	3.8	(1.0 to 8.5)	32	7.3	(4.0 to 13.5)	61	0.6	5	0.3	3
1 y	11.4	(6.0 to 17.5)	3.5	(1.5 to 8.5)	31	7.0	(4.0 to 10.5)	61	0.6	5	0.3	3
2 y	10.6	(6.0 to 17.0)	3.5	(1.5 to 8.5)	33	6.3	(3.0 to 9.5)	59	0.5	5	0.3	3
4 y	9.1	(5.5 to 15.5)	3.8	(1.5 to 8.5)	42	4.5	(2.0 to 8.0)	50	0.5	5	0.3	3
6 y	8.5	(5.0 to 14.5)	4.3	(1.5 to 8.0)	51	3.5	(1.5 to 7.0)	42	0.4	5	0.2	3
8 y	8.3	(4.5 to 13.5)	4.4	(1.5 to 8.0)	53	3.3	(1.5 to 6.8)	39	0.4	4	0.2	2
10 y	8.1	(4.5 to 13.5)	4.4	(1.8 to 8.0)	54	3.1	(1.5 to 6.5)	38	0.4	4	0.2	2
16 y	7.8	(4.5 to 13.0)	4.4	(1.8 to 8.0)	57	2.8	(1.2 to 5.2)	35	0.4	5	0.2	3
21 y	7.4	(4.5 to 11.0)	4.4	(1.8 to 7.7)	59	2.5	(1.0 to 4.8)	34	0.3	4	0.2	3

*Numbers of leukocytes are in thousands/mcL ($x10^9$/L), ranges are estimates of 95% confidence limits, and percentages refer to differential counts. Neutrophils include band cells at all ages and a small number of metamyelocytes and myelocytes in the first few postnatal days.

Reprinted with permission from Dallman PR. Blood and blood-forming tissues. In: Rudolph AM, ed. *Rudolph's Pediatrics*. 16th ed. New York, NY: Appleton-Century-Crofts/The McGraw-Hill Companies, Inc.; 1977:1178

Item 226

A 15-year-old boy is on a wilderness trip in the desert Southwest, United States, as part of a drug and alcohol rehabilitation program. He develops a fever and stiff neck and then has a generalized seizure. He is transported urgently to the nearest emergency department. On arrival, he has another generalized seizure and is given lorazepam 4 mg intravenously. Physical examination after lorazepam administration reveals a temperature of 39.1°C, blood pressure of 150/76 mm Hg, heart rate of 130 beats/min, and respiratory rate of 14 breaths/min. He is somnolent, there are no signs of trauma, and there are no rashes or insect bites. The remainder of his physical examination findings are normal. Computed tomography of the head without contrast is normal. Lumbar puncture is performed in the lateral recumbent position with legs extended. Cerebrospinal fluid (CSF) opening pressure is 380 mm H_2O; CSF protein is 182 mg/dL, and glucose is 8 mg/dL; and there are 900 white blood cells/µL (81% of which are polymorphonuclear leukocytes) and 190 red blood cells/µL.

Of the following, the MOST likely cause of this boy's symptoms is

A. Coccidioides immitis
B. Enterovirus
C. Neisseria meningitidis
D. Taenia solium
E. West Nile virus

Item 226 Preferred Response: C

Of the choices listed, Neisseria meningitidis is the most likely cause for this boy's meningoencephalitis. His risk factors for acquiring N meningitidis include adolescent age group and living in a group situation. The rapid onset of symptoms of meningoencephalitis, fever, headache, fatigue, lethargy, stiff neck, and seizure support a diagnosis of N meningitidis infection. In meningoencephalitis resulting from N meningitidis, a petechial rash may develop early in the course and lead to an early clinical diagnosis.

Cerebrospinal fluid (CSF) studies can also aid in early diagnosis. Although an elevated protein level is nonspecific, the very low glucose concentration suggests bacterial infection. However, this also can be seen in more rare infectious and inflammatory central nervous system processes such as tubercular and carcinomatous meningitis. In this case, the elevated WBC count, with predominance of polymorphonuclear leukocytes, gives additional support to a bacterial pathogen as a cause of symptoms. The CSF findings in different types of meningitis are listed (Item C226).

Coccidioides immitis is a fungus endemic to the desert southwestern United States, and typically has a more indolent clinical presentation. Enterovirus is a common cause of viral meningoencephalitis, but in this case, the CSF studies suggest a bacterial pathogen. Taenia solium infection causes neurocysticercosis, which typically presents with seizure and no other signs of meningoencephalitis. Computed tomography shows calcifications. West Nile virus can cause meningoencephalitis, and is particularly associated with multiple cranial neuropathies or flaccid limb paralysis; however, the CSF studies in this case are more consistent with a bacterial pathogen.

Item C226. Cerebrospinal Fluid Analysis*

	Glucose (mg/dL)	Protein (mg/dL)	White Blood Cell (µL)	Differential Count	Gram stain
Healthy child	40 to 80	20 to 40	<5	No PMNs	Negative
Bacterial meningitis	<1/2 serum Often <10 (0.6)	>100	>1,000	>50% PMNs Often > 90%	Variable
Enteroviral meningitis	>1/2 serum	40 to 60	50 to 500	>50% PMNs early (<48 h) <50% PMNs later (<48 h)	Negative
Lyme meningitis	>1/2 serum	50 to 150	50 to 500	Predominance of lymphocytes and monocytes	Negative
Tuberculous meningitis	<1/2 serum Often <10	>100	50 to 500	Lymphocyte predominance	Negative
Fungal memningitis	>1/2 serum	25 to 500	50 to 250	Lymphocyte predominance	Negative**

*Value should be used only as a guide, and none should be used in isolation because overlap between values in each of these categories is significant PMN=polymorphonuclear leukocytes. Adapted from Wubbel L, McCracken GH. Management of bacterial meningitis. *Pediatr Rev.* 1998;19:78-84. and Eppes, SC Nelson, DK, Lewis LL, Klein JD. Characterization of Lyme meningitis and comparison with viral meningitis is children. *Pediatrics*. 1999;103:957-960

**May be positive if *Candida*

PREP Pearls

- Cerebrospinal fluid (CSF) study results can aid in a clinical diagnosis of meningoencephalitis.
- With CSF showing a predominance of polymorphonuclear leukocytes, a bacterial cause of meningitis must be considered and treated emergently.

American Board of Pediatrics Content Specification(s):

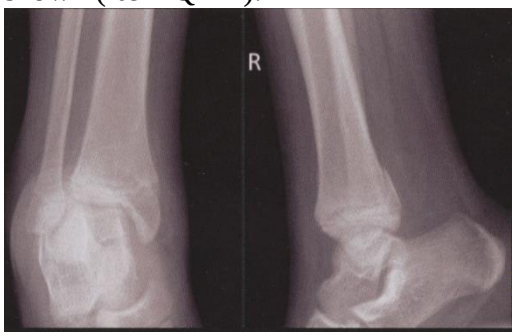
- Distinguish among cerebrospinal fluid findings in bacterial, fungal, and viral meningitis

Suggested Reading:

- Mann K, Jackson MA. Meningitis. Pediatr Rev. 2008;29:417. doi:10.1542/pir.29-12-417
- Prober CG, Dyner L. Central nervous system infections. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:2086-2098.el

Item 227

A 12-year-old girl is seen for evaluation of an injury to her right ankle. She reports that she twisted the ankle while skateboarding earlier that day. She has been unable to bear weight since her injury. On physical examination, she is sexual maturity rating 2-3, and you note significant, diffuse right ankle swelling with tenderness to palpation over the anterior aspect of the ankle and over the medial malleolus. Ankle radiograph findings are shown (Item Q227).



ITEM Q227: Radiographs for the girl described in the vignette.

Of the following, the MOST likely diagnosis is a

A. distal tibia fracture involving the physis
B. distal tibia shaft fracture sparing the physis
C. grade 3 ankle sprain involving the anterior talofibular ligament
D. high ankle sprain (syndesmosis sprain) with widening of the ankle mortise
E. talar dome fracture

Item 227 Preferred Response: A

The patient's lateral radiograph reveals a distal tibia metaphysis fracture that extends to the physis. There is also a subtle lucency in the epiphysis. This fracture pattern with the fracture occurring in the coronal plane should raise suspicion for a triplane fracture. A triplane fracture, as the name suggests, occurs in 3 planes and involves the distal tibia metaphysis in the coronal plane, the physis in the transverse plane, and the epiphysis in the sagittal plane. Triplane fractures are a type of transitional fracture, a fracture that occurs as the growth plate is in the process of closing. Because triplane fractures have complicated geometry, a computed tomography scan is often required to delineate the exact fracture pattern. Children with triplane fractures often require surgical open reduction and fixation.

Radiographs are typically normal in individuals with syndesmosis or high ankle sprains; however, widening of the space between the distal fibula and tibia indicates an unstable injury that should prompt orthopedic referral. Isolated anterior talofibular ligament injuries cannot be visualized on plain radiographs. A fracture of the talar dome can be difficult to identify radiographically and is often mistaken for an ankle sprain. Magnetic resonance imaging or computed tomography scan, to look for this type of injury, should be considered in the patient with an apparent ankle sprain that does not improve with 4 to 6 weeks of conservative treatment.

PREP Pearls

- Transitional fractures are physeal fractures with unique patterns that occur in individuals approaching skeletal maturity.
- Following an acute ankle injury, the presence of widening of the mortise on ankle radiographs in the absence of bony injury suggests an unstable syndesmosis sprain. An individual with this type of injury should be referred to an orthopedic surgeon.

American Board of Pediatrics Content Specification(s):

- Recognize the possibility of an epiphyseal injury in an apparent ankle sprain in a child whose growth plates have not closed

Suggested Reading:

- Sarwark JF, LaBella CR, eds. Pediatric Orthopaedics and Sports Injuries: A Quick Reference Guide. Elk Grove Village, IL: American Academy of Pediatrics; 2010
- Schnetzler KA, Hoernschemeyer D. The pediatric triplane ankle fracture. J Am Acad Orthop Surg. 2007;15(12):738-747

Item 228

The mother of a 3-year-old boy brings him in for a health supervision visit. Her son was diagnosed with hemophilia A shortly after birth. The child did well with only minor bleeds until 12 months of age, when he had his first of several large joint bleeds (hemarthroses). He is managed by a pediatric hematologist in a hemophilia treatment center and receives recombinant factor VIII concentrate infusions 3 times per week. The family history is significant for a maternal uncle who died of complications of human immunodeficiency virus infection, which he acquired from tainted factor VIII infusions. The mother tells you she is currently in her 12th week of pregnancy and is concerned about the risks of hemophilia and clinical consequences for her next child.

Of the following, you are MOST likely to tell her that

A. circumcision may be performed shortly after administration of vitamin K
B. a daughter will be at risk for excessive bleeding problems at birth or in the future
C. minor surgical procedures are safe in children with hemophilia without factor replacement
D. prenatal diagnosis for hemophilia A is possible through amniocentesis
E. there is a 25% risk for a son to be affected with hemophilia A

Item 228 Preferred Response: D

The mother in the vignette is 12 weeks pregnant, and it is important for her to know that prenatal diagnosis for hemophilia A (factor VIII deficiency) is possible through amniocentesis. This will enable her to make a decision about whether she wishes to pursue prenatal diagnosis. However, before such testing can be done, it is necessary to identify the specific mutation responsible for hemophilia in this family. Molecular testing can identify up to 98% of causative mutations and is best done first on an affected family member. This pregnant woman is an obligate carrier of this X-linked recessive condition based on her family history, so the mutation identified in her affected son will be the one used for prenatal diagnosis. Because molecular testing is time- and labor-intensive, testing should be initiated as soon as possible. Ideally, molecular testing is performed before conception for couples who are interested in using this information during a pregnancy. Many couples may not choose to pursue prenatal diagnosis, since it would not influence their decisions prenatally, and instead wait for diagnostic testing at birth. However, prenatal ultrasonography may be helpful to a certain degree if the fetal gender can be determined, since only male fetuses would be at risk for clinical hemophilia.

Early identification and modern therapies today enable affected boys to lead a near normal life. For previously untreated patients, therapy consists of on-demand or, more recently, prophylactic recombinant factor VIII infusions. The typical age at diagnosis in the absence of a family history (20%-30% of cases) as well as severity of bleeding depends on the factor VIII levels. Boys with severe hemophilia A are typically diagnosed between the ages of 1 year and 2 years and often experience joint or deep muscle bleeding; boys with moderate hemophilia A are more often diagnosed between the ages of 5 years and 6 years and have prolonged bleeding with minor trauma; finally, boys with mild hemophilia A tend to be diagnosed following dental extraction or surgery when prolonged bleeding occurs unexpectedly. Boys with hemophilia A should avoid intramuscular injections, activities where risk for trauma is high (especially blows to the head), and aspirin or products containing other nonsteroidal antiinflammatory drugs.

Male newborns who have hemophilia A (or who are at risk for this condition) should not undergo circumcision or elective surgical procedures until the diagnosis is made or excluded. Once a diagnosis is made, pretreatment with factor VIII concentrate is necessary for all surgical procedures, even minor ones. Vitamin K does not prevent or control bleeding in boys with factor VIII deficiency. Girls or women who are carriers of hemophilia A would generally have about 50% factor VIII clotting activity and would not experience any excessive bleeding. However, in some instances, due to unfavorable lyonization, a woman may have clotting activity less than 35%. In this case, she may have symptoms similar to boys with mild hemophilia A. For a woman who is an obligate carrier of hemophilia A, each son has a 50 % chance of being affected because he would receive the X chromosome with the normal gene or the one with the hemophilia (factor VIII mutation).

PREP Pearls

- Prenatal diagnosis is available via amniocentesis for women who are carriers of hemophilia (both A and B) but requires identification of the specific mutation prior to prenatal testing.
- For couples who desire prenatal diagnosis, mutation analysis is best done on an affected family member prior to conception.
- Boys with hemophilia A or B are treated with regular infusions of recombinant factor to prevent major bleeding events.

American Board of Pediatrics Content Specification(s):

- Know that Factor VIII and IX deficiencies can be diagnosed prenatally

Suggested Reading:

- Konkle BA, Josephson NC, Nakaya-Fletcher SM, Thompson AR. Hemophilia A. In: Pagon RA, Bird TD, Dolan CR, eds. GeneReviews. Seattle, Washington: University of Washington; 2013
- Konkle BA, Josephson NC, Nakaya-Fletcher SM, Thompson AR. Hemophilia B. In: Pagon RA, Bird TD, Dolan CR, eds. GeneReviews. Seattle, Washington: University of Washington; 2013

Item 229

A 12-year-old boy is brought to your office by his parents for evaluation of a painful lesion on his right ankle. He noticed the lesion 4 hours ago, shortly after helping his parents move some old lawn furniture out of the family garage. He denies any trauma, but he states that he noticed several shiny black spiders scurrying about the garage as he was moving the furniture. The boy is healthy and has no known allergies. He takes no medications and his immunizations are current. The boy's father took a photograph of one of the "shiny black spiders" with his cellphone; he shows it to you to see if you can identify the spider (Item Q229).

ITEM Q229: Spider as photographed by the patient's Father.

On physical examination, the boy is afebrile, and his vital signs are normal. He appears well. On his right ankle is a 1.5-cm blanched circular patch with a surrounding erythematous perimeter and a central punctum. The boy's right ankle is tender to palpation at the site of this lesion, but you note no other areas of tenderness. The remainder of his physical examination findings are unremarkable. A nurse in your office has cleansed the lesion with mild soap and water.

Of the following, the BEST next step in the management of this patient is

A. prescription of a short course of oral corticosteroid therapy
B. prescription of oral analgesics to be taken as needed
C. prescription of oral antibiotic therapy, including coverage for methicillin-resistant Staphylococcus aureus
D. referral to a pediatric surgery clinic for debridement of the lesion
E. referral to the local emergency department for Latrodectus (black widow) antivenom

Item 229 S Preferred Response: B

The boy described in the vignette presents with a painful lesion on his ankle that most likely resulted from a black widow spider bite. Given his clinical status, prescription of an oral analgesic is the best next step in management at this time.

Approximately 30 species of widow spiders are found worldwide. Most adult widow spiders are shiny black with red markings on the body. Like the spider photographed in the vignette (Item Q229), the American species (Latrodectus mactans) has a red hourglass or anvil-shaped mark located on the ventral part of the abdomen.

Widow spiders typically live outdoors in the clutter surrounding homes and garages, in woodpiles, and in infrequently used garden equipment, pots, and tools. Patients presenting with widow bites typically have a history of recent at-risk activity, such as using outdoor furniture, cleaning out a garage, or gardening. Roughly three-fourths of bites involve the extremities, most commonly the lower extremities. Typical black widow bites are minor lesions that consist of a blanched circular patch with a surrounding red perimeter and central punctum. Most bites are either asymptomatic or cause localized pain at the site of the lesion.

The clinical manifestations of widow spider bites are known as latrodectism. Significant Latrodectus envenomation may lead to acute muscle pain, rigidity, and diaphoresis in the affected extremity. These symptoms usually begin 1 to 8 hours after the spider bite occurs. Pain may extend to the abdomen for lower extremity bites and to the chest for upper extremity bites. The pain may be associated with tremor, weakness, myoclonus, and local paresthesias. Children with mild to moderate latrodectism typically have normal vital signs, whereas children with more severe envenomation often display signs of systemic illness-including tachycardia, nausea, vomiting, diaphoresis, headache, chills, facial swelling, urinary retention, and respiratory distress—in addition to generalized pain. Severe abdominal pain with abdominal wall rigidity but normal bowel sounds, is a characteristic finding of severe black widow envenomation; these findings have been mistaken for various abdominal surgical emergencies.

Death from latrodectism is rare, with overall mortality in published series ranging from 5% to 10% (although some experts believe that these may be overestimates). Children are thought to be at higher risk for mortality from black widow spider envenomation than adults, although this is controversial. The literature on pediatric mortality from black widow envenomation is sparse. The mortality rate from latrodectism in small children has been estimated to be as high as 50% by some experts; however, one case series of 12 children aged 15 months to 18 years treated at an urban pediatric tertiary care hospital for black widow spider envenomation found no deaths or long-term complications among these children.

A diagnosis of a black widow spider bite is most often made according to the patient's history and clinical findings. A Latrodectus bite should be presumed in any child with severe pain and muscle rigidity after a spider bite. In infants, latrodectism may present as a distressed, inconsolable child with generalized erythema and decreased feeding. A

classic history would be of an infant who manifests these symptoms after sleeping in a crib or cot that had been stored in a garage.

Many children who are bitten by black widow spiders require only local wound care and oral analgesics. Initial management after a presumed black widow bite should include cleansing of the bite wound with mild soap and water, administration of analgesics as needed, parenteral benzodiazepines in cases of severe muscle spasm, and tetanus prophylaxis as indicated.

Corticosteroids are not indicated in the management of black widow spider bites. Antibiotics should be prescribed only if there are signs of secondary infection, which include increasing erythema and warmth, fluctuance at the site of the lesion, purulent drainage from the wound site, and fever.

Referral to a pediatric surgeon for wound debridement would not be indicated for the boy in this vignette. Black widow spider bites generally do not become necrotic. Bites of the brown recluse spider (Loxosceles reclusa)—the other species of spider in the United States capable of producing severe manifestations—may become necrotic and require surgical management.
Although antivenoms are available for several widow spiders, including the American black widow spider (Latrodectus mactans), administration is not recommended for all patients. Antivenom treatment should be strongly considered in small children (<40 kg) with confirmed black widow spider bites and those with symptoms of moderate to severe envenomation. Consultation with a toxicologist or other physician who has specific expertise in managing widow spider bites is recommended before administration of antivenom.

PREP Pearls

- Patients presenting with widow bites typically have a history of recent at-risk activity, such as using outdoor furniture, cleaning out or playing in a garage, or gardening.
- Initial management following a presumed spider bite, including a black widow bite, includes cleansing of the bite wound with mild soap and water, administration of analgesics as needed, and tetanus prophylaxis as indicated.
- Antivenom treatment should be strongly considered in small children with confirmed black widow spider bites and those with symptoms of moderate to severe envenomation. Consultation with a toxicologist is recommended before antivenom administration.

American Board of Pediatrics Content Specification(s):

- Plan the management of a spider bite

Suggested Reading:

- Hodge D. Bites and stings. In: Fleisher GR, Ludwig S, eds. Textbook of Pediatric Emergency Medicine. 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2010:635-642
- Jelinek GA. Widow spider envenomation (latrodectism): a worldwide problem. Wilderness Environ Med. 1997;8:226-231. doi:10.1580/1080-6032(1997)008[0226:WSELAW2.3.00;2. www.ncbi.nlm.nih.gov/pubmed/11990169.
- Vetter RS, Swanson DL, White J. Bites of widow spiders. UPtoDate. Available online only for subscription
- Woestman R, Perkin R, VanStralen D. The black widow: is she deadly to children? Pediatr Emerg Care. 1996;12: 360-364

Item 230

The mother of a 4-year-old boy brings him into your office for evaluation of nasal congestion that has persisted over the last 4 weeks. He has been previously healthy and had no fevers during the course of this complaint. His appetite and activity level are normal. He has some difficulty sleeping at night due to the stuffy nose. His growth has been normal. Physical examination reveals unilateral purulent nasal discharge that is malodorous.

Of the following, the BEST next step is

A. computed tomography of the sinuses
B. nasal irrigation with saline
C. nasal speculum examination
D. oral antibiotics
E. otolaryngology referral

Item 230 SBP Preferred Response: C

Nasal congestion or "stuffy nose" is one of the most common symptoms in patients presenting to the pediatric health care professional. The causes are multiple. It is important to conduct a thorough history and physical examination to investigate for causes that may need intervention or that could lead to significant morbidity. The boy in this vignette presents with the classic findings of a nasal foreign body: unilateral, purulent, malodorous, sometimes bloody discharge. To make the diagnosis, the clinician must have a heightened suspicion based on these findings and perform a careful examination of the nares with a speculum.

It is common for young children (1-6 years of age) to insert small objects, such as beads, toy parts, berries, beans, seeds, or paper, into the nares. Usually, these foreign bodies are initially asymptomatic except for some "congestion." The longer these bodies remain lodged, occluding the nasal passage, the more complications may develop. Complications of nasal foreign bodies include local irritation, unilateral sinusitis or periorbital cellulitis, necrosis and perforation of the nasal septum (especially in the case of magnets or alkaline button batteries), or posterior migration of the foreign body into the trachea.

Bacterial rhinitis and sinusitis typically present with bilateral purulent nasal drainage. It is critical to differentiate these infections from a nasal foreign body because simply treating with an antibiotic might mask symptoms by partially treating the secondary infection and delay the recognition and removal of the foreign body. Likewise, computed tomography of the sinuses is not warranted because a thorough physical examination would reveal the foreign object.

Most nasal foreign bodies can be removed using simple techniques in the office or the emergency department without referral to the otolaryngologist. However, successful extractions require immobilization or cooperation of the child, adequate visualization, and proper equipment. In general, small soft objects may be grasped with alligator forceps. In an older child, the clinician or parent can occlude the unaffected nostril and ask the child to exhale forcibly through the nostril that contains the foreign body or the parent may apply positive pressure through mouth-to-mouth or mouth-to-nose. A suction catheter may be applied to smooth, round foreign bodies that are difficult to grasp. Irrigation of the nares when a foreign body is present should not be performed because some foreign bodies may expand when wet and there is a risk of posterior displacement of the object into the nasopharynx. Complications may result from multiple manipulations so an otolaryngology consultation is required once the diagnosis is made and when simple attempts at removal- fail. Referral is also indicated if the proper equipment and environment are not available; when the foreign body is a battery, magnet, or sharp object; or anytime there is concern for maintaining a secure airway.

PREP Pearls

- The classic findings of a nasal foreign body are unilateral, purulent, malodorous, sometimes bloody discharge.
- The diagnosis of a nasal foreign body is made by careful examination of the nares with a speculum.
- Most nasal foreign bodies can be removed using simple techniques in the office. Irrigation should not be performed and care should be taken to protect the airway.

American Board of Pediatrics Content Specification(s):

- Know the clinical presentation of a nasal foreign body

Suggested Reading:

- Gargiulo KA, Spector ND. Stuffy Nose. Pediatr Rev. 2010;31:320-324. DOI: 10.1542/pir.31-8-320
- Ojo A. Foreign bodies: ear and nose. UptoDate. 2012. Available online only for subscription.

Item 231

A medical student you are supervising during a summer rotation is evaluating a 3-year-old who has asthma. The child has been well this summer with no use of albuterol. During the past year, the child had 4 episodes of wheezing: I in the fall, 1 in the winter, and 2 in the spring. Each episode responded to albuterol and an oral corticosteroid. The last episode occurred 6 weeks ago; there have been no hospitalizations. During the fall, winter, and spring, the child has daytime symptoms about 3 times a week and nighttime symptoms about twice a month. You refer to the 2007 National Institutes of Health Guidelines for the Diagnosis and Management of Asthma summary.

Of the following, you are MOST likely to categorize this child's disease as

A. intermittent asthma, no impairment, and mild risk
B. intermittent asthma, no impairment, and moderate risk
C. mild persistent asthma, mild impairment, and moderate risk
D. moderate persistent asthma, mild impairment, and moderate risk
E. seasonal asthma, mild impairment, and moderate risk

Item 231 PBLI Preferred Response: C

The child described in the vignette has mild persistent asthma, mild impairment, and moderate risk. The child has had more than 2 wheezing episodes requiring steroid bursts, daytime symptoms more than twice per week, and nocturnal symptoms approximately twice a month. The National Heart, Lung, and Blood Institute (NHLBI) guidelines 2007 (Item C231, page C-182) clearly outlines the classification of asthma severity, impairment, and risk in children from birth to 4 years of age.

Asthma is a complex, chronic, inflammatory disorder of the airways characterized by variable and recurring symptoms, airflow obstruction, and bronchial hyperresponsiveness. The key elements of assessment and monitoring in the 2007 NHLBI guidelines include the concepts of severity, control, and responsiveness to treatment. Severity refers to the intrinsic intensity of the disease process assessed at the initial encounter. Control refers the degree to which the manifestations of asthma (symptoms, functional impairment, and risks of untoward events) are minimized and the goals of therapy are met. Classifying severity enables initiation of appropriate therapy; assessing control is emphasized for monitoring and adjusting therapy. Asthma severity and control are defined in terms of 2 domains: impairment and risk. Impairment reflects the frequency and intensity of symptoms and functional limitations the patient is experiencing. Risk is related to the likelihood of asthma exacerbations, progressive decrease in lung function (or, for children, reduced lung growth), or risk of adverse effects from medication. The impairment domain therefore focuses on the current and ongoing quality of life and functional capacity whereas risk pertains to long-term and future events and consequences, such as exacerbations and progressive loss of pulmonary function. These domains of asthma may respond differentially to treatment and need to be factored in while striving to achieve goals of therapy.

PREP Pearls

- Assessment of severity of asthma enables institution of appropriate initial therapy; assessment of control facilitates ongoing titration of therapy to achieve asthma goals.
- The impairment domain pertains to current and ongoing functional asthma status; the risk domain reflects future untoward events such as asthma exacerbations or decrease in lung function.

American Board of Pediatrics Content Specification(s):

- Know the classifications of asthma severity and their definitions

Suggested Reading:

- National Asthma Education and Prevention Program. Expert Panel Report 3 (EPR-3): guidelines for the diagnosis and management of asthma—summary report 2007 [published correction appears in J Allergy Clin Immunol. 2008;121(6):1330]. Allergy Clin Immunol. 2007;120(5 suppl):S94-S138. doi:10.1016/j.jaci.2007.09.029
- National Heart, Lung, and Blood Institute. Expert Panel Report 3: Guidelines for the Diagnosis and Management of Asthma. National Asthma Education

and Prevention Program, Third Expert Panel on the Diagnosis and Management of Asthma. Bethesda, MD: National Heart, Lung, and Blood Institute; 2007

- National Heart, Lung, and Blood Institute. Guidelines for the Diagnosis and Management of Asthma (EPR-3). http://www.nhlbi.nih.gov/guidelines/asthma/

Components of Severity		Classification of Asthma Severity (Children 0–4 years of age)			
			Persistent		
		Intermittent	Mild	Moderate	Severe
Impairment	Symptoms	≤2 days/week	>2 days/week but not daily	Daily	Throughout the day
	Nighttime awakenings	0	1–2x/month	3–4x/month	>1x/week
	Short-acting $beta_2$-agonist use for symptom control (not prevention of EIB)	≤2 days/week	>2 days/week but not daily	Daily	Several times per day
	Interference with normal activity	None	Minor limitation	Some limitation	Extremely limited
Risk	Exacerbations requiring oral systemic corticosteroids	0–1/year	≥2 exacerbations in 6 months requiring oral steroids, or ≥4 wheezing episodes/1 year lasting >1 day AND risk factors for persistent asthma		
		Consider severity and interval since last exacerbation. Frequency and severity may fluctuate over time.			
		Exacerbations of any severity may occur in patients in any severity category			

- Level of severity is determined by both impairment and risk. Assess impairment domain by caregiver's recall of previous 2–4 weeks. Assign severity to the most severe category in which any feature occurs.
- At present, there are inadequate data to correspond frequencies of exacerbations with different levels of asthma severity. For treatment purposes, patients who had ≥2 exacerbations requiring oral corticosteroids in the past 6 months, or ≥4 wheezing episodes in the past year, and who have risk factors for persistent asthma may be considered the same as patients who have persistent asthma, even in the absence of impairment levels consistent with persistent asthma.

- **Classification of severity in patients after asthma becomes well controlled, by lowest level of treatment required to maintain control.***

Lowest level of treatment required to maintain control	Classification of Asthma Severity			
	Intermittent	Persistent		
		Mild	Moderate	Severe
	Step 1	Step 2	Step 3 or 4	Step 5 or 6

Key: EIB, exercise-induced bronchospasm

*Notes:

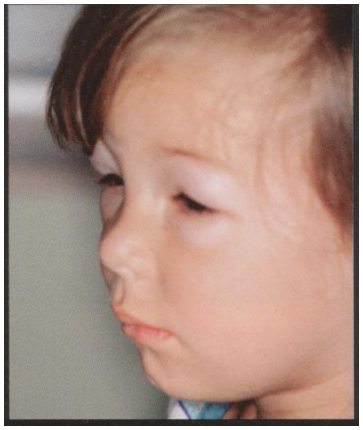
- For population-based evaluations, clinical research, or characterization of a patient's overall asthma severity after control is achieved. For clinical management, the focus is on monitoring the level of control , not the level of severity, once treatment is established.

Courtesy of the National Heart, Lung, and Blood Institute and the National Asthma Education and Prevention Program. *Expert Panel Report 3 (EPR3): Guidelines for the Diagnosis and Management of Asthma*. Bethesda, Md: U. S. Department of Health and Human Services. 2007.

ITEM C231: Classification of asthma severity in children 0-4 years of age who are currently taking long-term control medication.

Item 232

A 4-year-old boy is seen in the emergency department because of recurrent facial swelling. The mother reports that the boy has been evaluated by her pediatrician on several occasions with a similar complaint. Each time the boy was treated with 3- to 5-day courses of an antihistamine or oral steroid. The mother maintains full adherence with these treatment recommendations. Physical examination shows a healthy- appearing boy who has normal growth parameters. He is afebrile with a respiratory rate of 18 breaths/min, heart rate of 84 beats/min, and blood pressure of 90/60 mm Hg. The only finding of significance is facial puffiness and periorbital edema (Item Q232).



ITEM Q232: Periorbital edema as described for the child in the vignette.

Of the following, the MOST appropriate next step is to

A. obtain CI esterase concentration
B. obtain a specimen for urinalysis
C. prescribe a 5-day course of diphenhydramine and prednisone
D. reassure the mother and discharge the patient home
E. refer the patient for an allergy evaluation

Item 232 TE Preferred Response: B

Nephrotic syndrome should be considered in a patient presenting with facial puffiness as in the boy described in the vignette. Nephrotic syndrome is characterized by edema (facial puffiness or generalized anasarca), proteinuria, and hypoalbuminemia. Initially these patients are often mistaken as having an allergic reaction. Therefore they are often treated with a short course (3-5 days) of oral steroids or antihistamine. The initial episode of nephrotic syndrome is treated with oral steroids (60 mg/m2 per day for 4-6 weeks followed by weaning over 4-6 weeks). The short course of steroids as used for treating an allergic reaction may be associated with some or no improvement in symptoms in a patient with nephrotic syndrome. The patient in the vignette who has progressively increasing swelling needs further evaluation and the next step in the evaluation is urinalysis. Presence of proteinuria (2+ or more on dipstick test) and hypoalbuminemia will confirm the diagnosis of nephrotic syndrome.

This patient could have nephrotic syndrome as the underlying cause of his symptoms; therefore, treating him with another short course of steroids or referring him to an allergist without checking his urine is not appropriate at this time.

Hereditary angioedema (HA) is an inherited condition associated with low levels of Cl esterase inhibitor and patients typically present with recurrent episodes of nonpruritic edema without urticaria. Nearly 40% of patients with HA have the first attack before age 5 years; however, repeated attacks in preadolescent children are uncommon. Differences between nephrotic syndrome and HA are summarized (Item C232).

Item C232. Physical and Laboratory Findings in Nephrotic Syndrome and Hereditary Angioedema

Nephrotic Syndrome	Hereditary Angioedema
Pitting edema	Non-pitting edema
Periorbital swelling, generalized anasarca	Edema of lips, mouth, throat, extremities, and genitalia
Swelling worse in the morning and improves during the course of the day	Swelling is usually associated with a prodrome (tightness or tingling in the area) followed by acutely increasing swelling (over hours to days) followed by resolution
Screening; urine dipstick analysis	Screening; blood tests for both C4 and C1 esterase inhibitor.

PREP Pearls

- Nephrotic syndrome is characterized by edema (facial puffiness or generalized anasarca), proteinuria, and hypoalbuminemia.
- Patients with facial puffiness due to nephrotic syndrome are often mistaken as having an allergic reaction.

- Urinalysis is a quick and easy method to establish the diagnosis of nephrotic syndrome.
- Patients with idiopathic nephrotic syndrome are initially treated with oral steroids (60 mg/m2 per day for 4-6 weeks followed by weaning over 4-6 weeks).

American Board of Pediatrics Content Specification(s):

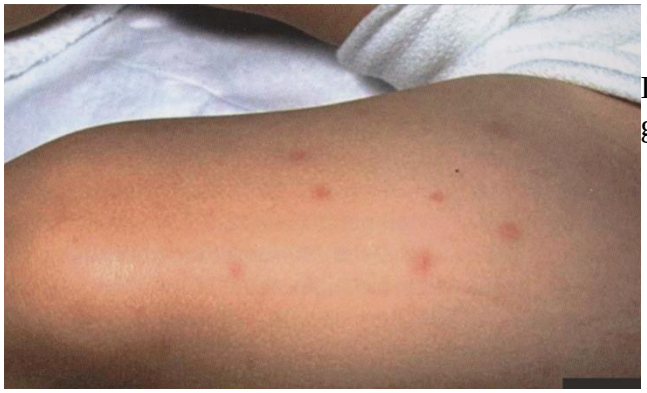
- Plan the initial treatment for a child with an initial episode of nephrotic syndrome

Suggested Reading:

- Atkins D, Frank MM, Dreskin SC, Leung DYM. Urticaria (Hives) and Angioedema. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Elsevier; 2011:811-816
- Pais P, Avner EA. Nephrotic syndrome. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:1827-1829
- Roth KS, Amaker BH, Chan JCM. Nephrotic syndrome: pathogenesis and management. Pediatr Rev. 2002;23(7):237-248. doi: 10.1542/pir.23-7-237

Item 233

A 4-year-old girl is seen in the emergency department with a 5-hour history of fever and vomiting. She tells you she "hurts all over." On physical examination, her temperature is 39°C, heart rate is 130 beats/min, respiratory rate is 40 breaths/min, and blood pressure is 90/45 mm Hg. The girl is ill-appearing and tired but answers questions appropriately. Her mucous membranes are dry, but her nares and oropharynx are normal. She is tachycardic but has no murmur, and her capillary refill is 2 to 3 seconds. Examination of the lungs, abdomen, and extremities is unremarkable. You note a rash (Item Q233, page Q-61). The peripheral white blood cell count is 25,000/pL (25.0 x 109/L), with 70% polymorphonuclear neutrophils, 20% lymphocytes, and 10% monocytes. The girl's hemoglobin level is 10 g/dL (100 g/L) and platelet count is 160 x 101/pL (160 x 109/L).



ITEM Q233: Rash as described for the girl in the vignette.

Of the following, the factor MOST closely associated with death caused by this infection is

A. leukocytosis
B. meningitis
C. pericarditis
D. serotype
E. young age

Item 233 Preferred Response: E

The ill-appearing girl in the vignette presents with fever, myalgias, rash, tachycardia, tachypnea, hypotension, and delayed capillary refill time concerning for evolving shock. The papular skin lesions and the polymorphonuclear leukocytosis suggest a bacterial infection. Given the rapid onset of illness and lack of focal findings besides the rash, early meningococcemia is likely. Mortality from meningococcemia occurs in 10% of cases and is associated with young age, hypotension, leukopenia, absence of meningitis, thrombocytopenia, and coma. Pericarditis can occur but is not associated with death caused by Neisseria meningitidis. There is no association between serotype and mortality. Invasive infection due to N meningitidis is rapid in onset and characterized by fever, chills, malaise, myalgias, and a rash that initially can be macular, maculopapular, petechial, or purpuric. Many patients are initially thought to have a viral infection. Invasive pneumococcal infection also may present in a clinically similar manner, but its incidence has notably decreased with the widespread use of pneumococcal conjugate vaccine in infants and young children.

Cultures of blood and cerebrospinal fluid (CSF) are recommended for patients with suspected invasive meningococcal disease. A Gram stain from cultures of blood or CSF typically reveals gram-negative (pink) diplococci. Empirical therapy with cefotaxime or ceftriaxone is recommended for patients with presumed sepsis due to meningococcemia because coverage for Streptococcus pneumoniae is warranted, pending culture results. Once the diagnosis of meningococcemia is established, intravenous penicillin G (250,000- 300,000 U/ kg per day divided every 4-6 hours) is the drug of choice. Meningococcal isolates with decreased susceptibility to penicillin have been identified in the United States but mostly occur in Spain. Because these isolates remain moderately susceptible to penicillin, penicillin at high doses for 5 to 7 days is adequate for the treatment of invasive meningococcal disease. Prompt treatment of shock with intravenous fluids, and inotropic or ventilatory support when needed, may be critical in improving survival.

PREP Pearls

- Invasive infection due to N meningitidis is rapid in onset and characterized by fever, chills, malaise, myalgias, and a rash that initially can be macular, maculopapular, petechial, or purpuric.
- Mortality from meningococcemia occurs in 10% of cases and is associated with young age, hypotension, leukopenia, absence of meningitis, thrombocytopenia, and coma.
- High-dose intravenous penicillin G for 5 to 7 days is the preferred treatment for infection due to N meningitidis.

American Board of Pediatrics Content Specification(s):

- Plan the treatment of a Neisseria meningitidis infection

Suggested Reading:

- American Academy of Pediatrics. Meningococcal infections. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:500-509
- Centers for Disease Control and Prevention. Meningococcal disease. www.cdc.gov/meningococcal

Item 234

A 16-year-old girl reports a history of irregular menses. Her periods have become progressively less frequent since menarche at 12 years of age. Her last menstrual period was 4 months ago. She is not sexually active and has had no galactorrhea. She is taking college-level classes in school, preparing for her college-entrance exams, and helping care for her sick mother. On physical examination, her vital signs are normal and her body mass index is 31. There are inflammatory acne lesions on her face and back, hyperpigmented velvety-appearing skin at the nape of her neck, and increased body hair. Her thyroid gland is not enlarged, and the diameter of her clitoris is 4 mm.

Of the following, the condition MOST likely responsible for this girl's symptoms and signs is

A. dysfunctional uterine bleeding
B. emotional stress
C. hypothyroidism
D. pituitary adenoma
E. polycystic ovary syndrome

Item 234 Preferred Response: E

Secondary amenorrhea is defined as an absence of menses for 3 or more months that occurs at least 24 months after menarche, when an ovulatory menstrual pattern should be established. The girl in the vignette underwent menarche 3 years ago and has irregular menses, along with acanthosis nigricans, obesity (body mass index >30), acne, and some increase in body hair, suggesting the diagnosis of polycystic ovary syndrome (PCOS). In addition to exclusion of related conditions (eg, Cushing syndrome or adrenal tumors), the 2003 Rotterdam criteria for the diagnosis of PCOS require the presence of 2 of the following 3 criteria: (1) oligoovulation or anovulation,(2) clinical or biochemical signs of hyperandrogenism, and (3) polycystic ovaries on ultrasonography. Other conditions with similar signs (eg, hyperprolactinemia, congenital adrenal hyperplasia, or Cushing syndrome) must be excluded.

Oligoovulation and anovulation present as irregular menses, and hyperandrogenism may present as acne, increased body hair, and rarely, clitoromegaly (a transverse clitoral diameter >3 mm). The extent of hirsutism can be documented using the Ferriman-Gallwey scoring system, with a score from 0 (no hair) to 4 (extensive hair growth) for each of 9 body areas most sensitive to androgens. These sites include the upper lip, chin, chest, abdomen, suprapubic region, arms, thighs, upper back, and lower back. A score of 8 or more is considered significant and suggestive of an increased androgen concentration.

The term dysfunctional uterine bleeding is reserved for postmenarchal irregularity or prolonged bleeding that occurs as a result of immaturity of the hypothalamic-pituitary axis with resultant anovulatory cycles. It lasts, on average, for 2 years. Etiologic factors for amenorrhea can be grouped under 3 anatomical compartments. The first compartment includes genital tract outlet problems. These are most frequently congenital and cause primary amenorrhea (eg, imperforate hymen). Surgical interventions causing cervical stenosis or uterine synechiae (Asherman syndrome) can result in genital tract outlet obstruction and secondary amenorrhea. The second compartment includes the ovaries. Ovarian causes may be chromosomal (eg, Turner syndrome) or nonchromosomal (eg, after chemotherapy or radiation damage). With ovarian causes, the gonadotropin (follicle-stimulating hormone and luteinizing hormone) levels are elevated, and the condition is referred to as hypergonadotropic hypogonadism. Although most commonly associated with primary amenorrhea, these patients may have some functioning ovarian tissue and present with secondary amenorrhea. The last compartment is the hypothalamic-pituitary axis and is associated with normal or lowered gonadotropin levels (ie, hypogonadotropic hypogonadism).

The most common conditions causing amenorrhea in an adolescent, after ruling out pregnancy, include stress, excessive exercise, and weight changes. Organic causes include those that are reversible, such as chronic illness associated with stress and weight loss, endocrinopathies (eg, thyroid disease and prolactin and cortisol excess), medications, and drugs. Irreversible organic causes include head trauma, radiation, and tumors. Pituitary adenomas are often microadenomas and are asymptomatic. They do not cause hirsutism or acanthosis nigricans but can be a cause of secondary amenorrhea

without galactorrhea. Therefore, a prolactin level should be part of the evaluation. Extensive laboratory studies are warranted in patients who present with signs of virilization. Treatment of secondary amenorrhea requires addressing the underlying condition.

PREP Pearls

- Irregular menstrual bleeding that persists for 2 years after menarche is unlikely to be the result of physiologic anovulation.
- PCOS, the most common endocrine condition in adolescents, is often the cause of irregular menstrual bleeding.
- PCOS increases the risk of type 2 diabetes mellitus and cardiovascular disease in the future.

American Board of Pediatrics Content Specification(s):

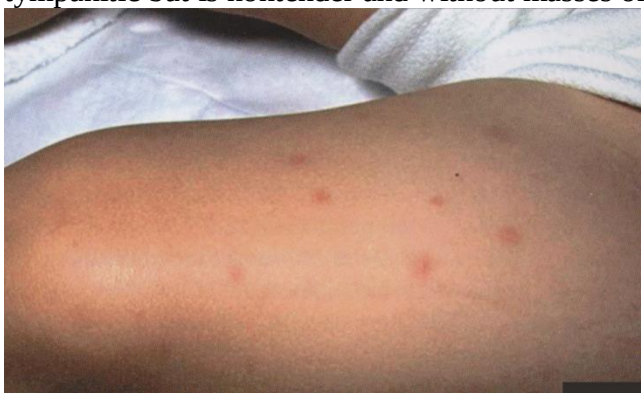
- Know the etiologies of secondary amenorrhea and their respective treatments

Suggested Reading:

- Bloomfield D. Secondary amenorrhea. Pediatr Rev. 2006;27:113-114. doi:10.1542/pir.27-3-113
- Emans SJ. Amenorrhea in the adolescent. In: Emans SJ, Laufer MR, Goldstein DP, eds. Pediatric and Adolescent Gynecology. Philadelphia, PA: Lippincott Williams & Wilkins; 2005:214-269
- Fleishman A, Gordon CM, Neinstein LS. Menstrual disorders: amenorrhea and the polycystic ovary syndrome. In: Neinstein LS, Gordon CM, Katzman DK, Rosen DS, Woods ER, eds. Adolescent Health Care: A Practical Guide. 5th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2008:691-705
- Goldstein MA, Dechant EJ, Beresin EV. Eating disorders. Pediatr Rev. 2011;32:508-521. doi:10.1542/pir.32-12-508

Item 235

A 4-year-old boy is referred to your office from a nearby summer day camp. He has been complaining of crampy abdominal pain, flatus, and intermittent loose bowel movements for the past 2 weeks. His height and weight are at the 50th percentile, and a review of his medical records indicates that he has not lost any weight since camp began. While at camp, he and his fellow campers take a nap together in one cabin and use a common bathroom. None of his other bunkmates have experienced similar symptoms. His recent dietary history includes 3 to 4 glasses of apple juice per day. He reluctantly drinks 1 glass of milk every day. On physical examination, his abdomen appears mildly distended and tympanitic but is nontender and without masses or organomegaly



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Of the following, the MOST likely cause of this boy's symptoms is

A. cryptosporidiosis
B. excessive sorbitol intake
C. giardiasis
D. lactase deficiency
E. sucrase-isomaltase deficiency

Item 235 Preferred Response: B

The boy described in the vignette presents with symptoms suggestive of carbohydrate malabsorption. Because he consumes large amounts of apple juice daily, the most likely cause of his symptoms is sorbitol-induced osmolar diarrhea. Item C235 lists the most common dietary products that contain significant quantities of this poorly absorbed sugar alcohol.

All orally ingested nutrients (eg, fats, carbohydrates, proteins, vitamins, and minerals) are absorbed in the small intestine, and the bulk of this absorption takes place in the proximal jejunum. Specific absorptive processes vary, depending on the nutrient class, and involve the contribution of pancreatic hydrolases (amylase for starch, proteases for proteins, and lipase for triglycerides), bile salts (micellar solubilization of long-chain fats and fat soluble vitamins), and intestinal microvillus membrane hydrolytic enzymes (disaccharides and some oligosaccharides and oligopeptides) and transport proteins (monosaccharides, amino acids, dipeptides, and tripeptides). When carbohydrates are malabsorbed, the osmolar load produced by the high amount of osmotically active carbohydrate moieties, most commonly disaccharides (eg, secondary to disaccharidase deficiency) and/or partially digested starches (hypoamylasemia), results in symptoms of intestinal distention, rapid peristalsis, and diarrhea. Bacterial fermentation of unabsorbed sugars produces excess quantities of volatile short chain fatty acids and gases, including carbon dioxide, hydrogen, and methane. The colon will salvage some of the short chain fatty acids. The remainder exert an additional osmolar load, contributing to diarrhea. Symptoms comprising meal-related abdominal pain, flatulence, and diarrhea secondary to carbohydrate malabsorption are more common in children than in adults.

Although symptomatic carbohydrate malabsorption may be the consequence of impaired or incomplete microvillus membrane uptake of monosaccharides (eg, sorbitol, fructose, or glucose), it most frequently results from either absent or decreased small intestinal disaccharidase activity. In all cases, symptoms occur within a few hours of ingesting the offending carbohydrate and include nausea, abdominal pain and distension, vomiting, diarrhea, and flatulence. Loss of enzyme activity may result from intestinal injury (eg, prolonged gastroenteritis, celiac disease, radiation, or starvation) or from a genetically programmed deficiency. Carbohydrate (and fat) malabsorption also occurs in conditions that impair adequate nutrient intraluminal and/or mucosal hydrolysis and uptake, including motility disorders and other disease states, leading to bacterial overgrowth.

Of the digestive hydrolases, lactase activity demonstrates the greatest sensitivity to small intestinal villus epithelial cell damage. Late primary lactase deficiency, a genetically preprogrammed phenomenon, occurs in up to 85% of individuals with non-Northern European ancestry. In patients with hypolactasia, loss of enzyme activity is usually detected in the second half of the first decade of life. In the vignette, the boy's age of 4 years, his excellent nutritional status, and absence of any chronic disorder that might predispose him to hypolactasia make this disorder unlikely. The other major disaccharidase is sucrase-isomaltase (SI). Unlike lactase, SI activity is relatively well preserved under conditions of intestinal villus injury. Accordingly, sucrose may be tolerated as a dietary substrate in many patients with prolonged or severe gastrointestinal

illnesses. However, of the disaccharidase deficiency states, SI represents the most common congenital disorder, despite a prevalence in North America of only less than 0.2%. Symptoms develop during early infancy in individuals whose diet includes formulas that contain sucrose or glucose oligosaccharides. In other affected individuals, symptoms develop with the introduction of these carbohydrates to the diet, commonly in the form of starches, fruits, and fruit juices.

In terms of other possible diagnoses in this clinical setting, infectious disorders should also be considered. Both cryptosporidiosis and giardiasis are potential pathogens associated with chronic, nonbloody diarrhea. Although infection with Cryptosporidium was once thought to be limited to those who were immunosuppressed, the disorder has emerged with increasing frequency in immunocompetent individuals. In almost all cases, this infection arises from a contaminated water supply. Giardiasis, as well, develops most commonly as the consequence of consuming contaminated drinking water, often from streams and lakes. The fact that no other children residing with the boy in the vignette are similarly affected, however, makes these infections unlikely.

Item C235. Sorbitol Content of Various Dietary Products

• Sugar-free gum	• 1.3–2.2 g per piece
• Sugar-free mints	• 1.7–2.0 g per piece
• Pears	• 4.6 g/100 g
• Pear juice	• 2.1 g/100 mL
• Prunes	• 2.4 g/100 g
• Prune juice	• 12.7 g/100 mL
• Peaches	• 1.0 g/100 mL
• Cherry juice	• 1.4 g/100 mL
• Apple juice	• 0.5–0.9 g/100 mL

PREP Pearls

- Consumption of large amounts of sorbitol-containing foods (particularly fruit juices) is a major cause of diarrhea in young children.
- Lactose intolerance is extremely uncommon in the first few years of life, unless it is associated with intestinal injury.
- In previously well infants presenting with watery diarrhea, a careful dietary history will suggest whether sucrase-isomaltase deficiency (SID) is a likely diagnosis (Remember: patients with SID will also maldigest glucose polymers).

American Board of Pediatrics Content Specification(s):

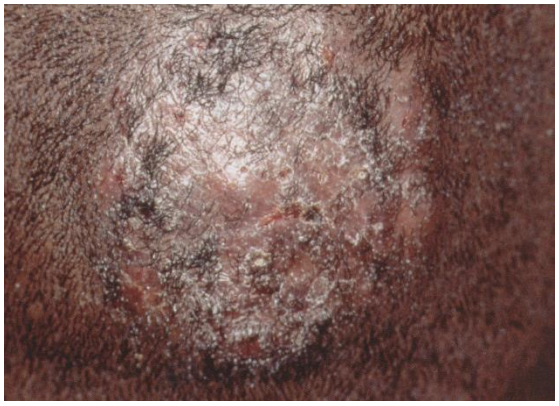
- Recognize the symptoms of a carbohydrate malabsorption disorder

Suggested Reading:

- Born P. Carbohydrate malabsorption in patients with non-specific abdominal complaints. World I Gastroenterol. 2007;13:5687-5691
- Guandalini S, Dincer AP. Nutritional management in diarrheal disease. Baillieres Clin Gastroenterol. 1998;12:697-717. doi:10.1016/S0950-3528(98)90004-5
- Gudmand-Hoyer E, Skovbjerg H. Disaccharide digestion and maldigestion. Scand I Gastroenterol Suppl. 1996;216:111-121
- Kneepkens CM, Hoekstra JH. Chronic nonspecific diarrhea of childhood: pathophysiology and management. Pediatr Clin North Am. 1996;43:375-390
- Robayo-Torres CC, Quezada-Calvillo R, Nichols BL. Disaccharide digestion: clinical and molecular aspects. Clin Gastroenterol Hepatol. 2006;4:276-287. doi:10.1016/j.cgh.2005.12.023

Item 236

You are evaluating an 8-year-old for hair loss and scalp swelling. The hair loss has been progressing over the past month; the swelling was noted last week. He is otherwise well and has no chronic medical conditions. Physical examination is notable for postauricular and occipital lymphadenopathy and a 5-cm area of hair loss with a boggy swelling (Item Q236) that produces pus when pressure is applied. The surface of the area is covered with scale.



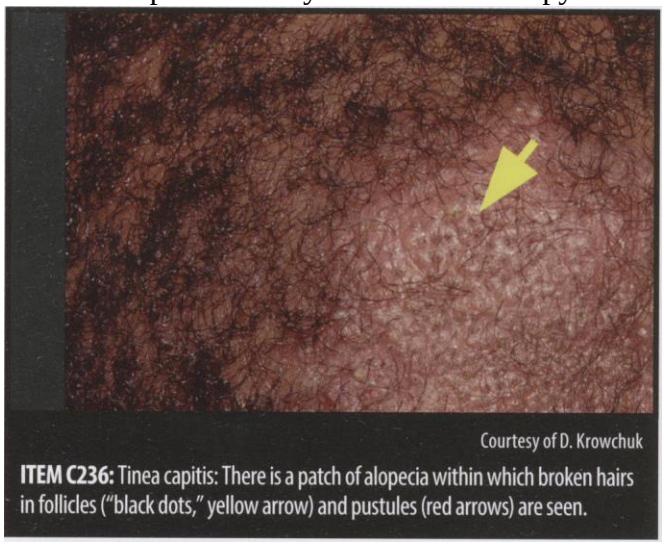
ITEM Q236: Findings as described for the boy in the vignette.

Of the following, the MOST appropriate treatment for this patient is

A. oral clindamycin
B. oral griseofulvin
C. oral ketoconazole
D. topical selenium sulfide
E. topical terbinafine

Item 236 Preferred Response: B

The patient in the vignette has a kerion, one of the manifestations of tinea capitis. Tinea capitis occurs primarily in children 3 to 9 years old. It presents in multiple ways: (1) diffuse scaling with or without hair loss; (2) "black dot" tinea with well-defined areas of alopecia and black dots on the skin where hair follicles have been broken off at the surface (Item C236); (3) scattered pustules or excoriations, often without hair loss; (4) "grey patch" areas of scaly alopecia with short hairs in the patch that provide a frosted appearance; (5) tinea favus (seen in geographic areas with malnutrition and high poverty) with yellow crusts around hairs; and (6) kerion, a boggy swelling of the scalp with alopecia, sometimes productive of pus. Most of these presentations are accompanied by occipital, posterior auricular, or cervical adenopathy, and some experts suggest that the combination of alopecia, scale, and adenopathy is diagnostic of the condition. Because treatment is prolonged, however, other experts recommend confirmation of the diagnosis with either potassium hydroxide microscopy or culture.



Courtesy of D. Krowchuk

ITEM C236: Tinea capitis: There is a patch of alopecia within which broken hairs in follicles ("black dots," yellow arrow) and pustules (red arrows) are seen.

In the Americas and Europe, Trichophyton tonsurans is the most frequent causative organism accounting for more than 90% of cases. T tonsurans is an endothrix, residing and producing spores in the hair shaft and root rather than on the surface. Therefore topical treatment alone is ineffective. Topical therapies, including terbinafine, ketoconazole, and selenium sulfate, may be helpful adjunctive therapies and may be appropriate for asymptomatic carriers. However, symptomatic patients such as the child in the vignette require systemic therapy.

Currently the Food and Drug Administration has approved 2 antifungals for use in tinea capitis. Griseofulvin has been the drug of choice for many years and has a long record of safe and effective use. However, it is poorly absorbed in the gastrointestinal tract, has a bitter taste, and requires long duration of treatment (8-12 weeks). Microcrystalline and ultramicrocrystalline preparations improve absorption, and higher dosing regimens are used to overcome resistance. Terbinafine has also proven to be an effective oral treatment for children older than 4 years. It has been generally safe with a single case of reversible neutropenia reported. A potential for hepatic toxicity has been found, and many experts recommend monitoring liver function and complete blood counts before and during treatment and avoiding use in patients with any history of liver or renal disease.

Treatment duration varies among studies, but average terbinafine treatment duration is about 4 weeks. Systemic fluconazole and itraconazole have been used successfully for treatment of tinea capitis, but oral ketoconazole is not recommended because of its high potential for hepatotoxicity.

Kerions, as seen in the patient in the vignette, are inflammatory lesions, and many practitioners are concerned about bacterial involvement. However, alopecia is rarely seen in bacterial infections, and bacterial cultures from kerions are almost always sterile. Therefore, antibiotics such as clindamycin are not recommended.

PREP Pearls

- Ninety percent of tinea capitis in the United States is caused by: T tonsurans.
- Topical therapy alone is insufficient to treat tinea capitis
- The 2 oral medications approved by the United States Food and Drug Administration for treatment of tinea capitis are griseofulvin and terbinafine.
- Kerion, a boggy, inflammatory form of tinea capitis, does not result from bacterial infection and antibiotics are not indicated.

American Board of Pediatrics Content Specification(s):

- Know that systemic therapy (eg, griseofulvin, itraconazole) is necessary to eradicate tinea capitis

Suggested Reading:

- Kelly BK. Superficial fungal infections. Pediatr Rev. 2012;33:e22-e37. doi: 10.1542/pir.33-4-e22
- Moriarty B, Hay R, Morris-Jones R. The diagnosis and management of tinea. BM/ 2012;345:e4380. doi: 10.1136/bmj.e4380
- Tey HL, Tan AS, Chan YC. Meta-analysis of randomized controlled trials comparing griseofulvin and terbinafine in the treatment of tinea capitis. I Am Acad Dermatol. 2011;64(4):663-670. doi: 10.1016/j.jaad.2010.02.048
- Tinea capitis (ringworm of the scalp). In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:712-714

Item 237

You are working in the emergency department when paramedics arrive with a 10-year-old boy who was struck in the right side of his head with a bat during a baseball game. He was initially groggy but is now awake and complaining of pain. The boy has a temperature of 37.6°C, a heart rate of 100 beats/min, and a respiratory rate of 25 breaths/min. He has no noticeable bone depression. His pupils are equal and reactive, and a small amount of blood is noted behind his right tympanic membrane. He also complains of a salty taste in his mouth. His neurological examination is nonfocal.

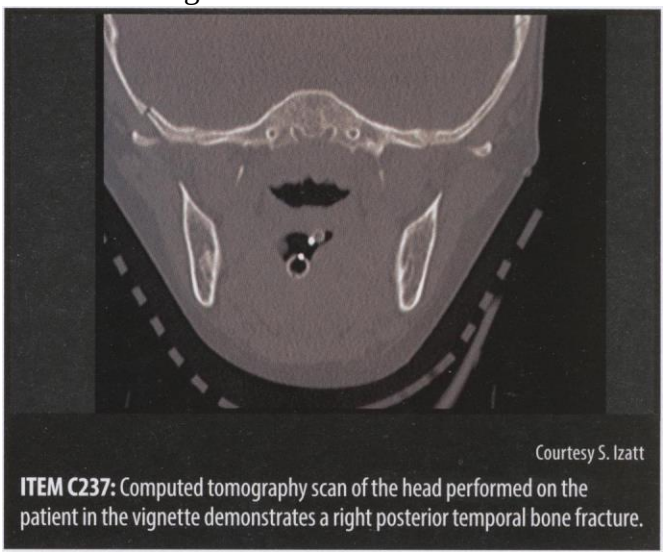
Of the following, computed tomography of the head in this boy is MOST likely to demonstrate a(n)

A. epidural hematoma
B. frontal skull fracture
C. orbital bone fracture
D. subdural hematoma
E. temporal bone fracture

Item 237 **Preferred Response: E**

A basilar skull fracture is defined as a fracture of 1 of the 5 bones that make up the base of the skull: cribriform plate of the ethmoid bone, orbital plate of the frontal bone, petrous and squamous parts of the temporal bone, sphenoid bone, or occipital bone. Owing to its relative fragility, the temporal bone is affected most often, as was the case for the boy in the vignette (Item C237).

In nearly one half of cases of basilar skull fracture, a tear occurs in the dura, resulting in cerebrospinal fluid (CSF) leak through the ear or nose. The salty taste experienced by this boy likely is the result of CSF leaking into the nose. Most leaks resolve without treatment within a week but some persist for months. The dural defect places patients at risk for bacterial entry into the CSF system and subsequent meningitis, a complication that occurs in less than 5% of those affected. Unfortunately, the risk of meningitis is not reduced with the use of prophylactic antibiotics. Other complications of basilar skull fracture include hearing loss and cranial nerve deficits.



Courtesy S. Izatt

ITEM C237: Computed tomography scan of the head performed on the patient in the vignette demonstrates a right posterior temporal bone fracture.

In addition to CSF leak, clinical signs of a basilar skull fracture include ecchymosis behind the ear (Battle sign) or around the eyes (raccoon eyes) arising a few days after injury. Hemotympanum, the presence of blood behind the tympanic membrane, is usually a sign of a temporal bone fracture and arises within a few hours of injury. Epidural and subdural hematomas can occur in association with basilar skull fractures, especially epidurals in the case of temporal bone fractures arising from the close proximity of the middle meningeal vessels. They are not specifically associated with a hemotympanum or CSF leakage.

PREP Pearls

- A basilar skull fracture is defined as a fracture of 1 of the 5 bones that make up the base of the skull.
- Complications of a basilar skull fracture include cerebrospinal fluid (CSF) leaks, hearing loss, and cranial nerve deficits.
- The risk of meningitis in a CSF leak is estimated to be less than 5% and is not reduced by prophylactic antibiotics.

American Board of Pediatrics Content Specification(s):

- Understand the significance of blood behind the tympanic membrane

Suggested Reading:

- Caviness AC. Skull fractures in children. UptoDate. Available online only for subscription
- Heegaard WG, Biros MH. Skull fractures in adults. UptoDate. Available online only for subscription

Item 238

A 5-month-old male infant presents to your office for evaluation of a 1 -week history of constipation, poor feeding, loss of appetite, and copious drooling. The mother also feels he has been weaker and more "floppy" over the past few days. She first noted the weakness in the arms and then in his legs. His birth history and past medical history are unremarkable. The child has been breastfed, and, over the past few weeks, they have introduced carrot puree prepared at home to his diet. He is afebrile, and examination is notable for decreased tone, weak cry, and a soft abdomen with distension and palpable stool.

Of the following, the MOST likely diagnosis for this patient is

A. Guillain-Barre syndrome
B. Hirschsprung disease
C. infant botulism
D. intussusception
E. spinal muscular atrophy

Item 238 S Preferred Response: C

Constipation, poor feeding, drooling, progressive descending weakness, and hypotonia, as described in the infant in the vignette, are signs and symptoms of infant botulism. Infant botulism resulting from intestinal colonization with Clostridium botulinum is presently the most common form of botulism reported in the United States with approximately 75 cases per year. Although previously associated with ingestion of raw honey, most cases in the United States are now associated with soil containing high botulism spore concentrations or home preparation of food contaminated with C botulinum. Infant botulism presents in the first year after birth with a median age of 3 to 4 months at presentation.

Guillain-Barre syndrome is extremely rare at this young age and generally presents with ascending paralysis. Hirschsprung disease may be associated with constipation but the other findings of progressive weakness, hypotonia, and drooling are not consistent with this diagnosis. The infant with intussusception may present with lethargy but typically in a more acute time frame and without systemic weakness or drooling. Spinal muscular atrophy is a genetic disorder that presents with chronic diffuse muscle weakness, not descending weakness, and would not be rapidly progressive as noted in the vignette. The diagnosis of infant botulism is confirmed by the identification of botulinum toxin from stool specimens. Isolation of C botulinum spores on stool culture supports the diagnosis. Findings on electromyography suggestive of infant botulism include abnormal incremental response to repetitive nerve stimulation and short duration, low-amplitude motor potential. Such findings may be helpful in supporting the diagnosis of infant botulism while awaiting the results of stool studies.

Meticulous supportive care is the mainstay of successful treatment of infant botulism. Human botulism immune globulin administered intravenously as early as possible may be beneficial. Although antibiotic therapy with penicillin or metronidazole is frequently administered for infant botulism, its benefits are unproven.
Other forms of botulism include foodborne disease secondary to ingestion of food contaminated with preformed botulinum toxin, wound botulism as described with illicit intravenous drug injection, and adult enteric infectious botulism, which is similar to infant botulism with gastrointestinal colonization with C botulinum. Aerosolized botulinum toxin or spores could theoretically be used as an agent of bioterrorism.

PREP Pearls

- Infant botulism occurs after intestinal colonization with Clostridium botulinum and is characterized by constipation, poor feeding, progressive descending weakness, and hypotonia.
- Diagnosis of infant botulism is made by identification of botulinum toxin in stool specimens.
- Meticulous supportive care is the mainstay of treatment of infant botulism. Human botulism immune globulin administered early in the course of disease may be beneficial.

American Board of Pediatrics Content Specification

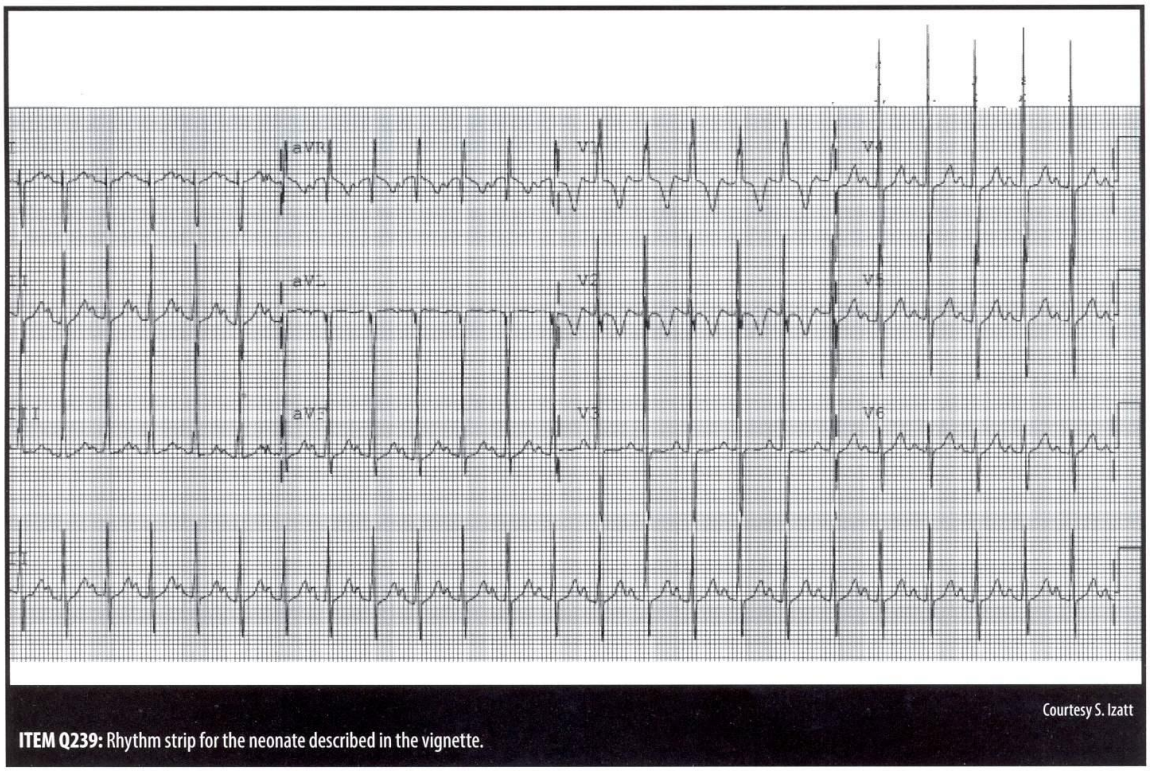
- Recognize the clinical manifestations of botulism

Suggested Reading:

- American Academy of Pediatrics. Botulism and infant botulism
- (Clostridium botulinum). In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:2E284
- Arnon SS, Schechter R, Maslanka SE, et al. Human botulism immune globulin for the treatment of infant botulism. N Engl I Med. 2006;354:462471. doi:10.1056/NEJMoa051926
- Pegram PS, Stone SM. Botulism. UptoDate. 2013. Available online only for subscription

Item 239

You walk into a mother's room to perform a newborn discharge examination at 72 hours after birth to find a mottled infant with cool extremities. The term newborn is appropriate for gestational age and was born by scheduled repeat cesarean section following an unremarkable pregnancy. Vitals signs include a temperature of 36.6°C, heart rate of 180 beats/min, respiratory rate of 74 breaths/min, blood pressure of 50/35 mm Hg (mean blood pressure, 38 mm Hg), and oxygen saturation of 90% on room air. The examination is notable for diffuse mottling, a capillary refill of 5 to 6 seconds, tachycardia with a single S2 and no murmur, liver down 2 centimeters below the right costal margin, and weak pulses. A rhythm strip is obtained (Item Q239).



ITEM Q239: Rhythm strip for the neonate described in the vignette.

Of the following, the MOST likely cause of this infant's presentation is

A. cardiac tamponade
B. cardiomyopathy
C. hypoplastic left heart syndrome
D. myocardial infarction
E. supraventricular tachycardia

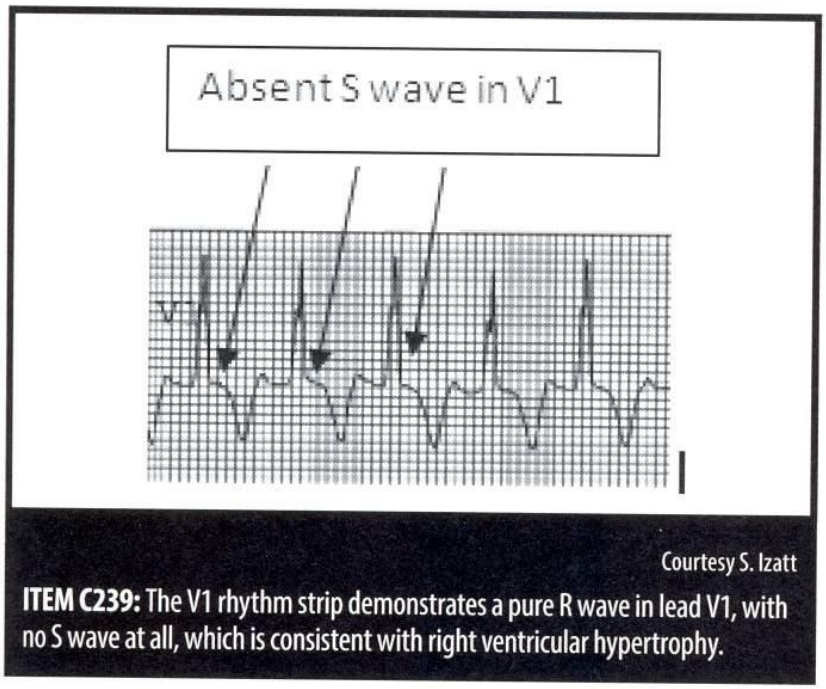
Item 239 Preferred Response: C

A neonate who presents within the first week after birth in cardiogenic shock with an echocardiogram lacking reciprocal forces in lead V1 (ie, no S wave in lead VI) most likely has hypoplastic left heart syndrome. Cardiogenic shock is caused by primary failure of the heart, which leads to a reduction in cardiac output and systemic hypotension. The clinical findings in neonates with cardiogenic shock include mottling, weak pulses, delayed capillary refill, low blood pressure, tachycardia, tachypnea, and hepatomegaly. Causes of cardiogenic shock in the neonate include left-sided critical congenital heart disease (hypoplastic left heart syndrome, critical coarctation of the aorta, interrupted aortic arch, critical aortic valve stenosis), cardiac muscle disorders (intrapartum asphyxia, myocarditis), dysrhythmias (supraventricular tachycardia, complete heart block), and rare metabolic conditions. Rapid cardiorespiratory stabilization is necessary, often before a diagnosis is made.

Forms of critical congenital heart disease that present with cardiogenic shock often have inadequate left ventricular development. These neonates rely on right ventricular function to eject blood through the patent ductus arteriosus to maintain systemic blood flow. The patent foramen ovale at the atrial level allows oxygenated blood returning from the lungs to shunt left to right and subsequently mix with the desaturated systemic blood returning from the body. This mixture allows relatively high oxygen saturation in the blood leaving the right ventricle and passing through the patent ductus arteriosus, preventing the infant from appearing cyanotic. Closure of the ductus decreases blood flow to the entire circulation (hypoplastic left heart syndrome, critical aortic valve stenosis) or to the lower body (critical coarctation of the aorta, interrupted aortic arch), leading to hypotension and poor perfusion. If ductal-dependent critical congenital heart disease is suspected in a neonate with cardiogenic shock, prostaglandin El should be given during the resuscitation to re-establish ductal patency.

Affected neonates have very few clinical examination findings until the ductus arteriosus closes. Careful cardiac examination may reveal a single and loud S2, an active precordium, and normal femoral pulses. Because ductal closure may not occur until after the neonate has been discharged from the hospital, congenital heart disease screening is now recommended before discharge. Screening of oxygen saturations in the right hand (preductal) and a lower extremity (postductal) permits identification of neonates with hypoplastic left heart syndrome and interrupted aortic arch as well as some with coarctation of the aorta.

Neonates with cardiac tamponade, cardiomyopathy, myocardial infarction, and supraventricular tachycardia can all present with cardiogenic shock. The presence of a single S2 in the neonate in the vignette suggests hypoplastic left heart syndrome. In addition, the V1 rhythm strip demonstrates a pure R wave in lead V1, with no S wave at all, which is consistent with right ventricular hypertrophy (RVH) (Item C239).
Absent S wave in VI.



ITEM C239: The V1 rhythm strip demonstrates a pure R wave in lead V1, with no S wave at all, which is consistent with right ventricular hypertrophy.

This particular pattern of RVH is typical in hypoplastic left heart syndrome, unlike the upright T wave seen in patients with RVH for other reasons. The neonate described in the vignette does not have muffled heart sounds or ST elevation on the V1 rhythm strip, which makes cardiac tamponade unlikely. The clinical examination and V1 rhythm strip of the neonate in the vignette are not consistent with supraventricular tachycardia, because the heart rate of a neonate with supraventricular tachycardia is typically greater than 220 beats per minutes. The V1 rhythm strip also does not demonstrate ST wave changes suggestive of myocardial ischemia that can be seen with myocardial infarction and cardiomyopathy. Both are extremely rare in the neonatal period.

PREP Pearls

- Neonates with left-sided congenital heart disease, including hypoplastic left heart syndrome, critical coarctation of the aorta, Interrupted aortic arch, and critical aortic valve stenosis, may present with cardiogenic shock.

American Board of Pediatrics Content Specification(s):

- Know that cardiogenic shock may be the initial finding in a newborn infant with congenital heart disease

Suggested Reading:

- Balakrishnan PL, Juraszek AL. Pathology of congenital heart disease. NeoReviews. 2012;13:e703-e710. doi: 10.1542/neo.13-12-e703.
- Lees MH, King DH. Cardiogenic shock in the neonate. Pediatr Rev. 1988;9:258-266. doi: 10.1542/pir.9-8-258
- Silberbach M, Hannon D. Presentation of congenital heart disease in the neonate and young infant. Pediatr Rev. 2007;28:123-131. doi:10.1542/pir.28-4-123

Item 240

A 4-year-old boy is brought to your office for leg pain. Over the past month, he has been more irritable and frequently asks to be carried. He has also been taking longer naps. The mother reports that he had intermittent, low-grade fevers (up to 38.6°C) with no upper respiratory or gastrointestinal symptoms. In the office, his oral temperature is 37.7°C, pulse rate is 100 beats/min, respiratory rate is 22 breaths/min, and blood pressure is 90/60 mm Hg. On examination, the child is in no apparent distress. There are scattered petechiae on his face and neck. There is no swelling, warmth, or tenderness in his joints or deformity in his legs. He has no hepatosplenomegaly or lymphadenopathy. The remainder of the physical examination is normal. The following are the results of the child's complete blood cell count:

- White blood cell count, 3,200/µL (3.2×10^9/L), with 5% polymorphonuclear leukocytes, 88% lymphocytes, 5% monocytes, and 2% eosinophils; peripheral blood smear showed normal morphology
- Hemoglobin, 9.0 g/dL (90 g/L)
- Mean corpuscular volume, 90/μm^3 (90 fL)
- Platelet count, 18×10^3/µL (18×10^9/L)

Of the following, the MOST appropriate next step in management is to

A. administer γ-globulin intravenous
B. obtain bone marrow studies
C. obtain radiographs of lower extremities
D. prescribe a course of prednisone
E. reassure parents that the findings are likely related to a viral illness

Item 240 Preferred Response: B

The boy described in the vignette who has fatigue, fever, leg pain, and pancytopenia should be evaluated for leukemia, despite the absence of blasts in the peripheral blood. Leukemia accounts for 30% of childhood cancers, making it the most common pediatric malignant disease. It results from a proliferation of malignant hematopoietic cells that occurs within the bone marrow and can involve other organs. Leukemic cell expansion in the bone marrow can produce severe bone pain, which is reported in 25% of patients with newly diagnosed leukemia. Malignant leukemic blasts are often present in the peripheral blood but may not be in some cases. Marrow infiltration by the leukemia cells leads to impaired production of normal leukocytes, erythrocytes, and platelets, resulting in pancytopenia. Leukemia can present with leukopenia or leukocytosis. Leukemic infiltration can occur in the liver, spleen, and lymph nodes, leading to organomegaly and lymphadenopathy.

The prognosis for pediatric leukemia has improved tremendously during the past 50 years. The 5-year survival rate for the most common pediatric leukemia, acute lymphoblastic leukemia, is now greater than 80%. Patients receive risk-stratified treatment to achieve comparable outcomes with decreased toxic effects. Those who have high-risk disease or have failed conventional chemotherapy may require hematopoietic stem cell transplantation.

This child presents with pancytopenia with no malignant blasts reported on the peripheral smear. The patient also has leg pain and constitutional symptoms. After obtaining the complete blood cell count, the evaluation for leukemia requires a bone marrow aspiration and biopsy, chest radio-graph (to evaluate for a mediastinal mass), and laboratory evaluation for possible disease complications, such as tumor lysis syndrome or coagulopathy. Intravenous γ-globulin and corticosteroids would be treatment options for immune-mediated cytopenias but not in cases of suspected leukemia. Furthermore, pretreatment with corticosteroids may cause a partial remission, which could complicate the diagnostic and treatment plan for leukemia. The leg pain is likely bone pain secondary to marrow infiltration by the leukemic cells; therefore, radiographs of the legs are not necessary. Viral infections can cause transient cytopenias; however, it is a diagnosis of exclusion that can only be made after serious conditions, such as leukemia, are ruled out.

PREP Pearls

- Leukemic expansion in the bone marrow can produce severe bone pain.
- Malignant leukemic blasts are often present in the peripheral blood but the absence of peripheral blasts does not rule out a diagnosis of leukemia.
- The evaluation for leukemia requires a complete blood cell count, bone marrow aspiration and biopsy, chest radiograph (to evaluate for a mediastinal mass), and laboratory evaluation for possible disease complications, such as tumor lysis syndrome or coagulopathy.

American Board of Pediatrics Content Specification(s):

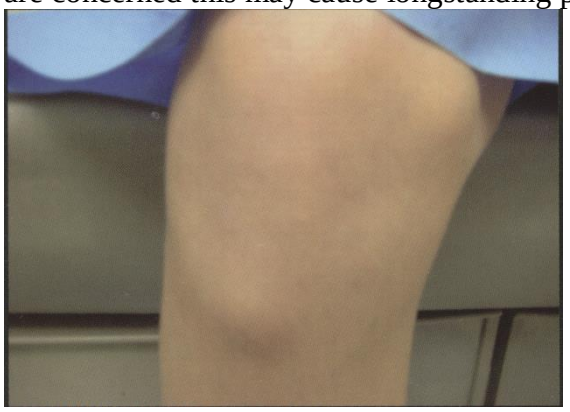
- Know that the absence of blasts in the peripheral blood of a patient with pancytopenia does not rule out the diagnosis of leukemia

Suggested Reading:

- Horton TM, Steuber CP. Overview of the presentation and classification of acute lymphoblastic leukemia in children. UptoDate. Available online only for subscription
- Hutter JJ. Childhood leukemia. Pediatr Rev. 2010;31:234-241. doi:10.1542/pir.31-6-234

Item 241

During a routine health supervision visit, a 13-year-old boy reports a 3-week history of left anterior knee pain that began during basketball practice and without any specific traumatic event. The patient reports localized swelling but denies any bruising, redness, catching, or locking. On physical examination, you note swelling and point tenderness over the tibial tuberosity (Item Q241). He does not have bruising, warmth, or erythema. He has full range of motion of the knee but reports mild pain when he actively extends the knee. The boy would like to continue his participation in basketball, but his parents are concerned this may cause longstanding problems with his knee.



ITEM Q241: Swelling of the tibial tuberosity as described for the boy in the vignette.

Of the following, the BEST next step for management is

A. complete rest from all physical activities
B. cylinder cast immobilization with unlimited weight-bearing
C. hinged knee brace with protected weight-bearing
D. home exercise program and physical activities as tolerated
E. referral to an orthopedic surgeon for arthroscopic debridement

Item 241 Preferred Response: D

In children and teenagers, the cartilaginous growth centers, the physes and apophyses, are especially vulnerable to skeletal injury. A physis is a major growth plate that contributes to long bone growth. Apophyses are "minor" growth plates or secondary centers of ossification and are located where tendons attach to bone. The boy described in the vignette has Osgood-Schlatter disease (OSD), an irritation of the tibial tubercle apophysis. The quadriceps tendon attaches to the top of the patella and the patellar tendon connects the inferior patella to the tibial tubercle. Thus, quadriceps muscle contraction puts tension on the tibial tubercle apophysis. A patellar strap, positioned over the patellar tendon, can alleviate some of the traction forces on the tibial tubercle. Quadriceps stretching may also relieve symptoms by decreasing the tension on the tibial tubercle. Ice and nonsteroidal anti-inflammatory drugs can also contribute to symptom relief. Young athletes who experience significant pain or gait alterations despite use of symptomatic treatments, stretching, and use of a patellar strap may need to take a break from physical activities. However, many children with OSD do not need to change patterns of sports participation.

A randomized controlled trial of 54 athletes with OSD compared the efficacy of hamstring stretching and quadriceps strengthening with injection of hyperosmolar dextrose (prolotherapy) at the tibial tubercle; athletes who received injections returned to sports sooner. Because most children experience symptom relief with less invasive methods, prolotherapy is not widely used in the sports medicine community. A knee immobilizer may benefit patients who do not respond to ice, nonsteroidal anti-inflammatory drugs, and stretching. Cylinder casting should not be used for OSD because of the risk of quadriceps atrophy and skin break-down that can occur if the cast slips.

Osgood-Schlatter disease typically occurs in girls between the ages of 10 and 13 years and in boys between the ages of 12 and 15 years. A prevalence of 10% to 20% in young athletes is reported. Although most patients with OSD report gradual onset of activity-related tibial tubercle pain, some have an acute onset of pain with activities such as running or jumping or with a fall onto the tibial tubercle. On physical examination, affected children generally have tenderness over the tibial tubercle and may exhibit localized swelling. Children with OSD may have pain with resisted knee extension; however, inability to actively extend the knee suggests a more severe injury, such as a tibial tubercle avulsion fracture.

The diagnosis of OSD is made clinically. Radiographs are indicated in patients with atypical signs and symptoms and are used to rule out other conditions. Fragmentation of the apophysis and soft-tissue swelling may be seen on lateral knee radiographs in individuals with OSD. While OSD symptoms typically abate within 1 to 2 years when the apophysis closes, approximately 10% of patients experience prolonged symptoms owing to development of an ossicle over the tibial tubercle. For patients with severe, persistent pain, surgical removal of the ossicle may provide relief.

PREP Pearls

- Most children and teenagers with OSD can continue to participate in sports and physical activities as pain allows.
- Some individuals with OSD have persistent symptoms even after they have completed growth at the tibial tubercle.

American Board of Pediatrics Content Specification(s):

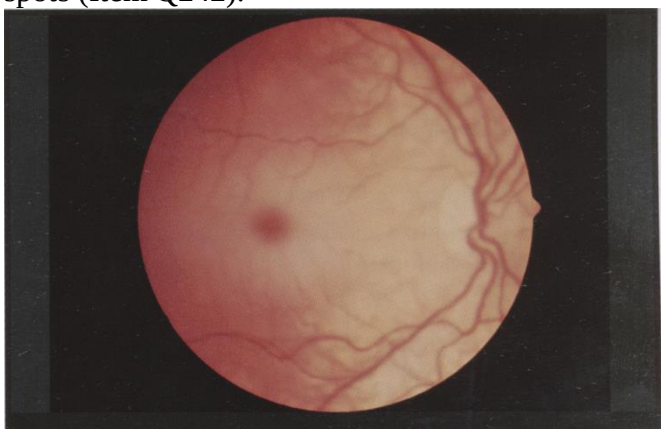
- Know the clinical manifestations and clinical course of Osgood Schlatter disease

Suggested Reading:

- Atanda A Jr, Reddy D, Rice JA, Terry MA. Injuries and chronic conditions of the knee in young athletes. Pediatr Rev. 2009 2009;30(11):419-430. doi:10.1542/pir.30-11-419
- de Lucena GL, dos Santos Gomes C, Guerra RO. Prevalence and associated factors of Osgood-Schlatter syndrome in a population-based sample of Brazilian adolescents. Am I Sports Med. 2011;39(2):415-420. doi:10.1177/0363546510383835
- Topol GA, Podesta LA, Reeves KD, Raya MF, Fullerton BD, Yeh HW. Hyperosmolar dextrose injection for recalcitrant Osgood-Schlatter disease. Pediatrics. 2011;128(5):e1121-e1128. doi:10.1542/peds.2010-1931

Item 242

A 4-month-old girl is brought in by her mother with the complaint of progressive weakness and poor head control. She has also noted that her daughter appears less attentive and has an exaggerated startle response. Results of examination are unremarkable except for diffuse hypotonia. She has a good suck and swallow, and she fixes and follows intermittently. As part of a comprehensive evaluation, she is seen by a pediatric ophthalmologist who notes slightly impaired visual acuity as well as cherry-red spots (Item Q242).



ITEM Q242: Macular spot, as described for the patient in the vignette.

Of the following, the MOST likely diagnosis for this child is

A. infantile Gaucher disease
B. Krabbe disease
C. neuronal ceroid lipofuscinosis
D. Niemann-Pick type A
E. Tay-Sachs disease

Item 242 Preferred Response: E

The infant described in this vignette has typical clinical features of infantile Tay-Sachs disease (TSD), including hypotonia, inattentiveness, an exaggerated startle response, evidence of visual dysfunction, and cherry-red spots on retinal examination. Whereas babies with infantile TSD appear normal at birth, progressive weakness may be noted between 3 and 6 months of age. Simultaneously, myoclonic jerks and an exaggerated startle reaction to sound may be seen. Between 6 and 10 months of age, there is plateauing of motor skill acquisition and even some loss of milestones. Following this period, the neurologic progression of infantile TSD is fairly rapid, with blindness, decrease in purposeful movements, and seizures noted prior to death. Macrocephaly often appears by 18 months of age but is not secondary to hydrocephalus. The so-called cherry-red macular spots result from diffuse retinal pallor secondary to accumulation of metabolic storage products in ganglion cells. The red color represents the normal foveal cells, which appear redder because of the retinal pallor in an area (ie, the fovea) that has no ganglion cells. In addition to infantile TSD, cherry-red spots are seen in many lipid storage disorders, including GM1 gangliosidosis, Sandhoff disease, some of the mucopolysaccharidoses, Niemann-Pick disease, and mucolipidoses.

The diagnosis of TSD is made by hexosaminidase A assay. Molecular genetic testing may also uncover disease-causing mutations but is not essential for making an accurate diagnosis. Three common disease-causing mutations account for more than 90% of mutations in the Ashkenazi Jewish population, but the mutations cannot be used for identification of affected individuals or carrier detection in other ethnic groups. Although TSD is found in about 1 in 3,600 individuals of Ashkenazi Jewish ancestry, it is also seen with increased frequency in individuals of French Canadian ancestry, Cajuns from Louisiana, and the Old Order Amish in Pennsylvania. Infantile TSD can actually be seen in any population, but with a frequency about 100 times less than in individuals of Ashkenazi Jewish ancestry. At present, there is no cure or enzyme replacement therapy for infantile TSD. Therefore, management is symptomatic and supportive for progressive complications as they occur.

Although cherry-red spots may also be seen in some infants with infantile Gaucher disease and Niemann-Pick type A, a child with infantile Gaucher disease or Niemann-Pick type A would typically exhibit hepatosplenomegaly. Infantile Gaucher disease is also frequently associated with pancytopenia, cranial nerve palsies, hyperreflexia, and spasticity. While neurologic regression is consistent with Krabbe disease and neuronal ceroid lipofuscinosis, in addition to infantile Gaucher disease and Niemann-Pick type A, infants with Krabbe disease often have very early onset of seizures and irritability, as well as unexplained fevers and vomiting. An infant with neuronal ceroid lipofuscinosis would typically develop microcephaly and myoclonus as well as visual failure due to retinitis pigmentosa (without cherry-red spots).

PREP Pearls

- Infants with Tay-Sachs disease appear normal at birth, but progressive weakness and an exaggerated startle response are typically noted by 4 to 6 months of age followed by a progressive deterioration.
- Tay-Sachs disease is most often seen in the Ashkenazi Jewish population, but it can and does occur in any ethnic group. Screening for Tay-Sachs carrier status in individuals who are not of Ashkenazi Jewish descent is best done through enzyme assay.

American Board of Pediatrics Content Specification(s):

- Recognize the signs and symptoms of Tay-Sachs disease

Suggested Reading:

- Bley AE, Giannikopoulos OA, Hayden D, Kubilus K, Tifft CJ, Eichler FS. Natural history of infantile GM2 gangliosides. Pediatrics. 2011;128(5):e1233-e1241. doi:10.1542/peds.2011-0078
- Kaback MM, Desnick RJ. Hexosaminidase A deficiency. In: Pagon RA, Bird TD, Dolan CR, eds. GeneReviews. Seattle, Washington: University of Washington; 2013

Item 243

A 2-year-old, previously healthy boy is rushed to the emergency department by his mother. Fifteen minutes ago, the child accidentally pulled a full pot of hot cooking oil down from the kitchen stove and onto himself, sustaining burns to his face, anterior neck, shoulders, and chest. His mother remarks that his cry sounds more hoarse than usual. On physical examination, the child is alert and crying in pain.
You note extensive second-degree burns to his face with some sloughing of his oral mucosa, in addition to second-degree burns involving a significant portion of his anterior neck and upper torso. His respiratory rate is 22 breaths/min; there are no signs of respiratory distress. You note that the boy has inspiratory stridor that persists even after he stops crying. He has normal central and peripheral pulses, with good perfusion of all of his extremities.

Of the following, the MOST appropriate next step in your management of this patient is to

A. administer a dose of nebulized racemic epinephrine
B. administer intravenous methylprednisolone
C. begin fluid resuscitation with 20 mL/kg of intravenous normal saline
D. irrigate the child's burns with cool sterile saline solution and cover with gauze
E. perform endotracheal incubation at this time

Item 243 S Preferred Response: E

The boy described in the vignette presents with signs of partial airway obstruction after sustaining significant burns to his face, the anterior aspect of his neck, torso, and oral mucosa. Securing a definitive airway by means of endotracheal intubation should be the immediate priority in his management.

Improper airway management has been implicated in the deaths of some children with severe burn injuries. Although the larynx generally protects the subglottic portion of the airway from direct thermal injury, the upper airway is extremely susceptible to obstruction due to heat exposure. In burns resulting from house fires, inhalation of hot gases can lead to progressive upper airway edema and ultimately complete obstruction. Direct exposure of airway structures to burning fluids can also result in thermal injury that may be progressive in nature and result in airway obstruction. Airway edema generally worsens in the first 24 to 48 hours after exposure to smoke or burning fluids. Patients with thermal airway injuries may not manifest signs of obstruction immediately; furthermore, early signs of partial airway obstruction from thermal injury may be subtle. Clinical signs indicating thermal injury to the airway may include hoarseness, carbon deposits and acute inflammatory changes in the oropharynx, cough, carbonaceous sputum, varying degrees of stridor, dyspnea, singed facial hairs, and altered mental status, indicating hypoxia. Burn patients displaying even subtle signs of airway compromise generally require prompt intubation due to the high potential for worsening edema and complete obstruction of the airway. Stridor is an indication for immediate endotracheal intubation. Early intubation of burn patients displaying signs of airway injury reduces the likelihood of a difficult intubation or, even worse, the need for a surgical airway later in the clinical course. Endotracheal tubes of a smaller diameter than would normally be used given a child's age should be available in anticipation of a narrowed airway. Transfer to a burn center is indicated for burn patients with inhalation injury. A definitive airway should be secured before transport.

The administration of nebulized racemic epinephrine is not routinely indicated in the treatment of patients with thermal airway injuries. Airway edema from thermal injury can be rapidly progressive; nebulized racemic epinephrine will have little effect on counteracting this edema, and administration would delay the placement of a definitive airway, which is the immediate priority in management.

Corticosteroids do not have a role in the emergency treatment of children who are displaying signs of partial airway obstruction due to thermal injury.
Although maintaining circulation through the administration of intravenous fluids is an important element in the treatment of burn patients, securing a definitive airway is the most urgent priority.

Irrigation of the child's burns is not the most important step in management at this time given that he is displaying signs of airway injury. The child's burns should be covered with warm, clean, dry linens to help prevent him from developing hypothermia and to reduce the pain associated with air currents circulating over the burns while his airway is being managed and during transport to a regional burn center.

PREP Pearls

- Edema can progress rapidly over the first 24 to 48 hours after a thermal airway injury.
- Burn patients displaying even subtle signs of airway compromise generally require prompt intubation because of the high potential for worsening edema and complete obstruction of the airway. Stridor is an indication for immediate endotracheal intubation in patients with thermal airway injury.
- Early intubation of children with signs of thermal airway injury can be life-saving because this measure reduces the likelihood of a difficult or even impossible intubation and prevents the need for a surgical airway.

American Board of Pediatrics Content Specification(s):

- Recognize airway injury in a patient with an acute burn

Suggested Reading:

- American College of Surgeons Committee on Trauma. Thermal injuries. In: Advanced Trauma Life Support for Doctors Student Course Manual. 8th ed. Chicago, IL: American College of Surgeons; 2008:211-224
- Joffe MD. Burns. In: Fleisher GR, Ludwig S, eds. Textbook of Pediatric Emergency Medicine. 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2010:1281-1288

Item 244

The mother of a 10-year-old child with asthma calls you and requests a refill of her child's albuterol inhaler. You review the patient's medical record and observe that 3 refills have been authorized in the past 3 months as well as 1 brief course of prednisone.

Of the following, the MOST appropriate course of action is to

A. authorize 1 refill and schedule a follow-up visit this week
B. authorize 3 refills to limit future refill phone calls from the mother
C. call in nebulized albuterol to ensure adequate delivery of rescue albuterol
D. call in a prescription for ipratropium
E. switch the patient to a levalbuterol inhaler to minimize adverse effects of albuterol

Item 244 SBP S Preferred Response: A

For the child in the vignette, you authorize 1 refill and schedule a follow-up visit this week. This child is at risk of having a severe adverse outcome from asthma. In individuals with asthma, the frequency of short-acting β agonist (SABA) use can be a clinical gauge of disease activity, since increasing use of SABA has been associated with increased risk for death or near death. Use of 2 or more SABA canisters per month is a risk factor for death from asthma (Item C244, page C-193). Use of more than 1 SABA canister every 1 to 2 months is also associated with an increased risk of an acute exacerbation that requires an emergency department visit or hospitalization. SABA canisters typically have 200 puffs (or 100 doses of 2 puffs) each. The 2007 National Heart, Lung, and Blood Institute (NHLBI) guidelines define good asthma control as needing SABA less than 2 times a week. A SABA canister should theoretically last for at least 10 to 12 months if used at that rate. Use of 1 canister each month would imply multiple daily SABA use and should raise concern regarding overreliance on this drug despite deterioration in asthma control. In addition, literature reports that patients who have had a near-fatal asthma exacerbation are more likely to have poor perception of airflow obstruction.

Authorizing 3 refills to limit future pharmacy calls without assessing the child's asthma control status or ability to accurately perceive symptoms may carry the risk of missing deteriorating asthma control. Ipratropium bromide treatment along with SABA treatment has been shown to have a synergistic effect in improving the outcome of moderate acute exacerbations of asthma presenting to the emergency room, but in this case, calling in a prescription for ipratropium would carry the potential of further masking worsening inflammation.

Levalbuterol is the R-racemic isomer of albuterol. In vitro studies had hinted at a possible deleterious effect of the (S)-enantiomer of albuterol on airway smooth-muscle responsiveness and other airway cells. Therefore, a product containing only the active enantiomer of albuterol (levalbuterol) was developed and approved for clinical use. Some initial clinical studies suggested an improved efficacy of levalbuterol over racemic albuterol when administered in equal (R)-albuterol doses, although other trials failed to detect any advantage of levalbuterol over racemic albuterol. Although there may be occasional patient preference in the use of nebulized albuterol, studies have shown equivalent results with delivery of albuterol via metered dose inhalers with spacers and nebulized treatments. Regardless of delivery device, use of any of the bronchodilators mentioned above at this stage would still carry the risk of masking poorly controlled asthma and would not be appropriate.

PREP Pearls

- Use of more than 2 canisters of SABA per month is a risk factor for death from asthma.
- Use of more than 1 SABA canister every 1 to 2 months is also associated with an increased risk of an acute exacerbation that requires an emergency department visit or hospitalization.

- **Asthma history**
  - Previous severe exacerbation (e.g., intubation or ICU admission for asthma)
  - Two or more hospitalizations for asthma in the past year
  - Three or more ED visits for asthma in the past year
  - Hospitalization or ED visit for asthma in the past month
  - Using >2 canisters of SABA per month
  - Difficulty perceiving asthma symptoms or severity of exacerbations
  - Other risk factors: lack of a written asthma action plan, sensitivity to Alternaria
- **Social history**
  - Low socioeconomic status or inner-city residence Illicit drug use
  - Major psychosocial problems
- **Comorbidities**
  - Cardiovascular disease Other chronic lung disease Chronic psychiatric disease

Key: ED, emergency department; ICU, intensive care unit; SABA, short-acting R2-agonist
Sources: Abramson et al. 2001; Greenberger et al. 1993; Hardie et al. 2002; Kallenbach et al. 1993; Kikuchi et al. 1994; O'Hollaren et al. 1991; Rodrigo and Rodrigo 1993; Strunk and Mrazek 1986; Suissa et al. 1994

American Board of Pediatrics Content Specification(s):

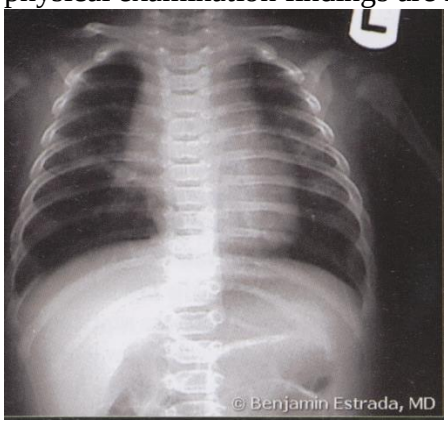
- Know that excessive daily use of beta adrenergic agonists has been associated with increased mortality and with diminished symptom control in asthma

Suggested Reading:

- Kikuchi Y, Okabe S, Tamura G, et al. Chemosensitivity and perception of dyspnea in patients with a history of near-fatal asthma. N Engl J Med. 1994;330(19):1329-1334. doi:10.1056/NEJM199405123301901
- National Heart, Lung, and Blood Institute. Expert Panel Report 3: Guidelines for the Diagnosis and Management of Asthma. National Asthma Education and Prevention Program, Third Expert Panel on the Diagnosis and Management of Asthma. Bethesda, MD: National Heart, Lung, and Blood Institute; 2007
- National Heart, Lung, and Blood Institute. Guidelines for the Diagnosis and Management of Asthma (EPR-3). http://www.nhlbi.nih.gov/guidelines/asthma/
- Qureshi F, Zaritsky A, Welch C, Meadows T, Burke BL. Clinical efficacy of racemic albuterol versus levalbuterol for the treatment of acute pediatric asthma. Ann Emerg Med. 2005;46(1):29-36. doi:10.1016/j.annemergmed.2005.02.001
- Rodrigo GJ, Castro-Rodriguez JA. Anticholinergics in the treatment of children and adults with acute asthma: a systematic review with meta-analysis [published corrections appear in Thorax. 2010;65(12):1118, Thorax. 2008;63(11):1029, Thorax. 2006;61(5):458, and Thorax. 2006;61(3):2741. Thorax 2005;60(9):740-746. doi:10.1136/thx.2005.040444

Item 245

A 2-month-old, formerly healthy term infant presents to the emergency department in December with 2 days of congestion and "breathing hard. The infant has not been eating well, but urine output—as measured by number of wet diapers—appears normal. The infant's stools have been yellow and seedy. On physical examination, his temperature is 37°C, heart rate is 140 beats/min, respiratory rate is 60 breaths/ min, and blood pressure is 80/45 min Hg. Oxygen saturation is 91% on room air. All growth parameters are at the 25th percentile for age. The infant is fussy but consolable. He has clear nasal discharge and mild nasal flaring. Auscultation of the lungs reveals coarse breath sounds bilaterally with scattered wheezing. There are mild subcostal retractions. The remainder of the physical examination findings are normal. You obtain a chest radiograph (Item Q245).



ITEM Q245: Chest radiograph for the infant in the vignette.

Of the following, the test MOST likely to establish the cause of the patient's illness is

A. acute and convalescent serologic testing
B. antigen detection in secretions
C. electron microscopy of stool for detection of viral particles
D. throat culture
E. viral culture from blood

Item 245 Preferred Response: B

The infant described in the vignette has bronchiolitis characterized by wheezing, tachypnea, and congestion, presenting during the winter months. The chest radiograph reveals hyperinflation of the lungs but no infiltrates. The most likely cause of the patient's illness is respiratory syncytial virus (RSV), which can be confirmed by rapid detection (using immunofluorescent or enzyme immunoassays) of viral antigen in nasopharyngeal secretions. Most commercially available rapid diagnostic assays are 80% to 90% sensitive.

Acute and convalescent serologic testing can be performed but is less sensitive in young infants. Electron microscopy of respiratory tract cells can reveal RSV, but this test is time-consuming and expensive. RSV would not be visualized by performing electron microscopy of stool. RSV can be isolated in culture from respiratory tract secretions, but viral culture of blood will not yield RSV. Performed appropriately, viral culture of nasopharyngeal secretions can result in viral isolation in 1 to 5 days; however, sensitivity depends on the laboratory performing the culture. A routine throat culture will not detect RSV.

Bronchiolitis most commonly occurs in children younger than 2 years, with a peak incidence between 2 and 6 months of age. More than half of RSV hospitalizations occur in infants younger than 6 months. Other than young age, risk factors for severe RSV disease include preterm birth, low birth weight, chronic pulmonary disease, cyanotic or complicated cardiac disease, neurologic disease, immunodeficiency or immunosuppression, and congenital defects of the airway.

Although RSV is the most common cause of bronchiolitis, other viruses, such as rhinovirus, parainfluenza, influenza, human metapneumovirus, and adenovirus, also cause this illness. Up to one-third of hospitalized patients have more than one viral pathogen that causes disease, but RSV is the most likely virus to be detected as a single pathogen. Respiratory syncytial virus occurs worldwide and is most common in the late fall and winter months in temperate climates.

PREP Pearls

- Rapid detection (using immunofluorescent or enzyme immunoassays) of viral antigen or viral culture performed on nasopharyngeal secretions are the best tests for diagnosing RSV.
- RSV is the most common cause of bronchiolitis.
- Bronchiolitis most commonly occurs in children younger than 2 years, with a peak incidence between 2 and 6 months of age.

American Board of Pediatrics Content Specification(s):

- Know the laboratory tests for the diagnosis of respiratory syncytial virus: culture, antigen detection

Suggested Reading:

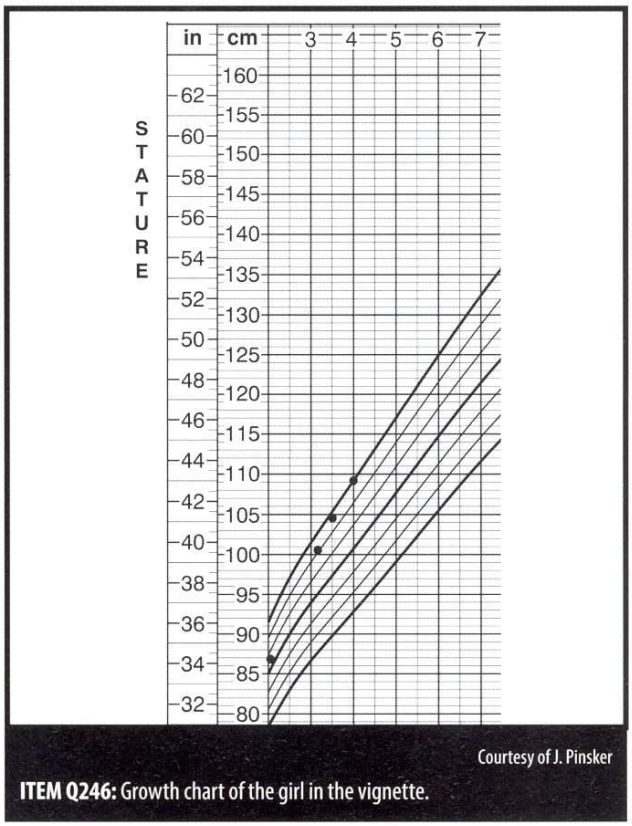
- American Academy of Pediatrics. Respiratory syncytial virus. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:609-618
- Barr FE, Graham BS. Respiratory syncytial virus infection: clinical features and diagnosis. UptoDate. Available online only for subscription

Item 246

A 4-year-old, previously healthy girl presents for evaluation of the appearance of pubic and axillary hair. Physical examination reveals Sexual Maturity Rating 3 for pubic hair but no breast development. Review of growth data shows she has had a rapid height acceleration (Item Q246). Her bone age is read as 7 years. Laboratory evaluation reveals a dehydroepiandrosterone sulfate (DHEA-S) level of 1,200 µg/dL (32.4 µmol/L), which is 10 times the upper limit of the normal range for a 4-year-old girl.

Of the following, the MOST likely cause of her early puberty is

A. adrenal carcinoma
B. benign premature adrenarche
C. hypothalamic hamartoma
D. McCune-Albright syndrome
E. ovarian granulosa cell tumor



ITEM Q246: Growth chart of the girl in the vignette.

Item 246 Preferred Response: A

The girl described in the vignette has evidence of peripheral precocious puberty due to an adrenal cortical tumor. She has rapid height acceleration, signs of androgen excess, and a very elevated dehydroepiandrosterone sulfate (DHEA-S) concentration.
Precocious puberty can be divided into 3 main categories: (1) central precocious puberty, (2) peripheral precocious puberty, and (3) normal variants.

In girls, the first sign of central precocious puberty (gonadotropin dependent) is usually breast budding before 8 years of age, which is not seen in the girl described in the vignette. In central precocious puberty, early activation of the normal pubertal system is manifested by pulsatile gonadotropin-releasing hormone (GnRH) secretion with subsequent luteinizing hormone (LH) and follicle-stimulating hormone (FSH) signaling and increased estradiol levels. A hypothalamic hamartoma is a classic lesion associated with premature activation of the central pubertal pathway. Other central nervous system (CNS) lesions associated with neurogenic dysfunction leading to central precocious puberty include pineal gland cysts or tumors and optic gliomas associated with neurofibromatosis type 1. However, any type of intracranial disturbance or underlying congenital CNS abnormality can disrupt GnRH inhibition and lead to central precocious puberty. In boys, the first clinical sign of central puberty is testicular enlargement. It is essential to recognize that human chorionic gonadotropin (HCG) can bind to the LH receptor and lead to testicular activation in the absence of pulsatile GnRH release. In boys who have signs of central precocious puberty, HCG levels should be measured and CNS imaging performed.

Peripheral precocious puberty (gonadotropin independent) is development of pubertal signs that are not from the central GnRH-mediated pathway. An example of this is androgen secretion from the adrenal glands (causing body odor, pubic hair, and signs of virilization), which is seen in congenital adrenal hyperplasia (CAH) or adrenocortical cancer (ACC), as seen in the girl in the vignette. With adrenal disease, most often there is no breast development because the adrenal glands are secreting androgens only. When children show signs of significant early androgen exposure, such as pubic hair with acne, clitoromegaly, voice deepening, or rapid height acceleration, as seen in the girl in the vignette, screening for DHEA-S (elevated in ACC), 17-hydroxyprogesterone (abnormal in the most common form of CAH), and a total testosterone level (to evaluate for an androgen-secreting ovarian tumor) is indicated. The patient in the vignette had a DHEA-S above 800 1.1g/dL (21.6 umol/L), essentially diagnostic for ACC. Any child diagnosed as having ACC should be evaluated for Li-Fraumeni syndrome (p53 mutation), and a careful family history of cancers should be obtained to consider other cancer syndromes. Other causes of peripheral precocious puberty include McCune-Albright syndrome, characterized by café au lait spots, fibrous dysplasia of the bone, and precocious puberty (often with rapid pubertal development in infancy and other signs of endocrine hyperfunctioning). A benign ovarian cyst or a granulosa cell tumor often secretes estrogens, causing suppression of LH and FSH, not seen in the girl in this vignette. Additional peripheral causes of early pubertal development include exogenous exposure to sex steroids, rarer forms of CAH, different types of ovarian or adrenal tumors, and

hepatic or CNS HCG-secreting tumors. Prolonged hypothyroidism is a very rare cause of early puberty.

Normal variants of puberty include premature thelarche in which girls younger than 2 years show some breast development but have no other signs of puberty and will not have rapid height acceleration consistent with a pubertal growth spurt. Generally, no workup is needed for premature thelarche and observation is appropriate. Benign premature adrenarche is a diagnosis of exclusion and typically causes isolated pubic hair development (without breast budding or other estrogen effects). Not all patients with signs of early adrenarche need extensive laboratory workup, but growth acceleration, other clinical signs of virilization, or markedly advanced bone age should prompt screening tests, as described above. Other normal variants include early menarche (due to an underlying genitourinary cause) and isolated pubic hair of infancy.

PREP Pearls

- Precocious puberty can be due to either a central or peripheral cause.
- DHEA-S (as opposed to DHEA) is specific for adrenal disease and is elevated with adrenal tumors.

American Board of Pediatrics Content Specification(s):

- Recognize the tumors that may produce precocious puberty (eg, in liver, CNS, ovary, testes, adrenal glands)

Suggested Reading:

- Muir A. Precocious puberty. Pediatr Rev. 2006;27: 373-381. doi:10.1542/pir.27-10-373
- Mul D, Hughes IA. The use of GnRH agonists in precocious puberty. Eur J Endocrinol. 2008;159(suppl 1):S3-S8. doi:10.1530/EJE-08-0814
- Nebesio TD, Eugster EA. Pubic hair of infancy: endocrinopathy or enigma? Pediatrics. 2006;117:951-954. doi:10.1542/peds.2005-1227
- Sperling M. Pediatric Endocrinology. 3rd ed. Philadelphia, PA: Saunders Elsevier; 2008:565-571

Item 247

You are asked to see a 3 1/2-week-old girl because of jaundice. The infant was the 3,200-g product of a full-term, uncomplicated pregnancy and delivery. She has been exclusively breastfed since birth, and "mild" jaundice was noted at the time of hospital discharge. The mother states that the infant has been feeding well but is concerned that her daughter's "eyes are still yellow." Physical examination demonstrates a vigorous infant whose weight is 3,520 g. Her skin appears jaundiced and the sclerae are icteric. Physical examination findings are normal except for a smooth liver edge palpated 1 cm below the right costal margin, with a total percussible span of 4 cm. Laboratory tests demonstrate the following:

- Hemoglobin, 13.5 g/dL (135 g/L)
- White blood cells, 9,500/µL (9.5×10^9/L)
- Total bilirubin, 9.0 g/dL (153.9 µmol/L)
- Direct bilirubin, 4.2 mg/dL (71.8 µmol/L)
- Reticulocyte count, 1%
- Aspartate aminotransferase, 85 U/L; reference range, ≤40 U/L
- Alanine aminotransferase 125 U/L; reference range, ≤30 U/L

Of the following, the MOST appropriate next test to order is

A. abdominal ultrasonography
B. hepatobiliary scintigraphy
C. percutaneous liver biopsy
D. serum αl-antitrypsin
E. serum immunoreactive trypsinogen

Item 247 Preferred Response: A

In the newborn, the presence of jaundice beyond the first 2 weeks of life, regardless of the method of feeding (ie, in the infant with suspected "breast milk jaundice"), should prompt investigation. The infant described in the vignette has direct hyperbilirubinemia, which is defined by a serum direct bilirubin concentration of more than 1.0 mg/dL (17.1 µmol/L) with a total bilirubin values of less than 5.0 mg/dL (85.5 µmol/L) or greater than 20% of the total bilirubin for values greater than 5.0 mg/dL (85.5 µmol/L). Direct hyperbilirubinemia is a sign of either obstructive or functional cholestasis and is always requires additional evaluation. The initial step in this evaluation should be to image the biliary tract by means of an abdominal ultrasound examination. This study is usually diagnostic for choledochal cyst and, in many cases, may suggest a diagnosis of extrahepatic biliary atresia (EHBA) (nonvisualization of the gall bladder and common bile duct; presence of a "triangular cord sign"). In addition to ultrasonography, laboratory investigation of the infant with suspected cholestasis will include the tests listed in Item C247A.

Cholestasis is typically a presenting feature of neonatal liver disease because of the immaturity of the hepatobiliary excretory system. Accordingly, the number of distinct disorders presenting with direct hyperbilirubinemia is greater in the neonatal period than at any other time of life. Cholestasis may result from perinatal infection, as a sign of genetic and metabolic disorders, and from biliary structural abnormalities. The major disease categories associated with prolonged neonatal cholestasis are listed in Item C247B.

Alpha 1-antitrypsin (A1AT) deficiency is an important cause of neonatal cholestasis, which may also be associated with progressive liver disease. Alpha 1-antitrypsin is the principal serum protease inhibitor. Deficiency results from a misfolding of the A1AT protein, preventing its secretion from the hepatocyte and resulting in intrahepatocellular accumulation of the mutant peptide. Clinical disease occurs in patients who have the homozygous ZZ phenotype (PiZZ) and demonstrate markedly reduced amounts of circulating A 1 AT, generally in the range of 10% to 15% of normal values. In the evaluation of neonatal cholestasis, AlAT deficiency is relatively more common than many other disorders, with the prominent exception of EHBA. In the presence of a (unequivocally) normal biliary tract ultrasonography, serum testing for A1AT deficiency is generally performed early in the evaluation of direct hyperbilirubinemia. Although the incidence of the PiZZ phenotype is approximately 1 in 2000 live births, cholestatic jaundice occurs in only approximately 10% of affected infants. These patients typically present with direct hyperbilirubinemia and hepatomegaly. Acholic stools may be present, and A1AT deficiency may be indistinguishable from the early stages of EHBA. The outcome of neonatal cholestasis relating to αl-antitrypsin deficiency varies. Although patients may develop cirrhosis in the first postnatal months, jaundice clears in most patients by 4 months of age. In children with the PiZZ phenotype who do not manifest neonatal cholestasis, signs or symptoms of liver dysfunction occur in as many as 50%, whereas cirrhosis and life-threatening disease only occur in approximately 5% before the age of 18 years. The risk of cirrhosis appears to increase with age in adults.

Evaluation for EHBA is paramount in the infant who has evidence of chronic cholestasis. Prognosis is directly related to age at diagnosis, with the best outcomes resulting when surgical correction (portoenterostomy) is performed between 30 and 45 days of age. Although some centers still use hepatobiliary scintigraphy in the evaluation of EHBA, this test is associated with a high rate of false-positive and false-negative results. Percutaneous liver biopsy remains the criterion standard in the evaluation, in advance of a definitive, intraoperative cholangiogram. Cystic fibrosis may also present with neonatal cholestasis, and a serum immunoreactive trypsinogen level would have been part of standard newborn screening tests. Further evaluation for cystic fibrosis should proceed only if the evaluation for more common causes of direct hyperbilirubinemia are excluded and if clinical suspicion exists.

Item 247A. Initial Laboratory Evaluation of the Neonate with Cholestasis

- Fractionated serum bilirubin concentration
- Liver chemical tests: alanine aminotransferase, aspartate aminotransferase, alkaline phosphatase, γ-glutamyl transferase
- Tests of liver function: serum glucose, albumin, cholesterol, coagulation studies
- Serum ammonia (if clinically indicated)
- Complete blood cell count
- Serum α1-antitrypsin level and phenotype
- Other tests if clinically indicated
    - Bacterial cultures of blood and urine
    - Paracentesis if ascites (bile leak, infection)
    - Serologic tests and cultures for viruses (TORCH agents, parvovirus B19, human herpesvirus type 6, human immunodeficiency virus)
    - Sweat chloride (check newborn screening for serum immunoreactive trypsinogen)
    - Metabolic screen (urine organic acids, serum amino acids)
    - Endocrine studies (thyroid, pituitary)
    - Urine and serum bile acid analysis
    - Specific enzyme assays (eg, galactose-1-phosphate uridyl-transferase activity)

Modified from Suchy FJ. Neonatal cholestatis. Pediatr Rev 2004;25:388-396

Item C247B. Major Diagnostic Categories of Neonatal Cholestasis

Disorder	Cases, %
Idiopathic neonatal hepatitis	15
Extrahepatic biliary atresia	25
α1-Antitrypsin deficiency	10
Genetic syndromes	25
• Progressive familial intrahepatic cholestasis • Alagille syndrome • Bile acid secretory defects	
TORCH syndrome	5
Metabolic disorders	20

PREP Pearls

- Direct hyperbilirubinemia is defined by a serum direct bilirubin concentration of more than 1.0 mg/dL (17.1 µmol/L) with total bilirubin values of less than 5.0 mg/dL (85.5 µmol/L) or greater than 20% of the total bilirubin for values greater than 5.0 mg/dL (85.5 µmol/L), and it is always an abnormal finding.
- Only 10% of infants with A1AT deficiency present with neonatal liver disease, which is associated with the Pi-ZZ phenotype.
- In the evaluation of direct hyperbilirubinemia of the neonate, the percutaneous liver biopsy remains the gold standard for suspecting biliary atresia (BA). However, since other cholestatic disorder may mimic the histologic findings of EHBA in the first month of life, this test should generally not be performed prior to 30 days after birth.

American Board of Pediatrics Content Specification(s):

- Recognize the signs and symptoms of liver disease due to alpha 1 antitrypsin deficiency

Suggested Reading:

- Nelson DR, Teckman J, Di Bisceglie AM. Brenne DA. Diagnosis and management of patients with al-antitrypsin (A1AT) deficiency. Clin Gastroenterol Hepatol. 2012;10:575-580. doi:10.1016/j.cgh.2011.12.028
- Sokol RJ, Shepherd RW, Superina R, et al. Screening and outcomes in biliary atresia: summary of a National Institutes of Health workshop. Hepatology. 2007;46:566-581. doi:10.1002/hep.21790
- Suchy FJ. Neonatal cholestatis. Pediatr Rev. 2004;25:388-396. doi:10.1542/pir.25-11-388

Item 248

The parents of a 3-week-old, formula-fed infant are concerned about increased fussiness and spitting up with every feed. The infant is growing and developing normally, has no skin or respiratory abnormalities, and has had no diarrhea or constipation. They would like to change the baby from cow milk-based formula to soy formula.

Of the following, the MOST appropriate advice about the proposed formula change is that soy formula

A. has been proven effective for the treatment of colic and spitting up
B. has been proven effective in the prevention of atopic (IgE-mediated) diseases
C. is beneficial when treating cow-milk enteropathy
D. provides better support for preterm infant growth
E. supports term infant growth in a manner equivalent to that of cow milk-based formula

Item 248 I-C Preferred Response: E

In the United States, approximately 20% of formula-fed infants consume soy formula despite limited indications for its use. It has a long history of safe use, and multiple studies demonstrate that soy formula supports normal growth and development in full-term infants and provides equivalent energy intake to cow milk-based formula. Soy formula is most appropriately used for infants who have galactosemia or primary lactase deficiency (extremely rare) who cannot tolerate the carbohydrate source (lactose) in cow milk formula. Soy formula also is recommended for families who prefer to avoid consuming animal-derived products (eg, vegan). Soy formula is not an acceptable choice for preterm infants because soy contains compounds called phytates that bind to calcium, phosphorus, iron, and zinc. Despite supplementation, these formulas are unable to meet the calcium and phosphorus needs of preterm infants, thus putting them at risk for osteopenia. Soy formulas also have markedly higher concentrations of aluminum compared with human milk, and aluminum competes with calcium for absorption. This may contribute further to osteopenia in preterm and low birth weight infants and infants with impaired renal function, but the levels are tolerated by full-term infants with normal renal function.

Many families and practitioners switch infants from cow milk to soy formula in hopes of preventing or alleviating various symptoms. Chief among these concerns are colic and immune-mediated conditions. Controlled trials do not demonstrate relief of colic with soy formula, but the increased fiber in most soy formula and sucrose in some varieties may be beneficial. Soy is not indicated for concerns of cow milk intolerance because of high cross-reactivity. Thirty percent to 64% of infants who demonstrate cow milk protein non-immunoglobulin E (IgE) enteropathy also experience soy-related enteropathy. A study of true allergic (IgE) responses revealed a rate of 0.5% for soy formula whereas cow milk allergy had a rate of 1.8%. In studies of infants with atopic dermatitis, soy allergy was present in approximately 5% of infants, and the incidence of eczema among infants at risk for atopic disease was not reduced when breastfeeding was supplemented with soy formula compared with cow milk formula. Soy formula also has been used for infants who have presumed transient lactase deficiency secondary to gastroenteritis, but its use has only limited benefit in decreasing the duration of diarrhea (6 days vs 4 days).
Soy formula contains phytoestrogens, a group of plant-derived nonsteroidal estrogens that include isoflavones. These compounds closely resemble 17-estradiol, raising concerns that they will interfere with endocrine function. Epidemiologic and animal studies are inconclusive and often contradictory. To date no definite evidence is seen of increased feminization, hypospadias, or major changes in menstrual flow among people who consumed soy formula as infants. There is concern, based on limited data, that soy formula may complicate treatment of congenital hypothyroidism. Infants fed soy formula have a prolonged increase in thyroid-stimulating hormone, and phytates may decrease uptake of exogenous thyroid hormone in these infants.

PREP Pearls

- For healthy full-term infants, soy formula supports normal growth and provides equivalent energy intake compared with cow milk formula.
- Soy formula is contraindicated for preterm infants because it does not provide sufficient calcium and phosphorus to support bone growth.
- The most appropriate indications for soy formula are in infants with galactosemia and congenital lactase deficiency (extremely rare), and when families desire to avoid all animal products in their diet.
- Because of cross-reactivity, soy formula should not be used by infants with non-IgE-mediated cow milk enteropathy.
- There are concerns that phyoestrogens in soy formula could cause endocrine abnormalities.

American Board of Pediatrics Content Specification(s)

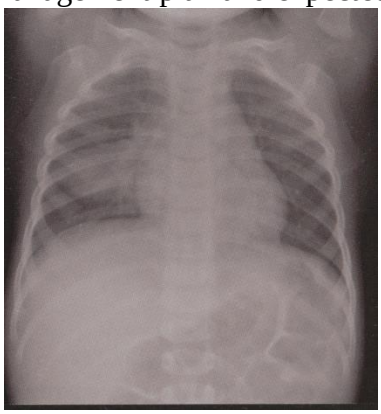
- Know the indications for the use of soy formula

Suggested Reading:

- Andres A, Cleves MA, Bellando, JB, Pivik RT, Casey PH, Badger TM. Developmental status of 1-year-old infants fed breast milk, cow's milk formula or soy formula. Pediatrics. 2012;129(6):1134-1140. doi: 10.1542/peds.2011-3121
- Bhatia J, Greer F, and the Committee on Nutrition. Use of soy protein-based formulas in infant feeding. Pediatrics. 2008;121(5):1062-1068. doi: 10.1542/peds.2008-0564
- Martinez JA, Ballew MP. Infant formulas. Pediatr Rev. 2011;32(5):179-189. doi: 10.1542/pir.32-5-179

Item 249

You are evaluating a 2-year-old boy who has a 2-day history of progressive cough, shortness of breath, and fever. His vital signs include a temperature 39.0°C, a heart rate of 120 beats/min, and a respiratory rate of 42 breaths/min. On physical examination, he has mild respiratory distress, prolonged expiration, and decreased breath sounds over the lower right side of his chest. You obtain a chest radiograph (Item Q249) and discuss the management plan and expected clinical course with the boy's parents



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ITEM Q249: Chest radiograph for the boy in the vignette.

Of the following, the MOST common complication of community-acquired pneumonia is

A. bronchopleural fistula
B. empyema
C. necrotizing pneumonia
D. pneumatocele
E. pulmonary abscess

Item 249 Preferred Response: B

Most pediatric patients who have pneumonia will recover uneventfully but a small number will develop pleural and parenchymal complications. Complications occur more commonly in patients who have bacterial pneumonia and include bronchopleural fistulas, effusion/empyema, necrotizing pneumonia, pneumatocele, and pulmonary abscess. Parapneumonic effusions are the most common complication occurring in up to 40% of bacterial pneumonias; progression to empyema occurs in more than 50% of these cases. Recent studies have shown a rise in the incidence of pediatric empyemas despite the decrease in pneumococcal pneumonia.

For patients suspected of having pneumonia, plain radiography usually is the first step in evaluation because it often confirms the clinical diagnosis and provides information about possible complications. Effusions are often seen on plain radiography with a blunting of the costophrenic angle as the initial radiographic sign. Decubitus views may better define the size of an effusion and determine if it is free-flowing or loculated, but ultrasonography is preferred because it is better able to distinguish thick purulent fluid from collapsed lung. Large pleural effusions, especially those that impair respiratory function, should be drained using video-assisted thoracoscopic surgery or thoracostomy.

Necrotizing pneumonia is associated most commonly with infections caused by Streptococcus pneumoniae, Staphylococcus aureus, and group A Streptococcus. Plain radiography may demonstrate an area of radiolucency but computed tomography is indicated to better define the area of involvement. Treatment generally consists of a prolonged course of antibiotics. Interventional procedures or surgical resection are rarely indicated and may produce additional complications such as a bronchopleural fistula.

Pneumatoceles, thin-walled cysts filled with air, normally arise as a complication of empyema and/or necrotizing pneumonia. Most pneumatoceles will resolve spontaneously with time but can rupture and produce a pneumothorax.

Pulmonary abscesses are usually associated with aspiration pneumonias but may also occur in patients who have congenital lung malformations. Symptoms are similar to those of uncomplicated pneumonia. Chest radiography may demonstrate a thick-walled cavity with an air-fluid level; computed tomography is used to confirm the diagnosis. Initial management consists of a prolonged course of antibiotics. For those who fail to respond, drainage is indicated.

Bronchopleural fistulas, abnormal connections between the bronchial tree and pleural space, typically arise from a pulmonary abscess or necrotizing pneumonia and rarely heal without intervention. The diagnosis may be suspected clinically but is confirmed with bronchoscopy. Chest computed tomography has shown promise in diagnosing and planning surgical correction of complicated cases. Management strategies include insertion or continuation of chest tubes, decreasing ventilatory pressure, differential lung ventilation, application of sealants during bronchoscopy, and surgical closure.

PREP Pearls

- Complications of pneumonia include bronchopleural fistulas, effusion/empyema, necrotizing pneumonia, pneumatocele, and pulmonary abscess, with higher complication rates seen in those patients with a bacterial etiology.
- Parapneumonic effusions are the most common complication of bacterial pneumonia, occurring in up to 40% of cases with progression to empyema in more than 50% of these cases.
- Necrotizing pneumonia is associated most commonly with infections caused by Streptococcus pneumonia, Staphylococcus aureus, and group A Streptococcus.

American Board of Pediatrics Content Specification(s):

- Know the sequelae of pneumonia and manage appropriately

Suggested Reading:

- Barson WJ. Clinical features and diagnosis of community-acquired pneumonia in children. UptoDate. Available online for subscription only.
- Barson WJ. Inpatient treatment of pneumonia in children. UptoDate. Available online for subscription only
- Bradley JS, Byington CL, Shah SS, et al. The management of community-acquired pneumonia in infants and children older than 3 months of age: clinical practice guidelines by the Pediatric Infectious Diseases Society and the Infectious Diseases Society of America. Clin Infect Dis. 2011;53(7):e2576. doi: 10.1093/cid/cir531
- Durbin WJ, Stille C. Pneumonia. Pediatr Rev. 2008;29:147-160. doi:10.1542/pir.29-5-147
- Li ST, Tanredi. Empyema hospitalizations increased in US children despite pneumococcal conjugate vaccine. Pediatrics. 2010;125:26-33. doi:10.1542/peds.2009-0184
- Winnie GB. Lossef SV. Pleurisy, pleural effusions, and empyema. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 19th ed. Philadelphia, PA: Saunders Elsevier; 2011:1505-1509

Item 250

A 3 year-old boy presents to the emergency department with a 1-day history of fever (up to 40°C), intermittent abdominal pain, and many small, bloody stools mixed with mucus. Over the last 6 hours, he has refused fluids and food and has vomited 3 times. He attends a child care center, where several other cases of gastroenteritis have been reported over the past several weeks. Two of the cases had confirmed Shigella on stool cultures. The child is assessed as 6% dehydrated and admitted for intravenous rehydration and further management. Stool and blood cultures are sent to the laboratory.

Of the following, the BEST treatment for this child is

A. ampicillin
B. ceftriaxone
C. metronidazole
D. no antibiotic therapy
E. trimethoprim-sulfamethoxazole

Item 250 Preferred Response: B

Shigella infection is characterized by high fever, diarrhea with bloody stools, abdominal cramping, and vomiting. Out-breaks in day care centers are common, and in the face of this exposure, shigellosis is the likely diagnosis for the child in the vignette. In addition to fluid resuscitation, antibiotic therapy is indicated to shorten the course of the child's illness and decrease secondary transmission.

Presently, because of increasing antimicrobial resistance in Shigella isolates, of the agents listed, ceftriaxone would be the drug of choice for treating this child. Ciprofloxacin is a potential alternative agent for treating Shigella infection but its use in children should be limited to indications for which a safer alternative is not available. Ampicillin and trimethoprim-sulfamethoxazole may be effective in treating a Shigella infection if the isolate is found to be susceptible, however, a number of isolates are now resistant to these drugs. Metronidazole is not effective against Shigella.

Antibiotic therapy in mild cases or in those whose symptoms have resolved by the time culture results become known is controversial, but in the face of the degree of acute symptoms described for the child in the vignette and the known exposure in day care, treatment would be indicated.

PREP Pearls

- Shigellosis is characterized by diarrhea, high fever, and abdominal cramping.
- Mild Shigella infection may not require antibiotic therapy but with severe symptoms antibiotics are indicated.
- Shigella is commonly spread in child-care settings.

American Board of Pediatrics Content Specification(s):

- Plan the treatment of Shigella infection

Suggested Reading:

- American Academy of Pediatrics. Shigella infections. In: Pickering LK, Baker CJ, Kimberlin DW, Long SS, eds. Red Book: 2012 Report of the Committee on Infectious Diseases. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:645-647
- Centers for Disease Control and Prevention. Shigellosis. www.cdc.gov/nczved/divisions/dfbmd/diseases/shigellosis/

Item 251

You are seeing a 1-year-old girl as a new patient in your practice. As you obtain the history, the father relates to you that his daughter was very yellow after birth and that she almost needed a "blood exchange" because her bilirubin was very high. She remained in the nursery for 1 week "under blue lights" She has done well since that time, and her immunizations are up to date. Developmentally, she is able to pull herself up to stand and has taken a few steps alone. She points at things and bangs things together but has not used any words. Her physical examination is unremarkable.

Of the following, the MOST appropriate next step in management is to

A. initiate physical therapy
B. obtain a brain magnetic resonance image
C. perform a hearing test
D. provide reassurance
E. refer to a pediatric ophthalmologist

Item 251 Preferred Response: C

Infants and children with a history of pathologic unconjugated hyperbilirubinemia during the neonatal period are at increased risk of developing auditory system abnormalities and should undergo hearing testing if speech delays are noted. The term auditory neuropathy/auditory dys-synchrony (AN/AD) is used to describe the hearing loss resulting from hyperbilirubinemia, perinatal asphyxia, and prematurity in infants and children. In AN/AD, the outer hair cells function normally in the cochlea but the transmission of the electrical information from the cochlea to the brain occurs dys-synchronously because of abnormal nerve function. Newborn hearing screening is often performed using otoacoustic emissions (OAE). OAE testing assesses function of the outer hair cells in the cochlea and may not identify an infant with AN/AD. Auditory brainstem response (ABR) testing measures the brainstem's response to sound and will identify affected infants and children.

Kernicterus describes the selective yellow staining of the basal ganglia, brainstem nuclei, and cerebellum associated with pathologic unconjugated hyperbilirubinemia. The areas affected explain many of the clinical findings seen in infants and children with classic kernicterus, which include (1) hearing loss or deafness, (2) dystonia, hypertonia, and athetosis, (3) oculomotor paresis of upward gaze, and (4) enamel dysplasia of deciduous teeth. Because the sequelae of bilirubin toxicity vary by severity, location, and type, Shapiro has proposed a new classification system of kernicterus that incorporates these varied findings. Clinical examination of muscle tone in combination with auditory brainstem response testing and a magnetic resonance imaging scan of the brain have been recommended to make the diagnosis of kernicterus in infants with a history of pathologic indirect hyperbilirubinemia.

The infant described in the vignette had pathologic unconjugated hyperbilirubinemia as a newborn and may be at risk for the clinical findings seen with kernicterus. She is meeting all of her developmental milestones except language and should be referred for hearing testing. Her neuromuscular examination findings are normal and physical therapy is not needed. She does not require a referral to a pediatric ophthalmologist, because she has no oculomotor paresis. If her hearing testing suggests AN/AD, imaging of the basal ganglia with magnetic resonance imaging of the brain could be considered next to assist in the diagnosis of kernicterus.

PREP Pearls

- Auditory neuropathy/auditory dys-synchrony (AN/AD) is the form of hearing loss resulting from hyperbilirubinemia, perinatal asphyxia, and prematurity in infants and children.
- Auditory brainstem responses can identify hearing loss caused by AN/AD, but otoacoustic emissions testing cannot identify such hearing loss.

American Board of Pediatrics Content Specification(s):

- Recognize the permanent sequelae of bilirubin toxicity

Suggested Reading:

- American Academy of Pediatrics Subcommittee on Hyperbilirubinemia. Management of hyperbilirubinemia in the newborn infant 35 or weeks gestation. Pediatrics. 2004;114:297-316. doi: 10.1542/peds.114.1.297
- Lauer BJ, Spector ND. Hyperbilirubinemia in the newborn. Pediatr Rev. 2011;32:341-349. doi: 10.1542/pir.32-8-341
- Shapiro SM. Chronic bilirubin encephalopathy: diagnosis and outcome. Semin Fet Neo Med. 2010;15:157-163. doi: 10.1016/j.siny.2009.12.004

Item 252

A 4-year-old Vietnamese boy who has no significant past medical history presents to your office with 2 days of high fever and cough. His sister is receiving chemotherapy for acute leukemia. A rapid diagnostic test is positive for influenza virus. The following are the results of his complete blood cell count:

- White blood cell count, 6,200 /µL (6.2 x 10^9/L with 10% polymorphonuclear leukocytes, 81% lymphocytes, 7% monocytes, and 2% eosinophils
- Hemoglobin, 11.8 g/dL (118 g/L)
- Mean corpuscular volume, 83/μm^3 (83 fL)
- Platelet count, 218x 10^3/µL (218 x 109/L)

Of the following, the MOST likely cause of the neutropenia in this boy is

A. cyclic neutropenia
B. ethnic neutropenia
C. familial leukemia syndrome
D. severe congenital neutropenia
E. transient (virus-associated) neutropenia

Item 252 Preferred Response: E

The child in the vignette has moderate neutropenia (absolute neutrophil count [ANC], 62041 [0.62 x 109/L]), most likely related to the viral infection.
Classification of severity of neutropenia in patients 1 year and older is shown (Item C252).

Item C252. Classification of severity of neutropenia in patients 1 year and older

Degree of severity	Absolute neutrophil count	Risk of infection
Mild neutropenia	1000-1500/µL (1.0-1.5 x 10^9/L)	Little or none
Moderate neutropenia	500-1000/ µL (0.5-1.0 x 10^9/L)	Less frequent, less severe
Severe neutropenia	<500/ µL (0.5 x 10^9/L)	Highly susceptible

Normal values for ANC vary with age. From 2 months to 1 year of age, neutropenia is defined as an ANC less than 1000/mL (1.0 x 109/L), whereas in children older than 1 year and adults, the lower limit of normal for ANC is 1500/mL (1.5 x 10^9/L). In addition, there is variation in normal neutrophil counts among different racial groups. In some blacks from South Africa, American Mexicans, Afro-Caribbeans, Yemenite Jews, and some Arabic populations, an ANC between 500 to 1000/mL (0.5-1.0 x 10^9/L) can be seen and is not associated with increased infections. The risk of infection is related to the severity, duration, and mechanism of neutropenia. Long periods of neutropenia without recovery lead to an increased risk of infection. Conditions in which there is adequate bone marrow reserve to mobilize neutrophils, as in immune neutropenia, are less likely to be associated with severe infection than in disorders of decreased marrow production, such as bone marrow failure syndromes or myelosuppression secondary to chemotherapy. In evaluating an infant or child with neutropenia, it is important to ask whether there is a history of recurrent infections in the patient or the family. A thorough physical examination for signs of neutropenia (eg, skin infections, mouth ulcers, and gingivitis) and congenital anomalies is necessary to evaluate for inherited syndromes. The presence of other cytopenias would suggest a generalized bone marrow disorder, such as aplastic anemia or leukemia.

In children, acquired forms of neutropenia are much more common than congenital neutropenia. The main causes of acquired neutropenia are infection, drugs, and immune disorders, with postinfectious neutropenia being the most common. The mechanisms are thought to be redistribution sequestration, aggregation, and destruction of neutrophils by circulating antibodies. In most cases, the neutropenia resolves quickly and does not lead to bacterial superinfection. Viral pathogens, such as respiratory syncytial virus, influenza, parvovirus, Epstein-Barr virus, and human herpes virus 6, are associated with transient mild to moderate neutropenia, which occurs during the first few days of the illness and then resolves in 3 to 8 days.

Children with transient neutropenia of short duration who are otherwise well-appearing are at low risk of infection. However, if fever occurs in a patient with moderate to severe neutropenia, a blood culture should be obtained and parenteral antibiotics given.
The boy described in the vignette, with no history of recurrent or severe infections, most likely has neutropenia secondary to the influenza infection. If the patient had recurrent

symptoms of neutropenia (eg, oral ulcers, gingivitis, or infections), serial complete blood cell counts twice weekly for 6 weeks should be obtained to look for the 21-day cycle consistent with cyclic neutropenia. The patient is not of the ethnic groups typically associated with having lower ANC values. This 4-year-old patient is not likely to have severe congenital neutropenia, which presents with recurrent infections during infancy. Familial leukemia syndrome is rare, and the patient does not have any other cytopenias to suggest leukemia.

PREP Pearls

- The main causes of acquired neutropenia are infection, drugs, and immune disorders, with postinfectious neutropenia being the most common.
- Viral pathogens, such as respiratory syncytial virus, influenza, parvovirus, Epstein-Barr virus, and human herpes virus 6, are associated with transient mild to moderate neutropenia, which occurs during the first few days of the illness and then resolves in 3 to 8 days.

American Board of Pediatrics Content Specification(s):

- Recognize that common viral infections may cause transient neutropenia that does not require specific treatment

Suggested Reading:

- Coates TD. Overview of neutropenia. UptoDate. Available on line for subscription only.
- Fioredda F, Calvillo M, Bonanomi S, et al. Congenital and acquired neutropenia consensus guidelines on diagnosis from the neutropenia committee of the marrow failure syndrome group of the AIEOP. Pediatr Blood Cancer. 2011;57:10-17. doi:10.1002/pbc.23108
- Segel GB, Halterman JS. Neutropenia in pediatric practice. Pediatr Rev.

Item 253

A 6-month-old male infant is seen for his routine health supervision visit. He is doing well at home, gaining weight, and meeting his milestones. The mother mentions some concerns about flattening of her son's head posteriorly, and you examine his head from above. While he has no evidence of torticollis, he has right-sided flattening of the occiput as well as flattening above the right orbit, giving a trapezoidal appearance to his head. His mother confirms that they have been placing him in a supine position to sleep at night. They also have been giving him "tummy time" at home during the day and, since his last checkup at 4 months of age, have been alternating his sleeping direction on a weekly basis so that his left and right sides are facing his bedroom door equally.

Of the following, you are MOST likely to recommend

A. continued tummy time and alternating head direction in bed with follow-up in the pediatric office in 1 month
B. referral to a pediatric neurosurgeon for evaluation for a possible right coronal craniosynostosis
C. referral to a physical therapist to work with the parents on neck stretching exercises
D. referral to the craniofacial team to fit a helmet to correct the positional head deformity
E. a switch to a prone sleeping position now that he is capable of rolling over and holding his head up

Item 253 Preferred Response: B

The infant described in this scenario has evidence of a unilateral lambdoidal craniosynostosis. Premature closure of a suture (ie, absence of a history and examination suggestive of torticollis and presence of a trapezoidal head shape as noted from above) will require neurosurgical evaluation.

Positional skull deformities may be present at birth and can be exacerbated by continued resting of the head on that side during early infancy. Postnatally acquired positional skull deformities may also be seen in infants whose cranial contour was normal at birth and in such instances are often associated with torticollis. Positional skull deformities typically exhibit a parallelogram cranial contour, with the ipsilateral ear displaced anteriorly. In contrast, a deformational defect due to premature fusion of a suture usually shows the trapezoidal contour, as seen in the infant in the vignette, with no anterior ear displacement. The differences in the ear position on the ipsilateral side can be compared and contrasted in Item C253.

Management strategies for positional skull deformities include supine positioning for sleep with the rounded (unaffected) side of the head positioned against the mattress and orienting the crib or sleeping direction so that the infant preferentially gazes away from the flattened side. In addition, parents should be encouraged to provide their infants with "tummy time" when they are awake and under observation. For infants with torticollis, parents should be instructed on neck stretching exercises. Most infants with positional skull deformities will respond to these simple interventions within 3 months. However, deformity progressing by 4 to 6 months of age despite conservative positioning measures may also prompt a referral to a neurosurgeon familiar with craniosynostoses.

Whereas some specialists promote skull-molding helmets for positional deformities, there is little evidence that these devices are more effective than conservative treatment for babies with mild to moderate deformities. Helmets would be completely ineffective for an infant with a true craniosynostosis, and neck stretching exercises would provide no benefit in the child in the vignette, who has deformational plagiocephaly and no evidence of torticollis. Prone positioning for sleep is not recommended for infants at 6 months of age and would provide no benefit to this child.

PREP Pearls

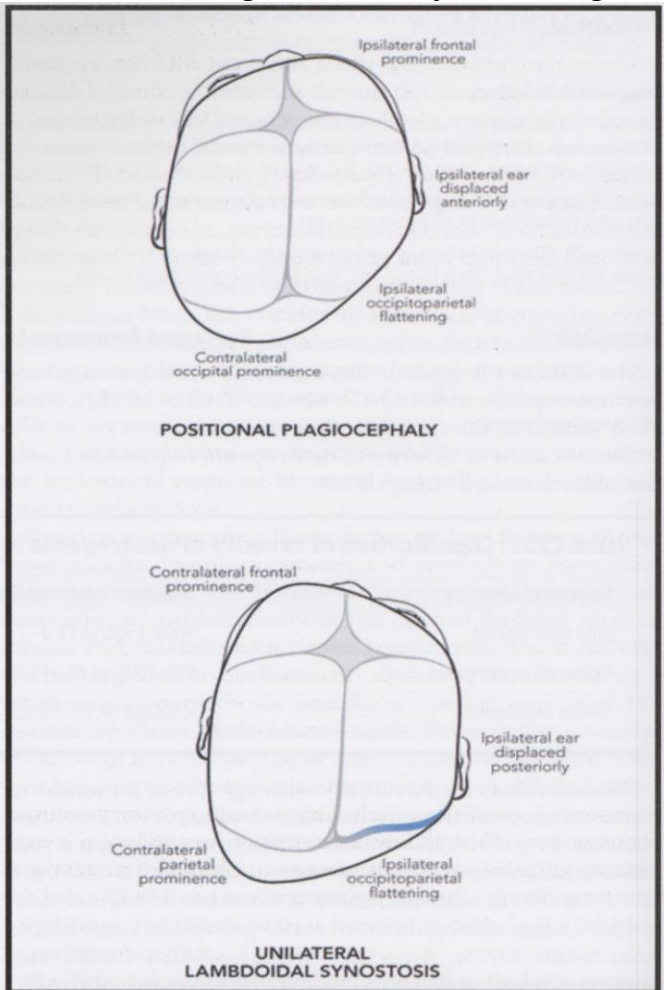
- An infant with plagiocephaly and a trapezoidal head contour is likely to have a significant craniosynostosis requiring neurosurgical consultation.
- If a suspected positional head deformity in an infant is not responding to conservative measures such as positioning by 4 to 6 months of age, a neurosurgical evaluation may be required.

American Board of Pediatrics Content Specification(s):

- Recognize the clinical findings of premature closure of a cranial suture

Suggested Reading:

- laughn I. Luerssen 1G, Dias MS; Committee on Practice and Ambulatory Medicine, Section on Neurological Surgery. Prevention and management of positional skull deformities in infants. Pediatrics. 2011;128(6):12361241. doi:10.1542/peds.2011.2220
- Robin NH. Falk Ml, Haldeman-Englert CR. rGFIL-relatedcraniosynostmis syndromes. In: Pagon RA, Bird TD, Dolan CR, eds. GencRervic". Seattle, Washington: University of Washington; 2013



ITEM C253: Positional plagiocephaly (top) when viewed from above, the head has a parallelogram contour with displacement of the ipsilateral ear anteriorly versus unilateral lambdoidal synostosis with a trapezoidal contour and posterior displacement of the ipsilateral ear.

Item 254

An intern you are supervising requests your assistance interpreting the results of spirometry performed on one of her patients who has persistent asthma. The intern would like to learn about the lung function parameters measured during spirometry that aid in the diagnosis of asthma. You review the interpretation of the spirometry results (ie, forced expiratory volume in 1 second [FEVI] to forced vital capacity [FVC] ratio, FEV, and post bronchodilator reversibility) with the intern.

Of the following, the findings MOST consistent with this diagnosis are

A. FEV, to FVC ratio < 85% and FEV, <80%; postbronchodilator reversibility of 12%
B. FEV, to FVC ratio < 85% and FVC < 80%; postbronchodilator reversibility of 12%
C. FEV, to FVC ratio > 85% and FVC < 80%; postbronchodilator reversibility of 20%
D. FEV, to FVC ratio > 85% and FEV, < 80%; postbronchodilator reversibility of 12%
E. FEV, to FVC ratio > 85% and FEV, < 80%; postbronchodilator reversibility of 20%

Item 254 Preferred Response: A

The spirometric findings most consistent with the diagnosis of asthma are forced expiratory volume in 1 second [FEV1] to forced vital capacity [FVC] ratio less than 85%, FEVI less than 80%, and postbronchodilator reversibility of 12%.

The 2007 National Heart, Lung, and Blood Institute (NHLBI) Guidelines for asthma recommend performance of spirometry as an essential objective measure to establish the diagnosis of asthma. The medical history and physical examination do not always provide sufficient information to exclude other diagnoses or to evaluate lung status. Spirometry can demonstrate obstruction and assess reversibility in patients at least 5 years of age. Spirometry is also helpful in monitoring the response to long-term therapy and changes in the degree of obstruction over time. Spirometry is generally recommended, rather than measurements by a peak flow meter, because of wide variability in peak flow meters and reference values. Peak flow meters are better used for monitoring asthma control or to gauge the severity of asthma exacerbations, as long as caretakers understand the limitations of peak flow meters.

The NHLBI Guidelines suggest that spirometry/lung function measurements be performed at the following times: at the initial assessment, after treatment is initiated and symptoms have stabilized, during periods of progressive or prolonged loss of asthma control, and at least every 1 to 2 years, more frequently if needed depending on response to therapy.

The important parameters derived from spirometry include indices of flow, including forced expiratory volume in one second (FEV_1), flow between 25% and 75% of the vital capacity (FEF_{25-75}), also known as the maximal midexpiratory flow rate, and peak expiratory flow rate (PEFR). FEV_1, FEF25-75, and PEFR are decreased in obstructive disorders. Indices of volume are also measured, including forced vital capacity (FVC). Forced vital capacity is the volume of air that can be forcibly blown out after full inspiration, measured in liters. FEV_1 is the volume of air that can be forcibly blown out in 1 second, after full inspiration. Values from 80% to 120% of the average value are considered normal. FEV_1/FVC (FEV_1%) is the ratio of FEV_1 to FVC. In healthy children, this should be 85% or greater.
In obstructive diseases FEV_I is diminished because of increased airway resistance to expiratory flow. In some cases, the FVC may be decreased as well, owing to the premature closure of airway in expiration, but the FEV_I is typically disproportionately affected. In restrictive diseases the FEV_I and FVC are both reduced proportionally and the ratio may be normal or even increased as a result of decreased lung compliance. An FEV_1/FVC ratio less than 85% in children 8 to 19 years of age therefore suggests that the observed decrease in airflow is due to airway obstruction rather than due to volume reduction. Reversibility is determined either by an increase in FEV_1 of at least 12% from baseline or by an increase of at least 10% of predicted FEV_1 after inhalation of a short-acting bronchodilator.

Recent research suggests that FEF25-75% or FEF25-50% may be a more sensitive parameter than FEV_1 in the detection of obstructive small airway disease. However,

owing to discrepancies in midrange expiratory flow, current practice guidelines recommend using FEV_1, FVC, and FEV_1/FVC as indicators of obstructive disease. Low FEV_1 indicates current obstruction (impairment) and risk for future exacerbations (risk). For children, the ratio of the FEVI/forced vital capacity (FVC) appears to be a more sensitive measure of severity and control in the impairment domain. FEV_1 is a useful measure of risk for exacerbations, although it is emphasized that even children who have normal lung function experience exacerbations.

PREP Pearls

- Spirometry is recommended as an objective measure to diagnose asthma and monitor control.
- An FEVI/FVC ratio less than 85% in children suggests the presence of airway obstruction.
- FEV, can be an indicator of current obstruction and predict risk for future exacerbations.
- Reversibility is determined either by an increase in FEU, of at least 12% from baseline or by an increase of at least 10% of predicted FEV, after inhalation of a short-acting bronchodilator.

American Board of Pediatrics Content Specification(s):

- Know what spirometry measures

Suggested Reading:

- National Heart, Lung, and Blood Institute. Guidelines for the Diagnosis and Management of Asthma (EPR-3)

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