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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

Question 1

A 9-month-old infant is referred to you by a community outreach program because of low weight. He was born at term after an uncomplicated pregnancy and delivery. He was exclusively breastfed until 6 months of age, when a few puréed baby foods were added to his diet. He continues to breastfeed 6 times daily. His development and physical examination are unremarkable. His current weight is 7.8 kg, length is 68 cm (Item Q1A), and head circumference is 44 cm (Item Q1B).

Item Q1A. Weight and length growth chart for the infant described in the vignette.
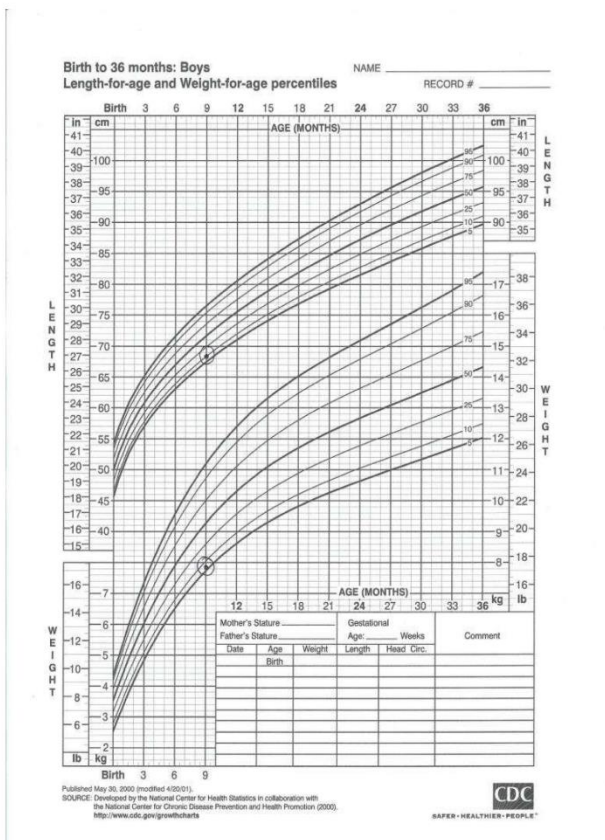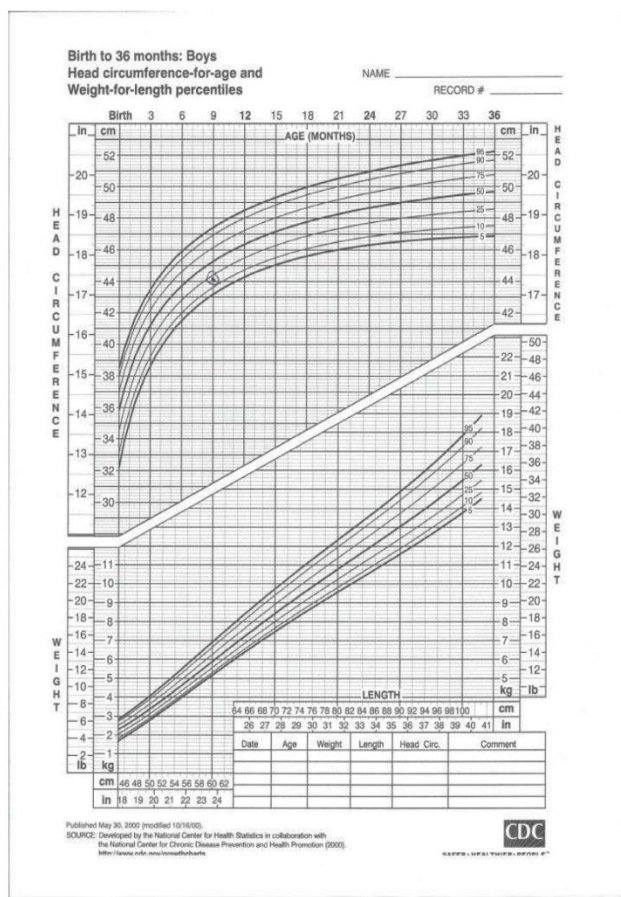
Of the following, the MOST appropriate statement about monitoring this infant's growth is

- A. body mass index is the best parameter for assessing this infant's growth
- B. children should follow a consistent growth percentile line throughout the first 2 years of life
- C. this infant's growth parameters indicate undernutrition
- D. US Centers for Disease Control and Prevention growth charts are the best option for monitoring growth of breastfed infants
- E. World Health Organization growth charts should be used for monitoring children younger than 2 years of age

Question 1   **Preferred Response: E**

Growth monitoring is one of the most important functions of pediatric care and should be performed at all health supervision visits. Currently, 2 growth charts predominate for use in assessing healthy term infants: US Centers for Disease Control and Prevention (CDC) and World Health Organization (WHO) growth charts. For children 24 months of age and younger, WHO charts should be used for monitoring growth because the WHO charts better reflect ideal growth for this age group. When the baby in the vignette is plotted on the WHO chart, both his weight and length are between the 10th and 25th percentiles for age, and therefore are unlikely to represent undernutrition.

The WHO infant charts are based on data collected longitudinally from 6 sites covering Europe, Africa, south Asia, North America, and the Middle East. The children in this study had no underlying disease and were born at term to nonsmoking mothers who had access to sufficient resources. These children were exclusively breastfed until 4 months of age and continued to be breastfed until at least 12 months of age, therefore the WHO growth charts are felt to represent "ideal growth." By contrast, CDC growth charts were derived from cross-sectional studies in the United States from 1963 to 1994, and the children from whom the data were derived were often formula fed. Breastfed children tend to gain weight faster than formula-fed infants in the first 3 months of life, then slow their weight gain velocity. These changes are reflected in the differences between the WHO and CDC growth charts. Therefore, the breastfed infant in the vignette is more likely to have a growth pattern consistent with the WHO chart than the CDC chart. Of note, both CDC and WHO growth charts for children older than 24 months of age are derived from cross-sectional data, and the CDC charts are acceptable for use in older children.

For both the WHO and CDC growth charts, changes over time are the best reflection of actual growth. Many children follow the same growth percentile lines throughout infancy, but it is also normal for some infants to cross 1 or 2 major percentile lines between 6 and 18 months of age. Growth parameters at birth reflect the intrauterine environment, but by later infancy, growth parameters are more consistent with genetic potential. In fact, growth at 2 years of age correlates well with mean parental height. In children younger than 3 years of age, the best assessment of acute undernutrition is a weight-for-height below the fifth percentile. Body mass index has not been validated for children younger than 2 years of age and would not be an appropriate parameter to assess this infant.

PREP Pearls
- World Health Organization (WHO) growth charts better reflect "ideal growth" of breastfed infants and are superior to US Centers of Disease Control and Prevention growth charts. The WHO charts should be used to assess growth of children younger than 2 years of age.
- Many normally growing infants cross 1 or 2 major percentile lines between 6 and 18 months of age as their growth changes from reflecting an intrauterine environment to reflecting genetic potential.
- For children younger than 3 years of age, the best assessment of acute undernutrition is a weight-for-height below the fifth percentile.

ABP Content Specifications(s)
- Differentiate between normal and abnormal growth velocity in neonates and infants
- Use a growth chart to monitor linear growth and weight gain
- Differentiate between normal and abnormal variations in linear growth and weight gain

Suggested Readings
- Danner E, Joeckel R, Michalak S, Philllips S, Goday PS. Weight velocity in infants and children. Nutr Clin Pract. 2009;24(1):76-79. doi: http://dx.doi.org/10.1177/0884533608329663 .
- Grummer-Strawn LM, Renold C, Krebs NF. Use of World Health Organization and CDC growth charts for children aged 0-59 months in the United States. Morbid Mortal Wkly Rep MMWR. 2010;59(RR09):1-15.http://www.cdc.gov/mmwr/preview/mmwrhtml/rr5909a1.htm.
- Keane V. Assessment of growth. In: Kleigman RM, Stanton BF, Schor NF, St Geme JW III, Behrman RE, eds. Nelson Textbook of Pediatrics. 20th ed. Philadelphia, PA: Saunders Elsevier; 2015: 84-89.

Question 2

A 17-year-old adolescent presents to the office with complaints of vaginal discharge for 3 days. She has no significant past medical history, but she is the mother of an 18-month-old child. She currently uses the etonogestrel implant for contraception. She is not on any other medications. She reports 2 sexual partners in the past 3 months, and was last sexually active about 2 weeks ago. Physical examination reveals a temperature of 37.2°C, heart rate of 72 beats/min, respiratory rate of 16 breaths/min, blood pressure of 110/68 mm Hg, and weight of 54.5 kg. She has no abdominal tenderness. On pelvic examination, she has a friable cervix with purulent cervical discharge, but no cervical motion or adnexal tenderness. You are concerned that she has gonococcal cervicitis.

Of the following, the BEST next steps in the diagnosis and management are

  A. culture and treatment with ceftriaxone 250 mg intramuscularly as a single dose and doxycycline 100 mg by mouth twice daily for 14 days
  B. Gram stain smear and treatment with ceftriaxone 250 mg intramuscularly as a single dose and azithromycin 1 g orally as a single dose
  C. nucleic acid amplification test and treatment with azithromycin 1 g orally as a single dose
  D. nucleic acid amplification test and treatment with ceftriaxone 250 mg intramuscularly as a single dose
  E. nucleic acid amplification test and treatment with ceftriaxone 250 mg intramuscularly as a single dose and azithromycin 1g orally as a single dose

Question 2   **Preferred Response: E**

The 17-year-old female adolescent depicted in this vignette has symptoms of gonococcal cervicitis that include mucopurulent cervical discharge, intermenstrual bleeding, cervical friability, and lower abdominal pain. According to the US Centers for Disease Control and Prevention Sexually Transmitted Diseases Treatment Guidelines, the recommended management for uncomplicated gonococcal cervicitis is ceftriaxone 250 mg intramuscularly, and either azithromycin 1 g orally in a single dose or doxycycline 100 mg twice a day orally for 7 days.

The azithromycin or doxycycline is included in the regimen because of the risk of co-infection with Chlamydia trachomatis as well as the growing concern for cephalosporin-resistant Neisseria gonorrhoeae. Azithromycin is preferred over doxycycline when possible because it is administered as a single dose. Oral cephalosporins are no longer recommended as first-line medications for treatment of N gonorrhoeae because of the concern for resistance.

Nucleic acid amplification tests have better overall sensitivity and specificity compared to other diagnostic tests for gonococcal infections. Nucleic acid amplification tests are therefore recommended for the detection of gonorrhea, except in cases of childhood sexual assault. Currently, data are insufficient to recommend nucleic acid amplification tests for testing rectal and oropharyngeal secretions in childhood sexual assault; instead, cultures should be used. Cultures should also be considered when there is a concern for treatment failure to allow for susceptibility testing.

PREP Pearls
- Gonorrhea is a common sexually transmitted disease. While patients can be asymptomatic, symptoms of gonococcal cervicitis include cervicovaginal discharge, intermenstrual bleeding, and lower abdominal pain.
- Nucleic acid amplification tests should be used for diagnosis in most situations.
- The current recommended treatment regimen is ceftriaxone 250 mg intramuscularly, and either azithromycin 1 g orally as a single dose (preferred) or doxycycline 100 mg orally twice a day for 7 days.

ABP Content Specifications(s)
- Plan appropriate management for a patient with Neisseria gonorrhoeae infection
- Recognize the major clinical features associated with Neisseria gonorrhoeae infection
- Plan the appropriate diagnostic evaluation for Neisseria gonorrhoeae infection

Suggested Readings
- Papp JR, Schachter J, Gaydos CA, Van Der Pol B. 4 Recommendations for the Laboratory-Based Detection of Chlamydia trachomatis and Neisseria gonorrhoeae. MMWR Morbid Mortal Wkly Rep. 2014;63(RR2).http://www.cdc.gov/mmwr/preview/mmwrhtml/rr6302a1.htm.
- Swygard H, Sena AC, Leone P, Cohen MS. Gonorrhea. In: Klausner JD, Hook EW III, eds. Current Diagnosis & Treatment of Sexually Transmitted Diseases. New York, NY: McGraw-Hill; 2007:99-107.

- US Centers for Disease Control and Prevention. Self-study STD modules for clinicians-gonorrhea. US Centers for Disease Control and Prevention website. http://www2a.cdc.gov/stdtraining/self-study/gonorrhea. Updated April 1, 2013.
- US Centers for Disease Control and Prevention. Sexually transmitted diseases treatment

Question 3

A 6-month-old infant presents for evaluation of a rash that developed 2 weeks ago. He has been well and is taking no medications. The physical examination reveals numerous erythematous papules located on the trunk and extremities, including the hands (Item Q3A), and feet (Item Q3B).

Item Q3A. Rash on the hand for the patient described in the vignette. Courtesy of D. Krowchuk.

Item Q3B. Rash on the foot for the patient described in the vignette. Courtesy of D. Krowchuk.

Of the following, the MOST appropriate treatment is

    A. cephalexin orally
    B. an emollient topically
    C. hydrocortisone topically
    D. hydroxyzine orally
    E. permethrin topically

Question 3   **Preferred Response: E**

The male infant described in the vignette has a generalized eruption composed of erythematous papules. Notably, the eruption involves the hands and feet, including the palms and soles. These findings suggest a diagnosis of scabies and he should be treated with permethrin 5% cream topically. Hydrocortisone and hydroxyzine could be used adjunctively to relieve pruritus, and cephalexin might be employed if there was evidence of secondary bacterial infection. However, none of these agents would eradicate the infestation. Although atopic dermatitis may produce a generalized eruption in infants, the presence of large papules and involvement of the palms and soles is uncommon.

Scabies is caused by infestation with the mite, Sarcoptes scabiei. Spread is primarily by direct contact with an infested individual, although fomites may be responsible. Two to 3 weeks following infestation, pruritus and rash develop. Lesions are erythematous papules, nodules (Item C3A), and burrows (Item C3B) located in the interdigital spaces, wrist flexures, axillae, and waist. In girls, the areolae may be involved; in boys, papules and nodules may affect the penis (Item C3C) and scrotum. In infants, such as the one in the vignette, the eruption may be generalized and vesiculopustules may be observed on the palms and soles.

Scabies is treated with permethrin cream 5% applied topically for 8 to 14 hours (ie, overnight). The cream is applied to the entire skin surface from the neck to the toes. In infants and possibly in young children and the elderly, the head (including the face) should be treated. Permethrin is

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not completely ovicidal and, for this reason, a second treatment is recommended 7 to 14 days later. The symptoms and signs of scabies represent a hypersensitivity reaction to the mite and its products, therefore 2 to 4 weeks may be required for the pruritus and rash to subside. Household or other close contacts may be infested, but not yet symptomatic, and as a result should receive a single application of permethrin at the time the index case is first treated. Bed linens and clothing should be laundered in hot water and dried at high temperature, although some advise a simpler strategy of placing items in a dryer at 60°C for 10 min. Items that cannot be treated in this manner may be stored in a sealed plastic bag for a minimum of 3 days.

An alternative to permethrin is oral ivermectin. It is US Food and Drug Administration-approved for use in adults with uncomplicated scabies (a single dose of 200 μg/kg repeated in 1 to 2 weeks) and crusted scabies (a single daily dose on days 1, 2, 8, 9, and 15). Ivermectin is not approved for the treatment of scabies in children and is not recommended for use in those younger than 5 years of age or those weighing less than 15 kg. Other topical therapies for scabies exist, but their role in treatment is limited. These include crotamiton (poor efficacy), lindane (poor efficacy, potential neurotoxicity), and benzyl benzoate (contact dermatitis, neurologic toxicity if ingested).

PREP Pearls
- In children and adolescents, the lesions of scabies typically are located in flexural areas (eg, between the digits, wrist flexors, etc). However, in infants, the eruption is generalized and the palms and soles often are affected.
- In boys, papules and nodules often are present on the penis and scrotum.
- Topical permethrin is the treatment of choice for scabies in children.
- Asymptomatic household contacts should be treated at the time the index case is first treated.

ABP Content Specifications(s)
- Recognize the clinical manifestations of scabies
- Plan the appropriate management of scabies

Suggested Readings
- Currie BJ, McCarthy JS. Permethrin and ivermectin for scabies. N Engl J Med. 2010;362(8):717-725. doi:http://dx.doi.org/10.1056/NEJMct0910329.
- Golant AK, Levitt JO. Scabies: a review of diagnosis and management based on mite biology. Pediatr Rev. 2012;33(1):e1-e12. doi: http://dx.doi.org/10.1542/pir.33-1-e1.

Question 4

A 6-year-old boy is brought to the emergency department after being hit by a car while walking across the street. He was thrown approximately 20 ft. His vital signs show a temperature of 37°C, pulse of 60 beats/min, blood pressure of 140/90 mm Hg, respiratory rate of 15 breaths/min, and SpO2 is 100% on 2 L of oxygen by nasal cannula. On neurologic examination, the boy's eyes are closed, but they open when his name is called. His right pupil is 2 mm and briskly reactive. His left pupil is 4 mm and sluggishly reactive. He purposefully bats at the examiner when a painful stimulus is applied. He moves all extremities equally. He does not say words, but rather incomprehensibly moans. There are abrasions on his face and a boggy area over his left temporal region. The boy is breathing comfortably on supplemental oxygen with equal breath sounds. His extremities are warm and well-perfused. He has bruising over the left upper quadrant of his abdomen, with no sign of chest or extremity trauma.

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

A.   administer 3% saline 5 mL/kg
B.   obtain magnetic resonance imaging of the brain
C.   perform diagnostic peritoneal lavage
D.   perform a focused abdominal ultrasonography of trauma (FAST)
E.   transfuse 15 mL/kg packed red blood cells

Question 4   **Preferred Response: A**

The boy in the vignette has suffered a traumatic brain injury. Based on evidence of acute encephalopathy, pupil asymmetry, hypertension, and bradycardia, it is likely that he has cerebral edema, increased intracranial pressure (ICP), and possibly an intracranial hematoma. Because of these findings and the severity of the mechanism of injury, he is at risk for clinical deterioration. Of the response choices listed, administering 5 mL/kg of 3% saline intravenously to decrease ICP is the best next step.

The Monroe-Kellie doctrine states that because the volume of the calvarium is constant, ICP will increase dramatically with an increase in the volume of its contents which include brain, blood, and cerebrospinal fluid. Therefore, ICP will rise in the event of an intracranial hemorrhage or cerebral edema without a compensatory decrease in another component. Hyperosmolar therapy, such as intravenous hypertonic saline or mannitol, is a mainstay of treatment as it causes water to shift from the intracellular and interstitial spaces of the brain into the serum, thus decreasing the volume of the brain component. Mannitol, a sugar alcohol commonly used in the management of increased ICP in both pediatric and adult traumatic brain injury, is a preferred therapy when acute herniation is suspected or anticipated. In addition to its osmotic effect, which occurs within 15 to 30 minutes of administration, mannitol immediately decreases blood viscosity, causing reflex vasoconstriction and thereby decreasing cerebral blood volume. The intravascular volume status of a trauma patient receiving mannitol must be closely monitored. Because mannitol is filtered in the glomerulus and not reabsorbed, the increased osmolality in the nephron leads to osmotic diuresis. In contrast, hypertonic saline can be used both for intravascular fluid expansion and to decrease ICP without causing osmotic diuresis, which is an added benefit in trauma cases. It should be noted that the 2012 pediatric traumatic brain injury guidelines state that hypertonic saline should be considered for the treatment of pediatric traumatic brain injury associated with intracranial hypertension, but mannitol is not mentioned. However, both therapies continue to be used by many major trauma centers.

The many causes of altered mental status in children, including coma, can be generally divided into direct structural derangements of the central nervous system such as hydrocephalus, trauma, and stroke, and medical causes such as hypoxia, infection, imbalances of metabolic supply and demand, seizure, and toxins (Item C4A). Although presentation is dependent on the age of the child, there is a continuum from normal mental status to coma, which includes confusion, delirium, lethargy, and stupor. An early sign of altered mental status is confusion and disorientation, in which the child cannot follow a conversation or lacks orientation to person, place, or time. Delirium is a state of mental or motor excitement that can include fear, irritability, and agitation. Lethargy is a sleepy state in which the child can be aroused with moderate stimulation with immediate relapse into sleep. Stupor is a more unresponsive state in which the child can only be aroused with vigorous or painful stimuli. It is important for the clinician to recognize a child experiencing progression of signs and symptoms toward impending coma, because this can be a harbinger of worsening illness or impending death (from herniation or loss of airway and breathing).

As ICP rises and compensatory mechanisms are exhausted, herniation syndromes can occur, leading to pupillary, extraocular, and motor derangements (Item C4B). Central herniation ensues if brain structures are forced caudally into the foramen magnum. In the case of an expanding temporal fossa lesion, such as an epidural hematoma, the medial temporal lobe (uncus) can herniate through the tentorium.

Item C4A. Differential Diagnosis of Altered Level of Consciousness.

Structural Causes
- Cerebral vascular accident
- Cerebral vein thrombosis
- Hydrocephalus
- Intracerebral tumor
- Subdural empyema
- Trauma (intracranial hemorrhage, diffuse cerebral swelling, shaken baby syndrome)

Medical Causes (Toxic-Infectious-Metabolic)
- Anoxia
- Diabetic ketoacidosis
- Electrolyte abnormality
- Encephalopathy
- Hypoglycemia
- Hypothermia or hyperthermia
- Inborn errors of metabolism
- Infection (sepsis)
- Intussusception
- Meningitis and encephalitis
- Postictal state
- Psychogenic
- Toxins
- Uremia (hemolytic-uremic syndrome)

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Item C4B. Progression of Herniation Syndromes.

Stage	Mental Status	Pupils	Extraocular Movements	Motor Response
Central Herniation				
Diencephalic (early)	Normal or decreased	Small, reactive	Normal	Appropriate
Diencephalic (late)	Decreased	Small, reactive	Normal	Decorticate
Midbrain, upper pons	Decreased	Midpoint, fixed	Asymetric	Decerebrate
Lower pons, medulla	Decreased	Pinpoint, fixed	Absent	Flaccid
Uncal Herniation				
Third nerve (early)	Usually normal	Unilateral, dilated, sluggish	Normal or asymetric	Appropriate or asymetric
Third nerve (late)	Decreased	Unilateral, dilated, fixed	Asymmetric or absent	Decorticate or decerebrate
Midbrain, upper pons	Decreased	Midpoint, fixed	Asymmetric or absent	Decerebrate

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Neither magnetic resonance imaging (MRI) nor diagnostic peritoneal lavage is the best next step in this case. Although MRI of the brain will detect bleeding, hydrocephalus, cerebral vasculature abnormalities, and tumors, it generally requires more than 1 hour spent in an inadequately monitored environment. Although it has been replaced by focused abdominal ultrasonography of trauma (FAST) in many trauma protocols, diagnostic peritoneal lavage can be used to detect abdominal trauma, but as noted, is not the first priority in this hemodynamically stable child with a mental status change. Similarly, a blood transfusion would not be indicated in this case without evidence of acute blood loss or hemodynamic instability.

PREP Pearls
- Hyperosmolar therapy, such as 3% saline or mannitol administered intravenously, can be used to treat an acute elevation in intracranial pressure.
- Altered mental status can be due to structural and medical causes.
- The clinician should be alert to the progression of signs and symptoms along the continuum from normal mental status to coma, especially if it is rapid.

ABP Content Specifications(s)
- Identify the signs of impending coma

Suggested Readings
- Avner J. Altered states of consciousness. Pediatr Rev. 2006;27:331-338. doi: http://dx.doi.org/10.1542/pir.27-9-331.
- Kochanek PM, Carney N, Adelson PD, et al. Guidelines for the acute medical management of severe traumatic brain injury in infants, children, and adolescents: 2nd edition. Pediatr Crit Care Med. 2012;13(suppl 1):S36-S40. doi: http://dx.doi.org/10.1097/PCC.0b013e31823f435c.
- Rubenstein JS. Consultation with the specialist: Initial management of coma and altered consciousness in the pediatric patient. Pediatr Rev. 1994;15:204-207. doi: http://dx.doi.org/10.1542/pir.15-5-204.

Question 5

An 8-month-old infant with asplenia presents to the emergency department for evaluation of irritability, fever, and rash. A physical examination reveals multiple purpuric lesions. Lumbar puncture reveals a cerebrospinal fluid white blood cell count of 1,800/µL. He is treated with vancomycin and cefotaxime, and admitted to the pediatric intensive care unit. His mother asks if anything could have been done to prevent this illness.

Of the following, the BEST response to the mother's question is that meningococcal conjugate vaccine is

- A. indicated for travelers starting at 6 months of age
- B. indicated for patients with human immunodeficiency virus infection
- C. indicated starting at 2 months of age
- D. licensed for children only older than 9 months of age
- E. protective against the most common serogroup in infants

Question 5   Preferred Response: C

The infant in the vignette with asplenia should have been vaccinated with meningococcal conjugate vaccine beginning at 2 months of age. Groups considered to have increased risk include those with anatomic or functional asplenia, such as the infant in this vignette, or those with complement component deficiencies. Of note, HIV infection is not an indication for infant immunization.

In the United States, there are 2 meningococcal quadrivalent polysaccharide protein conjugate vaccines licensed for young children that offer protection against serogroups A, C, W, and Y. An additional bivalent conjugate vaccine combined with Haemophilus influenzae vaccine protects against serogroups C and Y. Since 2005, vaccination has been routinely recommended in adolescents. In 2010, the recommendation for a booster dose at 16 years of age was made. Although 60% of meningococcal disease occurs in children younger than 5 years of age, the two licensed novel meningococcal serogroup B-specific vaccines available in the US are approved for use in persons 10 to 25 years of age. Serogroup B-specific vaccines have been used successfully in outbreak settings.

Meningococcal vaccination is recommended for travelers to endemic regions, such as the "meningitis belt" in sub-Saharan Africa or during the Hajj in Saudi Arabia. For children younger than 9 months of age who are travelling to endemic areas, a 3-dose primary series of conjugate vaccine at 2, 4, and 6 months of age should be completed prior to travel. Children 9 months to 23 months of age require 2 doses and those 24 months or older require a single dose.

In the United States, one of the commercially available quadrivalent vaccines is licensed for persons as young as 2 months of age and the bivalent vaccine that is combined with Haemophilus influenzae is licensed for infants starting at 6 weeks of age.

PREP Pearls
- High risk groups should be vaccinated with meningococcal vaccine starting at 2 months of age.
- Commercially available serogroup B-specific vaccines are licensed for persons 10 to 25 years of age
- Serogroup B is responsible for most infections in young children.
- For travel or residence in a hyperendemic region, meningococcal vaccination can begin at 2 months of age.

ABP Content Specifications(s)
- Know the indications and schedule for the meningococcal vaccine
- Know which serotypes are included in the meningococcal vaccine

Suggested Readings
- American Academy of Pediatrics. Updated recommendations on the use of meningococcal vaccines. Pediatrics. 2014;134(2):400-403. doi: http://dx.doi.org/10.1542/peds.2014-1383.

- Centers for Disease Control and Prevention. Vaccines and Immunization: Meningococcal Vaccination. Available at:http://www.cdc.gov/vaccines/vpd-vac/mening/default.htm. Accessed April 12, 2015.
- Cohn AC, MacNeil JR, Clark TA, et al. Prevention and control of meningococcal disease: recommendations of the advisory committee on immunization practices. MMWR Morbid Mortal Wkly Rep. 2013;62(RR02):1-22

Question 6

A 16-year-old adolescent presents to the emergency room with chief complaints of flank pain radiating to the groin and blood in the urine for the last 2 hours. There is no history of fever, burning on urination, or trauma. The mother mentions that the patient had a similar episode 1 year ago associated with passage of tiny particles in the urine. Vital signs show a temperature of 38.3°C, heart rate of 100 beats/min, respiratory rate of 28 breaths/min, and blood pressure of 144/80 mm Hg. On physical examination, the patient is bent over in pain and has mild dehydration. The patient refuses to let you examine her abdomen.

Of the following, the MOST sensitive imaging modality for this patient is

- A.   computed tomography
- B.   intravenous urography
- C.   magnetic resonance imaging
- D.   radiograph (kidney, ureter, bladder)
- E.   ultrasonography

Question 6   **Preferred Response: A**

The patient in the vignette has typical symptoms of kidney stones. Flank pain radiating to the groin, hematuria (gross or microscopic), and passage of tiny particles in the urine are indicative of kidney stones.

Noncontrast (unenhanced) helical computed tomography (CT) is the most sensitive imaging modality for evaluating patients with suspected urinary tract stones (adults and children). Computed tomography can detect stones in the ureters that may not be detected by ultrasonography, radiolucent stones (such as pure uric acid stones) that are not detected by plain radiography, and very small stones (around 1 mm in diameter) that are often missed on plain radiography or ultrasonography. Studies have also reported on the possible evaluation of the chemical composition of the renal calculi from the CT images. Computed tomography also provides better estimates of the size, number, and direction of the branches of a staghorn calculus, which may further help in treatment decisions.

Noncontrast CT avoids the risk of intravenous contrast material and can be completed in less than 5 minutes (with the newer multidetector CT machines). Therefore, the imaging can be done without anesthesia in most patients. A urine pregnancy test should be done in all female patients (of child-bearing age) prior to all radiation exposure.

The radiation exposure during CT is dependent on equipment and institutional protocols. In children, excessive radiation with conventional CT scanners calibrated for adults is a concern. However, radiation doses can be reduced significantly while adjusting scanning parameters based on the size and weight of the patient and at the same time maintaining adequate imaging quality. In institutions specializing in providing care for children, guidelines from the National Cancer Institute should be followed in establishing protocols to ensure effective and safe radiation doses for CT. If this is not the case, then another imaging modality, such as ultrasonography, could be used initially and the choice of imaging modality can be made based on the clinical scenario and discussion with a pediatric nephrologist or urologist.

Ultrasonography is a reasonable alternative to CT, especially in pregnant women and in institutions where radiation dose from CT cannot be reduced to safe levels (per the National Cancer Institute guidelines). Ultrasonography can detect radiolucent uric acid stones and urinary obstruction (dilatation of the renal collecting system proximal to obstruction). Ultrasonography is not a useful modality for identifying small stones and stones in the ureters, renal papilla, or the renal calyces. Ultrasonography is also operator-dependent and the experience and expertise of the ultrasonographer is an important factor in the sensitivity of this imaging modality. Ultrasonography, though not as sensitive as helical CT, is a reasonable alternative to CT to detect nephrolithiasis, especially when there are concerns about radiation exposure from CT.

A plain abdominal radiograph will detect radiopaque stones (such as calcium, struvite, and cystine kidney stones) and miss radiolucent uric acid stones. Plain radiographs will also miss small stones, stones overlying bony structures, and will provide no information on urinary

system obstruction. The current use of plain radiographs is limited to settings where renal ultrasonography and CT are not available for imaging children with suspected renal stones.

Intravenous urography has been used previously in patients with suspected renal stones following an initial plain film. Intravenous urography is a radiologic test for identifying the details of the urinary system, including kidneys and ureters, and identifying upper urinary tract obstruction. Delayed and prolonged excretion of the contrast medium and dilatation of the collecting system are indicative of obstructive calculi of the kidneys. The intravenous urography leads to less radiation exposure in comparison to a CT; however, it is also less sensitive in detecting kidney stones. The current availability of ultrasonography, CT, and magnetic resonance urography has replaced IVU in almost all diagnostic settings.

Magnetic resonance imaging, similar to plain radiography and ultrasonography, is not sensitive for detecting small calculi. Calculi can be detected by magnetic resonance imaging if their foci of signal void are large or they lie adjacent to tissues of high signal intensity. Signal defects on MRI are nonspecific and could be due to calculi, clots, debris, or tumors.

PREP Pearls
- Noncontrast (unenhanced) helical computed tomography (CT) is the most sensitive imaging test for evaluating patients with suspected urinary tract stones.
- Ultrasonography, though not as sensitive as helical CT, is a reasonable alternative to CT to detect nephrolithiasis, especially when there are concerns about radiation exposure.
- Radiation exposure during CT can be reduced significantly while adjusting scanning parameters based on the size and weight of the patient with little compromise of the imaging quality.

ABP Content Specifications(s)
- Plan the evaluation of urinary tract stones in patients of various ages
- Recognize the signs and symptoms of urinary tract stones in patients of various ages

Suggested Readings
- Mostafavi MR, Ernst RD, Saltzman B. Accurate determination of chemical composition of urinary calculi by spiral computerized tomography. J Urol. 1998;159(3):673-675. doi: http://dx.doi.org/10.1016/S0022-5347(01)63698-X.
- National Cancer Institute. Radiation risks and pediatric computed tomography (CT): a guide for health care providers. National Institutes of Health website. http://www.cancer.gov/cancertopics/causes/radiation/radiation-risks-pediatric-CT. Updated June 7, 2012..

Question 7

You are caring for a 12-month-old boy with Down syndrome and short bowel syndrome secondary to duodenal atresia. Following resection, he has 25 cm of small bowel remaining, including 15 cm of duodenum and 10 cm of ileum, which includes the ileocecal valve. He receives enteral feeds by continuous infusion, but requires parenteral nutrition for most of his calories to maintain growth. His height and weight are currently at the 25th percentile for age. Recent weekly laboratory studies to monitor his parenteral nutrition show stable electrolytes and normal liver function tests. The results of his complete blood cell counts over time are shown in Item Q7.

Item Q7. Results of Patient's Complete Blood Cell Counts Over Time.

Age	5 months	7 months	9 months	12 months
White blood cells (/µL)	5,000	6,500	5,500	4,800
Hemoglobin (g/dL)	10.8	9.6	8.5	7.8
Hematocrit (%)	32	28.8	25.5	23.4
Platelets (x 10^3/µL)	250	300	175	180
Mean corpuscular volume (fL)	80	85	95	100

Of the following, the value MOST likely to be abnormal in this patient is

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

Question 7 **Preferred Response: B**

Due to his history of short bowel syndrome and given the laboratory findings described, the child in this vignette is most likely to have macrocytic anemia caused by a vitamin B12 deficiency. Vitamin B12 is absorbed primarily in the ileum, most of which has been resected. In addition, vitamin B12 is often deficient in children on long-term total parenteral nutrition (TPN) therapy.

Children on long-term TPN require close observation and frequent laboratory monitoring. Item C7 provides a summary of parameters to be measured during TPN use. Fluid balance is critical when initiating and continuing TPN. Children on TPN are at risk for hypoglycemia and hyperglycemia, particularly when TPN is initiated. Electrolyte supplementation must be tailored to the needs of each child, with special attention to the age and underlying disorders of the child. Calcium can be low in premature infants and in patients with renal losses. Phosphate can be elevated in premature infants and is vital to the prevention of metabolic bone disease. Liver function should be monitored to assess nutrition and to evaluate for evidence of parenteral nutrition-associated liver disease.

Item C7. Parameters to be Measure During Total Parenteral Nutrition Use.

Parameters to Monitor	Early Use	Moderate Use	Chronic Use
Weight	Daily	Weekly	Biweekly to monthly
Input and output	Daily	Weekly	Biweekly to monthly
Urine specific gravity	Every 6 to 8 hours	Daily	N/A
Blood glucose	Every 6 to 8 hours	Daily	N/A
Basic metabolic panel	3x/week	Weekly	Biweekly to monthly
Calcium, magnesium, phosphate	Weekly	Weekly	Biweekly to monthly
Serum protein /albumin	Weekly	Weekly	Biweekly to monthly
Transaminases	Weekly	Weekly	Monthly
Direct Bilirubin	Weekly	Weekly	Monthly
Triglycerides	2 to 3 times/week	Weekly	Monthly
Calories	Daily	Monthly	Monthly

N/A, not applicable
Courtesy of C. Waasdorp Hurtado

Lipids provide a good energy source in a small volume and are critical to avoiding essential fatty acid deficiency. Lipid utilization varies and underutilization can result in elevated triglyceride levels. The goal is to maintain triglycerides at less than 150 mg/dL (1.7 mmol/L).

Fat soluble vitamins are absorbed throughout the duodenum, jejunum, and proximal ileum. Fat soluble vitamins (vitamins A, D, E, and K) are less likely to be deficient in this child, as they are supplemented in the TPN and deficiency is rare. Iron is absorbed in the proximal small intestines. This child is at risk for chronic iron deficiency, but this is less likely than vitamin B12 deficiency.

PREP Pearls
- Close monitoring of glucose, fluid balance, and electrolytes is needed in the first days of total parenteral nutrition (TPN) initiation.
- Children on chronic TPN are at risk for parenteral nutrition-associated liver disease.

ABP Content Specifications(s)
- Monitor a patient receiving parenteral nutrition while considering the associated complications

Suggested Readings
- Kleinman RE. Pediatric Nutrition Handbook. 5th ed. Elk Grove Villlage, IL: American Academy of Pediatrics; 2004:369-389.

Question 8

You are called to the neonatal intensive care unit to evaluate a 24-hour-old newborn with progressive obtundation, seizures, and tachypnea. Pregnancy, labor, and delivery were uneventful. The newborn initially did well during the first several hours after birth, but then became lethargic, hypothermic, and developed poor feeding. Results of a comprehensive metabolic panel including glucose, complete blood cell count with differential, and C-reactive protein are all normal. The anion gap is normal. Blood gas results reveal a respiratory alkalosis. Intravenous antibiotics are started and cultures are pending.

Of the following, the BEST next laboratory test for diagnosis and management is

A.   serum ammonia
B.   serum galactose-1-phosphate
C.   serum toxicology screen
D.   serum very long chain fatty acids
E.   urine organic acids

Question 8   **Preferred Response: A**

The neonate in the vignette has a classic presentation of a urea cycle disorder, with decompensation in the first 24 to 72 hours of life with progressive respiratory alkalosis, obtundation, and hyperammonemia, in the presence of a normal anion gap. Urea cycle disorders are caused by mutations resulting in the absence or partial functioning of 1 of the first 4 enzymes in the biochemical pathway responsible for the breakdown of nitrogen (the urea cycle). Urea is a waste byproduct of protein catabolism in the body. In a normal individual, nitrogen is broken down into urea that is excreted through the urine. If nitrogen levels build up in the body because of inefficient breakdown, it accumulates quickly in the form of ammonia. Hyperammonemia is very toxic to the brain and can cause irreversible damage without immediate intervention. Neonates will appear normal at birth, but within 24 to 72 hours, they will develop cerebral edema. This typically manifests as poor feeding, obtundation, hypothermia, seizures, hyperventilation, hyporeflexia, unusual posturing, and ultimately, coma. Classic laboratory findings include elevated ammonia levels (> 210 μg/dL [150 μmol/L]) with a normal anion gap and glucose level in the presence of respiratory alkalosis on blood gas measurement. To distinguish between the specific types of urea cycle defects, one must order a plasma amino acid analysis and an urine orotic acid. Ornithine transcarbamylase deficiency (OTC) is associated with extremely high urinary orotic acid levels. Carbamoyl phosphate synthetase I (CPS I) deficiency is associated with low to undetectable urine orotic acid levels. Definitive diagnosis is dependent on either enzymatic analysis or molecular genetic testing of the genes involved. All of the urea cycle disorders are autosomal recessive, except OTC, which is transmitted by X-linked recessive inheritance. A family history of early infant death, presumably because of hyperammonemia, may also be seen. Specific urea cycle disorders include N-acetyl glutamate synthetase deficiency, carbamoyl phosphate synthetase I deficiency, OTC deficiency, citrullinemia type I, argininosuccinic aciduria, and arginase deficiency.

Urea cycle disorders may present as a metabolic emergency, necessitating immediate recognition and treatment to avoid irreversible brain damage. Severe hyperammonemia is treated with dialysis and hemofiltration to rapidly reduce the plasma ammonia concentration, along with the intravenous administration of arginine hydrochloride and nitrogen scavenger drugs to promote the excretion of excess nitrogen through alternative pathways. Restriction of protein for 12 to 24 hours is essential, with calories provided through carbohydrates and fat. Care must be taken to stabilize the patient with intravenous fluids and inotropic drugs if necessary.

Long term management mandates the use of specialized formulas, oral nitrogen-scavenging drugs, dietary restriction of protein, and avoidance of hyperammonemic episodes. Patients are at high risk of decompensation, necessitating hospitalization for close observation of clinical status and ammonia levels if they have gastrointestinal or respiratory illnesses. Most patients are routinely treated on a long term basis by biochemical or metabolic geneticists, in addition to their primary care provider. They should have emergency protocols in place at home, at the primary care provider's office, and at the local hospital.

Organic acidemias or organic acidurias are a group of disorders characterized by dysfunction of a specific step in amino acid catabolism, typically the result of a specific enzyme deficiency.

Classic patterns of unusual excretion of non-amino organic acids in urine are a first line test for this group of disorders. Newborns appear well at delivery and for the first few days, followed by metabolic decompensation. Symptomatology includes vomiting, poor feeding, neurologic symptoms, and lethargy progressing to coma. Laboratory findings include metabolic acidosis (not respiratory alkalosis as in the child in this vignette), ketosis, hyperammonemia, elevated liver function tests, low blood sugar, and neutropenia. These disorders require immediate recognition to optimize long term outcome via clinical interventions.

Tyrosinemia type 1 presents in infancy with significant liver involvement, and eventually renal tubular dysfunction, growth failure, and rickets. Untreated children may present with repeated neurologic crises involving a change in mental status, peripheral neuropathy, abdominal pain, and occasionally respiratory failure. If untreated, many die before 10 years of age. Laboratory abnormalities include increased succinylacetone concentration in the blood and urine; elevated tyrosine, methionine, and phenylalanine on serum amino acids; and elevated tyrosine metabolites on urine organic acids.

Galactosemia presents in the neonatal period with jaundice, hypotonia, scleral icterus, bruising, bleeding, and cataracts in the face of rapidly progressive liver failure. Classic laboratory abnormalities of galactosemia include positive urine-reducing substances, abnormal liver function studies, coagulation abnormalities suggestive of a progressive bleeding diathesis, elevated erythrocyte galactose-1-phosphate, and sepsis, especially due to Escherichia coli.

Prenatal drug exposure is typically best assessed by a combination of a maternal interview, maternal hair analysis, and meconium drug testing. Withdrawal symptoms are dependent on the type of drugs abused by the mother. A history of placental abruption should be a red flag for potential cocaine exposure. Special attention should be focused on clinical signs including microcephaly, intrauterine growth retardation, prematurity, congenital malformations, and congenital infections. Neonatal abstinence syndrome can include signs of central nervous system dysfunction (high-pitched cry, restlessness, hyperreflexia, jitteriness, tremors, seizures), respiratory symptoms (nasal flaring, tachypnea, apnea), and gastrointestinal dysfunction (frantic rooting, poor feeding, vomiting, loose stools). The Finnegan scoring system is commonly used for assessing for signs of neonatal abstinence syndrome. Meconium drug analysis is currently the best method for assessing prenatal drug exposure in neonates, not a serum toxicology screen, as stated in the answers in the vignette. Babies with drug withdrawal may be obtunded, but they will not have hyperammonemia.

Peroxisomal biogenesis disorders are typically screened with serum very long chain fatty acids. They can present in the neonatal period with hypotonia, poor feeding, dysmorphic facies, seizures, and liver cysts with hepatic dysfunction. Bony stippling may occur in the patella or long bones detectable by skeletal survey. Infants do not present with metabolic crises, but with slowly progressive neurologic deterioration, dying in the first year of life.

PREP Pearls
- Urea cycle disorders clinically manifest with immediate decompensation in the first 24 to 72 hours of life with progressive respiratory alkalosis, obtundation, and hyperammonemia in the presence of a normal anion gap.
- Hyperammonemia is very toxic to the brain and can cause irreversible damage without immediate intervention. It is very important to check the serum ammonia level and quickly implementation of measures to decrease the ammonia level as soon as possible (dialysis, hemofiltration, nitrogen scavenger medications, and protein restriction).

ABP Content Specifications(s)
- Plan the appropriate immediate and long-term management of urea cycle defects, while considering the long-term prognosis

Suggested Readings
- Mew NA, Lanpher BC, Gropman A, et al. Urea cycle disorders overview. GeneReviews.http://www.ncbi.nlm.nih.gov/books/NBK1217/.

Question 9

The parents of a healthy 3-year-old boy call the office to report that they found him playing in the "junk" drawer and saw him put his fingers in his mouth. They are concerned that he may have swallowed something. He is in no respiratory distress and is not complaining of any pain. He drank some milk, but has not had anything to eat since then. Various objects are present in the drawer, including buttons, coins, batteries, safety pins, and small toys that contain magnets. A radiograph is obtained and it is determined that the patient needs immediate intervention.
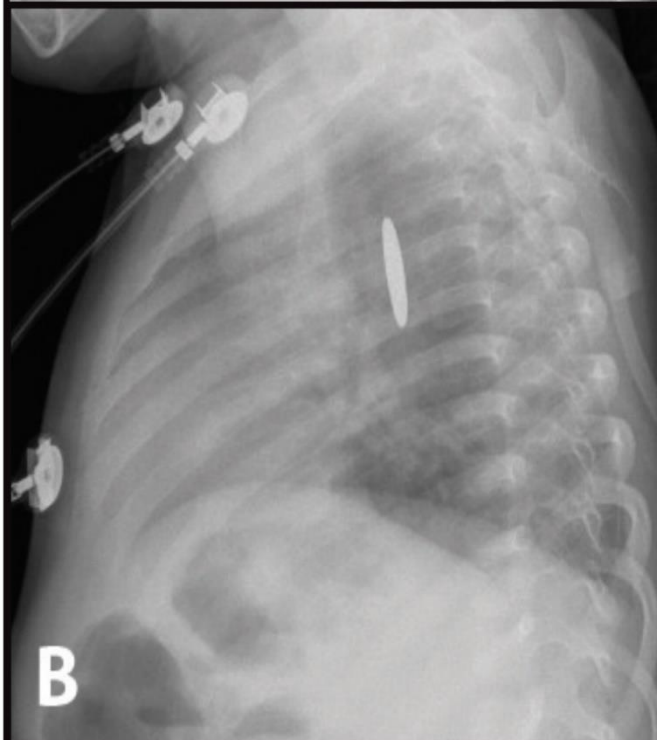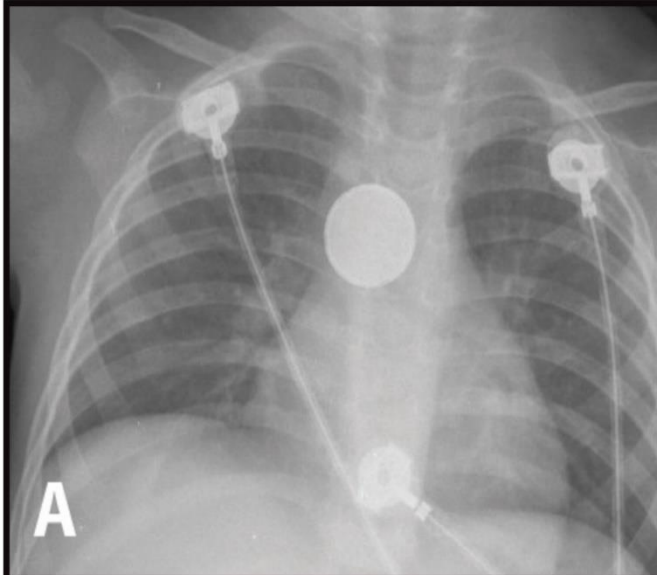
Of the following, the boy MOST likely ingested a

A.   button
B.   button battery
C.   closed safety pin
D.   magnet
E.   quarter

Question 9   **Preferred Response: B**

The radiograph obtained for the boy in this vignette reveals a round foreign body with a double rim appearance (ITEM C9AB). This is a

Courtesy of D. Mulvihill

Item C9A, Item C9B: A 2-year-old patient with a metallic foreign body in the esophagus that resembles a coin. However, on the anteroposterior radiograph (A), note that the periphery of the object is more radio-opaque than the center suggesting the double contour of a button battery.
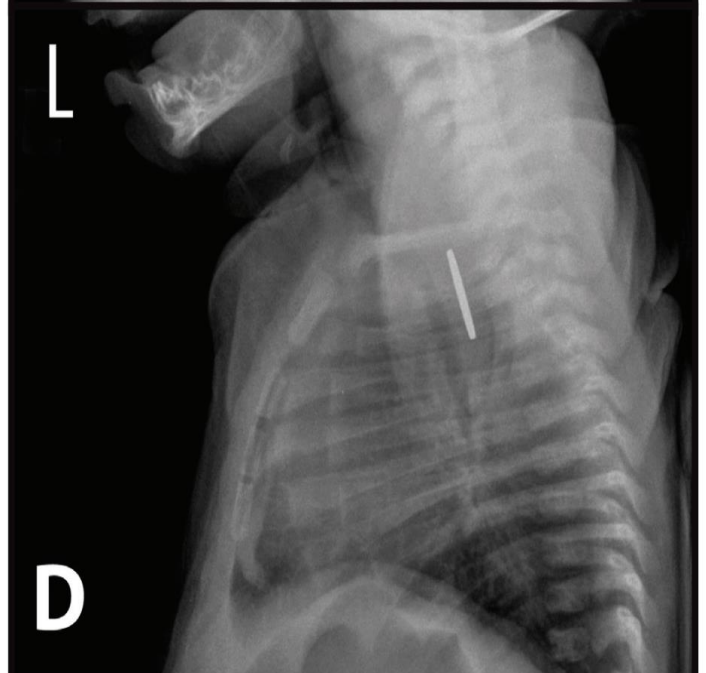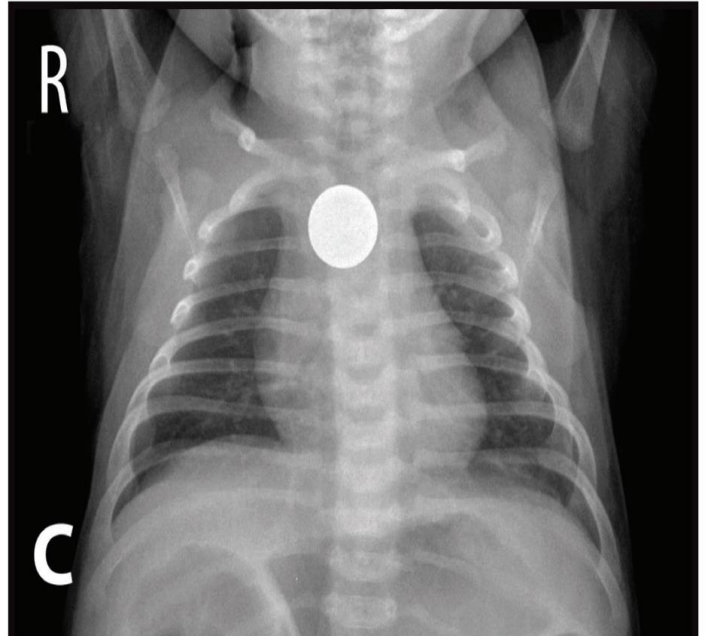
Courtesy of D. Mulvihill

ITEM C9C, Item C9D: Coin in the upper esophagus of a child who had been coughing for weeks. There is widening of the esophageal wall and compression of the trachea from the longstanding foreign body.

common feature seen with larger button batteries and helps to distinguish them from other round objects (ITEM C9CD). Among the response choices, the object that requires the most immediate attention is the button battery.

Foreign body ingestion is primarily a pediatric problem. Management of foreign body ingestions depends on the item ingested, anatomic location of the foreign body, and presence or absence of symptoms. Coins are the most commonly reported foreign bodies ingested and most will traverse the gastrointestinal (GI) tract without incident. Button batteries, sharp objects or toys, and magnets require action because of the increased risk for serious complications.

Button batteries contain toxic heavy metals and alkaline compounds that are caustic to the mucosa. Lithium cells are more likely to cause significant outcomes than the other chemistry types (manganese dioxide, zinc-air, or silver oxide). Complications of button battery ingestions have become increasingly frequent and devastating in parallel with increased household use of 20-mm lithium coin cells. Button batteries lodged in the esophagus, or at any point along the GI tract, may lead to significant mucosal injury, even perforation. Button batteries that have passed into the stomach may be monitored with serial abdominal radiographs every 12 hours. If there is no progress through the GI tract over 24 hours, then surgical removal is necessary.

Unfortunately, the degree of injury may worsen long after battery removal.

Potential complications include:
- perforation
- stricture
- fistula
- vocal cord paralysis (caused by recurrent laryngeal nerve damage)
- mediastinitis
- pneumothorax
- pneumoperitoneum
- aspiration pneumonia
- empyema
- lung abscess
- spondylodiscitis
- exsanguination from aortoesophageal fistulas
- death

Battery diameter is the most important predictor of a clinically significant outcome. All serious outcomes or fatal cases reported have occurred with batteries of 20 mm or larger. Severe burns can occur after only 2 to 2.5 hours. No clinically significant outcomes were observed with 15- to 18-mm battery cells. Young age (<4 years) and ingestion of more than 1 battery were also associated with worse outcome. New (fully charged) 20- to 25-mm batteries are 3 times more likely than spent cells to be associated with clinically significant outcomes. Unwitnessed ingestions are common and are at risk for misdiagnosis and delay in removal, leading to worse outcomes.

A plain anteroposterior (AP) radiograph that includes the neck, chest, and abdomen should be obtained immediately on all patients who present with a history of possible button battery ingestion, even if they are asymptomatic. The window of opportunity to remove an esophageal lithium cell battery before injury occurs is less than 2 hours, so timing is critical. Endoscopic removal is preferred because it allows direct visualization of tissue injury. Later complications must be anticipated, depending on the battery position and orientation, as well as time before removal. Patients should be monitored closely for the development of fistulas or perforations 1 to 3 weeks later and strictures weeks to months later.

Plain radiography of the neck, chest, and abdomen area should be performed in patients who present with a recent history of possible foreign body ingestion to confirm the item ingested and determine location. Symptoms that suggest esophageal impaction include feeding refusal, drooling, difficulty or painful swallowing, or emesis. In these cases, urgent endoscopic evaluation and removal of any impacted esophageal foreign body is mandatory. If the foreign body has passed the gastroesophageal junction, and the object does not pose a danger (magnets, sharps, button battery), then the patient's condition can be managed conservatively. Although most objects will pass through the intestinal tract without incident, entrapment can occur at the pylorus, the ligament of Treitz, or the ileocecal valve. Parents should be instructed to examine the child's stools closely for the object and if it is not recovered from the stools within 1 to 2 weeks after presentation, follow-up radiography should be performed. If the object is retained, then surgical intervention is indicated, and is the appropriate management for all of the other response choices given. If 2 or more magnets are ingested, there is a much greater risk for entrapment of mucosa between magnets leading to perforation.

Indications for immediate removal of an ingested foreign body either endoscopically or surgically include airway compromise; complete esophageal obstruction; esophageal location unchanged for more than 12 hours; button battery in the esophagus; ingestion of more than 1 magnet; sharp or pointed objects more than 4 cm in length, wider than 2 cm in diameter, or showing no movement on day 3 after ingestion; any symptomatic patient; and acute abdominal findings.

It is not recommended to induce vomiting or give cathartics for ingested foreign bodies, because their effectiveness has not been proven. Alternative techniques for removal of esophageal foreign bodies, such as use of a Foley catheter or advancement with bougienage, have been successful in experienced hands, but should not be used in the case of button battery ingestion.

PREP Pearls
- Larger button batteries may reveal a double rim on plain radiography, which distinguishes them from other round objects.
- Ingestions of lithium cell batteries with a diameter of 20 mm or greater have the worst outcomes, with severe burns occurring after just 2 to 2.5 hours.
- Indications for immediate removal of an ingested foreign body include
    - airway compromise

- esophageal obstruction or location unchanged for more than 12 hours
- any button battery in the esophagus
- lithium button battery 20 mm or greater in diameter
- button battery showing no progress through the intestinal tract over 24 hours
- ingestion of more than 1 magnet
- sharp or pointed objects more than 4 cm in length, wider than 2 cm in diameter, or showing no movement on day 3 after ingestion
- acute abdominal findings

ABP Content Specifications(s)
- Plan the management of a patient who has ingested a coin
- Plan the management of a patient who has ingested a button battery

Suggested Readings
- Litovitz T, Whitaker N, Clark L, White NC, Marsolek M. Emerging battery-ingestion hazard: clinical implications. Pediatrics. 2010125(6):1168-1177. doi: http://dx.doi.org/10.1542/peds.2009-3037.

Question 10

You are seeing an 18-month-old boy who has had 2 skin abscesses in the perianal region and 1 abscess in the scalp, recurrent oral ulcers, 5 episodes of otitis media, and 2 episodes of pneumonia in the previous year. Laboratory evaluation results are shown:

Test	Results
White blood cell count	11,000/µL (11.0 x 10^9/L)
% neutrophils	72
% lymphocytes	23
% monocytes	3
% eosinophils	1
% basophils	1
Platelet count	245 x 10^3/µL (245 x 10^9/L)
Hemoglobin	12.0 g/dL

Of the following, the MOST appropriate next steps in the management of this child include

    A.   testing for the phagocyte oxidative burst by flow cytometry and starting prophylaxis
          with allopurinol
    B.   testing for the phagocyte oxidative burst by flow cytometry and starting prophylaxis
          with trimethoprim and sulfamethoxazole
    C.   testing for the presence of lymphoblasts by flow cytometry and starting prophylaxis
          with allopurinol
    D.   testing for the presence of lymphoblasts by flow cytometry and starting prophylaxis
          with trimethoprim and sulfamethoxazole
    E.   testing for the presence of lymphoblasts by flow cytometry and starting treatment
          with prednisolone

Question 10   **Preferred Response: B**

The child in the vignette has had an excessive number of bacterial infections, raising concern for an abnormality of the innate immune system. Of deficiencies in the innate immune system, the most common are in the number or function of neutrophils. While frequent bacterial infections as described in the vignette could represent a clinical manifestation of severe neutropenia, such as that seen in severe congenital or cyclic neutropenia, the complete blood cell count reported has a normal number of neutrophils. Thus, consideration must be given to neutrophil dysfunction. The most common disorder of neutrophil function is chronic granulomatous disease (CGD), a defect caused by a failure to produce oxygen radicals to kill phagocytosed microorganisms. The gold standard for diagnosing and initial management of CGD is testing the phagocyte oxidative burst by flow cytometry and initiating prophylaxis with trimethoprim and sulfamethoxazole.

Chronic granulomatous disease is a genetic disorder that can be transmitted by X-linked or autosomal recessive inheritance. The X-linked form is by far the most common and is caused by a mutation in the CYBB gene that results in a dysfunctional gp91 protein. The most common autosomal form of CGD is caused by a mutation in the NCF1 gene on chromosome 7, resulting in a dysfunctional p47 protein. Chronic granulomatous disease most often presents with recurrent abscesses, most frequently caused by Staphylococcus aureus. It can also present with invasive fungal infections, usually with Aspergillus species.

Chronic granulomatous disease is the most common heritable defect of neutrophil function, although other rarer disorders of neutrophil function exist. One example is the leukocyte adhesion deficiency. Leukocyte adhesion deficiency is a group of genetic disorders that result in the failure to express proteins necessary for the normal trafficking of leukocytes to areas of infection. Without the ability to leave the blood stream and enter an area of infection, the leukocytes are unable to contain infections. The resulting phenotype can be very similar to CGD.

Frequent bacterial infections in a young child, especially of skin, lung, and sinus, should raise concern for an underlying immunodeficiency. The immune system can be divided into the innate immune system and the adaptive immune system. The innate immune system is comprised of barriers such as skin and mucosal membranes, and phagocytes such as the neutrophil, macrophage, and natural killer cells. The adaptive immune system is comprised of B and T lymphocytes and their subsets. Generally, abnormal viral infections suggest a deficiency of the adaptive immune system, while frequent bacterial or fungal infections suggest a deficiency of the innate immune system.

While children with leukemia are at risk for frequent bacterial infections, the complete blood cell count reported no lymphoblasts, and the normal hemoglobin and platelet numbers do not suggest bone marrow dysfunction. Thus, testing for lymphoblasts would not be warranted. Allopurinol is used to reduce the risk of end organ damage from the accumulation of uric acid in the setting of tumor lysis. As the presentation of the child in this vignette does not fit with a diagnosis of leukemia, starting allopurinol would not be appropriate.

PREP Pearls
- A defect of the innate immune system should be suspected in a child presenting with frequent bacterial infections.
- Chronic granulomatous disease is the most common disorder of neutrophil function.
- Chronic granulomatous disease is diagnosed through testing of the neutrophil oxidative burst by flow cytometry and the initial management should include prophylaxis with trimethoprim and sulfamethoxazole.

ABP Content Specifications(s)
- Recognize the clinical and laboratory findings associated with abnormal leukocyte function
- Recognize the clinical characteristics of phagocytic disorders
- Plan appropriate management of a patient with a leukocyte disorder

Suggested Readings
- Esfandbod M, Kabootari M. Images in clinical medicine: chronic granulomatous disease. N Engl J Med. 2012;367(8):753. doi: http://dx.doi.org/10.1056/NEJMicm1106350.

Question 11

You see a 3-year-old boy for a health supervision visit. He has been healthy other than an ear infection at 2 years of age. The mother reports no concerns about his health. His weight and height are consistently at the 25th percentile and his physical examination is unremarkable. His tympanic membranes appear normal and move appropriately with insufflation. You wish to assess his language development, but he is cautious and shy in your office and speaks only a few words quietly to his mother. Believing his mother to be an accurate and observant historian, you question her about the boy's speech.

Of the following, the finding that BEST supports the need for a speech evaluation in this child is

A.   he cannot follow a 3-step direction

B.   he cannot identify 5 colors

C.   he cannot use 3-word phrases

D.   he is unable to use words to talk about time

E.   strangers understand about 75% of his words

Question 11   **Preferred Response: C**

The 3-year-old boy described in this vignette has subtle findings that are suspicious for a language delay, including having had at least 1 early ear infection and a quiet or shy demeanor that may be masking a communication problem. His anxious behavior of only whispering a couple of words to his mother in the office presents an evaluation challenge in that he is not conversant enough to allow an assessment of the quality of his speech. In a case like this, the examiner should inquire about whether the parent has any concerns for their child's speech production, and if there are any specific speech milestones that raise red flags. If either of these are positive, then a referral for further speech and language assessment is warranted. Notably, a formal hearing evaluation will be a key component of any further speech and language evaluation.

Of the milestones listed, not being able to use a 3-word phrase would be of most concern for a 3-year-old child and would be a good reason to refer him for further assessment. There is a tremendous range of normal variation in speech, particularly between 1 to 2 years of age. Therefore, keeping in mind signs of when a referral would be needed can help this seem less confusing. The following red flag signs are far enough outside the norm that they would demonstrate a clear need for further assessment:
- 6 months of age: lack of turning to sound or voice
- 9 months of age: lack of babbling consonant sounds
- 18 months of age: does not say "mama," "dada," or other names
- 24 months of age: failure to use single words
- 30 months of age: failure to use 2-word phrases
- 36 months of age: failure to use 3-word phrases

The ability to follow a 3-step direction and the ability to point to 5 to 6 specific colors are expected normal range milestones for a 4-year-old child. The ability to use words to talk about time would also be a normal range milestone at 4 years of age. Intelligibility of speech to strangers typically follows this pattern: about 50% understandable at 2 years of age, about 75% understandable at 3 years of age, and 100% understandable at 4 years of age. It is not uncommon for a parent to feel they understand nearly everything their child says because of a high level of familiarity, while a stranger understands much less.

PREP Pearls
- A speech evaluation would be needed for a 2-year-old child who cannot use single words to communicate, or for a 3-year-old child who cannot use 3-word phrases.
- A 6-month-old infant who does not turn to sound or voice, and a 9-month-old infant who does not make babbling consonant sounds should have a careful hearing evaluation.
- The parents' ability to interpret meaning in their children's speech (that strangers cannot understand) can be a reason for delayed recognition of a speech problem.

ABP Content Specifications(s)
- Plan the appropriate initial management of speech and language disorders

American academy of pediatrics   38

- Plan the appropriate evaluation of language disorders in patients of various ages
- Identify the various etiologies of delayed language development

Suggested Readings
- McLaughlin MR. Speech and language delay in children. Am Fam Physician. 2011;83(10):1183-1188.

Question 12

A 2-month-old infant is seen in the emergency department for fever and respiratory distress. The baby had rhinorrhea 1 week ago, has been tachypneic for 3 days, and developed lethargy today. There is no history of a heart murmur. The baby is admitted for probable pneumonia, but after 2 days of antibiotics, is not improving as expected. She continues to fatigue easily with feeding, which her mother states has been the case for the past week. Her birth weight was 2.7 kg and her weight today is 5 kg (38th percentile). Her vital signs show a heart rate of 180 beats/min, respiratory rate of 80 breaths/min, and blood pressure of 65/45 mm Hg taken in the right arm. Her oxygen saturation is 92%. On physical examination, she has intercostal and subcostal retractions, but is not coughing and has no stridor. Her lungs are clear with breath sounds heard well to the bases. Her cardiac examination is significant for a soft S1 and S2 with no audible murmur. Her abdominal examination shows a liver edge that is 4 cm below the right costal margin. Her femoral pulses are palpable, but not strong. She has a capillary refill time of 3 seconds.

Of the following, the MOST likely etiology for the infant's clinical symptoms is

A.   aspiration
B.   group B streptococcal sepsis
C.   intussusception
D.   viral bronchiolitis
E.   viral myocarditis

Question 12   **Preferred Response: E**

The baby in this vignette had a viral infection 1 week ago and now presents with fatigue with feeding, an elevated heart rate, enlarged liver, and slightly decreased femoral pulses. She is in heart failure and, of the choices listed, viral myocarditis is the most likely cause of this combination of findings. Although bronchiolitis would make this baby's condition worse, the symptoms of quiet tachypnea are more consistent with pulmonary edema seen in cardiac failure. Myocarditis can present at any age and is often associated with viral infections such as parvovirus, coxsackievirus, or adenovirus. It can be mild or fulminant. When severe, it can lead to arrhythmia and death. The congestive heart failure (CHF) in this case is caused by poor cardiac muscle function, with subsequent inability to provide adequate cardiac output.

Congestive heart failure can present at any age, with some presentations that are age-specific. In the immediate newborn period, one may see a patient with critical (ductal dependent) cyanotic congenital heart disease, such as hypoplastic left heart syndrome. These newborns may develop signs of either pulmonary overcirculation with early CHF or poor systemic perfusion and shock. This occurs because the cardiac output shift s, sometimes very quickly, when the relative pulmonary and systemic vascular resistance changes. If the lungs become congested, the baby will become tachypneic. In this setting, the systemic output will decrease in proportion to the increase in pulmonary flow.

In the later newborn period, patients may become symptomatic with a large patent ductus arteriosus. This lesion causes excess pulmonary blood flow directly from the aorta to the pulmonary artery. This increases blood return to the pulmonary veins, the left atrium, and the left ventricle, and may result in volume overload of the left ventricle. Tachycardia and tachypnea may be seen.

At approximately 4 weeks of age, the pulmonary vascular resistance (PVR) will have dropped to that seen in later childhood. As the PVR drops, so does the right ventricular pressure. As this occurs, there will be increasing flow across any ventricular septal defect (VSD). The degree of left to right shunt in this case will depend on the size of the defect and the drop in PVR. Intracardiac shunts are described as the ratio of the pulmonary to systemic blood flow. In someone with a normal heart with no shunt, the ratio is 1:1. An infant with a large left to right shunt (such as 3:1 pulmonary flow to systemic flow) will have pulmonary vascular congestion and tachypnea. There will be a volume load to the right and subsequently to the left ventricle. Infants with this physiology are hyperdynamic and consequently have high caloric needs. They will often have failure to thrive.

A child with an atrioventricular canal or endocardial cushion defect will usually present with signs of CHF during the first few months of life. Those with a predominantly atrial level shunt may present later than an infant with both a large atrial septal defect (ASD) and VSD. The timing of presentation depends on when the PVR drops. The mechanism of CHF is similar to that described for a VSD.

Many patients with an isolated secundum ASD do not exhibit symptoms of CHF until the second or third decade of life (often during pregnancy when there is increased cardiac output).

Later in childhood, CHF may occur for many reasons, but it is a time when rheumatic heart disease may present. The mitral valve is the most commonly affected, with the finding of valve regurgitation. The mitral valve may become thickened and then will not close properly. This creates an increase in the volume load of the left atrium and the left ventricle (LV). As the regurgitation worsens, this large volume overload can dilate the LV. There will be both systolic (forward squeeze) and diastolic (ability for the LV to relax and fill) dysfunction.

An older child or adolescent with new onset CHF should be suspected of having cardiomyopathy. Often, the presentation is subtle and the decrease in function is tolerated until it becomes precipitously worse. This form of CHF is a LV failure rather than a volume overload issue. The symptoms of the infant in this critique are not of intermittent shock and there is no abdominal distension, making intussusception less likely. If the baby had worsening pneumonia as a result of aspiration, one would expect more severe hypoxemia and respiratory distress. For all the other choices, including group B streptococcal sepsis, one would not expect symptoms of heart failure such as the hepatomegaly and quiet tachypnea.

PREP Pearls
- Pediatric heart failure is often due to volume overload in infancy and early childhood.
- Heart failure can be easily mistaken for respiratory illness in infants.

ABP Content Specifications(s)
- Identify the causes of congestive heart failure in children of various ages

Suggested Readings
- Andréoletti L, Lévêque R, Boulangnon C, Brasselet C, Fornes P. Viral causes of human myocarditis. Arch Cardiovasc Dis. 2009;102(6-7):559-568. DOI: http://dx.doi.org/10.1016/j.acvd.2009.04.010.

Question 13

A 15-year-old adolescent girl presents to the emergency department with chest pain and shortness of breath. She describes the pain as a pressure in the center of her chest that is worsening. Initially, she had no difficulty breathing, but states that she is now unable to take a deep breath because of the pain. Her review of systems is positive for fevers up to 40°C for 1 week and generalized joint pain, with some swelling in the knees and wrists bilaterally.
The patient's pulse rate is 88 beats/min, respiratory rate is 17 breaths/min, and her blood pressure is 140/91 mm Hg. On physical examination, she appears pale. There is a confluent erythematous rash with raised borders and central clearing over her trunk. She has cervical lymphadenopathy, arthritis in the wrists and knees bilaterally, and a cardiac friction rub. Laboratory studies are significant for an erythrocyte sedimentation rate of 40 mm/hour and a urinalysis with 2+ protein and 3+ blood. The remainder of her laboratory studies are unremarkable. You obtain a chest radiograph (Item Q13).

Of the following, the test that would be the MOST specific for this patient's diagnosis is

    A. anti-double stranded DNA antibody
    B. antineutrophil cytoplasmic antibody
    C. antinuclear antibody
    D. antistreptolysin O titer
    E. rheumatoid factor

Question 13   **Preferred Response: A**

The patient in the vignette has signs and symptoms of systemic lupus erythematosus (SLE). The most specific diagnostic test associated with SLE is anti–double-stranded DNA antibody titer. Other antibodies associated with SLE are Smith antibody, anti-SSA, and anti-SSB antibody.

The range of signs and symptoms that can be seen in SLE is broad (Item C13A). The initial presentation of SLE can mimic several infectious diseases and cancers. Specific antibody studies are important to definitively diagnose the disease. To meet American College of Rheumatology criteria for the diagnosis of SLE, 4 of 11 criteria must be present (Item C13B), though pediatric patients with SLE often will not fully fit the adult defined criteria. It can be especially challenging to differentiate SLE from acute rheumatic fever (ARF). Although SLE and ARF share features such as fever, elevated acute phase reactants, rash, and arthritis or joint pain, they can be distinguished using the Jones criteria (Item C13C), antibody testing, organ function testing, and review of the patient's clinical presentation.

Item C13A. 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, alopecia
Endocrine	Hypothyroidism, hyperthyroidism, irregular menses, delayed puberty
Gastrointestinal	Abdominal pain, pancreatic, serositis, vasculitis, pancreatitis, hepatitis, hepatomegaly, enteritis
Hematologic	Cytopenia, leukopenia usually secondary to lymphopenia, anemia (normocytic normochromic or Coombs-positive hemolysis), thrombocytopenia, antiphospholipid syndrome, splenomegaly
Mucocutaneous	Malar 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, psychosis, acute inflammatory demyelinating polyradiculoneuropathy, autonomic disorder, mononeuropathy, myasthenia gravis, neuropathy, plexopathy, polyneuropathy
Pulmonary	Pleuritis, 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's phenomenon, nail fold capillary changes, livedo reticularis
Laboratory findings	Positive autoantibody studies including antinuclear antibodies (usually $\geq$1:160), anti-dsDNA, anti-Smith, anti-ribonuclear protein, anti-Ro, anti-La, hypo-complementemia, cytopenia, elevated erythrocyte sedimentation rate, elevated liver enzymes, elevated muscle enzymes, proteinuria, hematuria, urine casts

Courtesy of A. Brown

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Item C13B. American College of Rheumatology Revised Criteria for the Classification of Systemic Lupus Erythematosus.

Criterion	Definition
Malar 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) Pleuritis: Convincing history of pleuritic pain or rubbing heard by a physician, or evidence of pleural effusion OR b) Pericarditis: Documented by electrocardiography or rub or evidence of pericardial effusion
Renal disorder	a) Persistent proteinuria > 0.5 g/day or > 3+ if quantitation not performed OR b) Cellular casts: May be red blood 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: <4,000/μL total on 2 or more occasions OR c) Lymphopenia: <1,500/μL on 2 or more occasions OR d) Thrombocytopenia: <100,000/μL 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

Reprinted with permission from Tan EM, Cohen AS, Fries JF, et al. The 1982 revised criteria for the classification of systemic lupus erythematosus. *Arthritis and Rheumatism*. 1982;25(11):1271-1277

Item C13C. Jones Criteria.

Confirmation of preceding Group A *Streptococcus* by positive throat culture, positive rapid antigen test, or elevated (or rising) anti-streptolysin O titer
AND
2 major manifestations
OR
1 major and 2 minor manifestations

Major Manifestations	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)

Courtesy of A. Brown

The adolescent girl in the vignette has evidence of a pericardial effusion that can be life-threatening. It is important for practitioners to be aware that serositis can affect the heart and lungs of patients with SLE.

Many morbidities are associated with pediatric lupus, because of either the disease state or immunosuppressive therapy. These can include:
- Lupus nephritis, which can cause hypertension, and with poor disease control can lead to renal failure, need for dialysis, and even the need for renal transplant
- Premature atherosclerosis leading to myocardial infarction
- Increased risk of recurrent infection due to dysregulation of the immune system, from disease as well as immunosuppressive therapy
- Increased risk of malignancy
- Osteoporosis, compression fractures, and avascular necrosis resulting from steroid treatment
- Infertility, offspring with neonatal lupus, and increased fetal wastage
- Increased risk for diabetes, obesity, and growth failure

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- Increased risk of cataracts, glaucoma, and blindness
- Central nervous system manifestations, including neurocognitive disorder, seizures, and psychosis

Antineutrophil cytoplasmic antibody is positive in patients with glomerulonephritis, granulomatosis with polyangiitis (GPA) (also known as Wegener granulomatosis), microscopic polyangiitis (MPA), eosinophilic granulomatosis with polyangiitis (EGPA) (also known as Churg-Strauss syndrome), inflammatory bowel disease, and other nonautoimmune conditions. Antinuclear antibody (ANA) is positive at low titers in 10% to 30% of the population without any associated disease. Although 95% of patients with SLE have a positive ANA, this test only has 57% specificity for SLE. Antinuclear antibody can also be positive in the setting of infections such as chronic osteomyelitis, mononucleosis, hepatitis C, malaria, parvovirus B19, subacute bacterial endocarditis, and tuberculosis. It is important to note that leukemia and lymphoma can also cause ANA positivity. Antistreptolysin O titer is elevated after a streptococcal infection and can be helpful in diagnosing acute rheumatic fever and poststreptococcal reactive arthritis. The girl in the vignette has arthritis; however, she also has systemic signs and symptoms of disease that are associated with lupus and not seen with poststreptococcal disease. Rheumatoid factor may be positive in a patient with lupus, but is not specific for that diagnosis.

PREP Pearls
- An elevated anti–double-stranded DNA antibody titer is specific for the diagnosis of systemic lupus erythematosus (SLE).
- Antinuclear antibody (ANA) is positive in the majority of patients with SLE, but is not specific for the disease.
- The presence of Smith antibody is specific for SLE.

ABP Content Specifications(s)
- Recognize complications associated with systemic lupus erythematosus
- Differentiate the clinical findings of systemic lupus erythematosus from those of rheumatic fever
- Recognize the typical and atypical clinical findings associated with systemic lupus erythematosus in patients of various ages

Suggested Readings
- Levy D, Kamphuis S. Systemic lupus erythematosus in children and adolescents. Pediatr Clin North Am. 2012;59:345-364.

Question 14

At a health supervision visit, the parents of a 13-month-old boy express concern that their son's legs are very bowed. After reviewing the child's history and performing a physical examination, you diagnose physiologic genu varum.

Of the following, the feature MOST likely to be associated with this diagnosis is

A.   bilateral, symmetric bowing
B.   bowing that appears most pronounced at 3 years of age
C.   breech presentation at birth
D.   walking at an early age
E.   weight greater than the 95th percentile for his age

Question 14   **Preferred Response: A**

The child in the vignette has physiologic genu varum. Bowing of the legs is very common before 2 years of age. As children grow, the bowing gradually resolves. Most children subsequently develop physiologic genu valgum ("knock-knees"), which typically peaks around age 3 years and gradually improves by age 8 years. Adults typically have mild genu valgum, but many adults have straight legs or mild genu varum. Physiologic genu varum and genu valgum are generally bilateral and symmetric. Pediatricians should consider orthopedic referral for children older than 2.5 to 3 years with persistent genu varum, particularly if the bowing does not seem to be improving.

There are several pathologic causes of genu varum.

Vitamin D deficiency (rickets) can lead to a softening of the bones with resulting genu varum and, in most cases, short stature. Many of these children have radiographic findings in addition to genu varum, including widening of the growth plates and metaphyseal flaring.

Infantile tibia vara, sometimes referred to as infantile Blount disease, results from depression of the medial tibial physis. Risk factors include walking at an early age and obesity. Children with infantile tibia vara with progressive bowing may require surgical correction. Bracing for this condition is controversial, because no controlled trials have demonstrated efficacy.

Other causes of nonphysiologic genu varum include skeletal dysplasias, such as achondroplasia, and trauma-induced growth plate injuries, which lead to asymmetric bone growth resulting in unilateral genu valgum or genu varum, depending on the location of injury. Breech presentation is a risk factor for hip dysplasia but not for genu varum.

PREP Pearls
- Physiologic genu varum (bowing of the legs) generally resolves by age 2 years.
- Asymmetric bowing, and bowing that persists at age 3 years, should prompt evaluation for idiopathic tibia vara.

ABP Content Specifications(s)
- Recognize the clinical findings associated with various valgus and varus deformities, and understand when referral is appropriate

Suggested Readings
- Sarwark JF, LaBella C. Pediatric Orthopaedics and Sports Injuries: A Quick Reference Guide. 2nd ed. Elk Grove Village, IL: American Academy of Pediatrics; 2014:745 pp.

Question 15

A previously healthy 7-year-old boy presents to your office with a 4-day history of a "blister" on his heel that his mother believes started after he got new shoes. The lesion began as a "small bump," but now has grown in size, became red, and is draining yellow fluid. The boy reports that the lesion is increasingly painful. On physical examination, his temperature is 38°C, heart rate is 70 beats/min, respiratory rate is 20 breaths/min, and blood pressure is 100/60 mm Hg. You note a 1-cm circular lesion with surrounding erythema and swelling on the posterior aspect of the right heel that is draining purulent fluid. The remainder of the physical examination is unremarkable.

Of the following, in addition to local wound care, the BEST treatment to prescribe for this patient is oral
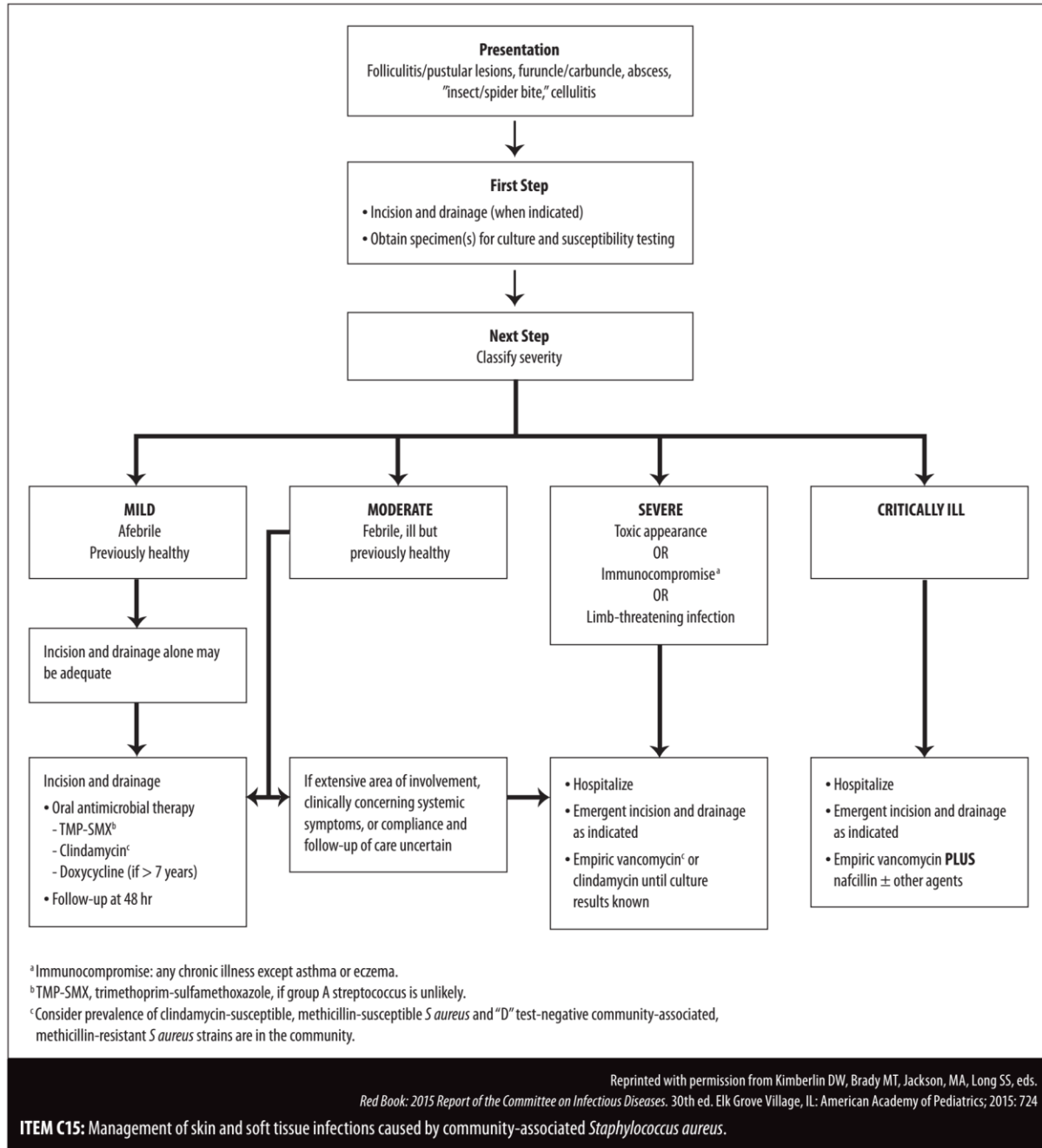
A.   cefixime
B.   clindamycin
C.   doxycycline
D.   penicillin
E.   trimethoprim/sulfamethoxazole

Question 15   **Preferred Response: B**

The patient in the vignette has a soft tissue infection after minor trauma (blister). The most likely organisms causing the infection include methicillin-resistant (MR) and methicillin-susceptible (MS)Staphylococcus aureus (SA) and Streptococcus pyogenes (group A Streptococcus, GAS). Therefore, clindamycin is the best empirical treatment. Clindamycin is a lincosamide antibiotic that disrupts bacterial protein synthesis by binding to the 50s ribosomal subunit. It has been approved by the Food and Drug Administration for the treatment of infections caused by staphylococci, streptococci, and anaerobes. Although clindamycin has been commonly associated with the development of antibiotic-associated diarrhea (including Clostridium difficile colitis) in adults, this adverse effect is relatively uncommon in children.

Culture of the fluid from the boy's lesion grew methicillin-resistant, clindamycin-susceptible S aureus. S aureus colonizes the skin and mucous membranes of approximately 30% to 50% of children and adults. More than 95% of staphylococcal disease manifests as localized infections such as cellulitis, impetigo, furuncles, abscesses, and lymphadenitis. Invasive infections such as bacteremia, pneumonia, pyomyositis, and osteomyelitis are less common. However, in the health care setting, S aureus is the second most common cause of health care–associated bacteremia and often causes pneumonia and surgical site infections. In addition, S aureus also can cause device-related (eg, intravascular catheter, pacemaker, ventriculoperitoneal shunt) infections as well as toxin-mediated (eg, toxic shock syndrome, scalded skin syndrome, food poisoning) illness.

The initial management of skin and soft tissue infections caused by community-associated S aureus is dictated by the disease severity and the prevalence and susceptibility pattern of MRSA isolates in the community (Item C15). For the boy in the vignette who was previously healthy with low-grade fever and soft-tissue infection, clindamycin is an appropriate initial choice of therapy because it has activity against streptococci as well as susceptible strains of MRSA and MSSA.

Presentation
Folliculitis/pustular lesions, furuncle/carbuncle, abscess,
"insect/spider bite," cellulitis

First Step
• Incision and drainage (when indicated)
• Obtain specimen(s) for culture and susceptibility testing

Next Step
Classify severity

MILD
Afebrile
Previously healthy

MODERATE
Febrile, ill but
previously healthy

SEVERE
Toxic appearance
OR
Immunocompromise[a]
OR
Limb-threatening infection

CRITICALLY ILL

Incision and drainage alone may
be adequate

Incision and drainage
• Oral antimicrobial therapy
  - TMP-SMX[b]
  - Clindamycin[c]
  - Doxycycline (if > 7 years)
• Follow-up at 48 hr

If extensive area of involvement,
clinically concerning systemic
symptoms, or compliance and
follow-up of care uncertain

• Hospitalize
• Emergent incision and drainage
  as indicated
• Empiric vancomycin[c] or
  clindamycin until culture
  results known

• Hospitalize
• Emergent incision and drainage
  as indicated
• Empiric vancomycin **PLUS**
  nafcillin ± other agents

[a] Immunocompromise: any chronic illness except asthma or eczema.
[b] TMP-SMX, trimethoprim-sulfamethoxazole, if group A streptococcus is unlikely.
[c] Consider prevalence of clindamycin-susceptible, methicillin-susceptible *S aureus* and "D" test-negative community-associated,
methicillin-resistant *S aureus* strains are in the community.

Reprinted with permission from Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds.
Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015: 724

ITEM C15: Management of skin and soft tissue infections caused by community-associated *Staphylococcus aureus*.

Cephalexin (first-generation cephalosporin) can be used for the treatment of MSSA, but cefixime (third-generation cephalosporin) lacks appropriate antistaphylococcal activity. Doxycycline has activity against susceptible strains of MSSA and MRSA but is not useful for treating GAS and generally is not recommended for children younger than 8 years. Although penicillin would be an appropriate choice of therapy for patients with cellulitis caused by GAS, it would be unlikely to be effective against S aureus infection which usually is penicillin-resistant. Trimethoprim-sulfamethoxazole also is effective against susceptible strains of S aureus but, like doxycycline, is not useful for treating GAS.

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PREP Pearls
- More than 95% of staphylococcal disease manifests as localized infection (eg, cellulitis, impetigo, furuncles, abscesses, and lymphadenitis).
- S aureus colonizes the skin and mucous membranes of 30% to 50% of children and adults.
- The initial management of skin and soft tissue infections caused by community-associated S aureus is dictated by the disease severity and the prevalence and susceptibility pattern of MRSA isolates in the community.
- Cephalexin is appropriate for treating MSSA, but is ineffective in treating MRSA.
- Clindamycin can treat susceptible strains of both MSSA and MRSA and also has activity against group A streptococci.

ABP Content Specifications(s)
- Understand the epidemiology of Staphylococcus aureus
- Plan the appropriate management of methicillin-sensitive and methicillin-resistant Staphylococcus aureus infection

Suggested Readings
- American Academy of Pediatrics. Group A streptococcal infections. In: Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:732-734.

Question 16

A 14-year-old male adolescent is brought to your office for a follow-up visit after being evaluated and discharged from a local emergency department the prior evening. The patient's parents took him to the emergency department after they found him stumbling around in the basement when they arrived home from work. At that time, he seemed confused, was unsteady on his feet, and his speech was slurred. His symptoms gradually resolved over approximately 1 hour. The emergency department physician ordered numerous "tests," including urine and blood drug screens, which the parents were told were "normal."

The adolescent's parents have noticed changes in his behavior over the past 3 months since he began spending time with a different group of friends. Recently, his grades have dropped and he is no longer interested in playing soccer. His mother found him and one of his friends "hanging out" in the family garage after they had told her they were going to the library. When she confronted them, the boys began to giggle uncontrollably and appeared "high." Both parents feel that their son is more irritable and withdrawn recently. Otherwise, his complete review of symptoms is unremarkable.

When you talk with the patient privately, he repeatedly denies drug or alcohol use. His vital signs are normal, and a complete physical examination reveals no abnormalities.

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

A.   obtain a random blood and urine drug screen within 1 week
B.   order a computed tomography of the brain
C.   reassure the parents that his behavior is normal for his age and developmental stage
D.   refer him to a mental health professional with expertise in substance abuse
E.   repeat his blood and urine drug screen in your office now

Question 16   **Preferred Response: D**

The ongoing symptoms reported for the adolescent in the vignette are highly suggestive of recurrent substance abuse, most likely inhalant abuse. Referral to a mental health professional with expertise in substance abuse is the best next step in management.

Substance abuse is an under-recognized cause of morbidity and mortality in children and adolescents, and is a public health priority. Pediatricians are on the front lines of preventing, recognizing, and treating drug abuse among children, so they must possess a comprehensive knowledge and understanding of this disorder.

In evaluating patients with suspected substance abuse, pediatric providers must fully understand the limitations of drug screening tests. Results of these tests are generally reported as simply "positive" or "negative," but these results can be misleading. To accurately interpret the results of drug screening tests, clinicians must understand the type of testing performed and carefully consider the clinical scenario in which the testing was ordered.

Urine drug screens are the most commonly used tests to identify substance use or exposure, though some institutions routinely perform serum drug screening as well. Certain drug assays can also be performed using hair, sweat, or saliva, but these assays are used much less commonly. Specific tests included in basic drug screening panels vary by region and institution; therefore, clinicians should know what drugs are included in the panel used by their laboratory. In the United States, basic urine screening panels for drugs of abuse typically test for amphetamines, cocaine, opioids, marijuana, and phencyclidine. Numerous drugs that are widely abused by adolescents, including designer amphetamines, such as MDMA (ecstasy), synthetic opioids (such as tramadol), synthetic marijuana, and inhalants, are not detected by many laboratories' routine drug screening panels. Thus, a negative result on drug screening cannot definitively exclude substance abuse.

Most routine drug screening tests are immunoassays that yield a positive result if a drug of abuse or its metabolite is present at or above an established threshold level at the time the sample is obtained from a patient. The time frame during which drugs are detected on screening tests varies, but for most drugs begins within minutes of exposure and lasts for a few days. A patient may have a negative drug screening result, despite recent substance use, if the level of the drug in the specimen tested falls below the threshold set for detection. As a result, patients may be able to intentionally achieve false-negative results by ingesting large amounts of water before giving a urine sample (diluting the concentration of a drug in their urine), ingesting masking agents, adding water or other adulterants to their urine samples, or even submitting a urine sample from another person or synthetic urine.

Positive drug screening results can also mislead clinicians. Many screening tests, especially immunoassays, can yield false-positive results if cross-reacting substances are present in a test specimen. For example, over-the-counter cold medications such as dextromethorphan can yield a false-positive result for phencyclidine, whereas ingestion of poppy seeds can lead to a positive result for opioids.

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Although a positive result to a drug screen may indicate that a patient has been exposed to a drug of abuse, routine screening tests generally do not indicate whether a patient is intoxicated or even symptomatic from the drug detected. Clinicians should not attribute signs and symptoms displayed by a patient to a positive result without considering and excluding other important differential diagnoses. For instance, in the scenario of a teenager presenting to an emergency department with fever and confusion and a urine drug screen that is positive for amphetamines, it would be essential to consider and exclude life-threatening conditions such as encephalitis and intracranial injury rather than immediately attributing these symptoms to acute amphetamine intoxication.

There is no clear consensus among clinicians regarding appropriate indications for drug testing to identify substance abuse among children, or follow-up for those undergoing treatment for substance abuse. A recent clinical report from the American Academy of Pediatrics regarding testing for drugs of abuse in children and adolescents provides clinicians with expert, evidence-based guidelines on this topic.

For the boy in the vignette, who has an ongoing history of behavioral changes, declining school performance, and symptoms suggestive of intermittent intoxication over the past few months, referral to a mental health professional with expertise in substance abuse is the best next step in management.

Considering their limitations, repeating the drug screening tests for this patient, either now or at a random time in the next week, will not likely aid in confirming his diagnosis and would only delay the most appropriate management, which is referral for substance abuse treatment. Pediatric providers should never disregard ongoing symptoms that are suggestive of substance abuse just because a drug screening test is negative.

Computed tomography of the brain would not contribute to the diagnosis or management of this boy's underlying problem. Given that his neurologic symptoms have been transient and self-limited, he has displayed no other "red flags" for intracranial pathology (such as persistent or worsening headaches, vomiting, or persistent neurologic deficits), his symptoms began shortly after he began spending time with a new peer group, and he had a completely normal neurologic examination at the time of emergency department follow-up, substance abuse is much more likely to be the etiology of his recent symptoms than intracranial pathology.

Finally, the progressive behavioral changes, irritability, and declining school performance displayed by this boy over the past 3 months are not consistent with normal behavior and development. Referral to a mental health professional should be made promptly to prevent the significant long-term morbidity and even mortality that can result from ongoing substance abuse.

PREP Pearls
• When interpreting results of drug screening tests, clinicians must understand the type of testing performed and carefully consider the clinical scenario that prompted the testing.

- Pediatric providers should never disregard ongoing symptoms that are suggestive of substance abuse just because a drug screening test is negative.
- Clinicians should not attribute signs and symptoms displayed by a patient to a positive drug test without considering and excluding other important differential diagnoses.

ABP Content Specifications(s)
- Understand the limitations of drug-screening tests
- Plan appropriate laboratory evaluation of substance use/abuse, including appropriate collection of test specimens and interpretation of results

Suggested Readings
- Crocetti M. Inhalants. Pediatr Rev. 2008;29:33. http://pedsinreview.aappublications.org/content/29/1/33.
- Kulig JW. Tobacco, alcohol, and other drugs: the role of the pediatrician in prevention, identification, and management of substance abuse. Pediatrics. 2005;115:816. http://pediatrics.aappublications.org/content/115/3/816.

Question 17

While making rounds in the newborn nursery, you are called emergently to the delivery room for the precipitous delivery of a neonate at 23 2/7 weeks gestation. You arrive to find a limp neonate with a heart rate of 40 beats/min and no respiratory effort. You are told the neonate weighs 470 g. As you prepare to begin resuscitation, you note microcephaly, cutis aplasia on the scalp, and a cleft lip and palate.

Of the following, the MOST appropriate indication for non-initiation of resuscitation for this neonate is

A.   birth weight less than 500 g

B.   extreme prematurity less than 24 weeks

C.   fetal aneuploidy

D.   parental request

E.   presumed birth asphyxia

Question 17   **Preferred Response: D**

The most appropriate indication for noninitiation of resuscitation in the neonate in the vignette is the request of the parents. Decisions surrounding delivery room resuscitation of neonates at 23 weeks' gestation presents a challenge to health care providers and families. Existing guidelines for delivery room resuscitation of extremely low gestation neonates remain limited and often quickly become outdated because advances in medical management lead to improved survival. A consistent recommendation is that the wishes of the parents about resuscitation should be respected when a neonate is born between 23 and 24 weeks of gestation. This is because of the high rate of mortality and high risk of diminished quality of life in surviving neonates born at this gestational age.

The ethical principles of autonomy, beneficence, nonmaleficence, and justice are tightly woven into care decisions at the edge of viability. No uniform agreement exists on the exact time of gestation when this occurs. Viability has been defined as the point at which there is a reasonable chance of survival with advanced medical support, with some ethicists arguing that resuscitation should be provided to all neonates who have at least a 50% chance of survival. Neonatal outcome data published in 2010 can be seen in Item C17, but these data may not reflect subsequent improvements in neonatal care or center-to-center variability.

Item C17. Neonatal Outcome of Extremely Premature Infants.

	Gestational Age			
	22 week	23 week	24 week	25 week
Survived	6%	26%	55%	72%
Survived without morbidity	0%	8%	9%	20%
Survived with morbidity	6%	18%	46%	52%

Adapted and reprinted with permission from Stoll BJ, et al. Neonatal outcomes of extremely preterm infants from the NICHD neonatal research network. *Pediatrics*. 2010;126(3):443-456

The sixth edition of the Neonatal Resuscitation Program of the American Academy of Pediatrics and the American Heart Association has outlined clinical situations in which noninitiation of resuscitation may be appropriate. These include confirmed gestation less than 23 weeks, birthweight less than 400 g, anencephaly, and a confirmed lethal genetic disorder or malformation. Ideally, ongoing discussions about care at the time of birth should be made jointly with the family before delivery.

The birthweight of 470 g, gestational age of 23 2/7 weeks, and stigmata of trisomy 13 aneuploidy in the neonate in the vignette may be indications for noninitiation of resuscitation, but only after discussion with the family. If the wishes of the family are unknown when a neonate with these

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findings is born, it is preferable to begin resuscitation pending discussion with the parents. Many extremely premature infants are born with bradycardia and apnea, so it would not be appropriate to assume that this indicates birth asphyxia and withhold resuscitation. If the neonate responds to resuscitation, further discussions and decisions about ongoing care may occur in the nursery with the family as active participants.

PREP Pearls
- The ethical principles of autonomy, beneficence, nonmaleficence, and justice are tightly woven into care decisions for neonates at the edge of viability.
- Parental wishes regarding resuscitation should be respected when a neonate is born between 23 and 24 weeks of gestation.

ABP Content Specifications(s)
- Recognize and apply ethical principles involved in decision making for imperiled newborn infants in the neonatal intensive care unit
- Recognize and apply ethical principles regarding imperiled newborn infants and delivery room resuscitation issues

Suggested Readings
- AAP Committee of the Fetus and Newborn. Noninitiation or withdrawal of care for high-risk infants. Pediatrics. 2007;119:401-403. DOI: http://dx.doi.org/10.1542/peds.2006-3180.

Question 18

A 14-year-old African American adolescent presents to your office for evaluation of severe headaches for the past 7 months. The headaches began shortly after the start of the school year. She had never experienced a headache before that. The headaches involve the left side of her head, are described as throbbing, and last 2 to 3 hours. During the headache, she feels nauseated and sometimes vomits. Her symptoms are somewhat relieved if she lies down in a dark room. The patient does not have any vision changes or weakness associated with the headaches, and they do not worsen with coughing, sneezing, lying down, or sitting up. They do not wake her up from sleep. She has not gained or lost weight recently. She does not take any medications other than occasional ibuprofen for the headaches. Her father has migraine headaches and her younger sister has sickle cell disease. Her blood pressure is 102/68 mm Hg, heart rate is 92 beats/min, respiratory rate is 22 breaths/min, and her body mass index is 21. Her physical examination, including neurological examination and fundoscopy, is unremarkable.

Of the following, the BEST imaging recommendation for this patient is

A.   computed tomography of the brain

B.   computed tomography venogram

C.   magnetic resonance imaging of the brain

D.   no imaging indicated at this time

E.   transcranial Doppler ultrasonography

Question 18   **Preferred Response: D**

The girl in the vignette has migraine headaches. Migraines are characterized by severe head pain, nausea, and phonophobia or photophobia. In younger children the location is typically bifrontal, and in adolescents and adults the headaches are often unilateral.

Although classic teaching is that morning headaches are suggestive of a brain tumor, in pediatric patient's migraines often occur in the morning. Time of day is not one of the diagnostic criteria for pediatric migraine, possibly because this criterion does not differentiate migraine from secondary headache very well. Red flags for pediatric headache include positional headache (worsening with lying down implies increased intracranial pressure) and headache that wakes the child from sleep.

In pediatric patients who have had migraine headaches for more than 6 months, with normal neurological examination findings, and without features suggestive of neurologic dysfunction, brain imaging is not needed. Family history of migraine is another reassuring factor that supports a diagnosis of migraine headaches. If there are red flags associated with headache (positional, sleep disrupting), the preferred brain imaging method is magnetic resonance imaging.

Magnetic resonance imaging of the brain shows the brain's structure. This is an ideal first imaging test if a structural malformation such as Chiari I malformation is suspected. Symptomatic Chiari I malformation presents with diffuse headache that is worse with a Valsalva maneuver, such as coughing, sneezing, laughing, or defecating. Symptomatic Chiari I malformation is treated with neurosurgical decompression.

If there is an urgent clinical indication, such as new encephalopathy, new focal deficit, or recent head trauma, then computed tomography of the head is the best imaging method because it is the quickest. Other imaging modalities may be indicated when specific diagnoses are being considered.

A computed tomography venogram is obtained to evaluate for cerebral sinus venous thrombosis. These cases present with headaches that have migrainous features, such as severe unilateral head pain, nausea, vomiting, and phonophobia or photophobia. The fundus examination will show papillitis and there may be a partial or full cranial nerve VI palsy (inability to abduct the eye). Treatment of cerebral sinus venous thrombosis is anticoagulation until the thrombus is resolved.

Transcranial Doppler ultrasonography is used to monitor flow velocity in the middle cerebral artery in people with sickle cell disease. Decreased flow velocity is associated with moyamoya disease. The headaches in moyamoya disease can be diffuse or unilateral, and are often provoked by exercise or activity or associated with hemiparesis. Treatment of moyamoya disease is surgical revascularization. The girl in the vignette does not have any signs of sickle cell disease or moyamoya disease, so a transcranial Doppler ultrasound is not indicated.

PREP Pearls
- Intermittent migraine headaches for at least 6 months, normal neurologic examination findings, and a family history of migraine are strongly suggestive of a diagnosis of migraine; brain imaging is not indicated if all these criteria are met.
- Headaches that wake a child from sleep or worsen with lying down are suggestive of a secondary cause, such as increased intracranial pressure, and would be an indication for neuroimaging.

ABP Content Specifications(s)
- Recognize the clinical findings associated with headaches of various etiologies
- Understand the appropriate use of neuroimaging in the evaluation of headache
- Plan the appropriate management of headache of various origins

Suggested Readings
- Bigal ME, Arruda MA. Migraine in the pediatric population: evolving concepts. Headache. 2010;50:1130-1143. DOI:http://dx.doi.org/10.1111/j.1526-4610.2010.01717.x.

Question 19

A 17-year-old adolescent with type 2 diabetes mellitus presents to the emergency department with altered mental status. He has had worsening polyuria and polydipsia for 10 days since starting perphenazine 8 mg orally once every evening for the diagnosis of schizophrenia. His only other medication is metformin 1,000 mg orally twice per day. On physical examination, his pulse is 130 beats/min and blood pressure is 108/75 mm Hg. Capillary refill is 3 seconds and extremities are cool.

Laboratory results show:
- Serum glucose, 955 mg/dL (53.0 mmol/L)
- Sodium, 150 mEq/L (150 mmol/L)
- Potassium, 4.2 mEq/L (4.2 mmol/L)
- Bicarbonate, 23 mEq/L (23 mmol/L)
- Urine ketones, negative

Of the following, the MOST important first treatment to initiate for this patient is an intravenous

A.   half-normal saline (0.45% saline solution) bolus of 20 mL/kg

B.   insulin bolus of 0.25 units/kg

C.   insulin drip at 0.05 units/kg per hour

D.   normal saline bolus 20 mL/kg, repeated as needed to restore circulatory support

E.   normal saline bolus limited to 10 to 20 mL/kg over the first 2 hours

Question 19   **Preferred Response: D**

The adolescent boy in this vignette with type 2 diabetes, very elevated glucose, and normal bicarbonate level is experiencing hyperglycemic hyperosmolar syndrome (HHS). Of reported cases of HHS in children, 20% have severe psychiatric conditions and may be taking medications associated with insulin resistance, such as the atypical antipsychotic agent taken by the adolescent in the vignette. Hyperglycemic hyperosmolar syndrome commonly occurs after prolonged and gradually increasing polyuria and polydipsia, resulting in profound dehydration, with fluid losses estimated to be twice those seen with diabetic ketoacidosis (DKA). Case fatality rates have been reported to be as high as 30%.

The mainstay of treatment for HHS is fluid resuscitation, as the degree of dehydration is more profound than that seen in DKA. The goal of initial fluid therapy is expansion of the intravascular and extravascular volume and restoration of normal renal perfusion. Vigorous fluid replacement is recommended, with rates of fluid replacement more rapid than those recommended for DKA. Fluid resuscitation that is not aggressive is associated with an increased risk of complications. Increased mortality is observed in patients with unreversed shock over the first 24 hours of admission who had received less than 40 mL/kg of intravenous fluids over the first 6 hours of treatment. Therefore, aggressive isotonic fluid replacement is recommended initially to restore perfusion, even if hypernatremia is present, followed by more hypotonic (0.45%-0.75% saline solution) fluids.

Ketosis in HHS is usually minimal. Although mild acidosis can occur, it is typically the result of hypoperfusion (lactic acidosis). Thus, in contrast to the recommended management of DKA, insulin administration should be withheld until initial fluid resuscitation has been accomplished and the serum glucose concentration is no longer decreasing from rehydration alone. When insulin treatment is begun, continuous administration at 0.025 to 0.05 units/kg per hour is recommended initially. The dose should then be titrated to achieve a decrease in glucose concentration of 50 to 75 mg/dL per hour (2.7-4.1 mmol/L per hour). Insulin therapy is not usually necessary for resolution of ketosis in HHS and should be suspended if the glucose concentration drops more than 100 mg/dL per hour (5.5 mmol/L per hour). Insulin administration as a bolus is never recommended. Treatment with an insulin drip is not the appropriate initial treatment for HHS, and unless preceded by adequate fluid resuscitation, may increase the risk of death.

PREP Pearls
- Fluid resuscitation with isotonic fluids is the mainstay of treatment for hyperglycemic hyperosmolar syndrome (HHS).
- In contrast to the management of diabetic ketoacidosis, insulin administration in HHS should be withheld until initial fluid resuscitation has been accomplished and the serum glucose concentration is no longer decreasing from rehydration alone.

ABP Content Specifications(s)
- Plan appropriate fluid therapy for a patient with hyperosmolar non-ketotic coma

Suggested Readings
- Rosenbloom AL. Hyperglycemic hyperosmolar state: an emerging pediatric problem. J Pediatr. 2010;156(2):180-184. DOI: http://dx.doi.org/10.1016/j.jpeds.2009.11.057.

Question 20

A 5-year-old boy is brought to the office for follow-up after an admission for loss of consciousness. His electrocardiogram suggested long QT syndrome and he was prescribed nadolol. When you see him for follow-up, it becomes clear that the family is not giving him the medication as prescribed. You express your concern and review the need for medication again with an explanation of the pathophysiology. The family is very concerned about adverse effects and does not believe that the medication is necessary. They agree to follow-up in a month and avoid medications that may worsen the QT prolongation. Two weeks later, a check for $1,000 arrives in your office from the family with a thank you card.

Of the following, the statement that is MOST accurate in this setting is

A.   accepting cash gifts may cause the family to expect preferential treatment

B.   cash donations should be confidential

C.   cash donations should not be sent to the development office

D.   cash gifts are permissible if they are less than $1,000

E.   cash gifts to the physician are permissible if they are used for research projects

Question 20   **Preferred Response: A**

The family in this vignette is not following medical advice. Acceptance of gifts may imply that you are in agreement with their choice when you are not, and may cause the family to expect preferential treatment. The American Medical Association suggests that each gift be evaluated case by case. Differing opinions have been expressed in the literature. General recommendations are that gifts that are too intimate, too expensive, or above the means of the family are not appropriate, and acceptance of them may create the wrong impression in the patient's or family's minds. In particular, monetary gifts of any size for personal use should not be accepted and the patient or family redirected to donate to a fund to benefit the facility or research. Any gift that makes a physician feel uncomfortable is likely one that should not be accepted. These gifts, in particular, may interfere in good patient care by influencing the physician to change their practice so as not to antagonize the family or patient. A monetary gift for personal use to the physician by patients is not recommended in any circumstance.

PREP Pearls
- Accepting monetary gifts for personal use from patients is not recommended in any circumstance.
- Expensive personal gifts are likely to disrupt the patient-physician relationship and create inappropriate expectations.

ABP Content Specifications(s)
- Understand the ethical principles that uphold professionalism and institutional ethics
- Recognize and apply ethical principles regarding professionalism and institutional ethics in the giving and receiving of gifts

Suggested Readings
- American Academy of Pediatrics. Appropriate boundaries in the pediatrician-family-patient relationship. Pediatrics. 1999;104(2):334-336. http://pediatrics.aappublications.org/content/104/2/334.full.pdf+html.

Question 21

During her health supervision visit, you note that a 7-year-old child's stature has fallen from the 25th percentile last year to less than the third percentile now. The family reports that the child has been fatigued and has had a poor appetite associated with nausea that began 6 months ago when she was hospitalized for a kidney problem in another state. She was treated with a 1-month course of oral prednisone at that time, but has not followed up since. Physical examination shows a small child for her age, with a blood pressure of 132/88 mm Hg; the remainder of the examination is unremarkable. Today, the urinalysis shows 4+ protein and 2+ blood. You have arranged an appointment with a pediatric nephrologist for tomorrow. The parents ask you about the child's growth (Item Q21).

Of the following, the MOST accurate statement regarding this child's linear growth is

A. growth failure reflects coincident hypothyroidism

B. her low growth rate can be fully explained by inadequate nutrition

C. the 1-month course of glucocorticoids was sufficient to suppress growth

D. renal failure frequently causes growth hormone resistance

E. renal transplantation results in good catch-up growth in this child's age group
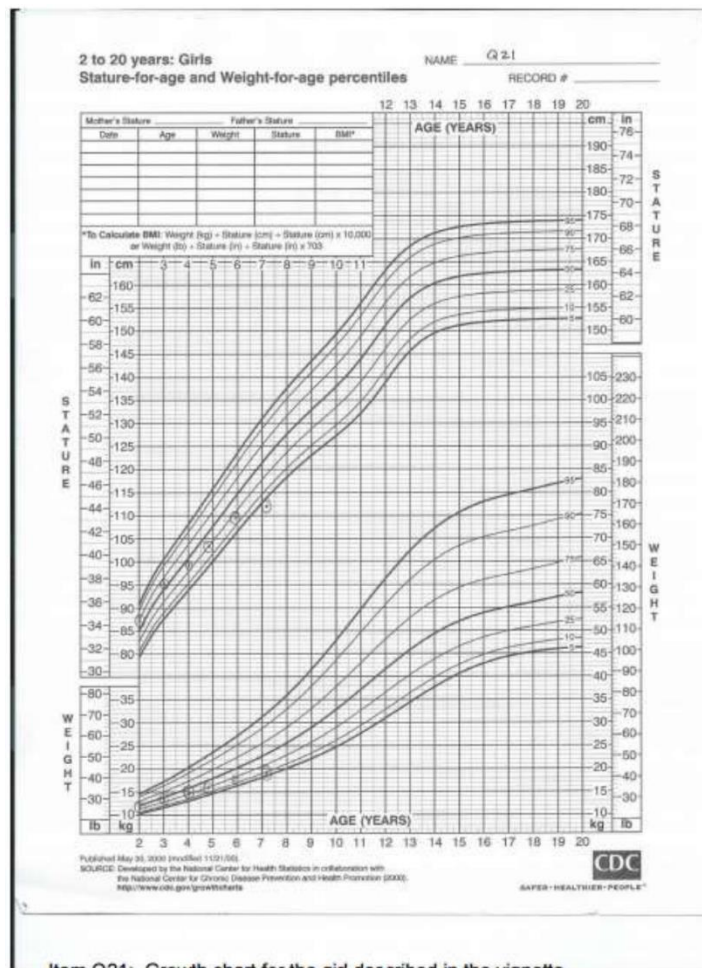
Item Q21: Growth chart for the girl described in the vignette

Question 21   **Preferred Response: D**

The patient in the vignette demonstrates impaired growth that is a common consequence of chronic kidney disease (CKD). More than one-third of children with CKD exhibit growth failure, with those younger than 1 year of age showing the greatest height deficit. Any degree of renal failure may be associated with short stature. The mechanisms producing short stature are complex and include nutritional, fluid, and electrolyte balance, and acidosis-related, hematologic, metabolic, and endocrine effects. Abnormalities of the growth hormone (GH) and insulin like growth factor-1 (IGF1) system are the primary endocrine causes of growth failure in children with CKD and involve a number of mechanisms. This is not caused by a GH deficiency; levels of GH are normal or even increased. However, there is resistance or insensitivity to GH effects as a result of decreased density of GH receptors on target organs, defective postreceptor GH signaling, and decreased bioactivity of IGF-1 caused by increased IGF-binding proteins.

Inadequate energy and protein intake is an extremely important cause of growth failure in children with CKD, and improvements in nutrition with supplementation to achieve adequate caloric intake result in improved linear growth rates. However, nutrition alone does not fully explain short stature related to CKD. Growth also improves with optimized fluid and electrolyte control, treatment of metabolic acidosis, and use of erythropoietin to prevent severe anemia. Metabolic bone disease related to CKD affects bone formation and growth plate structure, which also can lead to linear growth impairment. Prolonged steroid use affects GH secretion, decreases liver production of IGF-1, and contributes to abnormal bone formation, ultimately contributing to short stature. The child in the vignette, however, received a relatively short course of oral corticosteroids, so more complex mechanisms of growth impairment are likely to be involved in this patient. Acquired hypothyroidism is common in nephropathic cystinosis and contributes to inadequate growth, but coincident endocrine disorders are otherwise unlikely causes of growth failure in children with CKD.

Unfortunately for children with severe CKD, neither dialysis nor renal transplantation eliminates growth failure. The mean height 6 to 12 months after initiation of dialysis still shows standard deviation scores of -1.71 compared to the general population. After transplantation, there is an increase in mean height, but not enough to make up for the deficit. For children between 6 and 17 years of age at transplantation, there was no improvement in height standard deviation scores after transplantation.

PREP Pearls
• Linear growth impairment is common in chronic kidney disease and is the result of multiple, complex causes including inadequate nutrition, metabolic acidosis, fluid and electrolyte imbalance, metabolic bone disease, severe anemia, and growth hormone insensitivity.
• Dialysis and renal transplantation for children with chronic kidney disease improves growth, but will not make up for the deficits that already occurred.

ABP Content Specifications(s)
• Understand the effect of chronic disease on linear growth velocity

Suggested Readings
• Cooke DW, Divall SA, Radovick S. Normal and aberrant growth. In: Melmed S, Polonsky KS, Larsen PR, Kronenberg HM, eds. Williams Textbook of Endocrinology. 12th ed. Philadelphia, PA: Saunders Elsevier; 2011:935-1053 .

Question 22

An adolescent who is 13 years of age presents to the office with severe lower abdominal pain. Her mother reports that she has been complaining about this pain intermittently for the last 4 months. The patient and her mother deny any history of trauma. The patient has had no fever, nausea, vomiting, diarrhea, or dysuria. She has had constipation and took over-the-counter stool softeners. Her appetite is decreased when she has pain. She has had no weight loss. She is premenarchal. She describes the pain as sharp. She says that the pain usually lasts for a few days and is somewhat relieved by nonsteroidal anti-inflammatory medications. On physical examination, she is afebrile. Her blood pressure and heart rate are normal for her age and body mass index is at the 80th percentile. Her cardiovascular and pulmonary examinations are unremarkable. She has a midline suprapubic mass with associated tenderness. She is sexual maturity rating 4 for breasts and pubic hair. She has a bluish, bulging discoloration in her perineal region. Her abdominal ultrasonography is shown in Item Q22.
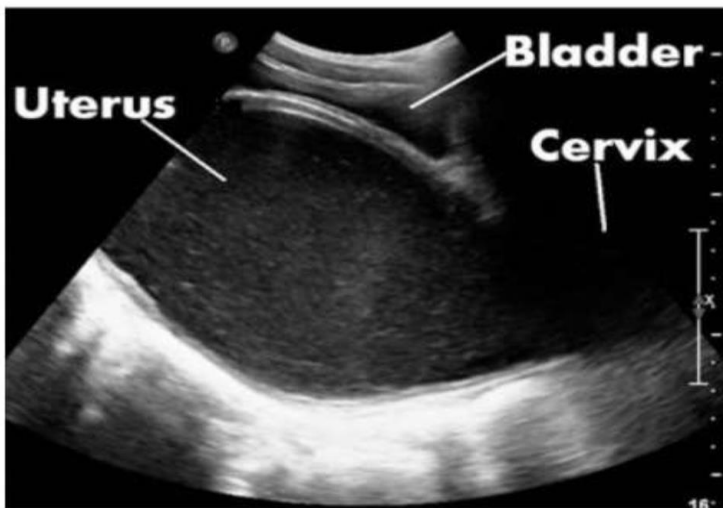
Item Q22. Abdominal ultrasonography of the patient in the vignette. Reprinted with permission from Domany E, Gilad O, Shwarz M, Vulfsons S, Garty BZ. Pediatrics. 2013;132(3):e768-e770.

Of the following, the MOST likely diagnosis for this patient is

A.   agenesis of vagina

B.   imperforate hymen

C.   labial adhesion

D.   low transverse vaginal septum

E.   uterine duplication

Question 22   **Preferred Response: B**

An imperforate hymen is the most commonly diagnosed congenital anomaly resulting from abnormal Müllerian duct development and is seen in 1 out of 2,000 females. Symptoms of imperforate hymen include lack of menses, cyclical abdominal or back pain, urinary retention, constipation, and lower extremity edema. On physical examination, an abdominal mass can be palpated, as well as a bluish bulging mass at the introitus from accumulation of menstrual bleeding. On ultrasonography, a pelvic hypoechoic mass can be visualized. Treatment of an imperforate hymen requires a hymenotomy, a surgical resection of the membrane.

A low transverse vaginal septum can present with similar clinical symptoms. However, a low transverse vaginal septum can be distinguished from an imperforate hymen with the Valsalva maneuver. During the Valsalva maneuver, bulging is seen with an imperforate hymen, and not with a low transverse vaginal septum.

Vaginal agenesis is typically characterized by absence of the proximal vagina and absence or hypoplasia of the uterus. In addition to those findings, ultrasonography may reveal additional abnormalities such as urinary tract anomalies.

Uterine duplication anomalies represent malformations related to failed fusion of the Müllerian duct structures. Uterine duplication anomalies are often asymptomatic unless an obstruction is present. Ultrasonography would reveal separate divergent uterine horns.

Labial adhesions occur typically in the prepubescent population before the production of endogenous estrogen, which starts at puberty.

PREP Pearls
- Imperforate hymen often presents during adolescence as lack of menses and cyclical abdominal pain with bluish bulging present at the introitus.
- Treatment of an imperforate hymen requires a hymenotomy, a surgical resection of the membrane.

ABP Content Specifications(s)
- Recognize the clinical findings associated with imperforate hymen

Suggested Readings
- Bacon JL. Prepubertal labial adhesions: evaluation of a referral population. Am J Obstet Gynecol. 2002;187(2):327-332. DOI: http://dx.doi.org/10.1067/mob.2002.126201.

Question 23

A 4-year-old girl presents for evaluation of bumps on the arm for several weeks. There have been no associated symptoms and the girl is otherwise well. Her physical examination is remarkable only for several papules located in the left antecubital fossa (Item Q23).
Of the following, the MOST likely diagnosis is

A.   closed comedones

B.   common warts

C.   flat warts

D.   milia

E.   molluscum contagiosum

Item Q23. Papules as described for the girl in the vignette. Reprinted with permission from Krowchuk DP, Mancini AJ, eds. Pediatric Dermatology. A Quick Reference Guide. Elk Grove Village, IL: American Academy of Pediatrics; 2012.

Question 23   **Preferred Response: E**

The girl described in the vignette has several "translucent" papules (ie, that mimic vesicles) located in the left antecubital fossa, some of which have a central umbilication (depression or dell) (Item C23A). The latter finding is typical of lesions of molluscum contagiosum. Molluscum contagiosum (MC) is a common poxvirus infection that is spread by direct contact including sexual contact, autoinoculation, or fomites. It is especially prevalent in children who have atopic dermatitis, with a disrupted skin barrier and impaired cutaneous immunity. Papules vary in number and range in size from 1 to 6 mm in diameter, and can affect most body surfaces, although involvement of the palms, soles, and mucous membranes is rare. Early lesions often are small and lack umbilication. Dermatitis surrounding the lesions of MC is common (Item C23B). It is not known whether this represents a host response to the virus or underlying atopic dermatitis. At times, individual MC lesions enlarge and become erythematous, suggesting possible secondary bacterial infection. In many cases, however, this represents an inflammatory reaction that heralds lesion resolution (the "beginning of the end [BOTE]" sign) and antibiotic treatment is unnecessary (Item C23B).

Item C23A. Lesions of molluscum contagiosum are white or translucent papules, some of which have a central umbilication (depression or dell, arrows). Reprinted with permission from Krowchuk DP, Mancini AJ, eds. Pediatric Dermatology. A Quick Reference Guide. Elk Grove Village, IL: American Academy of Pediatrics; 2012.
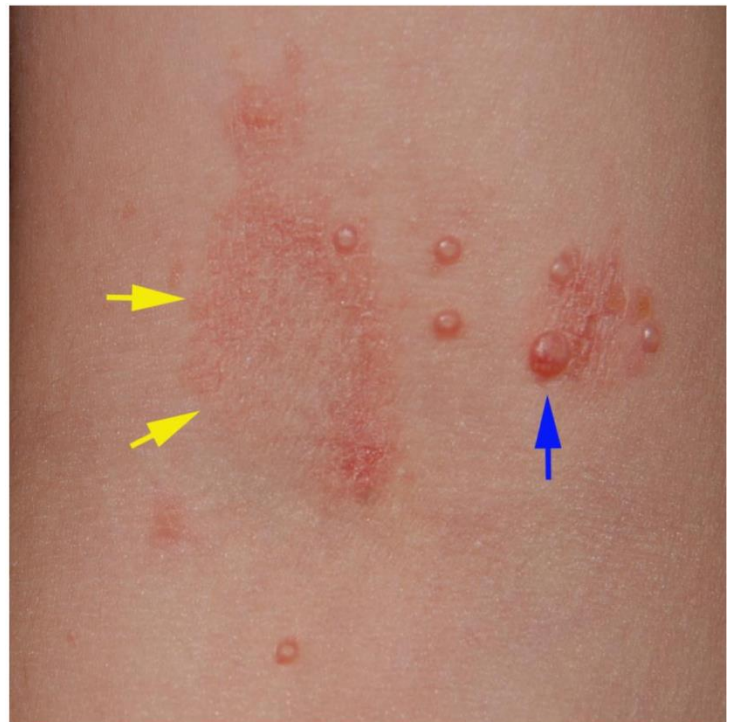
Item C23B. Dermatitis (yellow arrows) surrounding molluscum contagiosum lesions is common. One lesion (blue arrow) is enlarged and inflamed, consistent with the "beginning of the end sign." Courtesy of D. Krowchuk.

Item C23C. Differential Diagnosis of Molluscum contagiosum (MC).

Disorder	Differentiating features
Closed comedones	- Lesions are uniform in size and appearance and smaller than those of MC - Lesions often are limited to the face
Common warts	- Papules have a rough surface and usually are larger than those of MC - Dark specks (thrombosed capillaries) may be present
Flat warts	- Lesions are flat-topped papules or small, thin plaques
Milia	- Tiny white papules that are uniform in size

Adapted and 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.

Several disorders can mimic MC, but in each, the lesions lack umbilication. These entities and their differentiating features are summarized in Item C23C.

Individual lesions of MC may resolve in 1 to 2 months, but often are replaced by new papules. As a result, the condition may persist for months to years (average 13 months). Molluscum contagiosum is self-limited, therefore no intervention would be a reasonable choice for children who have only a few asymptomatic lesions. If treatment is desired, several options exist. Especially in young children who do not tolerate discomfort well, cantharidin (a blister beetle extract) may be applied in the office to individual lesions. Within several hours of application, a blister may form, after which the MC lesion resolves. Alternative painless topical agents that may be applied at home include salicylic acid and a topical retinoid (eg, tretinoin or adapalene). None of these topical therapies are US Food and Drug Administration-approved for the treatment of molluscum contagiosum. For children who can tolerate discomfort, cryotherapy and curettage are effective.

PREP Pearls
- Lesions that become enlarged and inflamed often represent the body's inflammatory response to the infection.
- Molluscum contagiosum is self-limited and does not require treatment.
- The lesions of molluscum contagiosum are skin-colored or "translucent" papules that have a central umbilication.

ABP Content Specifications(s)
- Recognize the clinical findings associated with molluscum contagiosum, and manage appropriately

Suggested Readings

- Butala N, Siegfried E, Weissler A. Molluscum BOTE sign: a predictor of imminent resolution. Pediatrics. 2013;131(5):e1650-e1653. DOI: http://dx.doi.org/10.1542/peds.2012-2933.

Question 24

A 15-year-old adolescent collapses while playing in a high school basketball game without any preceding trauma. He is unconscious and not breathing.
Of the following, the BEST sequence of actions is to

A.   administer a precordial thump and perform a jaw-thrust maneuver

B.   ask someone to retrieve an automatic external defibrillator and start chest compressions

C.   immobilize the cervical spine and attempt to palpate the carotid pulse

D.   perform a chin-lift maneuver and start rescue breathing

E.   perform rapid assessment of circulation/airway/breathing and call 911

Question 24   **Preferred Response: B**

The boy in the vignette most likely has suffered a sudden cardiac arrest. A less severe cardiovascular condition with a perfusing rhythm is not as likely because he is not breathing. Although a neurologic catastrophe (eg, trauma or spontaneous hemorrhage of a cerebrovascular malformation) should be considered as a possible cause of this event, it is significantly less likely without a supporting history. The best response choice is to ask someone to retrieve an automatic external defibrillator (AED) and start chest compressions.

Appropriate life support responses for children include the algorithms of basic life support, in which it is assumed that there is only 1 responder, and pediatric advanced life support, which takes place in an environment in which many rescuers are involved and actions can be undertaken simultaneously. Regardless of the environment, etiology, or age of the patient in cardiac arrest, the effectiveness of life support efforts depends on the quality of the cardiopulmonary resuscitation (CPR). Chest compressions and rescue breathing should be immediately started. For CPR in children it is recommended to push hard (greater than 1/3 of the anterior-posterior diameter of the chest), push fast (at least 100 compressions per minute), minimize interruptions between compressions, and rotate the person giving compressions every 2 minutes. If no advanced airway, ie, an endotracheal tube or laryngeal mask airway, is present, a 15:2 compression-ventilation ratio should be followed. If an advanced airway is in place, 8 to 10 breaths per minute should be given with continuous chest compressions. CPR recommendations for infants also include a 15:2 compression-ventilation ratio, however, newborns with cardiac arrest of cardiac origin in the delivery room or neonatal intensive care unit should receive a 3:1 compression-ventilation ratio.

Because the boy in the vignette presented with collapse and apnea, he is unlikely to have a perfusing rhythm. Palpation for a pulse in this setting may not be accurate, and could lead to a delay in definitive care. Even rapid assessment of circulation/airway/breathing is not necessary given the obvious gravity of the child's status and could lead to further delays in treatment. The cause of his collapse is likely cardiac, therefore jaw-thrust or chin-lift maneuvers would not be helpful. Although rescue breathing should be performed, it is not the best next step, because the therapy most likely to restore spontaneous circulation is cardioversion or defibrillation using an AED. The precordial thump is no longer recommended in the latest American Heart Association guidelines. In the absence of trauma, immobilization of the cervical spine is not recommended. Although 911 should be called, it is likely that an AED is available in the high school gymnasium and may be effective before the arrival of emergency medical services.

PREP Pearls
- Effective cardiopulmonary resuscitation includes pushing hard (to 1/3 of the anterior-posterior diameter of the chest), pushing fast (100 compressions per minute) with minimal interruptions, and rotating personnel performing compressions every 2 minutes.
- The first steps for a child with a sudden cardiac arrest are to call for an automatic external defibrillator (AED) and to start chest compressions immediately, especially in a public place where an AED is likely to be available.

ABP Content Specifications(s)
- Understand the correct method for cardiopulmonary resuscitation in patients of various ages

Suggested Readings
- 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:http://dx.doi.org/10.1161/CIRCULATIONAHA.110.971119.

Question 25

A 7-week-old term infant presents to the emergency department in July for evaluation of fever. He has had 3 days of diarrhea and yesterday was noted to have a rash. Vital signs show a temperature of 38.5°C, respiratory rate of 50 breaths/min, heart rate of 196 beats/min, blood pressure of 92/56 mm Hg, and his weight is 4.84 kg. On physical examination, he is irritable and has a faint erythematous maculopapular rash principally over the lower extremities. Laboratory data shows:

- White blood cells, 8,330/μL (8.33 x 10^9/L)
- Hemoglobin, 9.3 g/dL (93 g/L)
- Platelets, 479 x 10^3/μL (479 x 10^9/L)
- Differential count, 29% segmented neutrophils, 28% bands, 30% lymphocytes, 11% monocytes, 2% eosinophils
- Cerebrospinal fluid (CSF) white blood cells, 128/μL
- CSF red blood cells, 5/μL
- CSF glucose, 50 mg/dL (2.8 mmol/L)
- CSF protein, 63 mg/dL

Of the following, the CSF fluid study that is MOST likely to establish the diagnosis in this child is

A.   arbovirus panel

B.   enterovirus polymerase chain reaction

C.   herpes simplex virus polymerase chain reaction

D.   viral culture

E.   West Nile antibodies

Question 25   **Preferred Response: B**

The clinical presentation of febrile illness in a young infant occurring during the summer characterized by diarrhea and a maculopapular rash with the additional finding of cerebrospinal fluid (CSF) pleocytosis, while not pathognomonic for enterovirus, is suggestive of the diagnosis. Enterovirus polymerase chain reaction (PCR) on the CSF is most likely to confirm the diagnosis. A subset of patients with enterovirus infections will have a positive CSF PCR assay even in the absence of CSF pleocytosis.

While arboviral infections, including West Nile virus, can present with a nonspecific febrile illness or aseptic meningitis, they are a less common etiology in this age group, as they would require contact with the respective vector. Additionally, diarrhea is not usually associated with arboviral infections. The most common means of identifying arboviral infections is through the measurement of antibodies in either CSF or serum. Immunoglobulin M is usually detectable by 3 to 8 days in patients with arboviral infections. However, if an arboviral infection is strongly suspected and testing is negative within 10 days of the onset of illness, convalescent testing is recommended. Nucleic acid amplification tests for some arboviral infections are available only through select reference laboratories or the US Centers for Disease Control and Prevention.

While herpes simplex virus infection can present as a febrile illness with associated meningoencephalitis, an exanthem, if present, would be expected to be vesicular in nature. In patients with central nervous system (CNS) infection caused by herpes simplex virus, a PCR assay is considered to have greater sensitivity than cell culture.

While it is possible to grow some enteroviruses in culture, PCR assays have superior sensitivity. In the setting of CNS infection, the yield of CSF viral culture for enterovirus ranges between 0% to 80%, depending on the serotype and cell line used. Patients with neurologic disease caused by enterovirus 71 can have negative results with both CSF culture and PCR, and it is recommended that throat and rectal swabs be obtained to identify enterovirus at these sites. Echoviruses 22 and 23 have been reclassified as human parechoviruses. They can cause clinical illness indistinguishable from enteroviruses. Commercially available enteroviral PCR assays do not detect parechoviruses.

PREP Pearls
- Depending on geographic location and season, enteroviral infection should be considered a causative agent for acute febrile illness in a young infant.
- In enterovirus and herpes simplex virus central nervous system infections, cerebrospinal fluid (CSF) polymerase chain reaction assays have greater sensitivity than viral culture.
- Antibody testing in either CSF or serum is the preferred method for diagnosing arboviral infections.

ABP Content Specifications(s)
- Plan the appropriate diagnostic evaluation of meningitis of various etiologies

Suggested Readings

- American Academy of Pediatrics. Enterovirus (nonpoliovirus). In: Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:333-335.

Question 26

A 14-year-old adolescent presents to the emergency room (ER) with severe abdominal pain for the last 10 hours. There is no history of fever, burning on urination, or trauma. You note in the electronic medical record that the patient had 2 other ER visits in the last year, with complaints of abdominal pain and vomiting with spontaneous resolution of the symptoms.

On physical examination, the patient is anxious. The vital signs show a temperature of 37°C, heart rate of 110 beats/min, respiratory rate of 24 breaths/min, and blood pressure of 129/82 mm Hg. An abdominal examination reveals tenderness of the left upper abdomen and a mass on the left side.

Complete blood cell count, blood urea nitrogen, serum creatinine, electrolytes, and liver function tests all yield normal results. His urinalysis demonstrates a specific gravity of 1.035, pH of 6.0, 3+ blood, and no leukocyte esterase, protein, or nitrites. His urine microscopy shows 10 to 20 red blood cells/high power field, less than 5 white blood cells/ high power field, and no crystals and no bacteria. The significant finding on the patient's abdominal ultrasound is shown in Item Q26. Of the following, the MOST likely cause of the patient's symptoms is
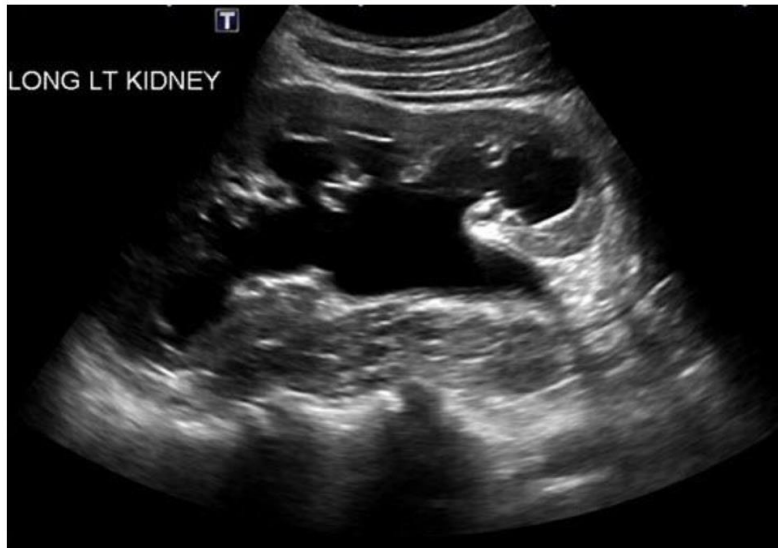
Item Q26. Abdominal ultrasonography for the patient described in the vignette. Courtesy of G. Kapur.

A.   autosomal dominant polycystic kidney

B.   multicystic dysplastic kidney

C.   uretropelvic junction obstruction

D.   vesicoureteral reflux

E.   Wilms tumor

Question 26   **Preferred Response: C**
The most common cause of congenital obstructive uropathy is ureteropelvic junction (UPJ) obstruction. Congenital UPJ obstruction is most commonly diagnosed upon postnatal evaluation of antenatal hydronephrosis (dilatation of the renal pelvis with or without dilatation of the renal calyces) detected on maternal ultrasonography screening. In the absence of antenatal screening, newborns with UPJ obstruction usually present with a palpable abdominal mass caused by an enlarged obstructed kidney. Other less common presentations include urinary tract infection, hematuria, or failure to thrive. Ureteropelvic junction obstruction rarely presents as renal failure. Renal failure secondary to UPJ obstruction is seen in patients with a single obstructed kidney or with bilateral UPJ obstruction. Older children with UPJ obstruction present with episodes of flank or abdominal pain (Dietl crisis). These symptoms may be accompanied by nausea and vomiting. Therefore, painful episodes secondary to UPJ obstruction may be confused with episodes of gastritis and managed as such. These patients may return to the emergency room with history of recurrent episodes of flank or abdominal pain, and may even have extensive negative evaluations for abdominal pain. The pain associated with UPJ obstruction may worsen during excessive diuresis, as seen after excessive water or caffeine intake. Children may also rarely present with:
1.   renal injury to the enlarged obstructed kidney after minor trauma
2.   hematuria
3.   renal calculi
4.   hypertension

In older patients, findings of hydronephrosis on ultrasonography are the clue to the possibility of UPJ obstruction as the underlying cause of the patients' symptoms (Item C26). It is important to perform an ultrasonographic examination during episodes of acute pain, as this may be normal once the pain subsides.

Diuretic renal scan (renal scan along with administration of a furosemide) is used to confirm the diagnosis of urinary tract obstruction. It measures the drainage time of the radioisotope (technetium-99m-mercaptoacetyltriglycine [Tc99mMAG3]) from the renal pelvis (evaluated as a washout curve). The timed excretion of the radioisotope correlates with the degree of obstruction. Administration of furosemide results in a prompt washout in nonobstructed kidneys, while a half-life greater than 20 minutes to clear the isotope from the kidney is indicative of obstruction.

Vesicoureteral reflux (VUR) is the retrograde passage of urine from the bladder to the kidneys. Vesicoureteral reflux may present prenatally as hydronephrosis on maternal ultrasonographic screening or be diagnosed in children after an episode of urinary tract infection (UTI) (usually before 6-7 years of age). Vesicoureteral reflux is less likely the cause of hydronephrosis in this patient, as the patient has no symptoms suggestive of current or prior UTIs.

Voiding cystourethrogram showing reflux of urine from the bladder to the upper urinary tract by either contrast or radioisotope is diagnostic for VUR. Normally, reflux of urine is prevented by compression of the intravesical ureter by the contracting bladder muscles. The shorter

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intravesical ureter (which may be genetically linked) has been implicated in the failure of the antireflux mechanism, thus leading to primary VUR. Secondary VUR occurs because of abnormally high pressures in the bladder, leading to incompetence of the ureterovesical junction and associated reflux. In more than 50% of patients, the reflux resolves spontaneously. Lower grade of reflux, unilateral reflux, prenatal hydronephrosis, and diagnosis before 1 year of age 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 I to grade IV VUR. Although patients with grade I and grade II VUR continue to have high rates of bilateral reflux resolution, only 10% to 20% of patients with grade III and grade IV bilateral reflux experience spontaneous resolution. Grade V reflux rarely resolves spontaneously, therefore these patients usually require surgical intervention.

Patients with multicystic dysplastic kidney are usually asymptomatic. Multicystic dysplastic kidney 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 VUR.

Autosomal dominant polycystic kidney disease (ADPKD) is considered an adult onset disease, with clinical manifestations developing later in life. Children with ADPKD are usually asymptomatic. Patients are diagnosed with ADPKD during ultrasonographic evaluation for other abdominal symptoms or by screening of children with strong family history. Infrequently, children with ADPKD may present with hematuria (gross or microscopic), flank or abdominal pain, infection (of the cyst), or asymptomatic hypertension. Patients with ADPKD have renal macrocysts that are visible on ultrasonographic imaging of the kidneys.

The classic presentation of Wilms tumor is abdominal swelling with or without associated symptoms, including abdominal pain, hematuria, and hypertension. Physical examination reveals a firm, nontender, smooth mass that usually does not cross the midline.

PREP Pearls
- In patients with antenatally detected hydronephrosis and postnatal genitourinary abnormalities, ureteropelvic junction (UPJ) obstruction is the most common and vesicoureteral reflux is the second most common diagnosis.
- Older children with UPJ obstruction present with episodes of flank or abdominal pain (Dietl crisis).
- Hydronephrosis on renal ultrasonography is the clue to the possibility of UPJ obstruction as the underlying cause of the abdominal or flank pain in patients.
- Ultrasonography should be performed during episodes of acute pain, as the examination may be normal once the pain subsides.

ABP Content Specifications(s)
- Recognize the clinical findings associated with hydronephrosis in patients of various ages
- Recognize complications associated with hydronephrosis

<u>Suggested Readings</u>

- Carmody JB, Carmody RB. Question from the clinician: management of prenatal hydronephrosis. Pediatr Rev. 2011;32(12):e110-e112. DOI: http://dx.doi.org/10.1542/pir.32-12-e110.

Question 27

You are caring for a 16-year-old adolescent diagnosed 1 year ago with Crohn disease, with fistulas affecting the ileum and colon. Despite aggressive medical management, she required resection of her ileum, including her ileocecal valve 4 months ago. Since her surgery, she has had persistent diarrhea. She is currently on a regular, low fiber diet without supplementation. Stool studies demonstrate normal bacterial flora, negative reducing substances, and are heme negative.

Of the following, the patient's diarrhea is MOST likely caused by malabsorption of

A.   bile

B.   fat

C.   fructose

D.   lactose

E.   sucrose

Question 27   **Preferred Response: A**

Crohn disease is a chronic inflammatory condition of the gastrointestinal tract. In addition to having a chronic disease, the child in this vignette has lost most of her ileum. Absorption of bile occurs in the ileum and malabsorption results in chronic diarrhea, as in the case in the girl in this vignette. Disaccharides (lactose, fructose, sucrose, etc) are digested and absorbed in the duodenum and jejunum. Small bowel inflammation may result in injury to villi, causing a secondary disaccharidase deficiency resulting in diarrhea. Malabsorption of carbohydrates results in increased stool reducing substances, not seen in the child in the vignette. Fat is digested and absorbed in the proximal intestine, and can be malabsorbed in severe chronic inflammation, however, this is fairly uncommon. Elevated fecal fat levels would identify this as a possible etiology.

Crohn disease may affect the small bowel and the colon. Malnutrition can occur in Crohn disease caused by chronic inflammation, as an adverse effect of medication or due to surgical resection. The most common cause of nutritional issues in Crohn disease is inadequate intake of calories or protein. Patients with active disease have a significant increase in their nutritional needs that exceed their ability to ingest sufficient calories. Additional nutritional deficiency concerns resulting from gastrointestinal disorders are listed in Item C27.

PREP Pearls
- Patients with Crohn disease are at increased risk for malnutrition and nutritional deficiency.
- Understanding the anatomy following gastrointestinal resection will help to identify nutrients that may require additional supplementation.
- Malabsorption of bile, disaccharides, and fats can result in worsening diarrhea.

ABP Content Specifications(s)
- Recognize the nutritional deficiencies associated with gastrointestinal disease

Suggested Readings
- Critch J, Day AS, Otley A, King-Moore C, Teitelbaum JE, Shashidhar H. Use of enteral nutrition for the control of intestinal inflammation in pediatric Crohn disease. J Pediatr Gastroenterol Nutr. 2012;54(2):298–305. DOI:http://dx.doi.org/10.1097/MPG.0b013e318235b397.

Item C27. Nutritional Deficiency Concerns Resulting From Gastrointestinal Disorders in Addition to Protein and Caloric Intake.

Nutritional Deficiency	Pathophysiology
Fat	Small bowel inflammation or short bowel syndrome due to resection can lead to fat malabsorption
Iron	Chronic blood loss, inadequate iron consumption
Vitamin B12	Significant ileitis or resection of the ileum results in inadequate absorption
Folic acid	Jejunal inflammation or resection of the jejunum results in inadequate absorption
	Sulfasalazine affects metabolism of folate requiring folic acid supplementation
Sodium and potassium	Both can be lost with excessive diarrhea
	Patients will require supplementation following colectomy as the colon is critical to salt regulation
Calcium	Inadequate intake
	Poor absorption can be a complication of chronic steroid use
Vitamins A, D, E, and K	Fat malabsorption
	Medications can interfere with absorption
Magnesium	Can be lost with chronic diarrhea
	Chronic inflammation of jejunum and ileum with poor absorption
Zinc	Chronic jejunal inflammation
	Chronic diarrhea with increased losses
	Medications may affect absorption

Courtesy of C. Waasdorp Hurtado

Question 28

You are called to the nursery to evaluate a male newborn who has intermittent cyanosis, increased work of breathing, and feeding problems. The cyanosis improves with crying. You are unable to pass an 8-French catheter through both nares. On physical examination, you note a heart murmur, left-sided facial droop, abnormal ears with no lobes, a left iris coloboma, and cryptorchidism.

Of the following, the MOST likely diagnosis is

A.   22q11.2 deletion syndrome

B.   branchiootorenal syndrome

C.   CHARGE syndrome

D.   renal coloboma syndrome

E.   VACTERL association

Question 28   **Preferred Response: C**

The newborn in this vignette has classic CHARGE syndrome. CHARGE is a mnemonic for coloboma, heart defects, choanal atresia, retarded growth and development, genital abnormalities, and ear anomalies. Neonates with this condition can present with multiple life-threatening conditions, necessitating immediate evaluation of the heart, airway, feeding, genitourinary tract, and hearing (Item C28A). A multidisciplinary team should be assembled to address the surgical correction of the choanal atresia, potential need for a tracheostomy if the airway is significantly compromised, cardiac evaluation for heart defects, feeding assessment due to the possibility of tracheoesophageal fistula and swallowing dysfunction, appropriate therapies, potential need for a gastrostomy placement, and hearing aids if hearing loss is noted.

Item C28A. Evaluations for Suspected CHARGE Syndrome and Types of Clinical Findings.

Evaluations for Suspected CHARGE Syndrome	Types of Clinical Findings
Dilated ophthalmologic examination	Colobomas, strabismus, refractive error, central vision impairment
Echocardiogram/electrocardiogram	Cardiovascular anomalies (conotruncal anomalies, atrioventricular canal defects, aortic arch anomalies, atrial septal defect, ventricular septal defect, and patent ductus arteriosus)
Nasal endoscopy or computed tomography scan for choanal anomalies	Unilateral or bilateral choanal atresia and/or stenosis
Ear, nose and throat and audiologic evaluation (BAER)	Hearing loss (both sensorineural and conductive), chronic recurrent otitis media
Computed tomography scan of the temporal bones	Middle ear and inner ear defects
Clinical examination for cleft palate	Palatal abnormalities including submucous cleft palate
Physical examination for facial palsy and swallowing studies	Cranial nerve dysfunction especially cranial nerve IX/X abnormalities, swallowing dysfunction, gastroesophageal reflux disease
Posterioranterior and lateral plain chest radiographs	Esophageal atresia or tracheoesophageal fistula, high risk for aspiration
Renal ultrasound and physical examination for genitourinary anomalies	Micropenis, cryptorchidism, labial hypoplasia, hypogonadotropic hypogonadism, renal anomalies
Genetics consultation	Formal examination and genetic counseling for families
Early intervention referral	Speech/language delays, motor delays, cognitive ranges from major learning disability to almost normal

Courtesy of L. Parsley

The diagnosis of CHARGE syndrome is based on clinical findings and imaging. The only known gene to be associated with CHARGE syndrome is the CHD7 gene. This gene mutation is detectable in 65% to 70% of people presenting with CHARGE syndrome. Less than 2% of patients with CHARGE syndrome are caused by a gene deletion. This disorder is autosomal dominant in inheritance. Clinical testing is available.

The diagnostic criterion for CHARGE syndrome is as follows:
- Definite CHARGE syndrome - A patient must have 4 major characteristics or 3 major and 3 minor characteristics (Item C28B shows major and minor characteristics)
- Probable CHARGE syndrome - A patient must have 1 or 2 major characteristics and several minor characteristics (Item C28B)

Item C28B. Diagnostic Criteria for CHARGE Syndrome.

Major Diagnostic Criteria
Ocular colobomas
Choanal atresia
Cranial nerve dysfunction or anomaly, including anosmia, facial palsy, auditory nerve hypoplasia, or swallowing problems
External ear anomalies, middle ear defects, abnormalities of the cochlea, temporal bone abnormalities
Minor Diagnostic Criteria
Genital hypoplasia
Developmental delay
Cardiovascular anomalies
Growth deficiency
Cleft lip and/or palate
Tracheoesophageal fistula or esophageal atresia
Facial dysmorphology (square face with prominent forehead, flat midface, broad nasal root)

Courtesy of L. Parsley

The patient in the vignette had all 4 major criterions and several minor criterions. Item C28C shows a patient with CHARGE syndrome.

22q11.2 deletion syndrome presents with congenital heart disease, palatal abnormalities, characteristic facial features, learning problems, hypocalcemia, immune deficiency, kidney abnormalities, and hearing loss. Patients do not have colobomas or choanal atresia.
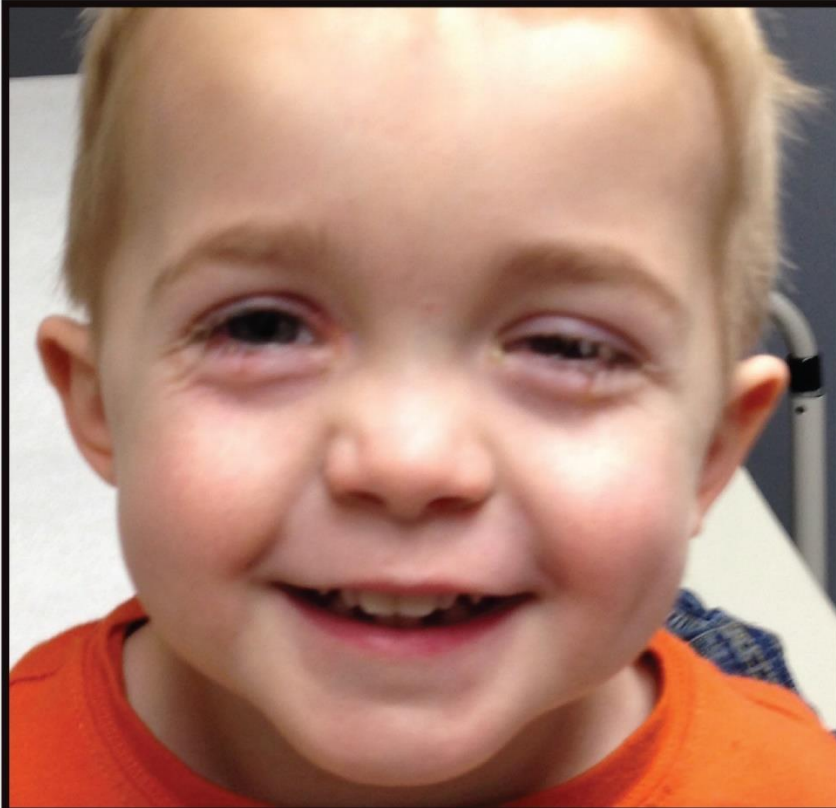
Courtesy of L. Parsley

ITEM C28C: Patient with CHARGE syndrome.

Branchio-oto-renal syndrome is characterized by deafness, external ear deformities, lateral semicircular canal hypoplasia, branchial arch anomalies, and renal malformations. The lack of other major criterion in CHARGE syndrome makes it easily distinguishable. It is caused by EYA1 gene mutations.

Renal coloboma syndrome presents with kidney abnormalities, retinal and optic nerve colobomas, and sometimes hearing loss. Other multiple congenital anomalies are not seen. It is caused by PAX2 gene mutations.

VACTERL association presents with a constellation of vertebral anomalies, anal atresia, cardiac anomalies, tracheoesophageal fistula or esophageal atresia, and renal and limb anomalies. The absence of colobomas, choanal atresia, ear deformities, and cranial nerve abnormalities distinguishes it from CHARGE syndrome.

PREP Pearls
- CHARGE syndrome is a mnemonic for a constellation of clinical findings including coloboma, heart defects, atresia choanae, retarded growth and development, genital abnormalities, and ear anomalies.
- Neonates with this condition can present with multiple life-threatening conditions, necessitating immediate evaluation of the heart, airway, gastrointestinal tract, genitourinary tract, and hearing to improve morbidity and mortality and optimize outcome.

ABP Content Specifications(s)
- Recognize the clinical features associated with genetic associations (eg, CHARGE, VACTERL, VATER)

Suggested Readings
- Lalani SR, Hefner MA, Belmont JW, Davenport SLH. CHARGE syndrome. GeneReviews.http://www.ncbi.nlm.nih.gov/books/NBK1117/ .

Question 29

A 12-year-old girl presents to your office with a 3-week history of nasal congestion and 2 days of ear pain. Her appetite has been diminished slightly, but she has not been febrile. She has not been sleeping well, but she has no other systemic complaints. On physical examination, you note pale purplish nasal mucosa, boggy nasal turbinates, clear nasal secretions, and cobblestoning of the posterior pharyngeal wall. The tympanic membranes are dull gray with air fluid bubbles present. Her lungs are clear to auscultation.

Of the following, the MOST likely cause for this girl's persistent nasal congestion is

A.   acute sinusitis

B.   allergic rhinitis

C.   nasal foreign body

D.   nonallergic rhinitis

E.   recurrent viral upper respiratory infection

Question 29   **Preferred Response: B**

The girl in the vignette has the classic physical examination findings of allergic rhinitis (AR): pale or bluish-purple nasal mucosa, swollen or boggy nasal turbinates, clear nasal secretions, and cobblestoning of the posterior pharyngeal wall. AR is common, yet it is often overlooked or misdiagnosed.

Allergic rhinitis typically begins in childhood and persists into adulthood, often improving in older adults. AR is an IgE-mediated hypersensitivity reaction to specific allergens in sensitized patients that results in inflammation of the nasal mucosa. It may be classified as seasonal or perennial, intermittent or persistent, and mild or moderate to severe. Patients with AR may present with various signs and symptoms, such as snoring, mouth breathing, sneezing, sniffing, congestion, rhinorrhea, nasal itching, postnasal drainage, and cough. The nasal symptoms may be associated with eye complaints, such as red, watery, itchy eyes. In addition, headaches, fatigue, poor sleep, plus decreased attention and daytime performance are common complaints.

There are several classic physical examination findings of AR. "Allergic shiners" is the descriptive term for venous congestion and suborbital edema that appears as dark discoloration under the eyes. Dennie-Morgan lines are accentuated lines or folds below the lower lids. The "allergic salute," pushing the tip of the nose up repeatedly, may lead to a transverse nasal crease. "Allergic facies" consists of an elongated face, high-arched palate, and open mouth breathing.

Seasonal or intermittent AR, often caused by pollen from trees, grasses, or weeds, has symptoms that are predictable and reproducible from year to year. Perennial or persistent AR may result from continual exposure to indoor allergens like dust mites, cockroaches, mold spores, and animal dander; outdoor allergens in subtropical regions with long pollinating seasons; or ongoing occupational allergen exposure.

Children with 1 component of the atopic triad (allergic rhinitis, asthma, eczema) are 3 times more likely to develop a second component. Comorbidities may exist with AR, such as poorly controlled asthma, sinusitis, or otitis media. Approximately 50% of children with chronic otitis media with effusion have AR and may present with symptoms of ear pain and air-fluid bubbles on examination like the girl in this vignette. Although she has complained of nasal congestion for 3 weeks, the girl does not have purulent rhinorrhea, headache, facial pain, or chronic cough, as would be expected in acute sinusitis. Patients with a nasal foreign body typically present with unilateral nasal obstruction with purulent malodorous rhinorrhea. The symptoms of nonallergic rhinitis are similar to AR, but triggers such as cold air, strong odors, or spicy foods can be identified in the patient's history. In this vignette, persistence of symptoms for 3 weeks and the girl's pale rather than erythematous turbinates are clues that this is not a case of recurrent upper respiratory infection.

PREP Pearls
- In cases of suspected allergic rhinitis, the clinician should attempt to elicit any history of exposure to allergens.

- Physical examination findings of pale bluish-purple nasal mucosa, boggy nasal turbinates, clear nasal secretions, and cobblestoning of the posterior pharyngeal wall are suggestive of allergic rhinitis.
- Allergic rhinitis, sinusitis, otitis media, and poorly controlled asthma are often comorbid conditions.

ABP Content Specifications(s)
- Understand the association between allergic rhinitis and sinusitis and/or otitis media
- Differentiate the historical and clinical findings of allergic rhinitis from those of nonallergic rhinitis

Suggested Readings
- Gargiulo KA, Spector ND. Stuffy nose. Pediatr Rev. 2010;31(8):320-324; DOI: http://dx.doi.org/10.1542/pir.31-8-320 .
- Mahr TA, Sheth K. Update on allergic rhinitis. Pediatr Rev. 2005;26(8):284-289.

Question 30

A 2-year-old boy is admitted to the hospital with newly diagnosed high risk neuroblastoma. He has had a documented 2 kg weight loss over the last 2 months and has been refusing to eat for several weeks. On physical examination, his weight is now 10 kg. He has evident muscle wasting and weakness. His laboratory evaluation is remarkable for hypoproteinemia and hypoalbuminemia. Nasogastric tube feedings are initiated and he has tolerated a gradual increase in calories.

Of the following, the BEST caloric goal for this patient is

A.   800 kcal per day

B.   900 kcal per day

C.   1,000 kcal per day

D.   1,100 kcal per day

E.   1,500 kcal per day

Question 30   **Preferred Response: E**

The child in this vignette currently weighs 10 kg and has experienced a 2 kg weight loss. A 12 kg child requires approximately 1,100 kcal per day for normal growth and development. The presence of malignancy increases caloric needs. This child would require more calories per day to maintain growth and development than other 12 kg children without a malignancy. Of the choices given, the best caloric goal for the child in the vignette is 1,500 kcal per day.

Children with cancer are at risk for malnutrition and weight loss. Multiple interacting factors are responsible for the malnutrition seen in children with malignancy (Item C30).

Item C30. Causes of Malnutrition in Children With Cancer.

Hypophagia / Anorexia	Increased Caloric Needs	Malabsorption
Malaise / stress / pain from illness or treatment	Cancer itself	Alterations in gut microbiome (eg, from antibiotics)
Alterations in taste and smell	Healing (eg, mucositis or marrow suppression)	Chemotherapy-induced diarrhea (eg, irinotecan)
Treatment-related nausea	Drug-induced (eg, corticosteroids)	Infection-related diarrhea (eg, *Clostridium difficile*)
Treatment-related mucosal changes (eg, xerostomia or mucositis)		
Decreased gastric emptying / treatment-related constipation (eg, vincristine)		

Courtesy of J. Fish

Despite the nutritional challenges faced by children with cancer, there are several methods to supplement nutrition during treatment. Oral caloric supplements can be added to the food or drink patients are already consuming; enteral nutrition can be provided via nasogastric, gastric, or gastro-jejunal tube; or nutrition can be provided parenterally through total parenteral nutrition (TPN). Despite TPN's ability to provide caloric supplementation, it has significant risks, including increased risk of infection and liver damage. Whenever possible, it is preferable to supplement caloric intake through enteral feeds. This allows for the administration of a balanced feeding formula providing adequate lipid, protein, and carbohydrate support, as well as permitting the child to eat as he wishes. For the child described in the vignette, nasogastric tube feeds providing more than the basic calories needed for growth and development would be most appropriate.

PREP Pearls
- Children with cancer experience malnutrition as a result of hypophagia and anorexia, increased caloric needs, and malabsorption.
- Children with cancer have higher caloric needs than children without cancer.
- There are multiple routes to provide nutritional support during cancer therapy, including oral caloric supplementation, enteral feeds, and parenteral nutrition.
- Enteral caloric supplementation is preferred over parenteral nutrition.

ABP Content Specifications(s)
- Recognize the specific nutritional problems in a child with a malignancy

Suggested Readings
- Bauer J, Jürgens H, Frühwald MC. Important aspects of nutrition in children with cancer. Adv Nutr. 2011;2(2):67-77.http://advances.nutrition.org/content/2/2/67.full .

Question 31

Item deleted

Question 32

A full term female newborn weighing 3,900 g is in the well-baby nursery and breastfed well on day 1 of life. You are performing the discharge physical on day 2 after birth, but the routine pulse oximetry screening has a saturation reading of 91%. The baby has a heart rate of 120 beats/min, respiratory rate of 40 breaths/min, and blood pressure is 65/45 mm Hg taken in the right leg. The baby is awake, alert, and fussy. The chest examination is unremarkable. The cardiac examination shows a single S2 and a 2/6 systolic murmur at the right upper sternal border. There is no hepatosplenomegaly. The femoral pulses are easily palpable but diminished. You are awaiting a call back from the neonatologist.

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

A.   follow serial 4 extremity blood pressures and allow normal feedings

B.   give fluid bolus of 20 mL/kg of normal saline

C.   provide 100% O_2 by facemask for 24 hours

D.   start low dose prostaglandin at 0.01 µg/kg per min

E.   stop feedings and start dopamine at 5 µg/kg per min

Question 32   **Preferred Response: D**

For the neonate in this vignette, the most appropriate next step is to start prostaglandin E (PGE) while you are awaiting the next specialty care provider's help. The patent ductus arteriosus (PDA) is a vascular structure that allows blood flow from the pulmonary artery to the aorta in utero, bypassing the lungs. After birth, there are many congenital heart disease variants that become critical when the PDA closes. The 2-day-old patient described in the vignette has physical examination findings of a single outflow tract (single S2) and intracardiac mixing with an oxygen saturation of 91%. Screening of all newborns on day 2 of life has been implemented in the United States to identify newborns with critical congenital heart disease who may be asymptomatic.

The decreased peripheral pulses in the neonate in the vignette point towards hypoplastic left heart syndrome with reduction in peripheral circulation. In this lesion, the PDA is functioning as if it is the aortic arch, much like it did in utero. Early recognition and initiation of PGE treatment will help to prevent the decompensation that occurs if the PDA is allowed to close. If the PDA closes, the baby will quickly become acidotic and hypotensive with resultant renal dysfunction. If you think about the entire cardiac output as the bubble in a carpenter's level, with the PDA being the level, then anything that causes the flow to shift toward one end will rob the flow to the other. Therefore, if the pulmonary vascular resistance (PVR) is acutely lowered by rapidly ventilating a newborn, the cardiac output will flow toward the lungs and leave the systemic circulation depleted. This will then cause decreased renal blood flow, decreased urine output, and poor pulses with acidosis. The lungs will become overcirculated and pulmonary edema will occur. This also happens naturally and gradually as the PVR drops during the first few days after birth and helps dictate when surgery is optimally done. If the PVR is acutely raised with a pneumothorax, for example, the baby will become quite hypoxemic.

Two other lesions that usually require the PDA for stabilization are pulmonary atresia and tricuspid atresia. In both of these instances where there is no or limited blood flow into the pulmonary veins, the PDA provides the pulmonary blood flow. The oxygen saturations in these lesions may be lower than the 91%, as in the patient in this vignette with hypoplastic left heart syndrome. In pulmonary atresia, a single S2 is audible. Blood flow in this lesion is from the right atrium through either an atrial septal defect to the left atrium and the left ventricle (LV), or through the tricuspid valve to the right ventricle and then through a ventricular septal defect to the LV. In either case, there is mixing in the LV and the blood leaving the LV will then get to the lungs via the aorta and PDA. Some patients may have collateral vessels that provide additional flow from the aorta to the pulmonary arteries. Prostaglandin E is needed to keep the PDA open and ensure pulmonary blood flow. In tricuspid atresia, A2 and a P2 may be audible if there is flow from the right ventricle to the pulmonary artery. In tricuspid atresia, there will need to be an adequate atrial level shunt, as well as a ventricular level shunt, to return blood to the right side of the heart and allow for blood flow into the pulmonary artery. If there is inadequate ventricular level shunting, then the PDA will be needed for pulmonary blood flow. Stabilization with PGE prior to transport for both types of patients will allow pulmonary flow to be maintained.

If a newborn has ductal dependent systemic circulation, they may decompensate quickly as the PDA closes. Following the 4 extremity blood pressures and allowing normal feedings may be dangerous to the baby if the mesenteric circulation has been diminished. A fluid bolus will not overcome the lack of flow in the descending aorta. One hundred percent O2 will cause the PDA to close and the pulmonary vasculature to dilate. Dopamine will increase the systemic vascular resistance, but without ductal patency in the setting of hypoplastic left heart syndrome, it will not improve distal perfusion or prevent acidosis.

PREP Pearls
- Cardiac decompensation secondary to outflow tract atresia can be emergently managed by maintaining the patency of the ductus arteriosus with prostaglandin.
- Screening of all newborns on day 2 of life has been implemented in the United States to identify newborns with critical congenital heart disease who may be asymptomatic.

ABP Content Specifications(s)
- Understand the role of the ductus arteriosus in cyanotic congenital heart disease, and manage appropriately

Suggested Readings
- Artman M, Mahoney L, Teitel D. Perinatal cardiovascular physiology. Neonatal Cardiology. 2nd ed. New York, NY: McGraw-Hill Professional; 2010:51-57.

Question 33
You are seeing a 4-year-old boy, diagnosed with juvenile idiopathic arthritis, for a health supervision visit. His arthritis, which affects his wrists and knees, has been well controlled with medication for the last 6 months. He has had 1 previous disease flare. He is meeting his developmental milestones. The boy's parents are concerned and ask for guidance about how his chronic condition will affect his health, development, and school performance.
Of the following, the MOST appropriate recommendation for this boy's family is to

A.   apply for a 504 modification plan when the boy starts school

B.   apply for an individualized educational plan when the boy starts school

C.   apply for supplemental security income

D.   enroll the boy in an early intervention program

E.   investigate services available through the Americans with Disabilities Act

Question 33   **Preferred Response: A**

The family of the boy in the vignette should apply for a 504 modification plan when he starts school. Practitioners should know and understand the available educational provisions, and which provisions to recommend to families of children with chronic conditions.

The provisions for education related services are under the Individuals with Disabilities Education Act (IDEA). Related services, as defined by IDEA, include speech-language pathology, audiology, psychology, physical and occupational therapy, recreation, social work, counseling, orientation, and mobility services. Educational services are based on the classification of the student. To qualify for an Individualized Education Plan (IEP), a student must qualify for special education. Conditions that qualify a student for special education include sensory deficits such as hearing or visual impairment, physical impairment, chronic illness, traumatic brain injuries, or cognitive disabilities. Students should have a medical referral for evaluation and services. Psychoeducational assessments can identify children with a specific learning disability, autism, emotional disturbances, or speech and language impairments for special education services.

If a child does not qualify for special education, accommodations can still be granted through a 504 modification plan. Section 504 allows for children with chronic diseases or disabling conditions to be provided with related services even when they do not qualify for special education.

An important role of the practitioner is the early identification of children with developmental delay and their referral to an early intervention program. To qualify for early intervention services, a child must have a documented developmental delay. Infants and toddlers up to age 3 years can be referred to this program, and should be referred as early as possible to gain maximum benefit. Children 3 years of age and older with developmental delay should be referred to the local school district for evaluation and treatment.

Publicly funded special education is available until age 22 years, as well as Supplemental Security Income (SSI) for those who qualify. For older individuals with chronic or disabling conditions, the Rehabilitation Act, the Americans with Disabilities Act, and SSI can provide financial assistance, employment assistance, and assistance with workplace accommodations.

PREP Pearls
- Early intervention programs provide education-related services to infants and toddlers with developmental delay up to 3 years of age.
- Individualized Education Plans address the needs of children who qualify for special education by authorizing appropriate education-related services.
- 504 modification plans provide education-related services to children with chronic or disabling conditions who do not qualify for special education.

ABP Content Specifications(s)
- Provide appropriate anticipatory and ongoing guidance to the parents of a child who has a chronic or handicapping condition

Suggested Readings
- American Academy of Pediatrics Council on Children With Disabilities. Provision of educationally related services for children and adolescents with chronic diseases and disabling conditions. Pediatrics. 2007;119:1218-1223. DOI:http://dx.doi.org/10.1542/peds.2007-0885.

Question 34

A 12-year-old girl presents to your office for a health supervision visit prior to attending a summer gymnastics camp. The girl is premenarcheal and there is no family history of scoliosis. On physical examination, you note a right thoracic spine prominence with the Adam's Forward Bend test. Measurement of her spine using a scoliometer reveals an 8-degree angle of thoracic rotation. History and physical examination are otherwise unremarkable.

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

A.   obtain magnetic resonance imaging of the thoracic spine

B.   obtain posteroanterior and lateral entire spine radiographs

C.   recommend use of a thoracolumbosacral spine brace

D.   repeat a clinical evaluation in 1 year

E.   withhold medical clearance for participation in gymnastics camp

Question 34   **Preferred Response: B**

Scoliosis is a vertebral rotation that leads to a curvature of the spine that is most pronounced in the coronal plane. Scoliosis that involves the thoracic vertebrae will also cause the ribs to rotate. A scoliometer is a tool designed to measure this rib rotation. Scoliometer measurements (expressed as angles of trunk rotation [ATR]) are an indirect measure of scoliosis. A 5- to 7-degree ATR corresponds to a scoliosis curve measuring approximately 20 degrees on radiographs using Cobb angle measurements. The girl in the vignette should undergo posteroanterior and lateral radiography of the entire spine to more accurately quantify the degree of curvature.

Scoliosis is categorized by underlying etiology. Idiopathic scoliosis is the most common type, affecting 1 in 50 individuals. Idiopathic scoliosis is generally diagnosed during preadolescence or early adolescence. With the exception of very mild cases, scoliosis shows a female predominance; girls are 10 times more likely than boys to have severe scoliosis that merits spinal fusion surgery. Idiopathic scoliosis tends to progress during years of rapid growth. If the curvature remains in the mild or moderate range by the time a teen reaches skeletal maturity, further progression of the scoliosis is unlikely.

The most common cause of nonidiopathic scoliosis is neuromuscular (eg, related to cerebral palsy, spinal muscular atrophy, or other neuromuscular conditions). Scoliosis is also associated with certain genetic syndromes such as neurofibromatosis and Marfan syndrome.

Occult spinal cord pathology, such as tethered spinal cord or Chiari I malformation with syrinx, can also cause scoliosis. "Red flags" for occult spinal cord pathology include young age at scoliosis presentation (<10 years), atypical curve pattern (eg, a thoracic curve toward the left side of the body), foot deformities (eg, high arch, toe contractures), and other neurologic signs and symptoms on history and physical examination.

Radiographs are the "gold standard" for the diagnosis of scoliosis. The curvature must measure 10 degrees or more on radiographs to meet the criteria for scoliosis. Providers taking the history of a patient with suspected scoliosis should ask about any history of worsening shoulder or back asymmetry, neurologic symptoms including bowel or bladder dysfunction, weakness or pain radiating into the extremities, and family history of scoliosis. Physical examination findings of scoliosis can include asymmetry of the shoulders or muscles of the upper back, or a shift of the pelvis. For the Adams forward bending test, the patient puts his/her palms together and attempts to touch the floor with his/her fingertips without bending at the knees. Scoliometer measurements can be performed with the patient in this position. Prominence of 1 side of the thoracic and/or lumbar spine should prompt radiographic evaluation, particularly if the ATR is 5 degrees or greater.

The treatment for idiopathic scoliosis depends on the severity of curvature and the years of growth remaining. Observation is the treatment of choice for mild scoliosis (10 to <25 degrees). Bracing is recommended for individuals with moderate scoliosis (25 to <45 degrees) with

remaining growth potential. For children and adolescents with scoliosis curves greater than 45 to 50 degrees, spinal fusion surgery is usually required.

The girl in the vignette has physical examination findings consistent with scoliosis. A magnetic resonance imaging scan is not warranted because she does not exhibit atypical features suggesting occult spinal cord pathology. Bracing would be considered only if radiography showed scoliosis in the moderate range. Observation would not be appropriate because radiography could demonstrate that this girl may be a candidate for bracing without which her scoliosis could worsen over the subsequent 12 months. Individuals with idiopathic scoliosis do not need activity restriction.

PREP Pearls
- Evaluation for occult spinal pathology is recommended for children with scoliosis presenting before age 10 years or those who present with a left-sided thoracic curvature.
- Scoliometer measures scoliosis indirectly, therefore radiographs should be obtained for individuals with a measurement of 5 degrees or greater.
- Occult neurologic disease can cause scoliosis.

ABP Content Specifications(s)
- Plan the appropriate clinical evaluation of scoliosis, and manage appropriately
- Recognize the various complications associated with scoliosis
- Understand the natural history and etiology of scoliosis

Suggested Readings
- Sarwark JF, LaBella C. Pediatric Orthopaedics and Sports Injuries: A Quick Reference Guide. Elk Grove Village, IL: American Academy of Pediatrics; 2014.

Question 35

A previously healthy, 6-month-old full-term female infant presents to your office for follow-up 3 days after a visit to the emergency department for a fever of 39°C, diarrhea, and poor oral intake. The infant's mother reports that "they did some laboratory tests, gave her an antibiotic shot, and said they thought she had a virus." On physical examination, her temperature is 37°C, heart rate is 110 beats/min, respiratory rate is 26 breaths/min, and blood pressure is 85/60 mm Hg. The infant is quiet and nontoxic appearing. Her mucous membranes are moist, capillary refill is 1 to 2 seconds, and you note clear rhinorrhea. The remainder of her physical examination is unremarkable.

Her laboratory results from the emergency department show:
- Electrolyte, calcium, serum urea nitrogen, creatinine, serum bicarbonate, and glucose levels are normal
- Aspartate aminotransferase, 35 U/L
- Alanine aminotransferase, 28 U/L
- White blood cell count, 5,600/µL (5.6 × 10^9/L) with 20% neutrophils, 68% lymphocytes, and 12% monocytes
- Hemoglobin, 11 g/dL (110 g/L)
- Platelet count, 180 × 10^3/µL (180 × 10^9/L)
- Urinalysis: specific gravity 1.024, pH 6.0, otherwise negative
- Urine culture: negative
- Blood culture: positive for Staphylococcus hominis

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

A.   administer intramuscular ceftriaxone

B.   admit to the hospital for intravenous antibiotic therapy

C.   prescribe oral antibiotic therapy

D.   recommend supportive care

E.   repeat a blood culture

Question 35   **Preferred Response: D**

The full-term infant in the vignette who has reassuring laboratory and physical examination findings requires only supportive care for a viral upper respiratory tract infection, and no further intervention for the finding of coagulase-negative staphylococci (Staphylococcus hominis) in the blood culture. S hominis is a coagulase-negative gram-positive coccus that typically exists as normal flora on the human skin. The genus Staphylococcus has more than 30 species, most of which are coagulase-negative staphylococci (CoNS). Isolation of CoNS in blood culture represents contamination in most cases. However, CoNS may cause true infection, especially in susceptible hosts. Factors suggesting that CoNS may be a pathogen include the following:
- repeatedly positive blood cultures documenting the same CoNS species
- symptomatic infection (eg, fever, hypotension, lethargy)
- an intravenous catheter (or implantable device) in place for more than 3 days
- susceptibility testing indicating multiple antibiotic resistance

Individuals most likely to acquire infection from CoNS include those with indwelling intravascular catheters or hardware such as cerebrospinal fluid shunts, pacemakers, vascular grafts, prosthetic heart valves, prosthetic joints, and other implantable devices. In addition, immunocompromised hosts are at increased risk of infection. Coagulase-negative staphylococci, most frequently S epidermidis, are the leading cause of hospital-associated bacteremia in neonates, especially those who are of less than 35 weeks' gestation, have low birthweight (< 2,000 g), have intravenous catheters, and are receiving total parenteral nutrition.

If the child described in the vignette had an indwelling vascular catheter or other risk factor(s) for infection because of CoNS, obtaining a repeat blood culture from the catheter and administering intravenous vancomycin would be recommended. In such cases, admission to the hospital is warranted for administration of intravenous antibiotics, repeat blood cultures, and determination of whether the central catheter can be retained. Ceftriaxone is not useful in treating CoNS, and oral antimicrobial therapy is not appropriate for bloodstream infections caused by CoNS.

PREP Pearls
- The genus Staphylococcus has more than 30 species, most of which are coagulase-negative staphylococci (CoNS)
- Isolation of CoNS in blood culture represents contamination in the majority of cases.
- Factors suggesting that CoNS may be a pathogen include:
  - repeatedly positive blood cultures with the same CoNS species
  - growth within 24 hours
  - symptomatic infection (eg, fever, hypotension, lethargy)
  - an intravenous catheter (or implantable device) in place for more than 3 days
  - susceptibility testing indicating multiple antibiotic resistance
  - immunocompromised host (including neonates)

ABP Content Specifications(s)
- • Understand that a positive culture for coagulase-negative staphylococci may represent specimen contamination or infection

Suggested Readings

- American Academy of Pediatrics. Staphylococcal infections. In: Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:715-732.

Question 36

You are seeing a 2-year-old girl brought to the emergency department after she was found unresponsive at her uncle's house. She had been playing normally and later was found lying unresponsive in the hallway. The uncle called 911 after his attempts to awaken her were unsuccessful. Upon arrival to the emergency department, the child is unresponsive. Her vital signs include a heart rate of 70 beats/min, respiratory rate of 8 breaths/min, blood pressure of 64/38 mm Hg, temperature of 35.6°C, and a pulse oximeter reading of 80% on room air. There are no obvious signs of traumatic injury. Her pupils are equal, 2 mm in diameter, with minimal responsiveness. No gag reflex is appreciated and you are unable to elicit deep tendon reflexes. Her glucose level is 70 mg/dL (3.9 mmol/L). The girl's mother expresses concern that the uncle keeps a lot of medicines in his house and she is worried the girl might have gotten into them. Of the following, the MOST likely cause of this girl's clinical findings is ingestion of

A.   digoxin

B.   diphenhydramine

C.   hydrocodone

D.   lithium

E.   verapamil

Question 36   **Preferred Response: C**

The girl in the vignette presents for emergency care because of an acute onset of somnolence accompanied by bradycardia, bradypnea, hypotension, hypothermia, hypoxia, and miosis, following suspected exposure to a relative's medications. Of the agents listed, hydrocodone is most likely to have caused these clinical findings.

Opioids are drugs with activity similar to that of opium or morphine that can be either naturally occurring or synthetic. Opioid receptors exist throughout the central and peripheral nervous systems and gastrointestinal tract. Opioids are prescribed for many indications including pain management, cough suppression, sedation or anesthesia, and diarrhea alleviation; they are widely available in multiple formulations for both prescribed and illicit use.

Over the past 25 years, opioid products available in the United States have increased threefold. Use of these products has risen drastically, along with resultant addiction and overdose-related fatalities. Because of their frequent and widespread use, opioids are accessible to many children and adolescents. All pediatric providers should recognize the clinical findings associated with acute opioid intoxication and understand how to manage this life-threatening condition appropriately.

In young children, opioid intoxication typically arises from exploratory ingestion of prescription or illicit opioids that are accessible in their environments. Some of these agents, such as codeine and methadone pills, powdered heroin, or fentanyl patches, may cause profound respiratory depression or even death with ingestion of only small quantities of pills or powder, or exposure to a single opioid patch. In addition, widely available over-the-counter medications containing diphenoxylate (eg, lomotil) or dextromethorphan may cause toxicity in young children and are sometimes even administered to children by well-meaning caregivers for the relief of diarrhea and cough, respectively.

The classic clinical triad of acute opioid toxicity consists of central nervous system depression, respiratory depression, and miosis. Other typical clinical findings include hypothermia, hyporeflexia, flushing, pruritus, bradycardia, hypotension, vomiting, and decreased bowel sounds. In the case of massive overdose, respiratory toxicity can lead to significant hypoxia, hypercarbia, and acute lung injury. Generalized seizure activity occurs less commonly, but has been reported after intoxication with propoxyphene, meperidine, tramadol, fentanyl, and pentazocine.

The initial management of opioid intoxication in both children and adults should focus on stabilization of the airway, support of breathing, and hemodynamic support. In cases of known or suspected opioid overdose, naloxone should be administered promptly. Naloxone, a pure opioid antagonist, can be given intravenously, subcutaneously, intramuscularly, via endotracheal tube, or by nebulizer. The onset of action of naloxone is rapid, usually within 1 minute of administration. In the setting of acute opioid overdose, for children up to 5 years of age and those weighing less than 20 kg, a dose of 0.1 mg/kg of naloxone should be administered. In older children, a rapid dose of 2 mg should be given. Repeat doses may be given every 2 to 3 minutes,

up to a maximum dose of 10 mg for children of all ages. If no clinical improvement occurs, alternative causes for the patient's symptoms should be considered. As the duration of action of naloxone is 20 to 30 minutes (shorter than that of most opioid agents), repeat doses or even a continuous intravenous infusion may be necessary, especially in patients experiencing the toxic effects of longer-acting opioids such as oxycodone, methadone, and diphenoxylate. Nalmefene is an alternate opioid antagonist with a longer duration of action than naloxone, but there is debate regarding indications for its use in the acute care setting. In addition to providing supportive care and prompt administration of an opioid antagonist, activated charcoal should be administered to children with toxicity arising from oral opioid ingestion within 1 hour after ingestion, provided that their airways have been adequately secured. In patients who have ingested large amounts of opioid (such as body "stuffers" or packers), or in the case of sustained-release opioid formulations, activated charcoal would still be recommended.

Any young child presenting with central nervous system and respiratory depression secondary to opioid toxicity should be hospitalized, even if symptoms improve with naloxone administration, because repeat dosing or continuous infusion of naloxone may be needed. Young children should be observed in a monitored setting for at least 24 hours after exposure because of the potential for delayed toxicity.

Children with acute toxicity from digoxin typically present with vomiting, bradycardia with atrioventricular heart block, hyperkalemia, and mental status changes. Although the girl in the vignette exhibits mental status changes and bradycardia, her findings of pinpoint pupils and respiratory depression are not typically seen with digoxin toxicity.

Children with toxicity from diphenhydramine, a widely used, over-the-counter antihistamine, generally present with symptoms reflecting an anticholinergic toxidrome. Symptoms may include agitation, confusion, hyperactivity, hallucinations, dry mouth and eyes, dry flushed skin, urinary retention, dilated pupils, tachycardia, tremor, and even seizures in severe cases. The patient in the vignette presented with constricted rather than dilated pupils, and bradycardia, rather than the tachycardia that would be expected from diphenhydramine overdose.

Children with acute toxicity from lithium may present with a range of clinical findings, including gastrointestinal upset (nausea, vomiting, and diarrhea), dehydration, tremor, weakness, hyperreflexia, slurred speech, visual disturbances, mental status changes, and even seizures with severe toxicity. Lithium toxicity would not explain the significant respiratory depression, miosis, or decreased reflexes observed in the girl in the vignette. Profound somnolence would also not be a characteristic early finding in a child with acute lithium toxicity.

Verapamil is a calcium channel blocker used (most commonly in adult patients) in the treatment of hypertension, coronary artery disease, and atrial fibrillation, and to prevent cerebral vasospasm. The predominant clinical findings in patients with calcium channel blocker toxicity are bradycardia and hypotension. Neurologic and respiratory system derangements can occur from calcium channel blocker overdose, but these generally arise secondary to cardiovascular toxicity and shock. Though the girl in the vignette presents with bradycardia and hypotension,

her constricted pupils, acute onset of somnolence, and profound respiratory depression are better explained by overdose of the opioid hydrocodone.

PREP Pearls
- The classic clinical triad of central nervous system depression, respiratory depression, and miosis should prompt clinicians to suspect opioid poisoning, even in the absence of a history of opioid exposure.
- Initial management of suspected or confirmed opioid toxicity should include stabilizing the patient's airway, ensuring adequate ventilation, and prompt administration of the opioid antagonist naloxone.
- Any young child presenting with central nervous system and respiratory depression secondary to opioid toxicity should be hospitalized, even if symptoms improve with naloxone administration, because repeat dosing or continuous infusion of naloxone may be needed.

ABP Content Specifications(s)
- Recognize the major behavioral consequences of opioid use/abuse
- Identify the major physiologic consequences associated with opioid use/abuse, including those associated with the various means of administration
- Recognize the clinical findings associated with an acute opioid overdose, and manage appropriately

Suggested Readings
- Erickson TB. Opioids. In: Erickson TB, Ahrens WR, Aks SE, Baum CR, Ling LJ, eds. Pediatric Toxicology: Diagnosis and Management of the Poisoned Child. New York, NY: McGraw Hill; 2005;chap 60:409-414.

Question 37

A mother presents to labor and delivery at 36 weeks' gestation with a chief complaint of decreased fetal movement for the past 6 hours. The pregnancy history is significant for a known fetal tachyarrhythmia that has been well controlled by maternal treatment with digoxin. Continuous monitoring reveals a fetal heart rate of 160 beats/min with rare variable decelerations seen with infrequent contractions. Further evaluation of fetal well-being includes a nonreactive nonstress test and a biophysical profile of 4 out of 10. Due to the late preterm status of the pregnancy and history of fetal arrhythmia, the obstetrician seeks your input in the subsequent management of the mother and fetus.

Of the following, the MOST appropriate next step in management of the mother and fetus is to

A.   continue to observe with continuous monitoring

B.   discharge home with outpatient follow-up

C.   increase maternal digoxin dosing

D.   initiate induction of labor

E.   perform an urgent caesarian delivery

Question 37   **Preferred Response: D**

The assessment of the fetus in the vignette suggests possible compromise of fetal well-being, with the induction of labor being the most appropriate step in management. Pregnancies in which maternal and/or fetal complications develop require increased surveillance to decrease the risk of fetal death. Methods of monitoring include maternal perception of fetal movement, nonstress testing, and biophysical profile evaluation.

Maternal perception of fetal movement has long been recognized as a measure of fetal well-being; with any concerns of decreased fetal movement prompting further evaluation. If the pregnancy is at 32 weeks' gestation or greater, a nonstress test (NST) is often performed to document the presence of fetal heart rate (FHR) accelerations with fetal movement. A reactive NST will demonstrate 2 or more FHR accelerations within a 20-minute period and is considered reassuring. Conversely, a nonreactive NST is nonspecific and should lead to further fetal assessment with a biophysical profile (BPP). A BPP uses fetal ultrasound evaluation along with an NST to assess 5 measures of fetal well-being: movement, tone, breathing, amniotic fluid volume, and heart rate (NST). Each is assigned either 0 or 2 points, with a composite score of 8 to 10 being normal, 6 being equivocal, and 4 or less being abnormal. For an abnormal BPP, induction of labor is recommended unless an obstetric contraindication necessitates caesarian delivery.

The presence of a fetal arrhythmia makes in utero assessment more challenging. The early identification of a fetal arrhythmia allows close fetal monitoring for complications such as hydrops fetalis. Some fetal arrhythmias are amenable to therapies delivered transplacentally, such as digoxin for supraventricular tachycardia. Undiagnosed arrhythmias presenting in the third trimester may be interpreted as fetal distress, leading to premature delivery.

The mother in the vignette presented with a complaint of decreased fetal movement. The subsequent nonreactive NST and biophysical profile of 4 out of 10 raise concern for fetal well-being and suggests the need to induce labor. Continued observation with intermittent monitoring or discharge from the hospital would not be recommended because of the increased risk of fetal death with these findings. The maternal digoxin dosing does not need to be increased in this case, because the fetal heart rate appears to be well controlled. At present, there is no evidence to support urgent caesarian delivery.

PREP Pearls
- Maternal perceptions of fetal movement have long been recognized as a measure of fetal wellbeing; any concerns of decreased fetal movement should prompt further evaluation.
- A reactive nonstress test (NST) will demonstrate 2 or more fetal heart rate accelerations within a 20-minute period and is considered reassuring.
- A biophysical profile, which uses both fetal ultrasound and NST, contains 5 measures of fetal wellbeing: movement, tone, breathing, amniotic fluid volume, and heart rate (NST). Each measure is assigned either 0 or 2 points, with a composite score of 8 to 10 being normal, 6 being equivocal, and 4 or less abnormal.

ABP Content Specifications(s)
- Know the factors used by obstetricians to evaluate fetal well-being
- Understand the appropriate use of stress and non-stress tests during fetal assessment
- Understand the significance of fetal arrhythmias, and manage appropriately

Suggested Readings
- Farley D, Dudley DJ. Fetal assessment during pregnancy. Pediatr Clin North Am. 2009; 56: 489-504. DOI:http://dx.doi.org/10.1016/j.pcl.2009.03.001.

Question 38

A 17-year-old high school junior presents for evaluation of migraine headaches. He has had infrequent migraine headaches since he was 10 years of age. Since school started 3 months ago, his headaches have increased in frequency. He experiences 2 to 3 migraine headaches per week and has to go home from school at least once a week. He has been taking ibuprofen 3 or 4 days every week for the past month and it no longer relieves the migraine symptoms. He takes isotretinoin for acne. His blood pressure is 118/68 mm Hg, heart rate is 86 beats/min, respiratory rate is 16 breaths/min, and body mass index is 18. His physical examination, including neurological examination and fundoscopy, is unremarkable.

Of the following, the BEST first step in treating his headaches is

A.   discontinue the ibuprofen

B.   discontinue the isotretinoin

C.   prescribe butterbur

D.   prescribe topiramate

E.   stress management skills training

Question 38   **Preferred Response: A**

The boy in the vignette has headaches that are interfering significantly with his ability to attend school. Frequent, excessive use of ibuprofen is a likely contributor by causing medication overuse headaches. The best first step in improving his headache severity and frequency is to discontinue the ibuprofen. He will then need appropriate migraine prophylaxis and abortive therapy plans.

A migraine prophylaxis plan addresses the multifactorial causes of migraine headaches. Migraines often worsen during times of stress, such as the start of an academic year. Improving stress management skills is an important part of a migraine prophylaxis plan. Prophylactic medications are indicated when the severity and frequency of migraines interfere with functioning. Other important elements of a migraine prophylaxis plan are the promotion of regular, restful sleep, and a regular, nutritious diet.

For the boy in the vignette, who is nearly an adult, migraine prophylactic medications used for adults would be effective. Butterbur and topiramate are both approved by the US Food and Drug Administration for migraine prophylaxis in adults. Medication choices should be tailored to individuals. For instance, topiramate can cause weight loss and should be used cautiously in a person who is already thin.

Isotretinoin has been associated with pseudotumor cerebri and should be stopped if a person develops signs of this condition. The boy in the vignette has typical migraines and his fundoscopic examination findings are normal, so he does not have signs of pseudotumor cerebri. Stopping isotretinoin is unlikely to help his headaches.

An abortive therapy plan for migraines starts with over-the-counter medications such as acetaminophen or ibuprofen. If these are ineffective, prescription medications such as one of the triptan class of medications can be tried. Caffeine is often helpful for migraines as well. Antiemetics should be considered if nausea or emesis are present.

Medications for abortive therapy should not be used more than 2 to 3 times a week; otherwise medication overuse headache can develop. The only treatment for medication overuse headache is to discontinue the inciting medication. Patients should know that the headaches will transiently worsen, but discontinuing the inciting medication is a necessary step as the migraines are unlikely to improve in the setting of ongoing medication overuse.

PREP Pearls
- Limit migraine abortive medications (eg, ibuprofen and acetaminophen) to 2 to 3 days per week, to prevent medication overuse headache.
- Isotretinoin has been associated with pseudotumor cerebri and should be discontinued if a person develops signs of this condition.

ABP Content Specifications(s)
- Plan appropriate abortive therapy for acute migraine
- Plan appropriate prophylaxis for recurrent migraine

Suggested Readings
- Blume HK. Pediatric headache: a review. Pediatr Rev. 2012;33(12):562-576. DOI: http://dx.doi.org/10.1542/pir.33-12-562.

Question 39

You are contacted by your state's newborn screening office about an 8-day-old male newborn with a positive newborn screen for congenital adrenal hyperplasia. In your office, the newborn's vital signs include a pulse of 175 beats/min and a blood pressure of 75/40 mm Hg. His body surface area is 0.2 m^2. He has descended testes bilaterally and a normal phallus. He has no hyperpigmentation.

Initial laboratory results include a serum sodium of 129 mEq/L (129 mmol/L), potassium of 6.6 mEq/L (6.6 mmol/L), glucose of 87 mg/dL (4.8 mmol/L), and bicarbonate of 16 mEq/L (16 mmol/L).

Based on the newborn's diagnosis, clinical status, and laboratory results, you administer a normal saline bolus of 20 mL/kg intravenously.

Of the following, the MOST important next step in the treatment of this patient is to administer

A.   betamethasone, 10 mg intramuscularly

B.   cortisone acetate, 20 mg intramuscularly

C.   dexamethasone, 10 mg intravenously

D.   hydrocortisone hemisuccinate, 20 mg intravenously

E.   methylprednisolone, 10 mg intravenously

Question 39   **Preferred Response: D**

The boy in the vignette has an abnormal newborn screen for 21-hydroxylase deficiency and presents in shock. As female patients would be expected to have some degree of ambiguous genitalia, the newborn screen is primarily designed to diagnose male infants before they can present in extremis with a salt-wasting crisis. Patients with a positive screen should have a confirmatory serum 17-hydroxyprogesterone level (sometimes written as 17-alpha-hydroxyprogesterone) as well as glucose and electrolyte levels obtained.

The infant in the vignette has abnormal vital signs, and his electrolytes show that he is already experiencing salt wasting and is dehydrated. Treatment with high-dose steroids (stress dose steroids) is needed to mimic the high doses of steroids normally produced under stress in patients with sufficient adrenal function.

The treatment of choice for adrenal crisis, of any cause, is fluid replacement, hydrocortisone hemisuccinate intravenously, and if hypoglycemia is present, intravenous dextrose. Hydrocortisone is quick acting and at an emergency stress dose (100 mg/m2) saturates all steroid receptors, causing a mineralocorticoid and glucocorticoid effect. 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 hospital or emergency department, intravenous hydrocortisone should be used because of its quick onset of action.

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. Dexamethasone and betamethasone can act quickly but have limited mineralocorticoid effect, and would be used in much smaller doses than those listed in the responses. It is helpful to be familiar with recommended hydrocortisone stress doses (Item C39).

Item C39. Stress Dosing Guideline for Hydrocortisone.

Physiologic replacement dosing, oral	6-10 mg/m^2 per day
Oral stress dosing (minor febrile illness, taking oral well)	30 mg/m^2 per day divided three times daily
Intravenous stress dosing (before surgery or major illness, but clinically stable)	50 mg/m^2 1 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)

Courtesy of J. Pinsker

PREP Pearls
- A newborn screen positive for congenital adrenal hyperplasia should prompt immediate testing for serum electrolytes, glucose and a confirmatory 17-alpha-hydroxyprogesterone level.
- High-dose intravenous hydrocortisone hemisuccinate (100 mg/m^2 up to 100 mg maximum per dose) is the steroid of choice for the treatment of adrenal crisis.

ABP Content Specifications(s)
- Plan the appropriate management of congenital adrenal hyperplasia, including that associated with an adrenal crisis
- Recognize the clinical features associated with congenital adrenal hyperplasia
- Plan the appropriate diagnostic evaluation of congenital adrenal hyperplasia, including during the perinatal period

Suggested Readings
- 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: http://dx.doi.org/10.1542/peds.2006-1612.

Question 40

A 7-year-old boy is brought to the emergency department with wheezing, tachypnea, and respiratory distress. He started to exhibit symptoms of an acute upper respiratory infection 2 days ago. He has had dry, spasmodic coughing, and now complains of chest tightness. There has been no loss of consciousness.

The child was born full term and has no prior history of hospitalization or respiratory failure. He plays soccer 5 days per week and receives β-adrenergic agonist therapy 4 to 6 times per week for exertional cough, chest tightness, and dyspnea; care providers report short term improvement with this intervention. On these occasions, he has used albuterol prescribed to an older sibling who has been treated for moderate persistent asthma and atopic dermatitis.

On physical examination, his heart rate is 130 beats/min and the respiratory rate is 24 breaths/min. Oxygen saturation is 98% in room air. His weight and height are at the 50th percentile. Mild suprasternal retractions are noted. There is no appreciable murmur on cardiac examination. Auscultation of lung fields reveals fair-to-moderate aeration with prolonged expiratory phase and diffuse expiratory wheezing. The abdomen is soft and nontender. Extremities are warm and well perfused, without cyanosis, clubbing, or edema.

You administer albuterol and ipratropium bromide via nebulizer, with only a modest improvement in the boy's respiratory signs and symptoms.

Of the following, the MOST appropriate next therapeutic intervention for this patient is administration of

A.   intravenous magnesium sulfate

B.   long acting β-agonist

C.   subcutaneous epinephrine

D.   systemic corticosteroids

E.   theophylline

Question 40   **Preferred Response: D**

The boy in this vignette is presenting with a moderate exacerbation of his chronically uncontrolled mild-to-moderate persistent asthma. His acute asthma exacerbation has not resolved with administration of a short-acting bronchodilator and inhaled anticholinergic therapy; the next most appropriate intervention is administration of a systemic corticosteroid.

Corticosteroids are an important component in the treatment of acute asthma as they reduce airway hyperresponsiveness, inhibit migration and activation of inflammatory cells, and prevent the late phase reaction to allergens. Assuming equal doses, systemic steroids are of similar efficacy when administered orally, intramuscularly, or intravenously. Systemic steroids may be given as prednisone, prednisolone, dexamethasone, or methylprednisolone. Although there is some controversy as to optimal dosing interval, it has been demonstrated that the treatment of severe acute asthma with systemic corticosteroids within 1 hour of presentation to the emergency department lowers hospitalization rate and improves pulmonary function.

According to the most recent National Asthma Education and Prevention Program Expert Panel Guidelines for the Diagnosis and Management of Asthma, the patient in the vignette demonstrates symptoms most consistent with mild persistent asthma (Item C40). He has required use of his sibling's short acting albuterol more than twice weekly with exertional symptoms, and demonstrates minor limitation in exertional activities.

Long acting β-agonists (LABAs) are contraindicated as a reliever for acute asthma exacerbations. They are also contraindicated as monotherapy in adult and pediatric asthma. The US Food and Federal Drug Administration has mandated that a "black box warning" label be added to products that contain salmeterol, formoterol, or indacaterol, as these compounds carry "an increased risk of severe exacerbation of asthma symptoms, leading to hospitalizations, in pediatric and adult patients, as well as death in some patients using LABAs for the treatment of asthma." The use of LABAs is indicated for treatment of moderate to severe persistent asthma (step 3 or higher), and only in combination with an inhaled corticosteroid in those patients whose asthma is not well controlled with use of low dose inhaled corticosteroid alone.

Subcutaneous epinephrine has α- and β-adrenergic activity and is an effective bronchodilator. However, the use of epinephrine has been limited by its significant adverse effects, which include hypertension and tachycardia. Moreover, the use of injected epinephrine has not been found to be superior to inhaled albuterol for the treatment of acute asthma in children.

Theophylline has not been demonstrated to have significant efficacy in the treatment of acute asthma in children. Other therapeutic steps, including intravenous magnesium sulfate and terbutaline, may be utilized in the treatment of status asthmaticus, but these therapies are classically employed after administration of a systemic corticosteroid.

Components of Severity		Classification of Asthma Control (5-11 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	≤ 2 times/month	3-4 times/month	>1 times/week but not nightly	Often 7 times/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
	Lung function	• Normal FEV$_1$ between exacerbations • FEV$_1$ >80% predicted • FEV$_1$/FVC >85%	• FEV$_1$ = >80% predicted • FEV$_1$/FVC >80%	• FEV$_1$ = 60-80% predicted • FEV$_1$/FVC = 75-80%	• FEV$_1$ = <60% predicted • FEV$_1$/FVC <75%
Risk	Exacerbations requiring oral systemic corticosteroids	0-1/year (see note)	≥2/year (see note)		
		Consider severity and interval since last exacerbation. Frequency and severity may fluctuate over time for patients in any severity category.			
		Relative annual risk of exacerbations may be related to FEV$_1$.			
Recommended step for initiating therapy		Step 1	Step 2	Step 3, medium-dose ICS option	Step 3, medium-dose ICS option, or step 4 and consider short course of oral systemic corticosteroids
		In 2-6 weeks, evaluation level of asthma control that is achieved, and adjust therapy accordingly.			

EIB, exercise-induced bronchospasm; FEV$_1$, forced expiratory volume in 1 second; FVC, forced vital capacity; ICS, inhaled corticosteroids

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. Available at http://www.nhlbi.nih.gov/health-pro/guidelines/current/asthma-guidelines/full-report.

ITEM C40: Classification of asthma severity in children 5-11 years of age.

PREP Pearls
- Systemic corticosteroids are indicated in the treatment of moderate-to-severe asthma exacerbations.
- Long acting β-agonists are contraindicated in the therapy of acute asthma and should not be used as chronic monotherapy in the treatment of asthma.
- Based on clinical history, a patient's asthma severity should be evaluated as recommended by the National Asthma Education and Prevention Program Expert Panel Guidelines for the Diagnosis and Management of Asthma; the most appropriate chronic preventive therapy may then be guided by treatment recommendations.

ABP Content Specifications(s)
- Provide appropriate treatment for a patient who has an acute exacerbation of asthma, including asthma that is not responsive to adrenergic agonist therapy

American academy of pediatrics   132

Suggested Readings

- Becker AB, Nelson NA, Simons ER. Inhaled salbutemol (albuterol) vs injected epinephrine in the treatment of acute asthma in children. J Pediatr. 1983;102(3):465-469 .

Question 41

A 7-month-old infant is referred to you for evaluation of failure to thrive. The baby has been breastfed since birth; puréed fruits and vegetables were started 2 months ago. The parents report that the baby stopped rolling over 2 months ago and he has not been able to sit, even with assistance. Last month, he received intravenous fluids in the emergency department when he became dehydrated after a day of vomiting. Physical examination is notable for a thin child who is alert but demonstrates poor tone. His liver is palpable 3 cm below the right costal margin. The remainder of the examination is unremarkable. His growth charts are shown in Item Q41A and Item Q41B.

Of the following, the MOST appropriate initial approach to this infant's failure to thrive is to
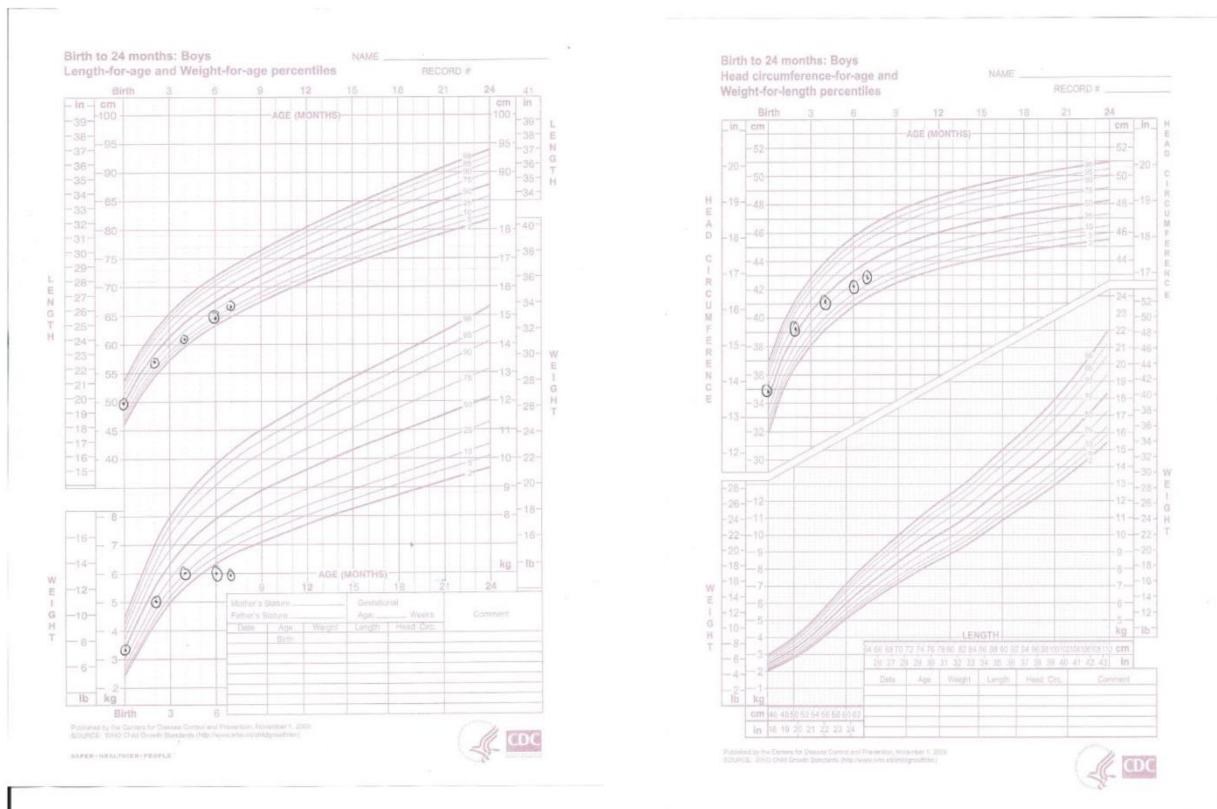
Item Q41A. Weight and length growth chart for the infant described in the vignette.

Item Q41B. Head circumference growth chart for the infant described in the vignette.

A.   fortify the breast milk to 22 cal/oz
B.   make a referral to child protective services
C.   obtain comprehensive metabolic panel, creatine kinase, and ammonia levels
D.   order noncontrast brain computed tomography
E.   refer him to a feeding specialist

Question 41   **Preferred Response: C**

Failure to thrive is a physical sign of a nutritional state that is inadequate to support normal growth and development. Often, the underlying causes of this growth failure are complex, involving organic, functional, and psychosocial components. The patient's history and physical examination are crucial and should alert the clinician to the possibility of a metabolic or genetic condition (Item C41). The patient in the vignette has several findings that raise suspicion of an underlying inborn error of metabolism including organomegaly, developmental regression, hypotonia, and vomiting with dehydration. Therefore, the next step in evaluation of this patient is to obtain laboratory studies to help elucidate an underlying cause, particularly because some metabolic errors can become life-threatening. Neuroimaging is appropriate if there are neurologic findings such as hypotonia, seizures, or stroke. The modality of choice is usually magnetic resonance imaging. In some cases, appropriate imaging may include magnetic resonance spectroscopy, a specialized test that detects metabolites in the brain, for example, elevated lactate in the basal ganglia found in mitochondrial disease. In the patient in this vignette, the priority is to evaluate the child's metabolic status and a computed tomography of the brain would not be the imaging study of choice.

For patients with failure to thrive but without abnormalities suggesting an underlying inborn error of metabolism, a multidisciplinary approach is appropriate. Involvement of an occupational or speech therapist to work on feeding techniques and texture tolerance is often a first step. Child abuse specialists and agencies may be necessary when there is concern for neglect or factitious disorder. Some infants, particularly those with known underlying conditions such as congenital heart disease, low birth weight, or chronic lung disease, may require a high caloric intake to support growth, and that intake can best be achieved by providing higher caloric density foods such as fortified breast milk. However, because the infant in the vignette has findings suggestive of underlying metabolic disease, evaluation for that takes precedence over changes in the approach to feeding the infant.

PREP Pearls
- Failure to thrive (FTT) is a physical sign of a nutritional state that is inadequate to support normal growth and development.
- An underlying metabolic cause of FTT is suggested by a history or physical examination finding of severe, life-threatening disease; recurrent vomiting and dehydration; developmental delay or regression; hypotonia, stroke, or seizure; organomegaly, particularly hepatomegaly; cardiomyopathy; visual or hearing deficit; dysmorphic features; or pancytopenia.
- For patients with FTT without a known or suspected underlying cause, a multidisciplinary approach to feeding is preferred.

ABP Content Specifications(s)
- Plan the management of an infant with failure to thrive
- Recognize and evaluate a patient with failure to thrive
- Differentiate among the possible causes of failure to thrive

Is there isolated failure to thrive? → YES → probably no underlying metabolic disorder

↓

NO

Suspect an IEM when any one of the following (or combination) is present

- History of acute, severe, and potentially life-threatening symptoms and signs (*recurrent ketoacidosis, hypoglycemia*)
- Recurrent attacks of vomiting, lethargy, dehydration
- Liver dysfunction
- Developmental delay, hypotonia, seizures, stroke, ataxia
- Cardiomyopathy, myopathy
- Hearing loss or visual impairment
- Organomegaly
- Mild dysmorphic or coarse facial features
- Pancytopenia

↓   ↓

Baseline screening tests for failure to thrive to rule out IEM   **If there are abnormal neurologic findings**

Sodium, potassium, chloride, CO_2, alanine aminotransferase, aspartate   Magnetic resonance imaging
aminotransferase, glucose, blood urea nitrogen, creatine, total protein, albumin,   Magnetic resonance spectroscopy
alkaline phosphatase, total bilirubin
Complete blood count
Urinalysis

If there is a suspicion for IEM*

Plasma amino acids
Plasma acylcarnitines
Ammonia**
Blood lactate, pyruvate**
Creatine kinase
Urine organic acids

*Not all IEM's can be ruled out by these tests. Some tests may be normal during well-state.
**Free flow blood sample and transfer to laboratory on ice for immediate analysis.
If an IEM is suspected, metabolic consult is essential.

Reprinted with pemission from Ficiciouglu C, An Haack K. Failure to thrive: when to suspect inborn errors of metabolism. *Pediatrics*. 2009;124(3):972-979.

ITEM C41: Approach to failure to thrive with suspected inborn error of metabolism (IEM).

Suggested Readings

- Ficiciouglu C, An Haack K. Failure to thrive: when to suspect inborn errors of metabolism. Pediatrics. 2009;124(3):972-979. DOI: http://dx.doi.org/10.1542/peds.2008-3724.

Question 42

A 21-year-old woman presents to the clinic for her first routine gynecologic examination. She states that her last menstrual period was about 3 weeks ago. She reports mild cramping during her cycle. She denies sexual activity. She has no other concerns or complaints. Upon physical examination, she is afebrile and has normal vital signs. Her pelvic examination is significant for a left-sided adnexal mass, but the rest of her examination is unremarkable. She is sent for a pelvic ultrasonography, which reveals a 4 cm anechoic fluid-filled mass of the left ovary.

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

A.   α-fetoprotein testing
B.   CA-125 testing
C.   culdocentesis
D.   cystectomy
E.   oral contraceptives

Question 42   **Preferred Response: E**

Functional ovarian cysts are the most common type of ovarian mass in the postpubertal female and are the result of ovulation. These cysts are often asymptomatic and found on routine pelvic examination or incidentally on imaging. Symptomatic patients may complain of pelvic pain or menstrual irregularity. Two types of functional cysts include simple or follicular cysts and corpus luteum cysts. Follicular cysts develop when the growing follicle does not open to release an egg. The corpus luteum is the natural result of ovulation. However, the area of the corpus luteum can fill with fluid and develop into an ovarian cyst.

Functional cysts that are less than 5 cm usually self-resolve within 2 to 3 menstrual cycles. Oral contraceptives can be used to prevent future cysts, but are not thought to aid in the resolution of a cyst.

α-fetoprotein is a tumor marker that is frequently elevated in patients with malignant ovarian germ cell tumors (MOGCT). On ultrasonography, an MOGCT would have a more complex and heterogeneous appearance than a simple cyst.

The tumor marker CA-125 is associated with epithelial ovarian cancer. However, it can also be elevated in young women with endometriosis. It is not indicated in the woman in this vignette because the ultrasonographic features of the mass suggest a benign process.
A simple cyst of less than 5 cm in a premenopausal woman can be followed for resolution and often does not require surgical intervention. Oral contraceptives can be used for prevention of additional cysts. Larger cysts, symptomatic cysts, or cysts that are increasing in size may require aspiration or cystectomy. Large cysts increase the risk of ovarian torsion because of their weight.

PREP Pearls
- Functional ovarian cysts are the most common type of ovarian mass in the postpubertal female and are the result of ovulation.
- Hormonal contraceptives may help prevent future ovarian cysts, but they are not thought to help with the resolution of existing cysts.

ABP Content Specifications(s)
- Recognize the association of small ovarian cysts with normal development
- Plan the appropriate diagnostic evaluation and management of ovarian cyst

Suggested Readings
- Hoffman BL, Schorge JO, Schaffer JI, et al. Ovarian germ cell and sex cord-stromal tumors. In: Hoffman BL, Schorge JO, Schaffer JI, et al, eds. Williams Gynecology. 2nd ed. New York, NY: McGraw-Hill; 2012:879-897.

Question 43

A 6-year-old boy presents for evaluation of a wart on his hand for several months. Parental attempts to "freeze" the wart with an over-the-counter preparation were painful and unsuccessful. The physical examination reveals a 4-mm papule with a rough surface located on the dorsum of the right hand.

Of the following, the MOST appropriate treatment for this boy is

A.   cimetidine orally

B.   cryotherapy

C.   imiquimod topically

D.   salicylic acid topically

E.   surgical excision

Question 43   **Preferred Response: D**

The boy in the vignette has a common wart, a skin-colored papule with a rough (ie, verrucous) surface. Since it is unlikely that he will tolerate painful interventions, the most appropriate initial treatment is salicylic acid. It is as effective as cryotherapy, although depending on the size of the wart, treatment may require several weeks. Directions for using salicylic acid preparations are provided in Item C43.

Item C43. Using Salicylic Acid Preparations to Treat Common Warts.

Apply 17% salicylic acid liquid or gel to the surface of the wart(s) • May substitute a pad or bandage impregnated with salicylic acid • For plantar warts may substitute a plaster impregnated with a higher concentration of salicylic acid (40%)
If using a liquid or gel preparation, allow to dry for 2 to 3 minutes (develops a white film)
Occlude the wart with duct tape or similar adhesive tape
Remove tape in the morning and debride the wart with an emery board
Repeat nightly until wart resolves
If the wart becomes erythematous or macerated, withhold treatment for a few days then resume

Adapted 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

For those children able to tolerate some discomfort, cryotherapy is useful, causing necrosis of wart tissue. Liquid nitrogen is the most effective cryogen, achieving a temperature of approximately -195°C. It is applied using a spray device or cotton-tipped applicator. The application is continued until a white ring extends 1 to 3 mm beyond the margin of the wart (typically 10 to 15 seconds). Some advise a second treatment after the wart thaws. Families should understand that a blister may form in 1 to 2 days. When the blister ruptures, the area may be cleansed twice daily, followed by the application of a topical antibiotic and bandage. If a residual wart remains (or if a blister did not form), salicylic acid treatment should be initiated as

described in Item C43. Repeat cryotherapy may be performed in 2 to 4 weeks. Cryotherapy devices may be purchased by patients without a prescription. These employ dimethyl ether and propane and are less effective than liquid nitrogen.

Cimetidine has immunomodulatory effects, enhancing T-cell function and cytokine production. Although not US Food and Drug Administration (FDA)-approved for the treatment of warts, it may be of benefit (used in conjunction with topical salicylic acid) in those who have multiple or resistant warts. Topical imiquimod has been used off-label, although its efficacy is limited by poor absorption through the highly keratinized skin characteristic of common warts. Surgical excision occasionally is considered for resistant warts, but carries a risk of scarring. Additional treatments include intralesional injection of skin test antigens (eg, Candida, Trichophyton); measles, mumps, and rubella vaccine or bleomycin; and immunotherapy with topical squaric acid. None of these therapies are FDA-approved for the treatment of warts.

PREP Pearls
- When treating young children who have common warts, salicylic acid is the preferred initial therapy. An important element of treatment is debridement of the wart using an emery board.
- Families should be informed that a blister may form 1 to 2 days after wart cryotherapy. When the blister ruptures, the area may be cleansed twice daily, followed by the application of a topical antibiotic and bandage.

ABP Content Specifications(s)
- Recognize the clinical findings associated with common warts, and manage appropriately

Suggested Readings
- Craw L, Wingert A, Lara-Corrales I. Are salicylic acid formulations, liquid nitrogen or duct tape more effective than placebo for the treatment of warts in paediatric patients who present to ambulatory clinics? Paediatr Child Health. 2014;19(3):126-127.

Question 44

A 10-year-old girl collided with a telephone pole while riding her bike. Upon presentation to the emergency department, she is awake, alert, and complaining of significant abdominal pain. Her vital signs are a temperature of 37°C, heart rate of 140 beats/min, respiratory rate of 30 breaths/min, and blood pressure of 120/65 mm Hg, with an oxygen saturation of 95% on room air. On physical examination, the girl's pupils are equal, round, and reactive. She does not have any cervical spine tenderness or signs of extremity trauma, and she is moving all extremities with no deficits. There is bruising over the anterior aspect of her abdomen tracking to her left flank. Computed tomography scans reveal no intracranial bleeding and no bleeding or contusion in the chest. There is a significant hematoma in the duodenal wall, hemorrhage involving the pancreas, and a grade IV splenic laceration. Over the next 24 hours, the girl's breathing becomes increasingly rapid and shallow, with grunting and hypoxia. She requires endotracheal intubation, mechanical ventilation, and 60% oxygen to maintain her arterial oxygen saturation above 90%. On chest radiography, she has bilateral infiltrates.

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

A.   aspiration pneumonia

B.   neurogenic pulmonary edema

C.   pancreatitis

D.   postobstructive pulmonary edema

E.   septic shock

Question 44   **Preferred Response: C**

The child in this vignette, who has a traumatic pancreatic injury, develops rapid, shallow breathing and subsequent hypoxemic respiratory failure. Because she has bilateral infiltrates on chest radiography, a noncardiogenic cause of pulmonary edema, and a low ratio of arterial oxygen tension to fraction of inspired oxygen (PaO_2/FiO_2), a clinical diagnosis of acute respiratory distress syndrome (ARDS) can be made. In trauma, pancreatitis is a common cause of systemic inflammatory response syndrome and ARDS.

The diagnosis of ARDS is made clinically based on criteria that include respiratory failure, a PaO_2/FiO_2 ratio of less than 200, bilateral pulmonary infiltrates on chest radiography, and a noncardiogenic cause of pulmonary edema. Acute lung injury results from the same pathogenic causes and uses the same clinical diagnostic criteria, except for the PaO_2/FiO_2 ratio, which is between 200 and 300. ARDS has both pulmonary and extrapulmonary causes. Direct lung injury can be caused by pneumonia, ventilator-induced lung injury, chest trauma, aspiration pneumonitis, acute chest syndrome, drowning, and smoke inhalation. Extrapulmonary causes include sepsis, transfusion-related lung injury, burns, fat embolism, pancreatitis, trauma, or systemic inflammation from numerous other causes.

The pathogenesis of ARDS involves the breakdown of the barrier between alveoli and pulmonary capillaries, which leads to debris in the airspaces, including blood, pus, and proteinaceous fluid. Debris in the airspaces and increased surface tension from low surfactant production and function exacerbate alveolar collapse. This leads to hypoxia in areas that are perfused but not ventilated, also known as V/Q mismatch, or shunt. In addition, fluid in the interstitium leads to decreased lung compliance and low tidal volumes. Repetitive and forceful opening and closing of lung units to maintain tidal volume can exacerbate the inflammatory cascade, leading to the secretion of proinflammatory cytokines, continuing the cycle of increased capillary permeability. Death can occur in severe cases of ARDS from worsening hypoxia, ventilator-associated pneumonia, pulmonary hypertension, end-organ damage, prolonged respiratory failure, or chronic lung disease.

For the girl in the vignette, the history does not support a diagnosis of aspiration pneumonia. Neurogenic pulmonary edema generally does not occur without a brain injury. Postobstructive pulmonary edema can occur after an airway obstruction is relieved, but this child did not have airway obstruction. There is no fever, infection, or end-organ perfusion compromise to suggest septic shock.

PREP Pearls
- Acute respiratory distress syndrome can be caused by direct lung injury or indirectly from nonpulmonary conditions.
- Acute respiratory distress syndrome can cause death from hypoxia, end-organ damage, or complications from prolonged ventilation.
- Acute respiratory distress syndrome is a clinical diagnosis based on respiratory failure, bilateral lung infiltrates on radiographs, a ratio of arterial oxygen tension to fraction of inspired oxygen of <200, and a noncardiogenic cause of respiratory failure.

ABP Content Specifications(s)
- Identify the etiologies of acute respiratory distress syndrome
- Recognize complications of acute respiratory distress syndrome that can lead to death

Suggested Readings
- Carlo WA, Ambalavanan N. Conventional ventilation: traditional and new strategies. Pediatr Rev. 1999;20(12):e117-e126.
  http://pedsinreview.aappublications.org/content/20/12/e117?related-urls=yes&legid=pedsinreview;20/12/e117 .

Question 45

You are seeing a 9-year-old boy for a health supervision visit. During a recent camping trip in Arkansas, one of his bunkmates was bitten by a tick and developed fever and a rash 1 week later. The bunkmate is currently hospitalized and has been diagnosed with ehrlichiosis. Your patient's mother is now very concerned about tick-borne infections and inquires as to the best means of protecting her children in the future.

Of the following, the BEST advice you can provide is

A.   avoid arid conditions favored by ticks

B.   use of dark-colored clothing in order to repel ticks

C.   use of DEET-containing insect repellants starting at 6 weeks of age

D.   use of permethrin-embedded bed nets when sleeping in high risk areas

E.   use long-sleeved shirts and long pants

Question 45   **Preferred Response: E**

Personal protection is critical in preventing tick-borne infections. Long sleeved shirts and long pants limit the vectors' access to the host and limit the ability to transmit infection. Prevention of tick-borne infections involves personal protection, environmental measures, and reducing the time a tick is attached to a human. It is recommended that light-colored clothing be used in order to more easily identify an attached tick. The longer a tick is attached, the more likely it is to transmit an infectious illness.

Similarly, prevention of mosquito-borne infections involves both personal protection and environmental measures to discourage mosquito habitats, including removal of standing water and cleaning of pools. Personal protection includes use of nets, covering exposed skin, and use of repellents. Repellents with good activity include those with diethyltoluamide (DEET), picaridin, oil of lemon eucalyptus, and IR3535 (3-[N-Butyl-N-acetyl]-aminopropionic acid, ethyl ester). In general, longer protection is provided by repellents with higher concentrations of their active ingredients. When used appropriately, DEET-containing products with concentrations of up to 30% are considered safe in children. However, the American Academy of Pediatrics does not recommend use of DEET products in children younger than 2 months of age because of increased skin permeability.

Generally, ticks require humid environments for survival. The use of permethrin-embedded bed nets when sleeping would protect a child from mosquito-borne infections but not tick-borne infections.

PREP Pearls
- Prevention of tick-borne and mosquito-borne infections involves both personal protection and environmental measures.
- To prevent tick-borne infections, avoid humid environments, use light-colored long-sleeved shirts and long pants, and perform full body inspections with removal of attached ticks as soon as detected.
- Diethyltoluamide (DEET)-containing products are considered safe for use in children 2 months of age and older.

ABP Content Specifications(s)
- Identify the measures to prevent tick- and mosquito-borne infections

Suggested Readings
- American Academy of Pediatrics. Prevention of mosquitoborne infections. In: Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:213-215 .

Question 46

A 6-month-old infant presents with fever, vomiting, and gross hematuria. He was born to a 17-year-old gravida 1, para 1 mother with no prenatal visits. His vital signs show a temperature of 39°C, heart rate of 130 beats/min, respiratory rate of 28 breaths/min, blood pressure of 90/62 mm Hg, and oxygen saturation of 97% by pulse oximetry on room air. A physical examination shows mild dehydration and a prominent suprapubic area that is dull on percussion.

The laboratory and ultrasonography results show:
Complete blood cell count:
- White blood cell count, 20,000/µL (20.0×10^9/L)
- Hemoglobin, 10.9 g/dL (109 g/L) Hematocrit, 33%
- Platelet count, 200×10^3/µL (200×10^9/L)
- Electrolytes, normal
- Blood urea nitrogen, 28 mg/dL (10.0 mmol/L)
- Creatinine, 1.0 mg/dL (88.4 µmol/L)

Urine test strip and microscopy:
- Specific gravity, greater than 1.030
- pH, 5.5
- 4+ blood
- 4+ bacteria
- Positive for leukocyte esterase and nitrites
- Red blood cells, 40 to 50 per high power field

Abdominal ultrasonography is significant for bilateral hydronephrosis with renal cortical thinning and a thickened bladder wall.

Of the following, the patient is MOST likely to have an obstruction at the

A.   intravesical level

B.   membranous urethra level

C.   prostatic urethra level

D.   ureteropelvic level

E.   ureterovesical level

Question 46   **Preferred Response: C**

Posterior urethral valves (PUV) are identified by prenatal ultrasonography in the majority of cases. Lack of prenatal care leads to patients with PUV presenting later with urinary tract (more common) or respiratory (less common) problems associated with PUV. Postnatally, patients with PUV usually present with urinary tract infection (UTI), failure to thrive, abdominal distension (from enlarged bladder), and poor urinary stream or voiding difficulty. Older boys may present with UTIs or voiding dysfunction (urinary frequency, day and nocturnal enuresis, and poor urinary stream). Some of these patients may present in the neonatal period with respiratory distress. Neonates with PUV can have oligohydramnios caused by decreased fetal urinary excretion associated with severe bladder outlet obstruction. This would lead to pulmonary hypoplasia because normal amniotic fluid levels are required for normal lung development. The outcome for neonates with lung hypoplasia caused by severe PUV is poor.

The patient described in the vignette has UTI (fever, pyuria, leukocytosis), enlarged bladder (prominent suprapubic area dull on percussion), and renal failure (serum creatinine of 1 mg/dL [88 µmol/L]). The ultrasonographic findings are suggestive of PUV as the underlying cause of the patient's symptoms.

Ultrasonographic findings of bilateral hydronephrosis (dilatation of the renal pelvis with or without dilatation of the renal calyces), dilated bladder, thickened bladder wall, and a dilated posterior urethra in male patients are highly suggestive of underlying PUV. Bilateral hydronephrosis in PUV is caused by urinary tract obstruction distal to the urinary bladder in the prostatic urethra from a persistent and obstructing urogenital membrane. Overgrowth of urethra-vaginal folds, persistence of the urogenital membrane with abnormal urethral canalization, or abnormal integration of the Wolffian duct into the cloaca resulting in a thicker, fused, and obstructing folds are the proposed mechanisms for development of PUV caused by disruptions of male urethral development. Voiding cystourethrogram demonstrates the characteristic findings of a dilated and elongated posterior urethra during the voiding phase (after catheter removal). Direct visualization of the PUV by cystoscopy confirms the diagnosis. Ablation of urethral valves during cystoscopy is the preferred initial surgical approach of PUV ablation.

Patients with PUV develop chronic renal failure caused by associated renal dysplasia and acquired renal injury caused by poor bladder function or infection. Therefore, such patients are regularly followed to monitor their renal function, blood pressure, and growth. Long term management is aimed at management of bladder dysfunction (to minimize increased urinary tract pressures), avoiding UTI and complications associated with chronic kidney injury.

The membranous urethra is the shortest, least dilatable, and the narrowest part of the urethral canal (except the external urethral orifice). Patients with PUV develop chronic renal failure caused by associated renal dysplasia and acquired renal injury caused by poor bladder function or infection. Therefore, such patients are regularly followed to monitor their renal function, blood pressure, and growth. Long term management is aimed at management of bladder dysfunction (to minimize increased urinary tract pressures), avoiding UTI and complications associated with chronic kidney injury. The membranous urethra is the shortest, least dilatable,

and the narrowest part of the urethral canal (except the external urethral orifice). It extends from the apex of the prostate to the urethral bulb and perforates the urogenital diaphragm behind the pubic symphysis. The membranous urethra is not obstructed in patients with PUV.

Unilateral hydronephrosis is more common in children with congenital or acquired uretropelvic or uretrovesical obstruction. Hydronephrosis without ureteral dilatation is seen in uretropelvic junction obstruction. Hydronephrosis with dilation of the distal ureter without bladder distension indicates obstruction at the ureteral orifice (uretrovesical). Ureteropelvic and ureterovesical obstructions are not associated with the thickened bladder wall or dilated posterior urethra.

PREP Pearls
- Posterior urethral valves (PUV) are identified by prenatal ultrasonography in the majority of cases.
- Postnatally, patients with PUV usually present with urinary tract infections, failure to thrive, abdominal distension, and poor urinary stream.
- Ultrasonographic findings of bilateral hydronephrosis, dilated bladder, thickened bladder wall, and a dilated posterior urethra in male patients are highly suggestive of underlying PUV.
- Voiding cystourethrogram demonstrates the characteristic findings of a dilated and elongated posterior urethra during the voiding phase (after catheter removal).
- Ablation of urethral valves during cystoscopy is the preferred initial surgical approach of PUV ablation.

ABP Content Specifications(s)
- Plan the appropriate long-term management of posterior urethral valves
- Recognize the clinical findings associated with posterior urethral valves in children of various ages

Suggested Readings
- Elder JS. Obstruction of the urinary tract. In: Kleigman RM, Stanton BF, St Geme JW III, Schor NJ, Behrman RE, eds.Nelson Textbook of Pediatrics. 20th ed. Philadelphia, PA: Saunders Elsevier; 2015:2567-2575.

Question 47

A 9 month-old infant who attends childcare presents with a 2-day history of vomiting and copious, nonbloody, watery diarrhea. He is breastfed and receives puréed food 3 times per day. He has not had any new food exposures in the week prior to developing the symptoms. Several other children in his childcare have similar symptoms. On admission to the hospital, his vital signs are a temperature of 38^{0}C, heart rate of 140 beats/min, and respiratory rate of 30 breaths/min. He is pale and has decreased energy. Abdominal examination demonstrates hyperactive bowel sounds. His mother is concerned by his decreasing interest in eating.

Of the following, the laboratory stool study MOST sensitive to diagnose the cause of this infant's diarrhea is

- A. enzyme-linked immunosorbent assay
- B. latex agglutination
- C. polymerase chain reaction
- D. reducing substance
- E. viral culture

Question 47   **Preferred Response: C**

Rotavirus is the leading cause of diarrhea worldwide. It affects most children before 5 years of age, although vaccination, initiated in 2006 in the United States, is changing the epidemiology. Prior to the availability of vaccination, rotavirus was the most common cause of diarrhea in the United States, especially in childcare centers. It was traditionally more prevalent during cooler months. This is less consistent following widespread vaccinations. Since the introduction of the vaccine, rotavirus gastroenteritis cases have been reduced by more than 80%. The primary mechanism of transmission of rotavirus is believed to be fecal-oral, although fomites and respiratory spread have been reported. The virus is present in highest titers several days before and after onset of clinical symptoms.

Rotavirus infection begins with acute onset of vomiting and fever, followed 1 to 2 days later with watery diarrhea. Diarrhea is typically mild to moderate. Symptoms last 3 to 8 days. Severe cases can develop significant dehydration, acidosis, and electrolyte abnormalities. A small number of children experience neurologic issues including seizures and less commonly, encephalitis, encephalopathy, and cerebellitis.

Rotaviruses are from the *Reoviridae* family and are segmented, double-stranded RNA viruses with at least 7 different antigenic groups (A through G). Group A is the most common worldwide. Testing is rarely completed, as it has no influence on the course of the disease. The virus can be detected by enzyme-linked immunosorbent assay (ELISA), latex agglutination, electron microscopy, electrophoresis, and reverse transcriptase polymerase chain reaction (RT-PCR). Polymerase chain reaction techniques have been shown to be more sensitive in detection of the virus at all stages of infection and are the primary tool used in research. Despite this, ELISA vs used most often in clinical practice because of the ease of testing and affordability, in addition to the high sensitivity and specificity. Reducing substances would be nonspecific as to the cause of the diarrhea. Viral cultures are time consuming and are not generally used to diagnose the cause of the acute vomiting and diarrhea seen in the patient described in the vignette.

PREP pearls
- Rotavirus is leading cause of diarrhea worldwide, with decreasing prevalence due to vaccinations.
- Vaccinations are safe and effective.
- Enzyme-linked immunosorbent assay testing is available clinically, is affordable, and has high sensitivity and specificity, although polymerase chain reaction is the most sensitive method to detect rotavirus, it is primarily a research tool.

ABP Content Specifications(s)
- plan the appropriate diagnostic evaluation for rotavirus infection, and recognize when diagnostic evaluation may not be necessary
- understand the epidemiology of rotavirus infection
- Recognize the clinical manifestations of rotavirus infection

Suggested Readings

- gernstein Di. Rotavirus overview. Pediatr Infect DuisJ. 2009 , http:/dx.doi.org/10.1097/INF.0b013e3181957bee.

Question 48

A 14-year-old adolescent presents to your office accompanied by her mother who is concerned about behavioral difficulties and mild intellectual disability. The mother reports that her child has attention problems, depression, impulsiveness, and occasional delusional thinking. She states that her daughter is paranoid that "people are always talking about her." The patient's medical history is significant for a large ventricular septal defect repaired at 4 months of age and recurrent infections. She had significant hypocalcemia and low parathyroid hormone with seizures in infancy. She has mild bilateral sensorineural hearing loss. Physical examination is remarkable for a thin teenager with slender, hyperextensible hands and fingers. She has a long narrow face with a narrow nose with a squared nasal root, short upward-slanting palpebral fissures, a bifid uvula, and a high arch palate. She has a mild pectus excavatum present with a linear vertical scar to her chest. She has a short, thick webbed neck with reduced range of motion and mild scoliosis. She does have unusual mannerisms and affect.

Of the following, the MOST likely diagnosis is

   A. 22q11.2 deletion syndrome
   B. Alagille syndrome
   C. CHARGE syndrome
   D. oculo-auriculo-vertebral syndrome
   E. VACTERL association

Question 48   **Preferred Response: A**

The patient in the vignette has 22q11.2 deletion syndrome, also known as velocardiofacial syndrome or DiGeorge syndrome. Major findings with this disorder include congenital heart disease, palatal abnormalities, characteristic facial dysmorphology (including facial asymmetry), learning difficulties, hypocalcemia, and immune deficiency. An example of a patient with these findings is shown in Item

The heart disease often manifests as conotruncal defects including tetralogy of Fallot, perimembranous ventricular septal defect, truncus arteriosus, or interrupted aortic arch. The palatal deformities can range from velopharyngeal incompetence to cleft palate. Additional clinical findings can include feeding and swallowing problems, gastrointestinal and laryngotracheoesophageal anomalies, hearing loss, growth hormone deficiency, seizures, central nervous system anomalies, skeletal abnormalities (club feet, scoliosis, vertebral anomalies), renal abnormalities, ophthalmologic problems, thyroid problems, psychiatric disorders, autism, and enamel hypoplasia.

Diagnosis of 22q11.2 deletion syndrome can be made with fluorescence in situ hybridization (FISH) analysis for the submicroscopic deletion of chromosome 22 or a chromosomal microarray. 22011.2 deletion syndrome is a contiguous gene deletion syndrome. A contiguous gene deletion syndrome is caused by a microdeletion that encompasses 2 or more genes in tandem position along a chromosome. By virtue of the fact that several genes are involved, contiguous gene syndromes often impact multiple systems of the body. Thus, one must assess the function of the specific genes involved within the deletion and thoroughly examine the patient for involvement for those specified regions (heart, kidney, brain, etc). Other common contiguous gene deletions include Williams syndrome, Ip36 deletion, Smith-Magenis syndrome, and Cri-du-chat syndrome.

Treatment for 22q11.2 deletion syndrome entails a multidisciplinary approach specific to the individual's needs by a combination of specialists that may include allergy, audiology, cardiology, cardiac surgery, developmental/behavioral specialists, otolaryngology, endocrinology, pediatric dentistry, gastroenterology, immunology, medical genetics, neurology, ophthalmology, orthopedics, plastic surgery, psychiatry, pulmonology, rheumatology, hematology, urology, speech pathology, and early intervention.

Calcium supplementation is sometimes warranted. Thyroid profile should be monitored as well. Growth hormone may be required if poor growth in association with growth hormone deficiency is present. Sixty percent of adults have a psychiatric disorder (schizophrenia, anxiety, and depression) that will require a psychiatrist.

Baseline evaluations at time of diagnosis are noted in Item C48B.

Alagille syndrome is an autosomal dominant disorder caused by JAG1 or NOTCH2 gene mutations or microdeletions that presents with liver cholestasis, butterfly vertebrae, congenital heart disease, ophthalmologic abnormalities, kidney abnormalities, and classic facial features.

CHARGE syndrome is an autosomal dominant disorder often caused by CHD7 gene mutations that can present with a combination of the following findings: coloboma, heart defects, choanal atresia, retarded growth and development, and genital and ear anomalies. This patient lacks the coloboma or choanal atresia findings commonly seen.

Oculo-auriculo-vertebral syndrome, otherwise known as Goldenhar syndrome or craniofacial microsomia, presents with facial asymmetry caused by maxillary or mandibular hypoplasia, ear anomalies (preauricular facial tags or pits), hearing loss, and vertebral defects. Cardiac and renal anomalies can occur less commonly. It is a sporadic condition with no known cause. VACTERL association presents with a constellation of vertebral anomalies, anal atresia, cardiac anomalies, tracheoesophageal fistula or esophageal atresia, and renal and limb anomalies. The causes largely unknown.

PREP Pearls
- 22q11.2 deletion presents with a combination of clinical findings including congenital heart disease, palatal abnormalities, characteristic facial dysmorphology, learning difficulties, hypocalcemia, and immune deficiency.

Question 49

The parents of a previously healthy 9-month-old male infant bring him to your office for follow-up after a 3-day hospitalization for bronchiolitis. In the hospital, the infant received oxygen and intravenous hydration. He is much better now, but still having a cough. On physical examination, you note diffuse rales, but no respiratory distress.

Of the following, this infant is MOST likely to be at increased risk for
   A. bronchiectasis
   B. failure-to-thrive
   C. recurrent pneumonia
   D. recurrent sinusitis
   E. recurrent wheezing

Question 49   **Preferred Response: E**

The most likely condition to occur in individuals who have a history of bronchiolitis during early infancy or childhood vs recurrent wheezing.

Bronchiolitis, the clinical syndrome of inflammation of the bronchioles, is usually caused by an acute viral infection in children younger than 2 years. The most common pathogens are respiratory syncytial virus (RSV), adenovirus, human metapneumovirus, influenza, and parainfluenza virus. Typically, upper respiratory symptoms (rhinorrhea) are followed by lower respiratory tract infection and inflammation, which may result in wheezing, rales, tachypnea, coughing, using accessory respiratory muscles, hypoxia, and even respiratory failure. Lower respiratory tract symptoms occur on day 2 to 3 of the illness, peak on day 5 to 7, and gradually resolve within 2 to 3 weeks. In healthy full-term infants, bronchiolitis is usually a self-limited disease and requires supportive care only.

Infants and young children with bronchiolitis are at increased risk for recurrent wheezing, particularly during the first decade of life. Studies have found this risk to range from 2 to 4 times that of controls. Also, some studies have shown a correlation between bronchiolitis, specifically RSV infection, in infancy or early childhood and the subsequent diagnosis of asthma. The nature of this association is uncertain and causality cannot be determined. This correlation may be reflective of the multifactorial nature of risk for asthma: genetic predisposition, environmental aeropollutants, atopy, immunologic of the multifactorial nature of risk for asthma: genetic predisposition, environmental aeropollutants, atopy, immunologic mechanisms, airway growth affected by prior infection, and inflammation, In one study, non-RSV etiology compared with RSV etiology of bronchiolitis before 2 years of age significantly increased asthma risk in adulthood.

Healthy children, without underlying cardiopulmonary disease or immunodeficiency, who have a single episode of bronchiolitis that follows the expected clinical course and time to resolution, should not have a greater risk of bronchiectases, failure to thrive, recurrent sinusitis, or recurrent pneumonia.

PREP pearls
- Infants who have an episode of bronchiolitis have a 2 to 4 times greater risk for recurrent wheezing than controls, particularly during the first decade of life,
- Some studies have shown a correlation between bronchiolitis in infancy or early childhood and the subsequent development of asthma.

ABP Content Specifications(s)
- Recognize the frequency of recurrent wheezing in infants who have bronchiolitis is caused by respiratory syncytial virus or rhinovirus

Suggested Readings

- Piippo-savolainen E, Korppi M, Korhonen K, Remes S. Adult asthma after non-respiratory syncytial virus bronchiolitis in infancy: subgroup analysis of the 20-year prospective follow-up study. Pediatr Int. 2007   001:

Question 50

A 3-month-old infant presents to your practice with a physical examination remarkable for the finding shown in Item Q50.

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

A. reassure her mother that this will self-resolve with time

B. refer her to a geneticist out of concern for Sturge-Weber syndrome

C. refer her to a pediatric oncologist out of concern for malignancy

D. refer her to a pediatric ophthalmologist out of concern for amblyopia

E. refer her to a pediatric ophthalmologist out of concern for strabismus

Item Q50. Findings for the infant described in the vignette. Courtesy of D. Krowchuk.

Question 50   **Preferred Response: D**

When the path of light from the pupil through to the retina is obstructed for an extended period of time in a young child, the brain will cull the visual pathways from that eye in favor of the other eye, resulting in decreased vision in the affected eye. This is amblyopia, which can be irreversible if the proper visual pathway is not restored while critical eye and brain development are taking place. The visual obstruction can be external or internal to the globe. The infant shown in the picture in this vignette has a bulging, red lesion of the left upper eyelid that is covering the upper lid and is likely to obstruct light flow through the iris and possibly the pupil, likely a hemangioma. The fact that the lesion covers part of the eye should prompt an ophthalmology consult out of concern for the development of amblyopia.

Although hemangiomas can regress with time, it can take many months or even years. As this lesion covers the eye and may obstruct the light pathway, waiting for spontaneous resolution may lead to permanent vision loss in that eye. Reassurance alone is therefore not the correct management.

Sturge-Weber syndrome, also called encephalotrigeminal angiomatosis, is a disorder characterized by a pronounced "port wine" facial birthmark and neurological deficits. Other organs may also be involved. Although the infant in this picture has a facial birthmark, it is not a traditional port wine stain birthmark, given its limited distribution to the eyelid and its bulge. While some hemangiomas may have malignant elements, these are extremely rare. A child with strabismus presents with either exotropia or esotropia that can impact visual development and lead to amblyopia.

PREP Pearls
• An obstruction of the pathway of light from the pupil to the retina in a young child can lead to permanent vision loss in that eye, called amblyopia.
• Strabismus occurs when a child develops exotropia or esotropia and can lead to amblyopia.
• Any time there is concern for the development of amblyopia, an immediate referral to an ophthalmologist should be considered.

ABP Content Specifications(s)
• Recognize the visual consequences of a tumor or hemangioma in the periorbital area

Suggested Readings
• Gunton KB. Advances in amblyopia: what have we learned from PEDIG trials? Pediatrics. 2013;131(3):540-547. DOI:http://dx.doi.org/10.1542/peds.2012-1622.

Question 51

You are seeing an 11-year-old girl in the sixth grade who has been having recent trouble in school. Her school performance was described as "okay" until she started fourth grade. Since that time, her grades are described as having been "poor." Her early motor skill development was normal, although the mother states that she was "a little slow" in acquiring language in the first 2 years after birth. Her mother adds that she has not been concerned about her speech since the girl started kindergarten. Last year, her parents arranged for the girl to have an IQ test, which revealed intelligence in the average range. She does not have many friends and is not a troublemaker. There are frequent conflicts around a rule that she completes her homework before she is allowed to watch television at night. In the office, the girl's speech sounds normal, she smiles, is pleasant, and seems to interact normally for her age.

Of the following, the evaluation MOST likely to reveal the cause of her poor school performance is

A.   audiology testing

B.   learning disability testing

C.   serum ferritin level

D.   serum lead level

E.   Wood lamp skin examination

Question 51   **Preferred Response: B**

A key etiology to suspect for this girl's past 2 years' worth of schooling difficulty (appearing after she started fourth grade work) would be a learning disability. Learning disabilities may not become apparent until children's academic demands increase, particularly for otherwise intelligent children who can initially find their own ways to compensate for learning challenges. The index of suspicion for this problem needs to be high because not recognizing the presence of a learning disability will likely lead to school and social failure. The benefit of intervention is increased with early identification, particularly if one intervenes at the point of "increased effort of learning" before there is school failure.

Children with learning disabilities can have normal range or high overall IQ. In the past, a lower academic performance relative to what one might predict from IQ was considered to be the definition of a learning disability, but that is no longer the case. Response to intervention (RTI) is now the standard approach because this helps to screen out those with poor performance due to a lack of adequate instruction. The RTI involves a psychoeducational assessment, followed by increasing levels of instruction in areas of deficit then reassessment. If a child's performance normalizes with minimal intervention, this would argue against having a learning disability.

Another common cause of schooling difficulties that may not appear until after academic demands increase would be inattentive type attention-deficit/hyperactivity disorder, but it is a bit less likely here because of a lack of parental reports of attention problems. It is also not one of the options listed.

An audiology test would be key to perform had her speech failed to normalize after the initial reported delay in speech development, and such testing may still be reasonable as part of health maintenance. However, it is not very likely to be the cause of her current poor school performance.

Serum ferritin level would be helpful in assessing for iron deficiency. Chronic iron deficiency does produce neuropsychiatric effects that include lower average math scores and impaired psychomotor development. Ten years of age is not a peak time for this diagnosis, however, and it is less likely to be found in this scenario than a learning disability. A serum lead level would also be appropriate to assess in a young child with developmental concerns because of its potential neurotoxicities. However, as with iron deficiency, 10 years of age is older than when one would typically expect to find this as an abnormality in the absence of persistent pica. A Wood lamp skin examination can help to detect ash leaf spots that are frequently found in tuberous sclerosis. While tuberous sclerosis is associated with intellectual disabilities, it is not a likely diagnosis.

PREP Pearls
- Children with learning disabilities can have normal range or high overall IQ.
- Response to intervention is now the standard approach for learning disability assessment because this helps to screen out poor performance due to a lack of adequate instruction.

- The benefit of learning disability intervention is increased with early identification, particularly if intervening at the point of "increased effort of learning" before school failure.

ABP Content Specifications(s)
- Understand the various etiologies of school-related difficulties
- Plan the appropriate diagnostic evaluation of poor school performance

Suggested Readings
- Dworkin PH. School learning problems and developmental differences. In: McInerny TK, Adam HM, Campbell DE, Kamat DM, Kelleher KJ, eds. American Academy of Pediatrics Textbook of Pediatric Care. Elk Grove Village, IL: American Academy of Pediatrics; 2009.

Question 52

A 13-year-old adolescent with trisomy 21 who underwent surgery at 4 months of age for an atrioventricular canal defect is brought to your office for a school physical examination. He has not been seen by you for 4 years. He has not been on any medications. He is in special education class and would like to participate in the Special Olympics. On review of symptoms, his mother states that he has not had the same level of energy for the last 2 weeks. He has been sleeping during the day on the weekend and has been too tired to do his school work.

On physical examination, his heart rate is 56 beats/min, his respiratory rate is 32 breaths/min, and his blood pressure is 92/45 mm Hg. He has mild intercostal retractions. His pulse oximetry is 90% on room air. He is alert but not talkative today. His chest examination shows decreased breath sounds bilaterally with crackles. His cardiac examination reveals a slow regular heart rate with a 2/4 diastolic murmur and a 2/6 systolic murmur at the left midclavicular line at the fourth intercostal space. No hepatosplenomegaly or lymphadenopathy is noted.

Given the combination of history and physical findings, the MOST likely etiology of his current fatigue is

A.   hyperthyroidism

B.   leukemia

C.   mitral stenosis

D.   pericardial effusion

E.   tricuspid valve regurgitation

Question 52   Preferred Response: C

The murmur described for the patient in this vignette is in the mitral position; it is both diastolic and systolic. The systolic murmur is consistent with mitral regurgitation, while the diastolic murmur is consistent with mitral stenosis. As both of these worsen, the patient will need repair or replacement of the mitral valve.

Patients with trisomy 21 have a 40% to 50% risk of congenital heart disease (CHD). Of the patients with CHD, 40% to 50% will have an endocardial cushion defect or atrioventricular septal defect (AVSD), commonly called atrioventricular (AV) canal defects. There are many variations on the anatomy of AV canal defects. Some children will have a large atrial septal defect (ASD) component (called a primum defect, where the lowest portion of the atrial septum is missing) and almost no ventricular component, with a cleft mitral valve. Other children will have a huge ventricular septal defect (VSD), as well as an ASD. The ASD is not the type that can close spontaneously. The VSD in an AV canal defect is classified as an inlet VSD and will not close spontaneously. The degree of flow or shunt across the defect can be very large or minimal, depending on the exact architecture of the AV valve or valves.

The natural history of infants with AVSDs is to develop symptoms of pulmonary overcirculation or high output heart failure within the first few months of life. The timing will be dependent on the size of the defect and the pulmonary vascular resistance. If no heart failure develops at several months of age, this would raise concern for pulmonary hypertension. Another confounding issue for patients with trisomy 21 is their increased risk for hypothyroidism. Follow-up with genetics and early intervention is very important for these patients. After initial repair of an AVSD, children require ongoing monitoring for development of atrioventricular valve dysfunction. The mitral valve often becomes regurgitant and may become stenotic, requiring subsequent repair or replacement later in life.

Children born with trisomy 21 may also have increased risk for leukemia and hypothyroidism. In this patient, there is no bruising, hepatosplenomegaly, or lymphadenopathy, making leukemia less likely as a cause of the child's fatigue. There is an increased risk of pericardial effusion, especially if there is hypothyroidism in patients with trisomy 21, but the physical examination is not consistent with that. The murmur suggests atrioventricular valve insufficiency and stenosis, rather than tricuspid valve findings.

Children born with congenital heart disease, even when initial repairs are done and are successful, require ongoing monitoring over their lifetime. Patients with AVSD repairs often require further surgery to address their mitral valve. Patients with tetralogy of Fallot after initial repair will require ongoing monitoring for pulmonary insufficiency and need for pulmonary valve replacement as they age.

Patients with single ventricle physiology such as hypoplastic left heart syndrome will require a minimum of 3 surgeries to complete palliation. The initial surgery for hypoplastic left heart syndrome is called the Norwood procedure to establish a systemic outflow tract utilizing the

pulmonary artery and then creating a source of pulmonary blood flow. The second surgery is the Glenn anastomosis, which creates an attachment of the superior vena cava to the right pulmonary artery. The third surgery is called the Fontan procedure, which creates (passive) circulation from the inferior vena cava to the pulmonary artery. This circulation is passive, therefore any disease process that increases the pulmonary vascular resistance, such as pneumonia, can cause a severe decrease in cardiac output in these patients.

PREP Pearls
- Atrioventricular canal defects are common in trisomy 21 patients.
- The natural history of infants with atrioventricular septal defect is to develop symptoms of pulmonary overcirculation or high output heart failure within the first few months of life.
- Ongoing monitoring is essential in any child who has undergone repair of congenital heart disease, especially for complex lesions. Additional procedures may be needed in later childhood or adolescence.

ABP Content Specifications(s)
- Recognize the major clinical findings associated with the various types of cyanotic congenital heart disease
- Understand the various etiologies of school-related difficulties

Suggested Readings
- American Academy of Pediatrics. Health supervision for children with Down syndrome. Pediatrics. 2001;107(2):442-449. DOI: http://dx.doi.org/10.1542/peds.107.2.442.

Question 53

You are seeing a 7-year-old boy in your office for a health supervision visit. The boy has asthma that is poorly controlled with his maintenance medication regimen of inhaled fluticasone and montelukast, with albuterol as a rescue medication. He has seasonal allergic rhinitis to grass that partially responded to nasal steroids last spring. The boy has no other medical problems. The allergist has recommended subcutaneous immunotherapy.

Of the following, the MOST appropriate advice regarding this recommendation is that the boy should

A.   be observed for at least 2 hours after each immunotherapy injection

B.   discontinue immunotherapy if there is a large local reaction at the injection site

C.   not receive immunotherapy while his asthma is poorly controlled

D.   not require injectable epinephrine for home if observed appropriately postinjection

E.   receive immunotherapy only during grass allergy season

Question 53   **Preferred Response: C**

The boy in the vignette should not receive immunotherapy while his asthma is poorly controlled. Most deaths associated with allergen-specific immunotherapy have been in patients with asthma. Although asthma is not a contraindication for immunotherapy, this treatment method should be used with caution in any patient with poor asthma control or a recent increase in asthma symptoms.

Specific immunotherapy (SIT) is useful for allergic rhinitis, and is most effective in a narrow range of allergens. Patients should be selected for SIT based on history and allergy testing, and after other causes of nasal symptoms have been ruled out. The effectiveness of SIT for allergic rhinitis in drug-resistant cases is well documented. Three years of treatment can provide long-lasting effects, whereas the benefits of only a single year of therapy diminish quickly. The risks and cost effectiveness of SIT for allergic rhinitis should be assessed on a case-by-case basis. Some patients with atopic dermatitis may benefit from SIT, with a decrease in the number and severity of atopic dermatitis events.

Specific immunotherpy affects the natural history of allergic disease. Allergic children start with a limited range of allergen sensitivities that progress over time. SIT may limit the tendency to acquire new sensitivities. A proportion of persons with allergic rhinitis will develop asthma every year, and SIT may be able to decrease this number.

Although current asthma treatments suppress inflammation and relieve bronchospasm, they are not curative. Allergen avoidance can be of benefit in allergic asthma, but is not always possible. SIT can be beneficial in allergic asthma, reducing symptom scores, the number of exacerbations, and the need to increase medications. However, it is important to recognize that the majority of fatal reactions with SIT have occurred in patients with asthma. Although asthma is not an absolute contraindication, SIT should be used with caution in patients with uncontrolled asthma or those with reduced peak flow rates.

Specific immunotherapy is contraindicated in patients with cardiac disease. Other conditions that should induce caution when using SIT include autoimmune conditions, immunodeficiency syndromes, and malignant disease. There is no direct evidence that SIT is harmful in these conditions, but manipulating the immune system in these patients could theoretically cause exacerbations or worsening of their underlying disease. ACE-inhibitors and β-blockers are contraindications to using SIT. β-blockers decrease the response to epinephrine that may be needed to combat adverse reactions to SIT. Angiotensin converting enzyme blockers can accentuate angioedema. A large local reaction may occur during SIT, but is not a contraindication to therapy. The most common intervention for a large local reaction is observation.

The medical indications for immunotherapy include venom hypersensitivity, atopic dermatitis, allergic rhinitis, and asthma. Approximately 40 deaths occur each year in the United States because of anaphylactic reactions to Hymenoptera venom. Although it is common to have some venom-specific IgE antibodies a few months after a sting, a few patients will have high levels of

IgE for years. This group of individuals, with persistently high IgE levels, is at risk for anaphylaxis with future envenomation. In addition, patients with systemic symptoms after a Hymenoptera sting are at higher risk for future systemic reactions, with this risk decreasing over time. The risk of systemic reaction is over 17% after 10 years in individuals with a previous systemic response compared with a 2% to 3% risk of systemic response in the general population. Desensitization with venom will reduce the risk of systemic reaction to approximately 10% after completion of therapy.

Patients should be observed for 30 minutes after each injection, longer observation is not required. All patients receiving SIT should have injectable epinephrine for home use in case of a delayed reaction, regardless of how long they are observed. Patients begin SIT with weekly injections until a maintenance dose is achieved, and then receive monthly or 6-week maintenance injections for 3 to 5 years. Use of SIT should not be limited to the allergy season.

PREP Pearls
- Specific Immunotherapy (SIT) is effective in treating allergic rhinitis, asthma, and venom hypersensitivity.
- All patients receiving SIT should have injectable epinephrine available for home use in case of a delayed hypersensitivity reaction.
- Although SIT is not contraindicated in asthma, it has been associated with fatal reactions in patients with asthma and should not be used in patients with poorly controlled asthma.

ABP Content Specifications(s)
- Understand the indications and limitations of immunotherapy, and manage associated side effects

Suggested Readings
- Frew AJ. Allergen immunotherapy. J Allergy Clin Immunol. 2010;125(2):S306-S312. DOI:http://dx.doi.org/10.1016/j.jaci.2009.10.064.

Question 54

A 6-year-old boy presents to your office for evaluation of a limp. His parents have noted a left-sided limp for the past 3 to 4 months. At times, the boy reports activity-related anterior thigh pain. There is no history of antecedent infection, trauma, recurrent fevers, change in appetite, or unusual rashes. On physical examination, you note decreased internal rotation and abduction of the left hip.

Of the following, the boy's presentation is MOST consistent with a diagnosis of

A.   juvenile idiopathic arthritis

B.   Legg-Calvé-Perthes disease

C.   septic arthritis

D.   slipped capital femoral epiphysis

E.   transient synovitis of the hip

Question 54   **Preferred Response: B**

This boy's age, chronicity of symptoms, lack of constitutional symptoms, and mild activity-related pain point to a diagnosis of Legg-Calvé-Perthes disease (LCP). Although LCP is often described as causing a painless limp, this condition can be associated with mild pain, particularly in the first 4 to 6 months.

Legg-Calvé-Perthes disease occurs when an inadequate blood supply to the femoral head epiphysis leads to synovitis and early necrosis (initial stage), resulting in collapse of the femoral head (fragmentation stage). The femoral head reossifies in the healing phase, but may not retain a spherical shape and therefore may not fit well in the acetabulum.

Legg-Calvé-Perthes disease has a male predominance and 90% of cases are unilateral. The peak incidence occurs in children aged 4 to 8 years. Children typically present with a history of limp and mild activity-related pain. On physical examination, there is decreased internal rotation and abduction of the affected hip(s).

In patients with LCP, anterior-posterior (AP) and frog-leg lateral radiographs show collapse of the femoral head. The degree of collapse along the lateral aspect of the femoral head is used to grade LCP; this is known as the lateral pillar classification. For patients with negative radiography findings, in the setting of a high degree of clinical suspicion for LCP, a technetium bone scan can be used to detect a deficient blood supply to the femoral head.

The treatment of LCP depends on a patient's age and the severity of radiographic findings. Children who present with symptoms before 6 years of age tend to have a better prognosis and treatment is often nonsurgical. Physical therapy can help maintain adequate hip range of motion, and nonsteroidal anti-inflammatory medications can be taken as needed for pain. Children older than 8 years who develop LCP tend to do poorly and generally require surgical intervention. Children with LCP should avoid high-impact physical activities.

Juvenile idiopathic arthritis (JIA) is an unlikely diagnosis for this boy, because he has no morning stiffness and his pain worsens with activity. Additionally, solitary hip involvement is uncommon with JIA. Septic arthritis is rapidly progressive and typically causes severe pain, fever, and refusal to bear weight. Boys with slipped capital femoral epiphysis typically present between the ages of 11 and 15 years, and the majority are overweight or obese. Transient synovitis has a short duration of symptoms, typically less than 1 week.

PREP Pearls
- Children with Legg-Calvé-Perthes disease generally present with chronic limp and may have activity-related pain.
- Pain or a decrease in range of motion with hip internal rotation and/or abduction generally indicates intraarticular hip pathology.

ABP Content Specifications(s)
- Understand the natural history of avascular necrosis (Legg-Calve-Perthes disease)
- Formulate a differential diagnosis of avascular necrosis (Legg-Calve-Perthes disease) in a patient with a limp

Suggested Readings
- Sarwark JF, LaBella C. Pediatric Orthopaedics and Sports Injuries: A Quick Reference Guide. Elk Grove Village, IL: American Academy of Pediatrics; 2014.

Question 55

A previously healthy 13-year-old adolescent presents to your office with a facial rash that began 2 days after the onset of fever and myalgias. He reports that the involved skin is swollen and painful. On physical examination, his temperature is 38°C, heart rate is 100 beats/min, respiratory rate is 20 breaths/min, and blood pressure is 115/68 mm Hg. He is ill appearing, but does not appear toxic. You note a large, raised, tender area of nonfluctuant induration and erythema with clear borders over the left cheek. His mucous membranes are moist. The remainder of the physical examination is unremarkable.

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

A.   intravenous ceftriaxone

B.   intravenous vancomycin

C.   oral amoxicillin

D.   oral doxycycline

E.   oral trimethoprim/sulfamethoxazole

Question 55   **Preferred Response: A**

The moderately ill adolescent boy in the vignette presents with erysipelas as manifested by a large, red, raised "butterfly" appearing rash on his face with a clear line of demarcation between the involved and uninvolved areas. Beta-hemolytic streptococci cause the vast majority of cases of erysipelas. Patients with systemic manifestations of erysipelas, as described for the boy in the vignette, should be treated with parenteral therapy. Ceftriaxone is convenient because it treats group A Streptococcus (GAS), can be administered once daily, and allows for outpatient therapy. Patients in whom oral antibiotic therapy is appropriate can be treated with penicillin or amoxicillin for 5 to 10 days depending on the clinical response.

In children, erysipelas is a relatively uncommon manifestation of GAS infection. Acute pharyngotonsillitis is the most common presentation of GAS disease; it occurs in all age groups but is most common in school-aged children. Complications of acute pharyngitis can include scarlet fever, cervical adenitis, peritonsillar and retropharyngeal abscesses, sinusitis, and otitis media. Nonsuppurative complications of untreated infection can include acute rheumatic fever and acute glomerulonephritis. In children 1 to 3 years of age, the most common manifestation of GAS infection is a febrile, protracted illness accompanied by rhinitis called streptococcosis. Other manifestations of GAS infection can include cellulitis, impetigo, pyoderma, bacteremia, endocarditis, septic arthritis, osteomyelitis, pyomyositis, pneumonia, necrotizing fasciitis, omphalitis, surgical wound infection, and streptococcal toxic shock syndrome. An association between GAS and the development of neuropsychiatric or tic disorders (pediatric autoimmune neuropsychiatric disorders association with streptococcal infection, PANDAS) is unproven.

Group A Streptococcus is transmitted through respiratory tract secretions or direct contact with the affected area (eg, impetigo). Environments with close crowding facilitate spread of the organism. Group A Streptococcus pharyngitis is more common in the late fall, winter, and spring in temperate climates, whereas pyoderma occurs more commonly in tropical regions and during warm seasons. The incidence of invasive GAS diseases is highest in infants and the elderly. In the United States, the development of acute rheumatic fever is rare, presumably because of the decreased circulation of rheumatogenic strains. The incubation period for GAS pharyngitis is 2 to 5 days, whereas impetigo develops 7 to 10 days after acquisition of the organism.

A swab of both tonsils and the posterior oropharynx for rapid antigen testing and bacterial culture are recommended for the diagnosis of GAS pharyngitis. The bacterial culture swab should be sent in all cases of negative rapid antigen testing, because of the possibility of false-negative results. For nonpharyngitis infections, routine bacterial cultures of blood and other sterile sites or tissues will isolate the organism. The diagnosis of acute rheumatic fever should be made using the Jones criteria (Item C55). Streptococcal toxic shock syndrome requires the isolation of GAS and a myriad of clinical findings to include hypotension and a minimum of two of the following additional presentations: an erythematous macular rash, severe soft tissue infection, adult respiratory distress syndrome, a coagulopathy, elevated liver enzymes or increased bilirubin, and increased creatinine levels.

Item C55. Jones Criteria for Diagnosis of Acute Rheumatic Fever.*

Major Criteria	Minor Criteria	Supporting Evidence
Carditis Polyarthritis Chorea Erythema marginatum Subcutaneous nodules	Clinical findings: Fever, arthralgia[†] Laboratory findings: Elevated acute phase reactants; prolonged PR interval	Positive throat culture or rapid test for Group A streptococcal antigen OR Elevated or rising streptococcal antibody test

* Diagnosis requires 2 major criteria or 1 major and 2 minor criteria with supporting evidence of antecedent group A streptococcal infection.

[†] Arthralgia is not a minor criterion in a patient with arthritis as a major criterion.

Reprinted with permission from Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. *Red Book: 2015 Report of the Committee on Infectious Diseases*. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015

Penicillin is the drug of choice for the treatment of GAS infection; amoxicillin is an appropriate oral alternative given its palatability. An infection caused by a strain of GAS resistant to either penicillin or a cephalosporin has never been documented. In patients with severe penicillin allergy, alternative agents for the treatment of GAS infection include clindamycin, azithromycin, erythromycin, and clarithromycin. Clindamycin is useful for severe infections because it has a long postantibiotic effect and inhibits the production of bacterial toxins; however, it should never be used alone in life-threatening infections because of the possibility of GAS-resistant strains. Vancomycin is appropriate for the empirical treatment of invasive infection caused by gram-positive organisms and for definitive therapy for severe infection (eg, endocarditis) caused by methicillin-resistant S aureus, but is unnecessary for the treatment of erysipelas or other infections caused by GAS. Tetracyclines (eg, doxycycline) and sulfonamides (eg, trimethoprim-sulfamethoxazole) are not useful for the treatment of GAS disease.

PREP Pearls
- Acute pharyngotonsillitis is the most common presentation of group A streptococcal disease.
- Erysipelas can present as a red, raised "butterfly" appearing rash on the face with a clear line of demarcation between involved and uninvolved areas. Ceftriaxone is used to treat erysipelas in moderately ill patients.
- Penicillin is the drug of choice for the treatment of infection caused by group A Streptococcus.

ABP Content Specifications(s)
- Understand the epidemiology of Streptococcus pyogenes
- Plan appropriate management for a patient with Streptococcus pyogenes infection
- Plan the appropriate diagnostic evaluation of suspected Streptococcus pyogenes infection

Suggested Readings
- American Academy of Pediatrics. Group A streptococcal infections. In: Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:732-734.

Question 56

A 17-year-old, previously healthy adolescent is brought to the emergency department with complaint of an acute change in mental status and abdominal pain. His father heard him entering the house at approximately 4 AM. Upon speaking with him, the father suspected that his son "was high on drugs."

Physical examination shows a diaphoretic, delirious, and agitated teenager, who is repetitively wringing his hands and writhing in bed. The patient can tell you his name, but when asked where he is, he states he is "at the concert." He does not consistently follow commands. His vital signs include a heart rate of 134 beats/min, respiratory rate of 24 breaths/min, blood pressure of 152/96 mm Hg, axillary temperature of 38.5°C, and pulse oximetry of 98% on room air. There are no obvious signs of traumatic injury. The boy's pupils are 5 mm in diameter with sluggish reactivity bilaterally. His mucous membranes are moist. His lungs are clear with no signs of respiratory distress. His cardiac examination reveals tachycardia with a regular rhythm. His pulses are bounding and symmetric. His abdomen is soft and nontender with no bruising and hyperactive bowel sounds. Neurologic examination reveals normal strength and brisk reflexes in all extremities. The patient is agitated, pulling at his intravenous tubing, and will not lie still, ignoring your attempts to calm and redirect his behavior verbally.

Of the following, the BEST option for treating this patient's symptoms is

A.   intramuscular haloperidol

B.   intravenous dantrolene

C.   intravenous diazepam

D.   intravenous sodium bicarbonate

E.   oral propranolol

Question 56   **Preferred Response: C**

The adolescent in the vignette presents with agitation, tachycardia, hypertension, and hyperthermia that can be explained by acute cocaine toxicity. Of the agents listed, the best option for treating his symptoms is intravenous diazepam.

Illicit cocaine use by adolescents and adults is relatively widespread, and small children may also be exposed inadvertently (or even intentionally) to cocaine by others. Pediatric providers must be able to recognize the clinical findings associated with acute cocaine intoxication and manage these cases appropriately.

Signs and symptoms in patients with cocaine toxicity arise from stimulation of the sympathetic nervous system. Characteristic findings include central nervous system (CNS) excitation (which may manifest as euphoria, agitation, delirium, hyperactivity), dilated pupils, tachycardia, hypertension, and diaphoresis. Patients may experience headache, myalgias, and abdominal pain; in the most severe cases, focal neurologic symptoms, intracranial hemorrhage, myocardial ischemia, seizures, and even coma may result.

Patients with mild toxicity from cocaine abuse or exposure generally require supportive therapy only. For patients with moderate or severe agitation, as well as for those with mild to moderate hypertension, benzodiazepines (including diazepam) are the agents of choice. Benzodiazepines would also be the first-line agents for initial treatment of seizures related to cocaine toxicity. Benzodiazepines are the first line treatment for both the CNS overstimulation and cardiovascular sequelae arising from cocaine's toxic effects. However, additional antihypertensive medications, such as phentolamine or sodium nitroprusside may be required in patients with severe or refractory hypertension. Active cooling measures should be initiated for those with severe hyperthermia. Fluid therapy with urine alkalinization may be necessary for patients with rhabdomyolysis precipitated by cocaine abuse. Activated charcoal should be strongly considered for gastric decontamination in cases involving oral ingestion.

Haloperidol is an antipsychotic medication that has long been used for sedation of patients with acute agitation or delirium, especially those with a history of psychiatric disorders including schizophrenia. Although haloperidol may effectively decrease agitation in the patient in the vignette, it can reduce the body's heat-dissipating capacity and lower the seizure threshold. Therefore, haloperidol would not be the safest agent for treating agitation in a patient with cocaine toxicity, which can also be complicated by hyperthermia and seizures. Benzodiazepines are generally the preferred agents for treating agitation in patients with drug overdose.

Dantrolene is used to treat muscular rigidity caused by malignant hyperthermia and neuroleptic malignant syndrome. It does not have an established role in the treatment of acute cocaine intoxication.

In the clinical scenario of drug intoxication, sodium bicarbonate is used to treat cardiovascular toxicity secondary to tricyclic antidepressant overdose. It may also be added to intravenous fluids to achieve urine alkalinization, which can increase the rate of elimination of many poisons from

the body. Sodium bicarbonate would not be the initial agent of choice to treat agitation and hypertension in a child with cocaine toxicity.

The use of β-blockers such as propranolol is generally not recommended in the treatment of cocaine-related cardiovascular sequelae, including hypertension. These agents may cause unopposed stimulation of alpha-adrenergic receptors, which could contribute to increased coronary vasoconstriction and end-organ ischemia.

PREP Pearls
- The predominant clinical effects seen in children and adolescents with cocaine intoxication include central nervous system excitation, hypertension, tachycardia, hyperthermia, diaphoresis, and dilated pupils.
- Benzodiazepines are the agents of choice for patients with mild to moderate cocaine toxicity, as well as for those with seizures related to cocaine overdose.
- For children and adolescents with acute cocaine intoxication, initial management must focus on immediate stabilization of the airway, adequate ventilation, and prompt establishment of vascular access to support circulatory function.

ABP Content Specifications(s)
- Recognize the major behavioral consequences of cocaine use/abuse
- Recognize the clinical findings associated with acute cocaine intoxication, and manage appropriately
- Identify the major physiologic consequences associated with cocaine use/abuse, including those associated with the various means of administration

Suggested Readings
- Gummin DD, Aks SE. Cocaine. In: Erickson TB, Ahrens WR, Aks SE, Baum CR, Ling LJ, eds. Pediatric Toxicology: Diagnosis and Mangement of the Poisoned Child. New York, NY: McGraw Hill; 2005;chap 57:392-397.

Question 57

A mother who received no prenatal care has just delivered a male newborn who weighs 2,100 g. Although her menses were irregular, the gestational age is estimated to be 39 weeks by her last menstrual period. The newborn's physical examination includes pink skin with some cracking pale areas with rare veins, thinning lanugo on the back, well-curved pinnae with soft but ready recoil, a stippled areola with a 1 to 2 mm breast bud, testes just below the inguinal ring, and an anterior crease on the plantar surface of the foot.

Of the following, the MOST appropriate description of this newborn is

A.   average for gestational age late preterm newborn

B.   average for gestational age term newborn

C.   large for gestational age late preterm newborn

D.   small for gestational age late preterm newborn

E.   small for gestational age term newborn

Question 57   **Preferred Response: A**

The description of the newborn in the vignette is consistent with an average-for–gestational age late preterm infant. Pregnancy dating traditionally has been based on the last menstrual period (LMP), with inherent error related to maternal recall. First trimester ultrasound is now often used to confirm the estimated date of delivery. In situations in which mothers receive limited care or have irregular menstrual cycles and do not have an early ultrasound, clinical assessment must be used to provide an estimate of gestational age.

The Dubowitz scoring system was developed in 1970 to estimate gestational age based on neuromuscular and physical findings. This scoring system required modification, with the increased survival of extremely premature infants, thus leading to the development of the New Ballard Score (NBS) (Item C57). The NBS has been validated in newborns between 20 and 44 weeks' gestational age and found to be accurate to within 2 weeks. Neurologic examination of posture and physical examination of the genitalia have the highest reliability.

Accurate assessment of gestational age and size for gestational age are important, because this information will guide the provision of safe and appropriate newborn care. Newborns born between 34 0/7 weeks and 36 6/7 weeks' gestational age are considered late preterm and require close monitoring for issues of respiratory control, feeding skills, and temperature regulation. Full-term small-for-gestational age newborns have an increased risk of hypoglycemia and hypothermia, whereas newborns who are large for gestational age should be monitored for hypoglycemia.

The infant in the vignette has the neuromuscular and physical findings of a late-preterm infant, including testes just below the inguinal ring, a single plantar crease, and immature posture with decreased flexion. His weight of 2,100 g is consistent with an appropriate-for–gestational age late preterm infant.

PREP Pearls
- Accurate assessment of gestational age and size for gestational age are important to guide safe and appropriate newborn care.
- The New Ballard Score is used to estimate gestational age based on neuromuscular and physical examination findings in newborns between 20 and 44 weeks' gestational age and is accurate to within 2 weeks.
- The neuromuscular examination of posture and physical examination of the genitalia have the highest reliability for estimation of gestational age.

ABP Content Specifications(s)
- Distinguish between small-for-gestational age and preterm gestation in low-birth-weight infants
- Recognize the physical and behavioral characteristics of infants born prematurely, at term, or post-term

Suggested Readings

- Ballard JL, Khoury JC, Wedig K, et al. New Ballard score, expanded to include extremely premature infants. J Pediatr. 1991;119:417. Abstract available at http://www.ncbi.nlm.nih.gov/pubmed/1880657.

Item C57. The New Ballard Score.

Neuromuscular Maturity							
	-1	0	1	2	3	4	5
Posture							
Square Window (wrist)	>90°	90°	60°	45°	30°	0°	
Arm Recoil		180°	140°-180°	110°-140°	90°-110°	< 90°	
Popliteal Angle	180°	160°	140°	120°	100°	90°	< 90°
Scarf Sign							
Heel to Ear							

Physical Maturity							
	-1	0	1	2	3	4	5
Skin	sticky friable transparent	gelatinous red, transluscent	smooth pink, visible veins	superficial peeling and/or rash, few veins	cracking pale areas, rare viens	parchment deep cracking, no vessels	leathery cracked wrinkled
Lanugo	none	sparse	abundant	thinning	bald areas	mostly bald	
Plantar Surface	heel-toe 40-50 mm: -1 < 40 mm: -2	> 50 mm no crease	faint red marks	anterior trans-verse crease only	creases over anterion 2/3 of sole	creases over entire sole	
Breast	imperceptible	barely perceptible	flat areola no bud	stippled areola 1-2 mm bud	raised areola 3-4 mm bud	full areola 5-10 mm bud	
Eye/Ear	lids fused loosely: -1 tightly: -2	lids open pinna flat stays folded	slightly curved pinna; soft; slow recoil	well-curved pinna; soft but ready recoil	formed and firm instant recoil	thick cartilage ear stiff	
genitals male	scrotum flat, smooth	scrotum empty faint rugae	testes in upper canal rare rugae	testes descending few rugae	testes down good rugae	testes pendulous deep rugae	
genitals female	clitoris prominent labia flat	prominent clitoris small labia minora	prominent clitoris enlarging minora	majora and minora equally prominent	majora large minora small	majora cover clitoris and minora	

Maturity Rating													
Score	-10	-5	0	5	10	15	20	25	30	35	40	45	50
Weeks	20	22	24	26	28	30	32	34	36	38	40	42	44

The New Ballard Score (Ballard JL, Khoury JC, Wedig K, et al. New Ballard Score, expanded to include extremely premature infants. *J Pediatr* 1991; 119:417.

Question 58

A 16-year-old adolescent is brought to the emergency department by her friends after she had a generalized tonic-clonic seizure at a house party where no adults were present. Her friends drop her off and leave immediately. She is arousal, but keeps falling asleep and cannot provide a coherent history. Her physical examination reveals a temperature of 36.9°C, blood pressure of 106/68 mm Hg, heart rate of 92 beats/min, and respiratory rate of 20 breaths/min. Her physical examination is unremarkable. Her neurologic examination is unremarkable, except for altered mental status. Her extraocular movements are normal and she can move all extremities equally. There is no arm dysmetria when she reaches for objects and she can sit up independently in bed. Gait examination is deferred because of her mental status. She has another generalized tonic-clonic seizure in the emergency department that stops before any medication is administered. To prevent further seizures, fosphenytoin (1,500 mg) is administered intravenously. A urine pregnancy test is negative and a computed tomography of the head is normal. Toxicology screen, complete blood cell count, and electrolytes are normal. She is admitted to the hospital for further monitoring. The next morning, her neurological examination shows bilateral nystagmus, brisk reflexes, and truncal ataxia, with a wide-based and unsteady gait.

Of the following, the MOST likely cause of the change in her neurological examination findings seen on the morning examination is

A.   cerebellitis

B.   fosphenytoin

C.   intoxication

D.   postictal state

E.   subdural hematoma

Question 58   **Preferred Response: B**

The girl in the vignette has acute ataxia resulting from the loading dose of intravenous fosphenytoin she received the night before in the emergency department. Acute ataxia, including truncal ataxia, limb dysmetria, and lateral nystagmus, is a common side effect of high doses of intravenous fosphenytoin or phenytoin, but can also happen if blood levels are in excess of the therapeutic range.

Acute cerebellitis due to infection can cause ataxia and nystagmus. It is usually associated with other infectious symptoms, such as fever or meningismus. In toddlers, acute cerebellar ataxia is a common neurologic cause of acute ataxia. This condition is thought to be postviral and therefore infectious symptoms can be absent. The girl in the vignette has no infectious symptoms of acute cerebellitis and is much older than the typical patient with postviral acute cerebellar ataxia.

Alcohol intoxication causes acute cerebellar dysfunction including acute ataxia. It is unknown whether the girl in the vignette ingested alcohol or other substances before her arrival in the emergency department; however, her neurologic examination in the emergency department did not show ataxia originally. A postictal state can cause ataxia, especially gait ataxia, but lateral nystagmus is not a common postictal phenomenon. Cerebellar stroke can cause ataxia and nystagmus, but typically the nystagmus is 1-sided (ipsilateral to the cerebellar stroke).

Development of a subdural hematoma while in the hospital overnight is very unlikely, and presentation with cerebellar signs is uncommon. Hemiparesis, headache, or signs of increased intracranial pressure are more common signs of a new subdural hematoma. Although each of these disorders is a possible cause, the most likely cause of acute ataxia for the girl in the vignette is the intravenous fosphenytoin.

PREP Pearls
- Acute ataxia can be caused by medications, including phenytoin.
- In toddlers, a common neurologic cause of acute ataxia is acute cerebellar ataxia.

ABP Content Specifications(s)
- Recognize the presentation of acute ataxia

Suggested Readings
- Dinolfo EA. Evaluation of ataxia. Pediatr Rev. 2001;22(5):177-178. DOI: http://dx.doi.org/10.1542/pir.22-5-177.
- Ramierz-Montealegr D, Mink, JW. Ataxias. In: Kliegman RM, Stanton BF, St. Geme JW III, Schor NF, Behrman RE. Nelson Textbook of Pediatrics. 20th ed. Philadelphia, PA: Saunders Elsevier; 2015: 2883-2888.

Question 59

You are called to the newborn nursery to evaluate a term newborn with ambiguous genitalia. On physical examination, you find a well-appearing newborn in no acute distress. The physical examination is unremarkable, except for the genital examination. The phallus is under 1 cm in length. There is hypospadias with the urethral meatus visible at the base of the penis, a bifid scrotum, and gonads palpable in the scrotum bilaterally (Item Q59). Family history reveals multiple maternal aunts who are infertile.

Genitalia of the infant showing small phallus, hypospadias with bifid scrotum, and scrotal gonads. Courtesy of Jordan Pinsker, MD.

Of the following, the MOST common cause of ambiguous genitalia in a newborn with this clinical presentation is

A.   5-α reductase deficiency

B.   21-hydroxylase deficiency

C.   Denys-Drash syndrome

D.   partial androgen insensitivity

E.   Turner syndrome

Question 59   **Preferred Response: D**

The classification of ambiguous genitalia has been simplified since the adoption of new terminology in 2006. Rather than using confusing terms such as "pseudohermaphrodite" or other vague terms, new terminology recognizes disorders of sex development (DSD), as shown below (Item C59).

Item C59. Updated Classification of Disorders of Sex Development (DSD)

Disorder of Sex Development	Older Terminology	Meaning
46,XY DSD	Male pseudohermaphrodite	Undervirilized male
46,XX DSD	Female pseudohermaphrodite	Overvirilized female
Sex chromosome DSD or Ovotesticular DSD	True hermaphrodite	Mixed gonadal findings
46,XX testicular DSD	XX sex reversal	Male phenotype with 46,XX
46,XY complete gonadal dysgenesis	XY sex reversal	Female phenotype with 46,XY

Courtesy of J. Pinsker

The infant in the vignette has descended testes that are palpable in the scrotum. This implies that he is an undervirilized male with a "46,XY DSD." In addition, there is a family history of maternal aunts who are infertile, implying the disorder is inherited in an X-linked fashion. Of the answer choices listed that are compatible with 46,XY DSD, only androgen insensitivity causes ambiguous genitalia and is inherited in this manner. Androgen insensitivity is the most common underlying cause of 46,XY DSD. When a child has partial androgen insensitivity due to incomplete expression of the mutation, the child can have ambiguous genitalia. When the mutation causes complete androgen insensitivity, the child will appear completely female, but will have a blind vaginal pouch and no Müllerian structures, resulting in infertility. Thus, the maternal aunts in this family who are infertile have complete androgen insensitivity and a 46,XY karyotype.

5-α reductase deficiency results in impairment of the conversion of testosterone to the more potent dihydrotestosterone. This disorder can also be a cause of 46,XY DSD. However, it is not inherited in an X-linked pattern, and occurs far less commonly than partial androgen insensitivity.

21-hydroxylase deficiency, the most common form of congenital adrenal hyperplasia (CAH), can affect both boys and girls, but would only cause ambiguous genitalia in a girl (46,XX DSD). Thus, in a child with palpable gonads in the scrotum (implying the presence of Y chromosome material leading to testicular formation), this would not be the underlying cause.

Denys-Drash syndrome results from a mutation in the WT1 gene. These children can have ambiguous genitalia (46,XY DSD) or in some cases appear completely female. However, in boys the testes will remain undescended (unlike the child in this vignette), and both boys and girls develop severe kidney disease. This syndrome occurs much less commonly than partial androgen insensitivity.

Turner syndrome, in which there is loss of all or part of 1 sex chromosome in a phenotypic female, is characterized by growth failure and pubertal delay, amongst other findings. It is not a cause of ambiguous genitalia.

PREP Pearls
- Ambiguous genitalia can be classified into disorders of sex development (DSD), with the terms 46,XY DSD representing an undervirilized male and 46,XX DSD representing an overvirilized female.
- The most common cause of 46,XY DSD is androgen insensitivity, which is inherited in an X-linked pattern.

ABP Content Specifications(s)
- Identify factors associated with virilization in female infants
- Understand the normal process of sex differentiation of a fetus
- Recognize the clinical features associated with androgen insensitivity syndrome

Suggested Readings
- Nabhan ZM, Lee PA. Disorders of sex development. Curr Opin Obstet Gynecol. 2007;19:440-445. Abstract available athttp://www.ncbi.nlm.nih.gov/pubmed/17885459.

Question 60

A 6-year-old child presents to your office for follow-up with a chief parental concern of persistent cough. The cough started 6 weeks ago in association with a viral respiratory illness. Associated fever and rhinorrhea have resolved, but the cough has persisted and worsened. The cough is mucousy and worse at night. The parents believe that sputum has been produced but swallowed. They report occasional vomiting after coughing, which has appeared mucousy. A chest radiograph series (posteroanterior and lateral views) was obtained 10 days ago and was read as normal without infiltrate, effusion, or air trapping.

The child is in preschool with multiple sick contacts. There is no prior history of chronic cough or exertional intolerance. The child's weight and height have consistently measured at the 75th percentile. Stools are normal without grease or malodor. There is no history of recurrent ear or skin infections. There is no history of headache or vomiting, other than as noted with the current cough. There have been no witnessed choking episodes.

On physical examination, the child is well developed and well nourished. The patient's respiratory rate is comfortable and nonlabored at 20 breaths/min. Tympanic membranes are clear without erythema. There is mucoid drainage in the posterior oropharynx. Inferior nasal turbinates are edematous with scant mucus. Lungs are clear bilaterally without wheezing, differential aeration, crackles, or rhonchi. Abdomen is soft and nontender. Extremities are without clubbing. Gait is normal.

Of the following, your MOST likely diagnosis is

A.   bronchiectasis

B.   cystic fibrosis

C.   immunodeficiency syndrome

D.   Kartagener syndrome

E.   sinusitis

Question 60   **Preferred Response: E**

The child in this vignette demonstrates symptoms of chronic cough. This child's cough is mucousy and began in the setting of a respiratory viral illness. The persistent nature of the cough, its wet quality, and the worsening of cough with supine positioning and sleep are most suggestive of postnasal drip and acute sinusitis.

The evaluation of the child with chronic cough is complex, but a complete history and physical examination are likely to assist with reaching the proper diagnosis in a timely manner without the need for extensive testing. The majority of coughs that last less than or equal to 8 weeks are postviral in nature and otherwise uncomplicated.

Most children experience multiple episodes of nasal congestion and cough each year; the most common etiology for these symptoms is an acute or resolving viral illness. Children may be expected to have 6 to 8 viral upper respiratory infections each year. Only 5% to 13% of these will be complicated by a bacterial superinfection. In an effort to prevent excessive antibiotic use for viral illness, the Infectious Diseases Society of America in 2012 provided guidelines to distinguish which clinical presentations best identify patients with acute bacterial, as opposed to viral, rhinosinusitis (RS):

1.   Onset with persistent symptoms or signs compatible with acute RS, lasting greater than or equal to 10 days without evidence of improvement

2.   Onset with severe symptoms/signs of high fever ($\geq$ 39°C) and purulent nasal discharge or facial pain for at least 3 to 4 days (consecutive) at beginning of illness

3.   Worsening symptoms or signs characterized by new onset fever, headache, or increase in nasal discharge following a typical upper respiratory infection that lasted 5 to 6 days and were initially improving

The young child with sinusitis may present only with a chronic cough and not with the more classic symptoms of nasal congestion, nasal drainage, postnasal drip, or fetid breath.
A child with recurrent or chronic sinusitis should be evaluated for conditions that predispose to recurrent infections or sinopulmonary disease, such as immune deficiency, cystic fibrosis, or ciliary dysfunction.

The patient in this vignette has experienced 6 weeks of cough, but has otherwise been well and thriving. The most recognizable etiologies for chronic bronchiectasis include suppurative lung diseases (cystic fibrosis, ciliary dyskinesia), immunodeficiency, chronic dysphagia, and aspiration. This child has a normal chest radiograph and has not demonstrated the signs or symptoms that would suggest a comorbid condition. A child with cystic fibrosis would likely have a chronic cough of longer duration. Failure to thrive and other symptoms of fat malabsorption, including malodorous or greasy stools, may predominate. The child with cystic fibrosis may also demonstrate digital clubbing.

A provider should suspect immunodeficiency in a child with recurrent skin, ear, sinus, or pulmonary infections. A child with immune defects may also present with poor weight gain or growth.

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The child with Kartagener syndrome not only has ciliary dysfunction, but also situs inversus totalis. Dextrocardia, as well as a right-sided stomach "bubble," will likely be noted on chest radiograph.

PREP Pearls
- Prior to initiating antimicrobials, a child with acute nasal congestion and cough should be evaluated by existing clinical criteria to differentiate viral from bacterial rhinosinusitis.
- Children with chronic cough should undergo a thorough clinical history and physical examination to evaluate for signs and symptoms of a chronic underlying condition.

ABP Content Specifications(s)
- Plan effective screening evaluation of chronic cough

Suggested Readings
- Chow AW, Benninger MS, Brook I, et al. IDSA clinical practice guideline for acute bacterial rhinosinusitis in children and adults. Clin Infect Dis. 2012;54(8):e72-e112. DOI: http://dx.doi.org/10.1093/cid/cir1043.

Question 61

A 14-year-old adolescent was recently evaluated in the emergency department for vomiting, which has now resolved. As part of his evaluation, he had a comprehensive metabolic panel, which was normal except for an alanine aminotransferase of 54 U/L and aspartate aminotransferase of 70 U/L. Abdominal ultrasonography obtained at that time showed fatty deposition in the liver. On physical examination today, you note a body mass index greater than 95th percentile for age and acanthosis nigricans. His parents ask how his condition can be most effectively treated.

Of the following, the MOST effective initial treatment approach for this adolescent is

A.   bariatric surgery

B.   diet supplementation with vitamins E and C

C.   family-based behavioral treatment

D.   Metformin

E.   ursodeoxycholic acid

Question 61   **Preferred Response: C**

The adolescent in the vignette has findings consistent with nonalcoholic fatty liver disease (NAFLD), a component of metabolic syndrome (MetS). The primary treatment is slow weight loss. Studies have shown this type of weight loss is best accomplished through family-based behavioral treatment. This type of intensive program requires both parent and child, and involves education about behavior therapy techniques, nutrition, and exercise. Unfortunately, access to such programs is often limited. Weight loss in the range of 5% to 10% reductions in body mass index can lead to clinically significant improvements in lipid levels and insulin resistance. Rapid weight loss such as what might occur with bariatric surgery may accelerate liver inflammation. Studies of the effect of bariatric surgery on NAFLD are inconclusive. Other interventions suggested for NAFLD treatment have included use of hepatoprotective agents, as well as agents to improve insulin action. Metformin may be useful, but the data are contradictory. One small study demonstrated improvement in transaminase levels, hepatic fat quantities, and insulin resistance with metformin, while a larger study in children did not find benefit. Further studies are needed before metformin can be recommended for treatment of NAFLD. The hepatoprotective agent ursodeoxycholic acid and antioxidants vitamin E and C have been suggested for NAFLD treatment, but studies have shown little benefit from these agents.

The differential diagnosis of obesity in children includes exogenous obesity, genetic conditions, and endocrine disorders. Exogenous obesity accounts for more than 95% of cases of excess weight. Height velocity and pubertal development are among the most important factors to differentiate exogenous obesity from other causes. Most children with exogenous obesity have normal to increased height velocity and a normal pubertal progression, although often at an earlier age than normal weight peers. The presence of short stature, dysmorphic features, and delayed puberty should cause the pediatrician to pursue other, much rarer, causes of obesity.

Nonalcoholic fatty liver disease is a common complication of exogenous obesity and represents a range of liver pathology from deposition of large fat droplets within hepatocytes all the way to cirrhosis and end stage liver disease, requiring transplantation. Inflammation in the presence of fatty deposition, with or without fibrosis, is called nonalcoholic steatohepatitis (NASH). The vast majority (90%) of patients with NAFLD are obese, and the condition is present in up to 80% of obese children. Boys are more commonly affected than girls, and the highest prevalence is among Hispanic children and youth, with Hispanic adolescents more likely to develop liver fibrosis than other ethnic groups. Despite higher rates of insulin resistance, African-American children have low rates of NAFLD compared to other racial and ethnic groups.

Current evidence suggests that the primary metabolic dysfunction in MetS is insulin resistance. Insulin resistance in the liver is fundamental to MetS pathogenesis. Two critical insulin-mediated liver pathways respond differentially in the face of insulin resistance. When presented with a glucose load, glucose homeostasis in the liver is impaired (ie, increased gluconeogenesis) and increased de novo lipogenesis (DNL) is seen. Increased DNL leads to increased intrahepatic lipid storage and increased release of triglyceride and apoprotein B as very low density lipoprotein, which in turn leads to MetS-associated dyslipidemia. At the same time in the liver, lipid metabolism is shifted toward mitochondrial β oxidation, which converts fatty acids into

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adenosine triphosphate and ketone bodies. This also leads to elevated production of free radicals, and the oxidative stress produced may promote inflammation characteristic of NASH.

Clinical findings in patients with NAFLD include acanthosis nigricans in 50% of children and hepatomegaly in 40% to 50%, although this is often difficult to detect. Laboratory studies show hyperinsulinemia and dyslipidemia, particularly hypertriglyceridemia. Elevated aminotransferase levels, particularly alanine aminotransferase, are commonly seen and may be twice the upper limits of normal. Ultrasonography of the liver reveals homogenously increased echogenicity, but does not differentiate fatty infiltration from inflammation or fibrosis. Other imaging modalities tend to be nonspecific and are not currently recommended. These biochemical and radiographic abnormalities are neither consistently present nor predictive of disease severity, therefore further evaluation may be required. The definitive diagnostic test for NAFLD is a liver biopsy, but the risks of the procedure often outweigh the benefits in otherwise well children. Research is underway to validate more specific and sensitive tests such as the pediatric NAFLD fibrosis index (PNFI) combined with the enhanced liver fibrosis test (ELF). The PNFI is determined from age, waist circumference, and triglyceride levels, and the ELF is made up of hyaluronic acid, aminoterminal propeptide of type III procollagen, and inhibitor of metalloproteinase 1 levels. A measure of hepatocyte apoptosis (caspase-generated cytokeratin-18 fragments) has been useful in adults with NASH; further work is needed to establish its validity in children. In children with persistently elevated transaminases, further evaluation to rule out other causes of liver disease should be undertaken, and consultation with a pediatric gastroenterologist is often recommended.

PREP Pearls
- The primary treatment of nonalcoholic fatty liver disease (NAFLD) is gradual weight loss. Studies show that the most effective method to achieve this is through family-based behavioral treatment.
- Findings of NAFLD include obesity, acanthosis nigricans, hepatomegaly, dyslipidemia (particularly hypertriglyceridemia), and elevated aminotransferase levels.
- Insulin resistance in the liver is critical to the development of metabolic syndrome.
- Nonalcoholic steatohepatitis is defined as liver inflammation in the presence of fatty deposition.
- The imaging study of choice for NAFLD is ultrasonography, although it does not differentiate fatty infiltration from inflammation or fibrosis.

ABP Content Specifications(s)
- Recognize the various complications associated with obesity
- Formulate a differential diagnosis in a patient who is obese

Suggested Readings
- Bremer AA, Mietus-Snyder M, Lustig RH. Toward a unifying hypothesis of metabolic syndrome. Pediatrics. 2012;129(3):557-570. DOI: http://dx.doi.org/10.1542/peds.2011-2912.

Question 62

You are seeing a 5-year-old girl in your office. Her mother reports that she first noticed a small bump in the child's "private area" about 2 months ago, and now the patient has several small bumps in the area. Her mother reports that the child has not complained of pain. The mother denies erythema or drainage from the lesions. Upon physical examination, the child is shy, but cooperative. A photo of the lesions in her perianal area is shown in Item Q62.

Of the following, the evaluation of this child should include

A.   an anal Papanicolaou test

B.   an assessment of immunologic function

C.   a biopsy of the lesions

D.   evaluation for sexual abuse

E.   high resolution anoscopy

Item Q62. Photo of the lesions in the perineal area of the girl in the vignette. Courtesy of D. Krowchuk.

Question 62   **Preferred Response: D**

The child described in the vignette has condylomata acuminata (genital warts) as a result of human papillomavirus (HPV) infection. These lesions often present as 1 to 5 mm flesh-colored, verrucous papules, which may coalesce into large plaques. While children can potentially autoinoculate (spread of HPV from other sites on the patient) or heteroinoculate (spread by nonsexual contact with lesions of a caregiver or others), condylomata acuminata have been associated with childhood sexual abuse. Therefore, consideration of sexual abuse is warranted in the evaluation of the prepubescent child in this vignette.

Condylomata acuminata is primarily a clinical diagnosis; biopsy is not typically indicated. Many cases self-resolve. Medical management includes the use of topical products such as imiquimod or podophyllotoxin. Surgical resection or laser therapy may be needed for lesions that do not resolve or cause symptoms. Genital warts are caused by low risk HPV types; however, in adults, they can be a marker for carriage of high risk types. The oncogenic potential of high risk types acquired prior to puberty is unknown. There are no current recommendations for Papanicolaou testing or high resolution anoscopy in this scenario.

While there are recommendations that all children with condylomata acuminata be tested for HIV, there is no recommendation for an assessment of immunologic function.

PREP Pearls
- Sexual abuse should be considered for any prepubescent child with condylomata acuminata.
- Condylomata acuminata (genital warts) is a result of human papillomavirus infection.
- Medical management includes the use of topical products such as imiquimod or podophyllotoxin.

ABP Content Specifications(s)
- Recognize the clinical findings associated with condylomata acuminata
- Understand the significance of condylomata acuminata in patients of various ages, including their association with sexual abuse
- Plan the appropriate management of condylomata acuminata

Suggested Readings
- Culton D, Morrell DS, Burkhart CN. The management of condyloma acuminata in the pediatric population. Pediatr Ann. 2009;38(7):368-372.

Question 63

You are evaluating a 15-year-old adolescent for management of her acne. She has used an over-the-counter salicylic acid wash without benefit. She has been in good health and is taking no medications. Her physical examination reveals 5 to 6 active inflammatory lesions on each cheek and the chin. There are several open comedones (Item Q63). There is no scarring. The forehead, chest, and back are free of acne lesions.
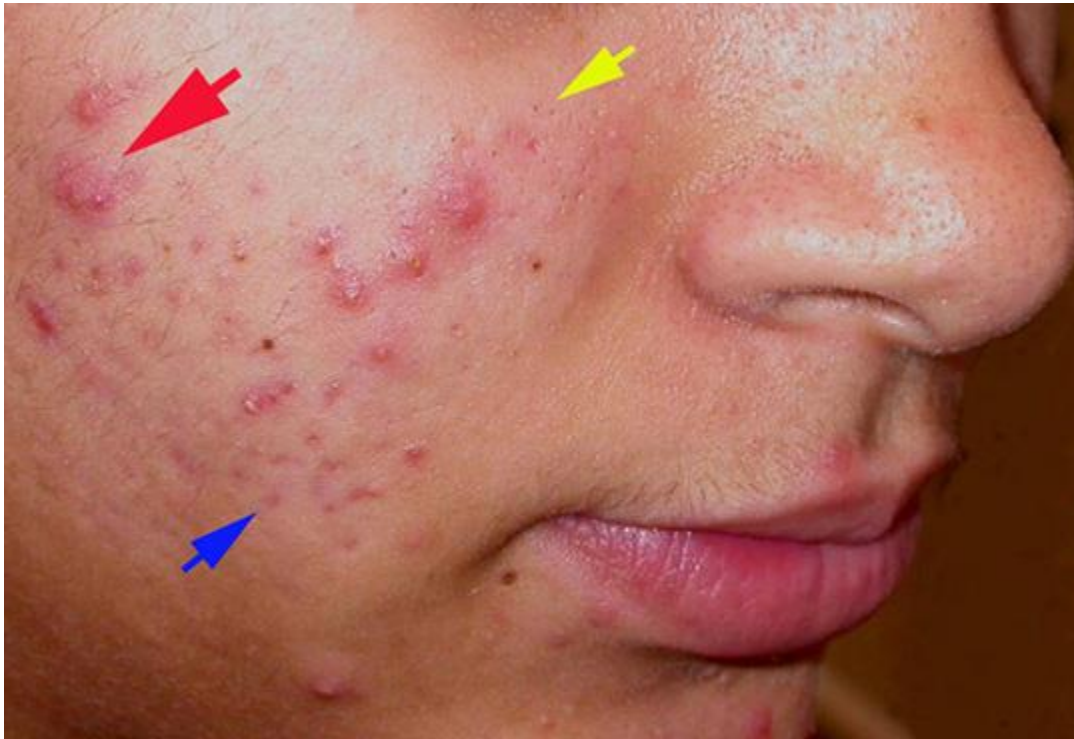

Adolescent described in the vignette. Reprinted with permission from Krowchuk DP, Mancini AJ, eds. Pediatric Dermatology. A Quick Reference Guide. Elk Grove Village, IL: American Academy of Pediatrics; 2012.

Of the following, the MOST appropriate treatment for this adolescent's acne is

A.   benzoyl peroxide topically

B.   benzoyl peroxide/clindamycin and a retinoid topically

C.   benzoyl peroxide/clindamycin topically

D.   doxycycline orally and benzoyl peroxide topically

E.   a retinoid topically

Question 63   **Preferred Response: B**

The adolescent in the vignette has moderate inflammatory and comedonal acne involving the face (Item C63A). There are several inflammatory papules (red arrow) and open comedones (yellow arrow). No scarring is evident, but there are several resolving inflammatory lesions (blue arrow). These lesions may remain erythematous or violaceous for months and are often confused with scars.

Moderate inflammatory and comedonal acne on the face. There are inflammatory papules and pustules (red arrow), open comedones (yellow arrow), and resolving inflammatory lesions (blue arrow). 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.

Several factors contribute to the development of acne. Key among these are disordered keratinization (leading to obstruction within pilosebaceous follicles), increased sebum production (which contributes to obstruction), and inflammation (due, in large part, to activation of the immune system by the bacterium, Propionibacterium acnes). As obstruction increases, follicles may rupture, contributing to the inflammatory process. In some patients, the inflammatory process results in scarring. On the face, scars appear as small pits, while on the trunk, they are hypopigmented macules.

Treatment plans, especially for those who have moderate or severe disease, should be designed to impact as many aspects of the disease pathophysiology as possible. In such cases, it is important to note that follicular obstruction is present, even if blackheads (open comedones) and whiteheads (closed comedones) are not observed. Accordingly, for the adolescent in the vignette, the most appropriate treatment is benzoyl peroxide (BPO)/clindamycin applied to the face each

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morning and a retinoid (eg, tretinoin or adapalene) applied at bedtime. Therapy with BPO, BPO/clindamycin, or doxycycline orally combined with BPO would not address the obstructive component of her disease. In addition, because the adolescent has moderate acne limited to the face, has no scarring, and is using no medication, an attempt to manage the inflammatory component of her disease with topical agents is reasonable. Although topical retinoids have some anti-inflammatory activity, the number of papules and pustules exhibited by the adolescent in the vignette indicates the need for specific treatment of this component of her disease.

Guidelines for acne management exist, but treatment plans should be individualized based on the patient's perception of disease severity, past experiences with medications, and the ability to adhere to therapy. Suggested treatment plans for mild, moderate, and severe acne are presented in Item C63B, Item C63C, and Item Item C63D. For more detailed information, refer to the suggested readings.

Item C63B. Options for Managing Mild Acne.

Only the face is involved

- Inflammatory or mixed
    - Benzoyl peroxide (BPO) 5% once daily
    - Alternatives (once daily):
        - BPO/antibiotic fixed-dose combination product
        - BPO/topical retinoid fixed-dose combination product
        - Topical antibiotic/topical retinoid fixed-dose combination product
- Comedonal
    - Topical retinoid (as a single agent) or topical retinoid-containing fixed-dose combination product once daily

Face and chest or back involved

- Inflammatory or mixed: BPO wash 5% or 10% once daily in the shower
- Comedonal: Salicylic acid wash 2% once daily in the shower

Item C63C. Options for Managing Moderate Acne.

Only the face is involved

- Topical retinoid at bedtime
    - Individual product (tretinoin cream 0.025% or adapalene cream 0.1%) OR
    - Fixed dose combination product (like benzoyl peroxide [BPO]/adapalene or clindamycin/tretinoin)
- Topical antimicrobial each morning
    - BPO (if topical retinoid alone prescribed) OR
    - BPO/topical antibiotic fixed-dose combination product (if topical retinoid alone prescribed)

Face and chest or back involved

- Topical retinoid at bedtime (as above), AND
- Oral antibiotic (like doxycycline or minocycline 50 mg to 100 mg once or twice daily)
- Consider adding BPO wash once daily in the shower to the chest and back

If no improvement, proceed to **Item C63D** or refer to a dermatologist.

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Item C63D. Options for Managing Severe Acne.

-Consider referral to a dermatologist, OR

-Maximize the treatment plan

- Topical retinoid for the face: If a low potency agent (like tretinoin 0.025% cream or adapalene cream 0.1%) was prescribed previously, consider increasing the potency (eg, tretinoin micro gel 0.04% or 0.1%)
- High-dose oral antibiotic (like doxycycline or minocycline 100 mg twice daily)
- Add BPO once daily to the face (if not being used as part of a fixed-dose combination product)
- For women, consider a combined oral contraceptive

If no improvement, consider referral to a dermatologist for possible oral isotretinoin therapy.

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PREP Pearls
- A topical retinoid should be included in the management of adolescents with moderate or severe acne. Obstruction within follicles is present and should be addressed, even if blackheads and whiteheads are not observed.
- Extensive inflammatory acne (ie, involving the trunk, as well as the face) requires treatment with an oral antibiotic.

ABP Content Specifications(s)
- Plan the appropriate management of acne

Suggested Readings
- Eichenfield LA, Krakowski AC, Piggott C, et al. Evidence-based recommendations for the diagnosis and treatment of pediatric acne. Pediatrics. 2013;131(Suppl 3):S163-S186. DOI: http://dx.doi.org/10.1542/peds.2013-0490B.

Question 64

A 2-year-old girl was found face down on the surface of the family's above ground pool. She was out of her caregiver's sight for 10 min before she was found. When she was pulled from the water, she was motionless, blue, and not breathing. Emergency medical services (EMS) were immediately called and when they arrived 5 min later, she was still not breathing. The EMS personnel performed cardiopulmonary resuscitation consisting of chest compressions and rescue breathing, with the return of spontaneous circulation within 5 min. As she was not spontaneously breathing in the emergency department, the girl was endotracheally intubated. Upon arrival in the intensive care unit, the girl's vital signs were a temperature of 36°C, heart rate of 150 beats/min, respiratory rate of 20 breaths/min, and blood pressure of 100/60 mm Hg. Pulse oximetry was 95% on 50% oxygen via mechanical ventilation. On physical examination, there was no spontaneous movement. On painful stimulus, the girl exhibited extensor posturing, but did not open her eyes. Her pupils were 4 mm, equal, and sluggishly reactive. There were no external signs of trauma. Her abdomen was soft, nontender, non-distended, with no organomegaly. Her extremities were cool with capillary refill time of approximately 3 seconds.

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

A.   hyperventilation to achieve arterial CO_2 tension of 30 to 32 mm Hg

B.   maintenance of arterial hemoglobin oxygen saturation greater than 93%

C.   mannitol 0.25 g/kg intravenously every 6 hours

D.   placement of an external ventricular drain

E.   placement of a strain gauge intracranial pressure monitor

Question 64   **Preferred Response:B**

The child in this vignette has suffered a cardiac arrest from a submersion injury, also known as drowning or near-drowning. No therapeutic modalities have been proven to prevent secondary injury after cardiac arrest from drowning in children, therefore therapy should consist of supportive care and maintenance of blood pressure, oxygenation, and ventilation. Of the response choices listed, maintenance of arterial hemoglobin oxygen saturation greater than 93% is most appropriate.

Drowning is an important cause of childhood morbidity and mortality. According to the Centers for Disease Control and Prevention, more than 3,500 fatal, unintentional, non–boating related drowning deaths occur in the United States annually. Approximately one in five people who die from drowning are age 14 years or younger. Children ages 1 to 4 years have the highest drowning rates, most occurring in home swimming pools. In that age group, drowning is the second most common cause of death (the first being congenital anomalies). Risk factors for drowning include poor swimming ability, inadequate barriers around the pool, lack of supervision, and alcohol use (for older children and adults). Pediatric health supervision visits should include anticipatory guidance that emphasize supervision, swimming skills, avoidance of alcohol, and installation of appropriate barriers and alarms around home pools.

When water enters the airway, the diving reflex is stimulated, causing apnea, bradycardia, and laryngospasm. Although laryngospasm can prevent further aspiration of water, it impairs oxygenation and ventilation. Water or aspirated vomitus in the airspaces can cause abnormal surfactant production and hypoxia from ventilation-perfusion mismatch, leading to intrapulmonary shunting, poor lung compliance, and acute respiratory distress syndrome. Ventilator management should be targeted toward recruitment of lung volume and maintenance of oxygenation and ventilation.

Hypoxia, hypercarbia, acidosis, and the resultant decreased myocardial contractility can lead to asphyxial cardiopulmonary arrest. Asphyxial cardiac arrest can cause death or long-term encephalopathy, resulting from both hypoxic-ischemic and reperfusion injury. Supportive critical care ensuring adequate oxygenation, ventilation, hemodynamics, and nutrition is recommended. Extensive clinical trials investigating various treatments for cardiac arrest after drowning were undertaken in the 1970s and 1980s including therapeutic hypothermia, hyperventilation, osmotherapy, and goal-directed therapy to limit intracranial pressure. However, no evidence of therapeutic benefit was proven. Although hyperventilation can lower intracranial pressure, it is not recommended after cardiac arrest because it can exacerbate cerebral ischemia. There are ongoing multicenter clinical trials investigating therapeutic modalities after pediatric cardiac arrest.

PREP Pearls
- Drowning is most common in children ages 1 to 4 years, and usually occurs in home pools.
- Hypoxia after drowning can occur from laryngospasm, aspiration of water or vomitus, and ventilation-perfusion mismatch.

- Neurologic outcome after cardiac arrest is dependent on the timing, quality, and duration of cardiopulmonary resuscitation.

ABP Content Specifications(s)
- Plan the appropriate management of near-drowning
- Understand the epidemiology associated with drowning deaths
- Counsel parents regarding safety measures for a home pool
- Understand the prognostic factors associated with near-drowning

Suggested Readings
- Lieh-Lai MW, Sarnaik AA, Sarnaik AP. Drowning. In: Fuhrman BP, Zimmerman JJ, eds. Pediatric Critical Care. 4th ed. Philadelphia, PA: Saunders Elsevier: 2011:1480-1488.

Question 65

An 18-month-old girl with chronic malnutrition caused by intestinal malabsorption presents to the emergency department with fever, irritability, and left arm weakness. Vital signs show a temperature of 38.2°C, respiratory rate of 36 breaths/min, heart rate of 138 beats/min, blood pressure of 110/50 mm Hg, and a weight of 9 kg. On physical examination, she is irritable, has a facial droop, and left-sided weakness and tremor.

Laboratory data shows:
- White blood cells, 8,500/µL (8.5 x 10^9/L)
- Hemoglobin, 11 g/dL (110 g/L)
- Platelets, 307 x 10^3/µL (307 x 10^9/L)
- Differential, 54% segmented neutrophils, 37% lymphocytes, 8% monocytes, 1% eosinophils

Cerebrospinal fluid (CSF) results:
- White blood cells, 147/µL (4% segmented, 83% lymphocytes)
- Red blood cells, 10/µL
- Glucose, 25 mg/dL (1.4 mmol/L)
- Protein, 179 mg/dL

The purified protein derivative skin test result is 14 mm of swelling. You suspect tuberculous meningitis.

Of the following, the immune defect that BEST explains this child's increased susceptibility to this infection is

A.   decreased phagocytic cell function

B.   impaired antibody specific responses

C.   increased lymphocyte anergy

D.   increased natural killer cell levels

E.   upregulation of regulatory T cells

Question 65   **Preferred Response: C**

Lymphocytes are an integral component of controlling mycobacterial infections, especially by way of T helper 1 cell (Th1) type responses. Malnutrition, specifically protein calorie malnutrition as described for the girl in the vignette, can alter Th1 immune responses, leading to lymphocyte anergy and thus increased risk for progression from latent tuberculosis infection to tuberculosis disease. Lymphocyte anergy limits the use of purified protein derivatives (PPD) for screening for tuberculosis infection in this population.

Overall, both lack of adequate macro- and micronutrients can be associated with immune dysfunction and infections. Protein-calorie malnutrition has been associated with varied immune dysfunction, including atrophy of lymphoid tissue, decreased cell-mediated immunity, decreased immunoglobulin and complement levels, and diminished phagocytosis. Vitamin D and zinc deficiencies have also been linked to impaired immune responses.

While malnutrition can be associated with altered innate immunity, such as decreased phagocytic cell function, adaptive immunity is felt to be more critical in responding to intracellular pathogens, such as mycobacteria.

Malnutrition can also alter antibody-specific responses. However, antibodies are not critical in the control of tuberculosis infection.

Natural killer cells are a component of the innate immune system and are critical in immunity against viral infections. Deficiency of natural killer cells is associated with increased susceptibility to infection, especially Herpesviridae.

Immune tolerance is mediated by regulatory T cells. Decreased regulatory T-cell function can be associated with increased autoimmune and atopic disease. Upregulation of regulatory T cells is not associated with malnutrition.

PREP Pearls
- Malnutrition can impair both innate and adaptive immune responses, increasing susceptibility to infections.
- Malnourished children are at high risk for progression from latent tuberculosis infection to tuberculosis disease.
- Malnourished children can have increased lymphocyte anergy, which can limit the use of purified protein derivatives (PPD) for screening for tuberculosis infection.

ABP Content Specifications(s)
- Understand the association of infections with malnutrition

Suggested Readings
- Jaganath D, Mupere E. Childhood tuberculosis and malnutrition. J Infect Dis. 2012;206(12):1809-1815. DOI:http://dx.doi.org/10.1093/infdis/jis608.

Question 66

A 14-year-old adolescent presents to the emergency room with the chief complaint of having cola-colored urine for 1 day. He had a mild sore throat without fever 4 weeks ago. Vital signs show a respiratory rate of 26 breaths/min, heart rate of 110 beats/min, and blood pressure of 138/90 mm Hg. He has normal growth parameters. On physical examination, he has facial puffiness, but the remainder of the examination is unremarkable. 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 MOST appropriate anti-hypertensive agent for this patient is

A.   chlorothiazide

B.   enalapril

C.   furosemide

D.   hydralazine

E.   nifedipine

Question 66   **Preferred Response: C**

The patient described in the vignette has acute glomerulonephritis (GN), as characterized by the triad of cola-colored urine, hypertension, and azotemia on serum chemistry. Edema and hypertension in patients with acute GN is due to sodium and water retention with associated renal failure. Suppression of the renin-angiotensin-aldosterone system and enhanced release of atrial natriuretic peptide in patients with acute GN are indicative of volume overload in such patients.

Facial puffiness, respiratory distress, and high blood pressure, as present in the patient in the vignette, are indicative of volume overload. Such patients are managed with volume restriction (two-thirds maintenance) and intravenous furosemide for achieving diuresis and net negative fluid balance. Loop diuretics (furosemide, bumetanide, torsemide) inhibit sodium absorption via the Na-K-2Cl channels in the medullary and cortical aspects of the thick ascending limb, leading to excretion of up to 20% to 25% of tubular sodium. All diuretics inhibit sodium reabsorption at different sites in the nephron, thereby increasing sodium and water losses in urine. Intravenous furosemide (onset of action: oral, sub-lingual: 30-60 minutes; intramuscular: 30 minutes; intravenous: approximately 5 minutes) has a rapid onset of action, and in patients with pulmonary edema symptomatic improvement, in 15 to 20 minutes prior to the onset of the diuretic effect has been reported.

The thiazide diuretics (chlorothiazide) have a decreased natriuretic and diuretic effect compared to loop diuretics and inhibit the reabsorption of 3% to 5% of filtered sodium in the distal tubule. Thiazide diuretics inhibit sodium entry via the Na-Cl cotransporter in the distal nephron. Thiazides are not the preferred diuretics for the patient in the vignette, in view of the decreased diuresis in comparison to loop diuretics and slower onset of action (oral, within 2 hours; intravenous, 15 minutes). However, thiazide diuretics are preferred over loop diuretics for chronic antihypertensive therapy and have been commonly used for management of primary hypertension, especially in adults. The antihypertensive effects of thiazide diuretics are incompletely understood. The initial decrease in systemic blood pressure (BP) is associated with the reduction in plasma volume and cardiac output secondary to the diuretic effect. However, the volume depletion is blunted because of the activation of the renin-angiotensin system in response to hypovolemia. Long term decrease in BP with chronic thiazide therapy is associated with partial reversal of the initial decrease in plasma volume and vasodilation, leading to decrease in systemic vascular resistance. The factors responsible for the chronic vasodilation with prolonged thiazide treatment remain unclear.

Angiotensin-converting enzyme (ACE) inhibitors (such as enalapril, lisinopril), dihydropyridine calcium channel blockers (such as nifedipine, amlodipine, isradipine), and hydralazine are vasodilators leading to decrease in systemic blood pressure. Angiotensin-converting enzyme inhibitors inhibit the conversion of angiotensin I (AT1) to angiotensin II, thereby blunting the effects of the renin-angiotensin-aldosterone system on BP via angiotensin II. Angiotensin II activates AT1 receptors leading to:
  1.   arteriolar vasoconstriction
  2.   sympathetic nervous system activation

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3. aldosterone secretion leading to sodium and water reabsorption
4. stimulation of vascular and myocardial fibrosis

All calcium channel blockers inhibit the L-type calcium channels, leading to decreased influx of calcium into cells during depolarization. The dihydropyridine calcium channel blockers act on the vascular smooth muscles and are potent vasodilators with minimal (or no) negative effect on cardiac contractility. Angiotensin-converting enzyme inhibitors or long acting calcium channel blockers can be used for management of persistently elevated BP despite edema resolution, as in high BP associated with prednisone or other immunosuppressive therapy (tacrolimus, cyclosporine) or in chronic kidney disease. Intravenous hydralazine is most frequently used for hypertensive emergencies in the emergency department, intensive care unit, or inpatient hospital settings. In patients with severe acute renal failure and oliguria or anuria resistant to aggressive diuretic therapy, intravenous hydralazine or oral nifedipine may be used for hypertensive emergencies. Oral hydralazine is used as a third or fourth antihypertensive choice (after ACE, calcium channel blocker, diuretics, or β-blockers) in patients with resistant chronic hypertension requiring multiple antihypertensives for BP control.

PREP Pearls
- Edema and hypertension in patients with acute glomerulonephritis (GN) is caused by sodium and water retention with associated renal failure.
- Loop diuretics are the treatment of choice for initially managing volume overload and hypertension in patients with acute GN.
- Angiotensin-converting enzyme inhibitors, dihydropyridine calcium channel blockers, and hydralazine are vasodilators, leading to decrease in systemic blood pressure.

ABP Content Specifications(s)
- Understand the mechanism of action of the different classes of antihypertensive drugs

Suggested Readings
- Brady TM. Hypertension. Pediatr Rev. 2012;33(12):541-552. DOI: http://dx.doi.org/10.1542/pir.33-12-541.

Question 67

A 4-year-old girl presents to your office because of nose bleeds on and off for the last few months. The child was diagnosed with extrahepatic biliary atresia at 7 weeks of age and underwent a hepatoportoenterostomy (Kasai procedure). She is currently awaiting evaluation at a regional liver transplant center.

The child consumes a regular diet for age, including a daily multivitamin supplement. Her height and weight are at the 20th and 25th percentile, respectively, and a review of her growth chart demonstrates both previously at the 25th percentile. Physical examination shows a small, active child in no distress, icteric sclera, small amounts of crusted blood in the nares, a mildly protuberant abdomen, a firm liver edge palpated 2 cm below the right costal margin, and a spleen tip palpated 3 cm below the left costal margin. Blood vessels are prominently visible on her abdomen. Laboratory studies include:

- Electrolytes, normal
- Prothrombin time, 18.0 seconds (reference 11-14 seconds)
- Partial thromboplastin time, 45 seconds (reference 23-40 seconds)
- Platelet count $150 \times 10^3/\mu L$ ($150 \times 10^9/L$)
- Hemoglobin, 10 g/dL (100 g/L)
- Calcium, 7.8 mg/dL (1.95 mmol/L)
- Bilirubin (total), 6.5 mg/dL (111.2 µmol/L)
- Bilirubin (direct) 4.0 mg/dL (68.4 µmol/L)
- Aspartate aminotransferase, 90 U/L (reference range 20-60 U/L)
- Alanine aminotransferase, 110 U/L (reference range 20-50 U/L)
- Alkaline phosphatase, 650 U/L (reference range 150-350 U/L)

Based upon these findings, the MOST likely cause of this child's symptoms is a deficiency of

A.   vitamin A

B.   vitamin C

C.   vitamin D

D.   vitamin E

E.   vitamin K

Question 67   **Preferred Response: E**

The child in this vignette has chronic liver disease caused by biliary atresia previously treated with a hepatoportoenterostomy (Kasai procedure). She now has chronic cholestasis following her surgery. Her physical examination and laboratory studies are consistent with chronic liver disease, with coagulopathy caused by vitamin K deficiency.

Chronic liver disease is often associated with malnutrition. This places the child at increased risk for complications and poor outcomes following liver transplantation. The malnutrition is multifactorial, including inadequate intake caused by anorexia associated with chronic liver disease and malabsorption of nutrients.

Chronic liver disease can result in an impairment of the production and secretion of bile. Malabsorption of fat in cholestatic liver disease is caused by decreased bile salts in the small intestine. Bile is required for fat emulsification and micelle formation. Without adequate bile salts, the digestion and absorption of fats is inadequate, resulting in gastrointestinal losses of fats. Portal hypertension-associated vascular congestion can result in gastropathy and decreased nutrient absorption. Small bowel bacterial overgrowth in the Roux-en-Y loop created by the surgical procedure can be associated with bile salt deconjugation, resulting in additional fat malabsorption.

Fat malabsorption also results in fat soluble vitamin deficiencies, each with a classic presentation. Vitamin E deficiency is the most common, presenting with peripheral neuropathy and hemolysis in severe cases. Vitamin D deficiency results in osteomalacia and rickets. Vitamin K deficiency causes coagulopathy. Vitamin A deficiency is less common and is typically associated with night blindness.

It is important to monitor the nutritional status of children with chronic liver disease. A thorough history and physical examination to include a complete nutritional history should be completed at every clinic visit. Growth parameters should be plotted and anthropometric measurements serially monitored. To evaluate for fat malabsorption, a spot stool fat may identify elevated fecal fat. Additional laboratory tests to investigate deficiencies seen in chronic liver disease are shown in Item C67.

Item C67. *Additional Laboratory Tests to Investigate Deficiencies Seen in Chronic Liver Disease.*

Laboratory	*Importance*
Serum retinol	*Vitamin A deficiency*
Triene/tetraene ratio	*Essential fatty acid deficiency*
Serum 25-OH vitamin D level	*Vitamin D deficiency*
Plasma tocopherol and tocopherol/cholesterol ratio	*Vitamin E deficiency*
PT/INR	*Vitamin K deficiency*

Modified and reprinted with permission from S Nightingale, VL Ng. Optimizing Nutritional Management in Children with Chronic Liver Disease. Pediatr Clin North Am. 2009 Oct;56(5):1161-83

Fat malabsorption can be treated with supplementation of medium chain triglycerides, which have better absorption than other fat sources and can be used to increase absorption of fats for added calories. Vitamins A, D, E, and K are typically supplemented to avoid fat soluble vitamin deficiencies; however, levels should be monitored to avoid toxicity.

The child in the vignette has a coagulopathy caused by vitamin K deficiency. She is also at risk for vitamin A, D, and E deficiency; however, these are not contributing to her epistaxis. Vitamin C is not a fat soluble vitamin and although severe deficiency can be associated with bruising or bleeding, it is not associated with a prolongation of the prothrombin or partial thromboplastin time.

PREP Pearls
- Chronic liver disease is often associated with malnutrition.
- Fat malabsorption is multifactorial and includes inadequate bile, malabsorption caused by vascular congestion, and small bowel bacterial overgrowth, resulting in bile deconjugation.
- The fat soluble vitamins are A, D, E, and K. Each has a unique presentation when deficient.

ABP Content Specifications(s)
- Plan the appropriate management of fat malabsorption

Suggested Readings
- Nightingale S, Ng VL. Optimizing nutritional management in children with chronic liver disease. Pediatr Clin North Am.2009;56(5):1161-1183. DOI: http://dx.doi.org/10.1016/j.pcl.2009.06.005.

Question 68

You are called to the nursery to evaluate a baby with multiple anomalies and respiratory distress. The family is from a rural community, but was told to deliver at a tertiary care center because of suspected congenital defects. The nurse reports that the mother's prenatal ultrasonography revealed that the baby had an absent right kidney, a single umbilical artery, and polyhydramnios, as well as absence of a fluid-filled stomach, a small abdomen, and intrauterine growth retardation suggestive of a swallowing dysfunction caused by obstruction. Shortly after birth, the baby had copious oral secretions, cough, vomiting, and intermittent respiratory distress. Physical examination shows a cardiac murmur, imperforate anus, tachypnea, grunting, and mild subcostal retractions. While consulting a pediatric surgeon to manage the obstructive anomaly, an echocardiogram demonstrated a moderate ventricular septal defect.

Of the following studies, the BEST next step to establish the diagnostic cause for the multiple anomalies is

A.   brain magnetic resonance imaging

B.   chromosomal microarray

C.   dilated eye examination

D.   spine radiograph

E.   TORCH titers

Question 68   **Preferred Response: D**

In the newborn in this vignette, a prenatal high risk ultrasonography was utilized to take a comprehensive look at the structures of the developing fetus, including the brain, lips, face, nose, spine, heart, abdomen, kidneys, bladder, extremities, and umbilical cord. This sonogram, which is also known as a genetic sonogram, level II sonogram, or targeted sonogram, is typically performed at the 15- to 22-week mark in a pregnancy to diagnose birth defects. If major concerns arise, the newborn should be delivered at a major medical center that is equipped to handle babies with multiple congenital anomalies.

The combination of polyhydramnios, absence of a fluid-filled stomach, a small abdomen, and intrauterine growth retardation was suggestive of a swallowing dysfunction caused by obstruction. Thus, the necessity of a tertiary care center at delivery is imperative. After birth, the additional identification of a cardiac defect and imperforate anus, along with a classic tracheoesophageal fistula, is highly suggestive of VACTERL association. VACTERL association is the mnemonic for a complex non-random association of vertebral, anal, cardiac, tracheoesophageal (TE) fistula, renal, and limb defects that concomitantly occur in a patient, but are not currently known to be caused by a unifying genetic defect. In order to be diagnosed with VACTERL association, a patient must have at least 3 of the characteristic features from the list of congenital malformations described later in this critique. Vertebral defects are present in 60% to 80% of people with VACTERL association. Therefore, the best next appropriate test for diagnostic purposes in this situation would be a spine radiograph to look for dysplastic vertebrae, fused vertebrae, or missing or extra vertebrae. These patients are also at high risk for scoliosis. The percent risk in a patient with VACTERL association for each of the defects is:
- Vertebral defects- 60% to 80%
- Anal atresia possibly accompanied by genitourinary anomalies- 60% to 90%
- Cardiac defects- 40% to 80%
- TE fistula/esophageal atresia- 50% to 80%
- Renal anomalies- 50% to 80%
- Limb abnormalities- 40% to 50%

Most cases are sporadic with no family history of the condition. Its frequency is 1 in 10,000 to 40,000 newborns. There has been some association with maternal hyperglycemia.
Since the constellation of findings is highly suggestive of VACTERL association, a brain magnetic resonance image and dilated eye examination would not be the best next tests. Some VACTERL patients can have associated hydrocephalus, which should be clinically suggestive on physical examination and would best be assessed by a head ultrasound in the neonatal period. The first steps in evaluation of a patient should involve a thorough clinical workup to determine the extent and type of congenital malformations. Additionally, a detailed family and medical history is necessary. TORCH infections do not typically manifest with this combination of findings. A chromosomal microarray and karyotype would be indicated in this situation, but would not be a first line test in assessing the degree of systemic involvement that would be most useful in this newborn at initial assessment for clinical management.

PREP Pearls
- VACTERL is the mnemonic for a complex nonrandom association of vertebral, anal, cardiac, tracheoesophageal (TE) fistula, renal and limb defects that concomitantly occur in a patient, but are not currently known to be caused by a unifying genetic defect.
- In order to be diagnosed with VACTERL association, a patient must have at least 3 of the congenital malformations listed in the mnemonic.
- The first steps in evaluation of a suspected VACTERL patient should involve a thorough clinical workup to determine the number and type of congenital malformations, as well as a detailed family and medical history.
- A prenatal high risk sonography or a genetic sonogram is utilized to take a comprehensive look at the structures of the developing fetus, including the brain, lips, face, nose, spine, heart, abdomen, kidneys, bladder, extremities, and umbilical cord to identify birth defects during pregnancy, ideally performed between 15- to 22-weeks of gestation.

ABP Content Specifications(s)
- Understand which genetic disorders can be diagnosed prenatally
- Understand the role of fetal ultrasonography in prenatal diagnosis

Suggested Readings
- Shaw-Smith CJ. Oesophageal atresia, tracheo-oesophageal fistula, and the VACTERL association: review of genetics and epidemiology. J Med Genet. 2006;43(7):545-554. DOI: http://dx.doi.org/10.1136/jmg.2005.038158.

Question 69

You are asked to speak to a group of new mothers about the importance of breastfeeding. You want to highlight that there are only a few absolute medical contraindications to breastfeeding. In the case of the rare maternal infection that requires temporary discontinuation of breastfeeding, expressed breast milk from the mother may be offered until feeding at the breast can be resumed. Of the following, an example of this situation would include a mother who

A.   develops varicella 4 days before delivery

B.   is human T-lymphotropic virus-1 seropositive and lives in the United States

C.   is recently diagnosed with brucellosis and has not been treated

D.   is seropositive for cytomegalovirus and her infant is born at 40 weeks of gestation

E.   receives the live attenuated rubella virus vaccine immediately after delivery

Question 69   **Preferred Response: A**

A limited number of medical contraindications to breastfeeding exist, including several maternal infections. With active maternal varicella, temporary interruption of feeding at the breast is warranted. Expressed breast milk may be offered in the case of maternal varicella because there is no concern that the infection will be passed through the breast milk. Mothers who develop varicella from 5 days before through 2 days after delivery should be separated from their infants, and expressed milk may be used for feeding. Similarly, if a mother has untreated active infectious tuberculosis or has active herpes simplex lesions on her breast, expressed breast milk should be offered. Breastfeeding may be resumed once tuberculosis has been treated for a minimum of 2 weeks and the mother is no longer considered contagious, or once the herpetic lesions have resolved.

Mothers who have untreated brucellosis or those who live in the industrialized world and are positive for human immunodeficiency virus (HIV), human T-lymphotropic virus (HTLV) 1, or HTLV type 2 should not breastfeed or provide expressed breast milk to their infants. Breastfeeding is not recommended for HIV-infected women in the United States where safe and affordable nutritional alternatives are available, even when adequate treatment with antiretroviral medications is assured. In developing countries, where mortality is increased in non-breastfeeding infants because of malnutrition or infectious diseases, the benefits of breastfeeding may outweigh the risk of acquiring HIV or HTLV infection.

Although cytomegalovirus (CMV) may be shed intermittently in human milk, there is no contraindication to breastfeeding for a mother who is seropositive for CMV whose infant is born at term. There is no evidence of sequelae of CMV infection acquired through breast milk in preterm infants, except in the case of very-low-birthweight preterm infants who are at greater risk for developing symptomatic disease.

Mothers who receive the live attenuated rubella virus vaccine after delivery may continue to breastfeed. Although wild type strains from natural disease and vaccine strains of rubella virus have been isolated from human milk, neither situation has been associated with significant disease in infants.

PREP Pearls
- Feeding at the breast should be temporarily interrupted and expressed breast milk offered in cases of perinatal maternal infection with varicella, untreated active infectious tuberculosis, or active herpes simplex lesions on the breast.
- Mothers with untreated brucellosis should not breastfeed or feed their infant expressed breast milk.
- In the industrialized world, mothers with HIV, HTLV 1, or HTLV 2 should not breastfeed or feed their infant expressed breast milk.

ABP Content Specifications(s)
- Recognize when breast-feeding should be interrupted because of maternal infection

Suggested Readings

- AAP Section on Breastfeeding. Breastfeeding and the use of human milk. Pediatrics. 2012;129(3):e827-41. DOI:http://dx.doi.org/10.1542/peds.2011-3552.

Question 70

A 2-year-old girl presents to your office for a follow-up visit after being seen in the local emergency department. One week ago, the girl fell down a flight of 5 stairs onto a tiled floor and hit her forehead. Her mother reported that her daughter cried immediately and was taken to the local emergency room. She had an unremarkable neurological examination, was observed for several hours without incident, and was discharged without any further workup.

At this visit, of the following physical examination findings, the one that MOST suggests the need for further evaluation with a complete blood cell count is

A.   2 to 3 cm nonpalpable ecchymoses over her dorsal elbows bilaterally

B.   a 4 cm palpable, nontender hematoma over her glabella

C.   diffuse palatal purpura on the mucosa of her hard palate

D.   nonpalpable ecchymoses over her shins bilaterally

E.   purple-green discoloration under both of her eyes

Question 70   **Preferred Response: C**

Bruising is the external appearance of extravasated, subcutaneous blood as it degrades and is resorbed by the body. Bruising is often unavoidable following trauma. Assessing whether the extent of bruising noted following trauma is excessive or pathologic requires an understanding of the trauma event itself and the parts of the body that would likely have experienced impact during the trauma. The girl in this vignette fell down 5 stairs and landed at the bottom on her forehead. In this scenario, it is likely that the forehead, elbows, and knees all experienced impact, as she would have instinctively raised her arms and curled her legs as she fell. It is unlikely that she experienced impact to the interior of her mouth, nor would the shearing from a sudden stop be expected to cause intraoral bruising. The appearance of purpura over the hard palate could not be explained by the described trauma and would require further investigations.

Palatal purpura or petechiae are typically seen in the context of thrombocytopenia. Thus, any presentation of palatal petechiae or purpura requires a complete blood cell count to assess platelet number. If bruising occurs in excess to what would be expected in the context of a given trauma or if bruising occurs in the absence of trauma, an assessment of the entire coagulation system, including platelet number and function and the components of the coagulation pathways leading to fibrin formation should be undertaken. In the assessment of nonaccidental trauma, it is important to demonstrate that bruising or bleeding that has occurred is not a result of abnormal coagulation, either through platelet (number or function) or fibrin deficiency or dysfunction.

The glabella is the area between the eyebrows and above the nose. It has a very loose subcutaneous tissue and very little anchored tissue constricting the compartment. Any bleeding into that area can lead to a significant accumulation of blood, as there is little pressure to stop the bleeding. It would be expected that if there is a significant impact to the glabella, a large, palpable hematoma may develop. Thus, this would be an expected finding in this scenario.

Given the large potential space of the glabella with little to constrict the movement of blood, any accumulation of blood would follow gravity to the lowest sealed location and would be expected to seep beneath the eyes over time. As the blood degrades, it would go through the color changes consistent with the degradation of hemoglobin, so purple-green discoloration under the eyes would be expected several days after the injury.

As stated previously, it is likely that the girl in the vignette would have instinctively raised her arms and legs, so there would be expected impact to the elbows and legs, making bruising in those locations explainable by the trauma.

PREP Pearls
- An assessment of bruising requires an understanding of the nature of the trauma, and the parts of the body that would have been expected to experience impact.
- The glabella is the area between the eyebrows and above the nose and has very loose subcutaneous tissue unable to easily staunch bleeding
- Any trauma to the glabella would be expected to result in a sizable hematoma.

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ABP Content Specifications(s)
- Distinguish clinical findings associated with thrombocytopenia from those caused by normal bruising

Suggested Readings
- Anderst JD, Carpenter SL, Abshire TC. Evaluation for bleeding disorders in suspected child abuse. Pediatrics. 2013;131(4):e1314-e1322. DOI: http://dx.doi.org/10.1542/peds.2013-0195.

Question 71

A 4-year-old girl with bilateral congenital visual impairment comes to your office for a health supervision visit. Other than her vision, she has not had any other significant health problems. Her hearing screens and her growth are normal. Some of her motor milestones have been delayed, such as acquisition of a pincer grasp, head raising, crawling, and walking. She started early intervention services at 2 years of age. At 3 years of age, she received services through the local school district's developmental services program. These services have focused on physical therapy to assist her with fine and gross motor skills. She is legally blind and can only identify light, dark, and large shapes and symbols while using corrective lenses. Her parents report getting mixed advice about whether she should be taught to read braille and would like your opinion.

Of the following, the BEST advice to offer is

   A.   academic and employment success is closely linked to the ability to read braille

   B.   audio reading machines of printed language allow one to read just as quickly as with using braille

   C.   braille is a unique language of its own that would take her many years to learn

   D.   literacy is more readily achieved through the use of audio reading machines than learning braille

   E.   reading magnified print will be a preferred skill for her to learn over braille

Question 71   **Preferred Response: A**

Due to advancements in technology, a debate has emerged regarding the best treatment and educational approach for children with a significant visual impairment. While in the past, learning to use braille was widely viewed as the only way for someone who is functionally blind to acquire the ability to read, many now promote using assistive technology for those with residual visual ability. These technologies include screen reader software for personal computers, handheld portable video magnifiers, or optical character recognition voice output reading machines in mobile devices. Due to the expense of providing braille reading instruction, schools may now push for the use of assistive reading devices instead. Well-meaning parents may view learning braille as something that further sets their child apart from the rest of society. Despite these other considerations, learning to read braille continues to be an essential life skill for children with significant visual impairments.

Braille is not a language, but rather a tactile coding system for letters, numbers, and punctuation. Standard braille print takes nearly 5 times as many pages to convey the same information as regular print, so there is also a braille contraction system called "braille 2" that is used to reduce paper volume. The benefit of reading braille is that it is significantly faster to use than an audio- or video-magnifying assistive technology. This reflects the difference in the time it takes for listening to a book on tape versus reading a book.

Unemployment in the visually impaired community is a major problem. Unemployment rates for visually impaired people who do not use braille is 77% compared to 44% for those who can read braille. Those who can read braille are also 3 times as likely to achieve an advanced degree. Technology has made it easy to actively convert printed or electronic information into a refreshable tactile braille output that can be read more quickly than what is possible through visual- or audio-assistive devices. Therefore, learning to read braille is important to promote as a way to improve future employment and academic success.

While learning to read with a braille code takes at least as much time with skilled instruction as it takes for anyone else to learn and read visually, it would not be correct to call braille a language of its own. This is because the same braille system is used to encode many different written languages. Audio reading machines translate text into spoken words, which is the skill of listening rather than reading (reading = literacy). For those with sufficient visual ability, reading magnified print is a helpful option, but again, people can read much more text in a unit of time by reading braille.

PREP Pearls
• Braille is a coding system for letters and numbers rather than a language of its own.
• Visually impaired people who can read braille have more academic success and employment success than those without that skill.

ABP Content Specifications(s)
- Understand the major approaches to education for visually impaired children
- Recognize the value and limitations of language, occupational, and physical therapy
- Understand the provisions of current legislation for patients of various ages who have educational or physical disabilities

Suggested Readings
- Massof RW. The role of braille in the literacy of blind and visually impaired children. Arch Ophthalmol. 2009;127(11):1530-1531. DOI: http://dx.doi.org/10.1001/archophthalmol.2009.295.

Question 72

A 7-year-old boy is seen for the first time in your office. He has just come from overseas to live with an aunt and uncle. On the flight, he became very dizzy and almost passed out. He has a history of a heart murmur as an infant and was thought to need surgery, but was not able to have that done. The murmur resolved by 4 years of age. He is not able to participate in gym class in his new school because he gets too short of breath. He is very drowsy in the morning.

On physical examination, you note that he is cyanotic, but not in any acute distress. His weight is 19 kg (sixth percentile) and his height is 122 cm (50th percentile). His heart rate is 100 beats/min, his respiratory rate is 30 breaths/min, his blood pressure is 85/45 mm Hg, and his oxygen saturation is 78% on room air. His jugular veins are distended. His chest examination shows intercostal retractions. The breath sounds are clear. The cardiac examination is notable for a loud P2. There is a 3/6 systolic murmur at the right midclavicular line. The liver is 2 cm below the right costal margin. Screening laboratory test results are significant for a hemoglobin of 17 g/dL (170 g/L) and a hematocrit of 51%.

Of the following, the MOST likely cause of the patient's symptoms and physical findings is

A.   aortic stenosis with a gradient of 60 mm Hg

B.   pulmonic stenosis with a gradient of 60 mm Hg

C.   tricuspid atresia with a large atrial septal defect

D.   ventricular septal defect with a gradient of 90 mm Hg and left to right shunt

E.   ventricular septal defect with no gradient across the defect

Question 72   **Preferred Response: E**

The child in this vignette has symptoms related to a large ventricular septal defect (VSD) with no gradient across it because of pulmonary hypertension (PHTN), resulting from long-standing pulmonary overcirculation secondary to his uncorrected VSD. The lack of the usual holosystolic murmur is consistent with equal pressures in the left and right ventricles. Both aortic stenosis and pulmonic stenosis with gradients of 60 mm Hg would cause loud systolic murmurs in the outflow tract locations at the upper sternal borders. Tricuspid atresia would not necessarily cause a murmur.

This child in the vignette has pulmonary hypertension and the murmur in this case is a result of tricuspid regurgitation. The right ventricle (RV) has systemic pressure, so there will not be any gradient across the defect. The airplane flight made this child worse because airplanes are pressurized only to what would be encountered at 8,000 ft elevation. This causes worsening hypoxic vasoconstriction in a patient with PHTN. If there were a gradient of 90 mm Hg across his VSD, there would be a loud systolic murmur.

On physical examination, a newborn with a VSD may have a loud murmur or no murmur. A small muscular VSD will cause a short systolic high-pitched murmur at the left mid-sternal border as soon as the RV pressure drops below the left ventricular pressure. Since it is small, the speed of the blood across the defect will be rapid and the murmur will be loud. This murmur may be heard within the first week of life. These small defects usually close spontaneously.
A perimembranous VSD, which may be large or small (or be physiologically small if there is tricuspid valve tissue within it), may or may not cause a murmur early in life (Item C72). Most will cause a murmur by 3 to 4 weeks of age as the pulmonary vascular resistance drops. A very large VSD that is part of an atrioventricular canal will often not cause a murmur because the defect is so large that there is no difference in pressure between the right and left ventricles. Blood may be flowing from the left to right ventricle, but it is not across a pressure gradient, so the speed of the flow is not rapid or audible.

If there is large flow across a VSD for a prolonged period of time, and therefore a large increase in blood flow to the lungs, damage may occur to the pulmonary vasculature with development of PHTN. Once this occurs, the RV pressure may increase to greater than the systemic pressure and the flow across the ventricular defect will reverse and become right to left. This is the characteristic of Eisenmenger syndrome. Repair of an isolated perimembranous VSD by 12 to 24 months of age has been shown to protect against this phenomenon. Results of surgery have become so reliable at younger ages that there is not usually any need to wait until after a year of age if closure is indicated.
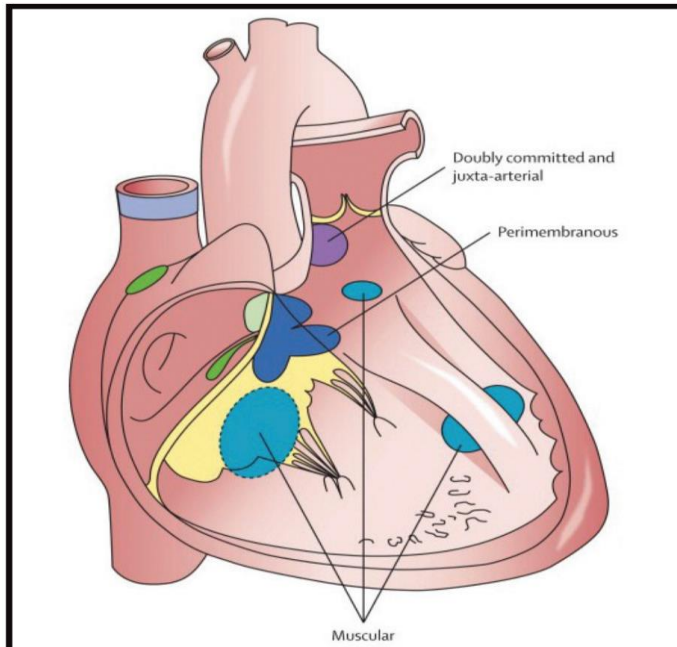
Reprinted with permission from Penny DJ, Vick GW 3rd. Ventricular Septal Defects. *Lancet.* 2011;377(9771):1103-1112

Item C72: There are several types of ventricular septal defect (VSD). The common perimembranous VSDs may be large in infancy, but close spontaneously if tricuspid valve tissue moves into the defect. The figure demonstrates the proximity of the tricuspid valve to this type of VSD.

PREP Pearls
- The clinical course of ventricular septal defects (VSD) can vary depending on the size of the defect and the size or degree of left to right shunt over time.
- If there is large blood flow across a VSD for prolonged period of time, and therefore a large increase in flow to the lungs, damage may occur to the pulmonary vasculature with development of pulmonary hypertension.
- If right ventricle pressure increases to greater than the systemic pressure, then the flow across a ventricular septal defect will reverse and become right to left, resulting in Eisenmenger syndrome.

ABP Content Specifications(s)
- Understand the natural history of ventricular septal defect
- Identify risks associated with an untreated large left-to-right shunt and pulmonary hypertension

Suggested Readings
- Engelfriet PM, Duffels MJ, Möller T, et al. Pulmonary arterial hypertension in adults born with a heart septal defect: the Euro Heart Survey on adult congenital heart disease. Heart. 2007;93(6):682–687.

Question 73

You are seeing a 7-year-old girl in your office for a health supervision visit. She has juvenile idiopathic arthritis that affects her right knee and ankle. The girl's parents state that she took steroids for 6 months when she was initially diagnosed. Her disease is now well controlled with methotrexate, folic acid, and etanercept. Her parents are concerned about the girl's bone health, specifically her risk of developing osteoporosis.

Of the following, the MOST accurate statement regarding this patient is

A.   her primary disease process is not a risk factor for osteoporosis

B.   her treatment regimen can lead to decreased bone mineral density

C.   she should avoid exercise, as her knee and ankle are affected with arthritis

D.   she should discontinue the methotrexate to avoid osteoporosis

E.   she should start magnesium supplementation

Question 73   **Preferred Response: B**

The girl in the vignette has been treated with long-term corticosteroids. This treatment regimen can lead to decreased bone mineral density, and is 1 of the major risk factors for osteoporosis in young patients.

Osteoporosis is defined as a decrease in bone mineral mass per volume of bone tissue without evidence of mineralization defects (ie osteomalacia or rickets). Primary forms of osteoporosis are rare in young patients. This can occur in idiopathic juvenile osteoporosis and genetic connective tissue diseases such as Ehlers-Danlos syndrome, Marfan syndrome, and osteogenesis imperfecta. More common in the pediatric population is secondary osteoporosis. Secondary osteoporosis may occur with neuromuscular disorders, inborn errors of metabolism, endocrine diseases, various chronic diseases, gastroenterologic and nutritional disorders, and certain medications or treatments (Item C73).

Item C73. Causes of Secondary Osteoporosis.

Neuromuscular Disorders and Inborn Errors of Metabolism	Chronic Diseases	Endocrine Disorders	Gastroenterologic and Nutritional Disorders	Iatrogenic
• Cerebral palsy • Muscular dystrophy • Prolonged immobilization • Spinal cord injury • Homocystinuria • Glycogen storage disease • Galactosemia • Gaucher disease	• HIV infection • Cystic fibrosis • Organ transplants • Thalassemia • Chronic inflammatory conditions • Inflammatory bowel disease • Systemic lupus erythematosis • Juvenile idiopathic arthritis • Rheumatoid arthritis • Chronic renal insufficiency • Nephropathy • Multiple myeloma • Lymphoma • Ectopic ACTH syndrome • Leukemia • Mastocytosis • Alcoholism	• Delayed puberty • Hypogonadism • Hyperprolactinemia • Growth hormone deficiency • Hyperthyroidism • Juvenile diabetes mellitus • Hypophosphatasia • Cushing syndrome • Hyperparathyroidism • Cushing disease	• Vitamin D deficiency • Calcium deficiency • Protein intolerance • Malnutrition • Chronic liver disease • Primary biliary cirrhosis • Anorexia nervosa • Malabsorption syndromes • Gastric bypass surgery	• Glucocorticoids • Methotrexate • Cyclosporine • Heparin • Anticonvulsants • Medroxyprogesterone injections • Diuretics • Radiotherapy • Chemotherapy

Modified and reprinted with permission from Bianchi, ML. How to manage osteoporosis in children. *Best Practice & Research Clinical Rheumatology.* 2005: 19 (6):991-1005

The best initial step in the management of osteoporosis is control of the underlying condition, and reduction of other risk factors for bone loss. Risk factors include low body mass, undernutrition, low levels of physical activity and/or weight bearing, hypogonadism, minimal sun exposure, and other lifestyle choices such as smoking or significant soft drink intake. Patients at risk for osteoporosis should be counseled regarding nutritional and lifestyle factors, including the importance of appropriate calcium, phosphorous, and vitamin D intake. Fruit and vegetable intake positively correlate with bone mineral density. Malnutrition and undernutrition are common in many chronic conditions and should be directly addressed. Physical activity including weight-bearing exercises can increase bone mineral density and is recommended for the patients who are able. When appropriate, the female athlete triad that includes malnutrition, intense exercise, and hypogonadism should be discussed. Calcium supplementation is

recommended for deficient individuals and children at high-risk for osteoporosis. The Institute of Medicine recommended daily dietary allowance is 700 mg for children age 1 to 3 years, 1,000 mg for age 4 to 8 years, and 1,300 mg for age 9 years and older. Children with malabsorption or on medications that induce osteoporosis may need higher doses. The recommended daily dose of vitamin D for children at low risk for osteoporosis is 400 IU.

Vitamin D supplementation is recommended for those at risk for osteoporosis, because low vitamin D levels affect bone quality. However, research on the effect of supplementation on bone mineral densities has not shown consistent results. Patients age 1 year and older who are deficient in vitamin D should be supplemented with 2000 IU daily or 50,000 IU weekly for at least 6 weeks. Children at risk for osteoporosis should be treated with 400 to 1,000 IU daily on a long-term basis. Patients with malabsorption may need higher doses. Either vitamin D2 or D3 can be used to treat vitamin deficiency, but some studies have demonstrated that the D3 form may be more beneficial in increasing bone mass and strength.

Juvenile idiopathic arthritis (JIA) is a risk factor for osteoporosis. Weight-bearing exercise can increase bone mineral density and is not contraindicated in JIA. Although methotrexate has been associated with decreased bone mineral density, the girl in the vignette should not discontinue the methotrexate to avoid osteoporosis, because uncontrolled arthritis can itself do significant bone and joint damage. The girl should not start magnesium supplementation, because this would have little effect on her bone density.

PREP Pearls
- Weight-bearing exercise is osteogenic.
- The recommended daily dose of vitamin D for children at low risk for osteoporosis is 400 IU.
- The recommended daily dietary allowance for calcium is 700 mg for children age 1 to 3 years, 1,000 mg for age 4 to 8 years, and 1,300 mg for children 9 years and older.

ABP Content Specifications(s)
- Counsel families and patients regarding the effects of diet, exercise, and smoking on the natural history of osteoporosis

Suggested Readings
- Ma N, Gordon CM. Pediatric osteoporosis: where are we now? J Pediatr. 2012;161(6):983-990. DOI:http://dx.doi.org/10.1016/j.jpeds.2012.07.057.

Question 74

A 9-year-old girl presents to your office for the evaluation of a 3-month history of activity-related bilateral heel pain. The pain worsens during and after soccer practice and games. She denies swelling, decreased ankle motion, or constitutional symptoms. Physical examination is remarkable for tenderness with medial and lateral compression (squeezing) of the heel.

Of the following, the BEST diagnostic modality for this patient is

A.   computed tomography of the heel

B.   an erythrocyte sedimentation rate

C.   history and physical examination

D.   magnetic resonance imaging of the heel

E.   radiographs of the heel

Question 74   **Preferred Response: C**

Young patients are particularly susceptible to injuries of the cartilaginous growth centers, the physes and apophyses. The term physis applies to a major growth plate that contributes to long bone growth. Apophyses are accessory growth centers or "minor" growth plates located at the point where tendons attach to bone.

The girl in the vignette has Sever disease, inflammation of the calcaneal apophysis. The diagnosis of Sever disease is based on history and physical examination findings. Patients often report insidious onset of activity-related heel pain. On physical examination, pain with simultaneous medial and lateral compression of the heel is the classic finding.
Sever disease typically affects patients between 8 and 13 years of age. The Achilles tendon, an extension of the gastrocnemius and soleus muscles, attaches to the calcaneus adjacent to the apophysis. With contraction of the calf muscles, the Achilles tendon puts tension on the apophysis, causing mechanical irritation. Direct force applied by high impact activities such as running and jumping also irritate the apophysis. Use of a soft heel cup in the shoe can blunt the force applied to the apophysis and appears to relieve symptoms. Ice and nonsteroidal anti-inflammatory drugs may help diminish pain. Activity modification or restriction is indicated for patients with significant pain or alteration in gait despite the use of symptomatic treatment. The symptoms of Sever disease typically abate when the apophysis closes, within 1 to 2 years of onset. This condition does not appear to have any long-term sequelae.

The diagnosis of Sever disease is made clinically. Radiography may be useful to rule out other conditions and may show fragmentation or sclerosis of the calcaneal epiphysis. Magnetic resonance imaging may be indicated if stress fracture or osteomyelitis of the calcaneus is suspected. Elevation of the erythrocyte sedimentation rate may be seen with infection or an inflammatory condition such as juvenile idiopathic arthritis. Computed tomography is not indicated for Sever disease.

PREP Pearls
- The diagnosis of Sever disease is based on history and physical examination findings; radiographic studies are generally not needed.
- Most children and adolescents with Sever disease may participate in sports and physical activities as pain allows.

ABP Content Specifications(s)
- Recognize the clinical findings and etiologic characteristics of Sever disease, and manage appropriately

Suggested Readings
- Sarwark JF, LaBella C. Pediatric Orthopaedics and Sports Injuries: A Quick Reference Guide. 2nd ed. Elk Grove Village, IL: American Academy of Pediatrics; 2014:744 pp.

Question 75

A 3-week-old, previously healthy full-term male newborn is brought to your office for evaluation of a rash on the bottom of his feet that began a few days ago. The mother has no other concerns. On physical examination, the newborn is afebrile with normal vital signs. The anterior fontanel is soft, open, and flat. There is minimal clear rhinorrhea. The liver and spleen are palpable 2 cm and 1 cm below the right and left costal margins, respectively. There are numerous copper-colored, circular lesions, each less than 0.5 cm in diameter on the plantar surfaces of both feet (Item Q75). The remainder of the physical examination is unremarkable.

Item Q75. Lesion for the newborn described in the vignette.

Of the following, the MOST likely cause of the newborn's rash is

A.   Candida albicans

B.   coxsackievirus

C.   cytomegalovirus

D.   Streptococcus agalactiae

E.   Treponema pallidum

Question 75   **Preferred Response: E**

The infant in the vignette has a copper-colored rash on the soles of the feet and hepatosplenomegaly consistent with congenital infection with Treponema pallidum (syphilis). Pregnant women typically are tested for syphilis with screening nontreponemal serologic tests (rapid plasma reagin [RPR]; VDRL) early in pregnancy, at the time of delivery, and occasionally, at the beginning of the third trimester. However, if this testing does not occur, an infant can present with symptomatic infection in the weeks after birth. Intrauterine infection with T pallidum can result in stillbirth, preterm birth, hydrops fetalis, or asymptomatic infection.

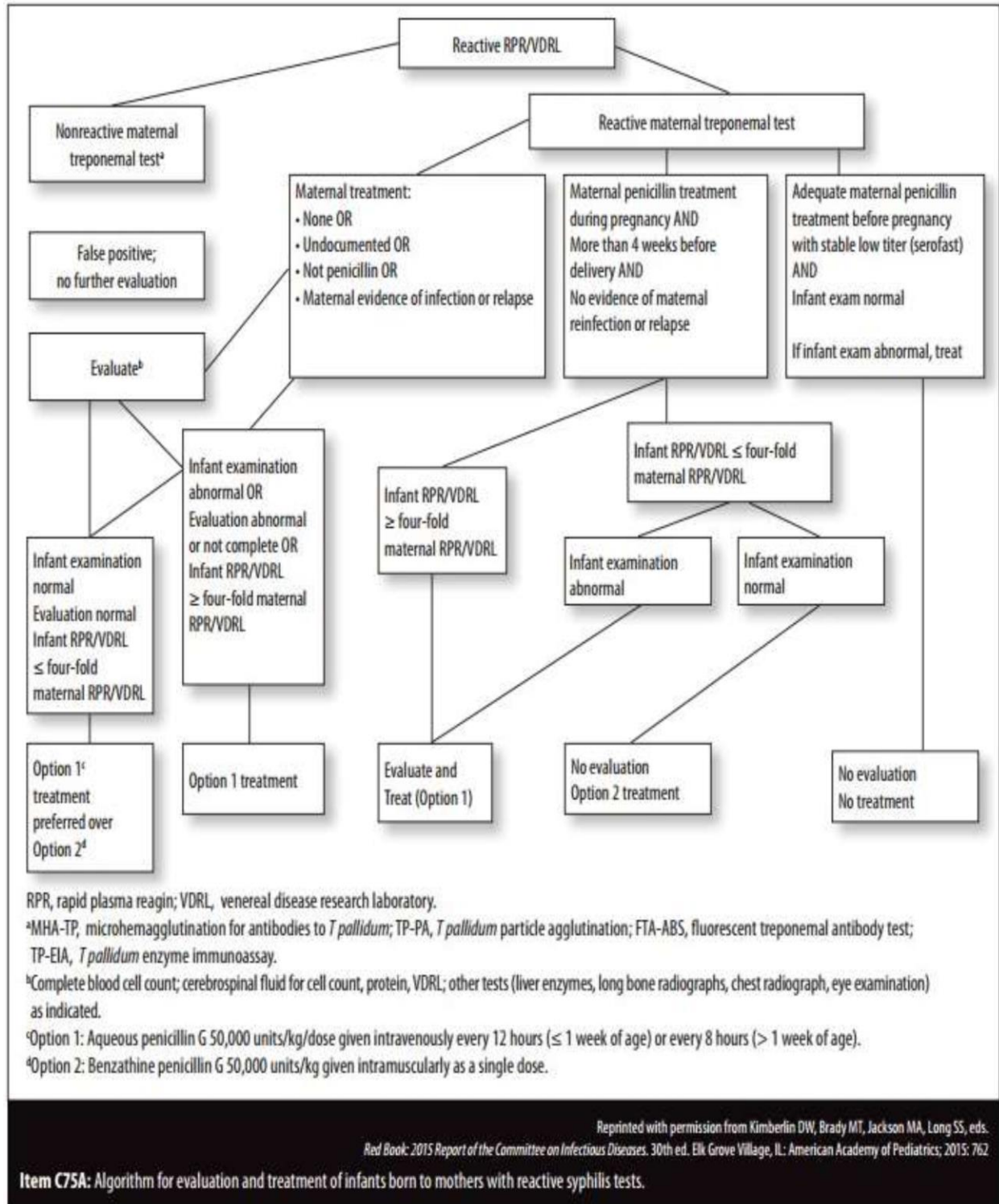Infants such as the child described in the vignette, can have:
- hepatosplenomegaly
- copious nasal secretions (snuffles)
- cutaneous lesions
- edema
- lymphadenopathy
- osteochondritis
- pneumonia
- pseudoparalysis
- hemolytic anemia
- thrombocytopenia

An untreated intrauterine infection can affect the:
- central nervous system (eighth cranial nerve deafness)
- eyes (interstitial keratitis)
- teeth (peg-shaped incisors [Hutchinson teeth], mulberry molars)
- bones (frontal bossing, saddle nose, tibial bowing)
- joints (swelling of knees [Clutton joints])
- skin (ulceration, desquamation, palpable lesions)

Acquired syphilis occurs in 3 stages: primary, secondary, and tertiary. The primary stage of infection is characterized by 1 or more painless ulcers (chancres) on the skin or mucous membranes at the initial site of inoculation that develop approximately 3 weeks after exposure. These lesions will spontaneously heal after a few weeks and can go undetected. The secondary stage of syphilis is characterized by lymphadenopathy, mucocutaneous lesions, and rash. The rash is generalized and typically involves the palms and soles. Patients may experience flulike symptoms such as fever, headache, sore throat, arthralgias, and malaise. This stage will spontaneously resolve in 1 to 4 months without treatment. The period following the secondary stage is called the latent period during which time patients are asymptomatic and seroreactive but may suffer recurrences of secondary stage symptoms. The tertiary stage of syphilis occurs 15 to 30 years after initial infection and can include neurosyphilis, cardiovascular symptoms, and gumma formation.

Penicillin is the treatment of choice for congenital or acquired infection. For patients with penicillin allergy and neurosyphilis, congenital syphilis, syphilis during pregnancy, or human immunodeficiency virus infection, desensitization is recommended. The recommended evaluation and treatment of neonates exposed to mothers infected with T pallidum is outlined in Item C75A. The recommended treatment for syphilis in patients older than 1 month of age is displayed in Item C75B.

Reactive RPR/VDRL

Nonreactive maternal treponemal test[a]

Reactive maternal treponemal test

False positive; no further evaluation

Maternal treatment:
• None OR
• Undocumented OR
• Not penicillin OR
• Maternal evidence of infection or relapse

Maternal penicillin treatment during pregnancy AND More than 4 weeks before delivery AND No evidence of maternal reinfection or relapse

Adequate maternal penicillin treatment before pregnancy with stable low titer (serofast) AND Infant exam normal

If infant exam abnormal, treat

Evaluate[b]

Infant examination abnormal OR Evaluation abnormal or not complete OR Infant RPR/VDRL ≥ four-fold maternal RPR/VDRL

Infant RPR/VDRL ≥ four-fold maternal RPR/VDRL

Infant RPR/VDRL ≤ four-fold maternal RPR/VDRL

Infant examination normal Evaluation normal Infant RPR/VDRL ≤ four-fold maternal RPR/VDRL

Infant examination abnormal

Infant examination normal

Option 1[c] treatment preferred over Option 2[d]

Option 1 treatment

Evaluate and Treat (Option 1)

No evaluation Option 2 treatment

No evaluation No treatment

RPR, rapid plasma reagin; VDRL, venereal disease research laboratory.

[a]MHA-TP, microhemagglutination for antibodies to *T pallidum*; TP-PA, *T pallidum* particle agglutination; FTA-ABS, fluorescent treponemal antibody test; TP-EIA, *T pallidum* enzyme immunoassay.

[b]Complete blood cell count; cerebrospinal fluid for cell count, protein, VDRL; other tests (liver enzymes, long bone radiographs, chest radiograph, eye examination) as indicated.

[c]Option 1: Aqueous penicillin G 50,000 units/kg/dose given intravenously every 12 hours (≤ 1 week of age) or every 8 hours (> 1 week of age).

[d]Option 2: Benzathine penicillin G 50,000 units/kg given intramuscularly as a single dose.

Item C75A: Algorithm for evaluation and treatment of infants born to mothers with reactive syphilis tests.

Status	Children	Adults
Congenital syphilis	Aqueous crystalline penicillin G, 200,000-300,000 units/kg/day, IV, administered as 50,000 units/kg, every 4-6 h for 10 days[a]	Penicillin G benzathine, 2.4 million units, IM, in a single dose **OR**
Primary, secondary, and early latent syphilis[b†]	Penicillin G benzathine,[c] 50,000 units/kg, IM, up to the adult dose of 2.4 million units in a single dose	*If allergic to penicillin and not pregnant,* doxycycline, 100 mg, orally, twice a day for 14 days **OR** Tetracycline, 500 mg, orally, 4 times/day for 14 days Penicillin G benzathine, 7.2 million units total, administered as 3 doses of 2.4 million units, IM, each at 1-wk intervals **OR**
Late latent syphilis[d] or latent syphilis of unknown duration	Penicillin G benzathine, 50,000 units/kg, IM, up to the adult dose of 2.4 million units, administered as 3 single doses at 1-wk intervals (total 150,000 units/kg, up to the adult dose of 7.2 million units)	*If allergic to penicillin and not pregnant,* doxycycline, 100 mg, orally, twice a day for 4 wk **OR** Tetracycline, 500 mg, orally, 4 times/day for 4 wk Penicillin G benzathine 7.2 million units total, administered as 3 doses of 2.4 million units, IM, at 1-wk intervals
Tertiary		*If allergic to penicillin and not pregnant,* same as for late latent syphilis Aqueous crystalline penicillin G, 18-24 million units per day, administered as 3-4 million units, IV, every 4 h for 10-14 days[f] **OR**
Neurosyphilis[e]	Aqueous crystalline penicillin G, 200,000-300,000 units/kg/day, IV, every 4-6 h for 10-14 days, in doses not to exceed the adult dose	Penicillin G procaine,[c] 2.4 million units, IM, once daily plus probenecid, 500 mg, orally, 4 times/day, both for 10-14 days[f]

IV, intravenously; IM, intramuscularly.

[a] If the patient has no clinical manifestations of disease, the cerebrospinal fluid (CSF) examination is normal, and the CSF Venereal Disease Research Laboratory (VDRL) test result is negative, some experts would treat with up to 3 weekly doses of penicillin G benzathine, 50,000 units/kg, IM. Some experts also suggest giving these patients a single dose of penicillin G benzathine, 50,000 units/kg, IM, after the 10-day course of intravenous aqueous penicillin.

[b] Early latent syphilis is defined as being acquired within the preceding year.

[c] Penicillin G benzathine and penicillin G procaine are approved for intramuscular administration only.

[d] Late latent syphilis is defined as syphilis beyond 1 year's duration.

[e] Patients who are allergic to penicillin should be desensitized.

[f] Some experts administer penicillin G benzathine, 2.4 million units, IM, once per week for up to 3 weeks after completion of these neurosyphilis treatment regimens.

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Item C75B: Recommended treatment for syphilis in people older than 1 month of age.

Mucocutaneous infection caused by Candida in the neonate can involve the oropharynx (eg, thrush) or vagina, digits and nails, and the intertriginous areas in the groin, axillae, and neck. It also can cause a mild but diffuse cutaneous infection (Item C75C). Cutaneous infection caused by Candida typically is described as an erythematous rash with satellite lesions; it typically would not be isolated to the feet as described for the patient in the vignette. Coxsackievirus can

cause a nonspecific viral exanthem or hand, foot, and mouth disease characterized by painful red blisters on the affected areas (Item C75D); it would not cause macular copper-colored lesions as described for the infant in the vignette. Similarly, acquired cytomegalovirus infection could present with a diffuse, nonspecific rash but not with localized findings. The classic "blueberry muffin" rash of congenital cytomegalovirus infection (Item C75E) would be present at or shortly after birth and not develop in the third week after birth as described for the patient in the vignette. Lastly, late-onset infection caused by group B Streptococcus (S agalactiae) may present as cellulitis but discrete copper-colored macular lesions are not characteristic findings.

Item C75C

Cutaneous infection caused by Candida. Courtesy of J. Brien. Reprinted with permission from Pickering LK, et al. Red Book® Online. Elk Grove Village, IL: American Academy of Pediatrics.

Item C75D

Viral exanthem caused by Coxsackievirus. Courtesy of L. Frenkel. Reprinted with permisson from Pickering LK, et al. Red Book® Online. Elk Grove Village, IL: American Academy of Pediatrics.

Item C75E

Characteristic rash seen in neonates with congenital cytomegalovirus infection. Reprinted with permission from Pickering LK, et al. Red Book® Online. Elk Grove Village, IL: American Academy of Pediatrics.

PREP Pearls
- Intrauterine infection with Treponema pallidum can result in stillbirth, preterm birth, hydrops fetalis, or asymptomatic infection.
- Manifestations of congenital syphilis include
  - hepatosplenomegaly
  - copious nasal secretions (snuffles)
  - cutaneous lesions
  - edema olymphadenopathy
  - steochondritis
  - pneumonia
  - pseudoparalysis
  - hemolytic anemia
  - thrombocytopenia
- Untreated intrauterine infection can affect the
  - central nervous system (eighth cranial nerve deafness)
  - eyes (interstitial keratitis)
  - teeth (peg-shaped incisors [Hutchinson teeth], mulberry molars)
  - bones (frontal bossing, saddle nose, tibial bowing)
  - joints (swelling of knees [Clutton joints])
  - skin (ulceration, desquamation, palpable lesions)
- The treatment of choice for all stages of syphilis is parenteral penicillin.

ABP Content Specifications(s)
- Recognize the clinical features associated with congenital and acquired Treponema pallidum infection
- Plan appropriate management for a patient with Treponema pallidum infection

Suggested Readings
- American Academy of Pediatrics. Syphilis. In: Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. Red Book: 2015Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:755-768.

Question 76

A 30-month-old girl is brought to the pediatric urgent care center for evaluation of fever, cough, and rhinorrhea. Her mother is concerned that her daughter has an ear infection because she has been crying frequently. The girl is a well-appearing, well-developed, active child in no distress. Her vital signs include a temperature of 38.4°C, heart rate of 115 beats/min, respiratory rate of 22 breaths/min, and pulse oximetry of 100% on room air. Physical examination is remarkable only for clear rhinorrhea from both nares and a swollen, tender right wrist with bruising over the dorsal aspect.

When you inquire about any history of trauma, the mother explains that the girl is extremely active and "very accident prone," but that she recalls no specific injury. You order a right wrist radiograph (Item Q76). A review of the urgent care records show that she was seen for a left humerus fracture at 9 months of age. The mother denies any family history of musculoskeletal disorders.

Item Q76. Right wrist radiograph for the girl i
the vignette. Courtesy of S. Sinal.

Of the following, the MOST likely explanation for her current radiologic finding is

A.   child abuse

B.   leukemia

C.   osteogenesis imperfecta

D.   osteomyelitis

E.   rickets

Question 76   **Preferred Response: A**

The girl in the vignette was brought to an urgent care center by her caregiver for evaluation of frequent crying and cold symptoms. She was found to have a metaphyseal corner fracture of her right radius on plain radiography (Item C76) performed to evaluate unexplained wrist swelling noted on physical examination. Child abuse is the most likely explanation for her radiologic finding. Further evaluation for child abuse and reporting the suspected abuse to child protective services is indicated.

Item C76. Metaphyseal corner fracture of the right radius on plain radiography performed to evaluate unexplained wrist swelling noted on physical examination of the girl in the vignette. Courtesy of S. Sinal

Metaphyseal corner fractures result almost exclusively from inflicted injury. These typically occur in young children (usually ≤3 years of age) and are often identified in the absence of a history of trauma. This injury typically results from traction or torsion forces that occur when a child's extremities are pulled or swung forcefully. For the girl in the vignette, in addition to the current injury, the history of a left humerus fracture during her first year after birth is a "red flag" for ongoing child abuse.

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Differentiating inflicted fractures from those associated with underlying musculoskeletal disorders can be a challenge for pediatric providers. Although many children with unexplained fractures are victims of inflicted injury, a number of underlying musculoskeletal diseases are associated with increased bone fragility; often these disorders can be subtle and difficult to diagnose. Pediatric health care providers should be aware of the clinical features and fracture patterns associated with child abuse. This distinction has major implications for the safety and well-being of children affected by abuse as well as those who have underlying musculoskeletal disorders.

No fracture, when considered in isolation, can definitively be said to result from an abusive versus a nonabusive cause. During the evaluation of individual bony abnormalities, the lesion site and type, age and developmental stage of the child, and associated reported history can help clinicians to determine the likelihood of inflicted injury. A number of studies, including a recent systematic review of the child abuse literature, have found that abusive fractures are detected most commonly in infants younger than 1 year and toddlers (1-3 years of age). When any infant or toddler presents with a fracture in the absence of a confirmed accidental cause, inflicted injury should be considered as a potential etiology. Specific fracture patterns that should further raise suspicion for child abuse include posterior rib fractures, metaphyseal corner fractures (also known as "bucket-handle" fractures, Item C76), humeral and femur fractures (especially those occurring in nonambulatory children), and complex skull fractures.

The presence of multiple fractures is also suggestive of abusive trauma, though this can certainly occur in accidental trauma and as a result of underlying musculoskeletal disorders. In addition, recurrent fractures at an unusual location for repeat injury, fractures in radiographic stages of healing that do not correspond to the reported clinical history, or multiple fractures in various stages of healing should raise suspicion of abuse. When suspicion of abuse is high, a radiographic skeletal survey should be obtained to evaluate the entire skeletal system for unrecognized injuries (new or old). Whenever possible, in cases of suspected nonaccidental injury, clinicians should consult with providers possessing specific expertise and experience in the diagnosis and management of child abuse. Physicians are mandated to report suspected child abuse to the appropriate child protective and/or law enforcement agencies.

Children with various forms of leukemia may present with fever or bone pain, or may even limp. However, the diagnosis of leukemia does not explain the radiographic finding of a metaphyseal corner fracture in the child in the vignette.

Osteogenesis imperfecta (OI) is a musculoskeletal disorder resulting in fractures that are most frequently mistaken for abusive fractures. OI is a heterogeneous group of disorders, usually caused by mutations in the gene involved in the production of type I collagen, a protein integral to the structural framework of bone. Although OI is an inherited disorder, the presentation of the disease can be quite variable even within the same family. Obtaining a thorough medical and family history, performing a physical examination, and interpreting the results of appropriate laboratory and radiographic studies can usually lead to a diagnosis of OI. Many children with OI

will have the classic diagnostic signs such as osteopenia, bony deformities, and "wormian bones" of the skull on radiography. Findings such as blue sclera, discolored teeth, limb deformities, and a triangular-shaped face may be apparent on physical examination in some children with OI, but these features may be subtle or absent. The girl in the vignette has no reported family history of OI or physical examination findings, other than the fracture, that would suggest the diagnosis of OI. This child's history of a humerus fracture at 9 months of age raises suspicion for child abuse, especially given that her newly identified fracture is a type that almost always arises from an abuse mechanism.

Osteomyelitis is an infection of the bone that typically presents with fever and localized musculoskeletal pain and tenderness. Although these symptoms are present in the girl in the vignette, metaphyseal corner fractures are not typically associated with osteomyelitis. Early radiographic findings suggestive of osteomyelitis include deep soft-tissue swelling with elevation of the muscle planes from adjacent bone, which can be seen 3 to 4 days after the onset of symptoms. Lytic bone changes and periosteal elevation may also be observed; however, these changes are generally not visible on plain radiography until at least 10 to 14 days after the onset of infection.

Rickets is an uncommon disorder that can occur in exclusively breastfed infants who are not receiving vitamin D supplementation, or in dark-skinned children who do not get adequate exposure to sunlight because of lifestyle or geographic location. Rickets can be diagnosed by characteristic changes on plain radiography, which include cupping and fraying of the costochondral junctions and epiphyses, demineralization, widened epiphyses, and cortical thinning. Children affected by rickets will have decreased serum concentrations of vitamin D metabolites, and serum alkaline phosphatase concentration is usually elevated. The radiographic finding in this child is not consistent with the diagnosis of rickets.

PREP Pearls
- Metaphyseal corner fractures result almost exclusively from inflicted injury. These fractures typically occur in young children, secondary to traction forces that occur when extremities are pulled or swung forcefully.
- In evaluating fractures, the site and type, age and developmental stage of the child, and associated reported history can help clinicians to determine the likelihood of inflicted injury.
- When child abuse is suspected, a radiographic skeletal survey should be obtained to evaluate the entire skeletal system for injuries. Providers with experience in the evaluation and management of child abuse should be consulted, and the suspected abuse must be promptly reported to appropriate child protection and law enforcement agencies.

ABP Content Specifications(s)
- Differentiate the findings associated with inflicted fractures from those of fractures related to other musculoskeletal disorders

- Differentiate the cutaneous findings associated with physical abuse from those of non-abusive skin conditions

Suggested Readings
- Jenny C. Evaluating infants and young children with multiple fractures. Pediatrics. 2006;118;1299. DOI:http://dx.doi.org/10.1542/peds.2006-1795.

Question 77

You are examining a small-for–gestational age newborn 8 hours after birth. She was delivered at 37 weeks of gestation because of worsening maternal pregnancy-induced hypertension. The pregnancy was unremarkable, except for increasing maternal blood pressure and proteinuria that were first noted 1 week before delivery.

Of the following, the MOST likely finding on physical examination of this newborn is

A.   clitoral prominence

B.   hepatosplenomegaly

C.   increased breast tissue

D.   jaundice

E.   microcephaly

Question 77   **Preferred Response: A**

The newborn in the vignette is small for gestational age (SGA) because of maternal pregnancy-induced hypertension (PIH) and is likely to have clitoral prominence on physical examination. Maternal PIH is associated with fetuses that have intrauterine growth restriction (IUGR). Histologic evaluation of placentas from mothers with PIH reveals hypertrophic decidual vasculopathy. This causes abnormal blood flow to the placenta and fetus, leading to decreased fetal nutrient supply. Affected fetuses are considered SGA if their birthweight falls below the 10th percentile for gestational age. Other conditions that are associated with SGA infants include certain chromosomal abnormalities and congenital infections, as well as constitutional small size. Newborns who are SGA because of maternal PIH appear thin and lanky. They have decreased muscle mass and subcutaneous tissue because of the diminished in utero nutrient supply. Decreased vernix leads to peeling, dry skin and increased plantar creasing on the feet. Additional findings include diminished breast tissue, underdeveloped ear cartilage, and clitoral prominence from decreased perineal adipose tissue covering the labia. The head circumference may be disproportionately large in some SGA newborns, leading to the term asymmetric SGA. In these cases, the nutrient supply is believed to decline later in pregnancy, allowing relatively normal length and head growth.

Small-for-gestational age newborns are at risk for hypothermia, hypoglycemia, and polycythemia. The combination of decreased subcutaneous fat and fewer nutrient substrates leads to difficulty with thermoregulation. Close temperature monitoring and the use of a thermal neutral environment, such as an isolette, are recommended. Hypoglycemia results from decreased glycogen stores in the skeletal muscle and liver. Glucose monitoring and management guidelines for SGA newborns are outlined in the American Academy of Pediatrics glucose homeostasis clinical report. Fetal hypoxia arising from abnormal placental blood flow is hypothesized to increase fetal erythropoietin production, leading to polycythemia. The risk of these complications increases with the severity of the growth restriction.

Hepatosplenomegaly may be found in SGA newborns with congenital infections, but the small size of the infant in the vignette is attributed to PIH rather than infection. SGA newborns typically have decreased breast tissue because of their undernourished state. Jaundice may develop in these infants due to polycythemia, but it is not expected to be present 8 hours after birth. The head circumference of a newborn who is SGA due to PIH is typically in the same or higher percentile as the weight, not lower as would be seen with microcephaly.

PREP Pearls
- Small-for–gestational newborns have decreased muscle mass and subcutaneous tissue, dry peeling skin, increased plantar creasing, diminished breast tissue, underdeveloped ear cartilage, and clitoral prominence.
- Small-for–gestational age newborns are at risk for hypothermia, hypoglycemia, and polycythemia.

ABP Content Specifications(s)
- Understand the physiologic and physical abnormalities that may be present in a small-for-gestational-age infant

Suggested Readings
- AAP Committee on Fetus and Newborn. Postnatal glucose homeostasis in late-preterm and term infants. Pediatrics. 2011;127:575-579. DOI: http://dx.doi.org/10.1542/peds.2010-3851.

Question 78

The father of a 3-year-old boy in your practice calls on Friday evening. The boy has previously been healthy, growing and developing well. The father reports that the boy has had nonbloody, nonbilious vomiting intermittently for 3 days. He is still drinking fluids, urinating his usual amount, and has no diarrhea, fever, or rash. He is not lethargic. There are no sick contacts. You advise supportive care. On Monday, the mother brings the boy in to your office for evaluation. He continues to vomit, is now drinking less, and continues to urinate. His mother reports the boy's eyes are sometimes "crossed," which is new for him. On physical examination, he has a temperature of 36.5°C, blood pressure of 96/50 mm Hg, heart rate of 91 beats/min, and respiratory rate of 36 breaths/min. He is tired appearing, but arouses with the examination. His abdominal examination is unremarkable. There is no rash. On neurologic examination, the boy is able to visually fixate on your face with both eyes, but when tracking to the left, his left eye cannot fully abduct.

Of the following, the BEST recommendation for this child is referral to the

A.   emergency department

B.   neurology clinic

C.   neurosurgery clinic

D.   ophthalmology clinic

E.   urgent care

Question 78   **Preferred Response: A**

The boy in the vignette has signs of increased intracranial pressure: persistent, unexplained emesis and cranial nerve VI palsy. This presentation is an emergency, and emergency medical services should transport him to the emergency department immediately. Other signs of increased intracranial pressure include papillitis (which can be difficult to assess in a toddler), head tilt, limb weakness, or gait abnormalities. These signs are not necessarily all present in the early stages. Increased intracranial pressure has many causes. In this case, computed tomography showed obstructive hydrocephalus and an intraventricular mass that was found to be a choroid plexus papilloma (Item C78).
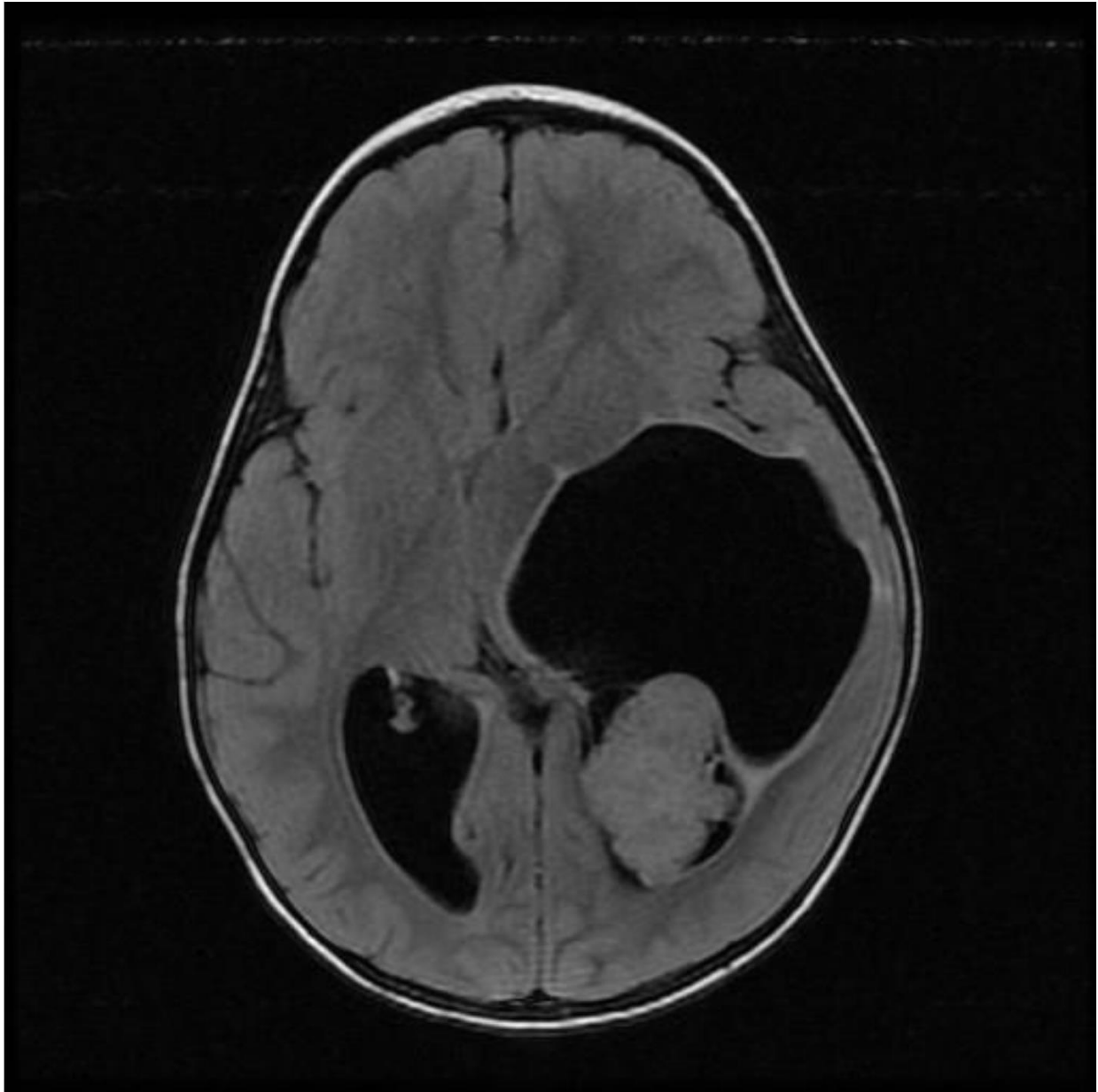
Item C78. Magnetic resonance imaging depicting choroid plexus papilloma and hydrocephalus. Courtesy of D. Morita.

When acute increased intracranial pressure is suspected, even when vital signs are normal, immediate emergency department evaluation is the most appropriate referral. This child is at risk for rapid decompensation. The boy in the vignette will require urgent imaging and neurosurgical consultation and intervention. A neurologist or ophthalmologist may be consulted in the hospital, but referral to an outpatient clinic or urgent care is not appropriate in this situation.

PREP Pearls
- A child with new-onset signs of increased intracranial pressure must be immediately transported to the emergency department.
- New ocular malalignment, (crossed eyes), should prompt evaluation for other signs of increased intracranial pressure.

ABP Content Specifications(s)
- Plan the appropriate diagnostic evaluation of increased intracranial pressure, and manage appropriately

Suggested Readings
- Ansell P, Johnston T, Simpson J, Crouch S, Roman E, Picton S. Brain tumor signs and symptoms: analysis of primary health care records from the UKCCS. Pediatrics. 2010;125:112-119. DOI: http://dx.doi.org/10.1542/peds.2009-0254.
- Crawford J. Childhood brain tumors. Pediatr Rev. 2013;34(2): 63-78. DOI: http://dx.doi.org/10.1542/pir.34-2-63.

Question 79

A 10-year-old boy has had poor growth over the past few years (Item Q79). He also complains of fatigue, muscle weakness, and worsening acne. On physical examination, the boy has a prominent fat pad on the back of his neck, central obesity, and stretch marks (striae) on his abdomen.

Of the following, the test MOST likely to determine the underlying cause of this patient's poor growth is a(n)

A.   24-hour urine-free cortisol

B.   AM growth hormone level

C.   fasting AM prolactin level

D.   random insulin like growth factor 1

E.   thyroid-stimulating hormone and free thyroxine levels

Question 79   **Preferred Response: A**

When linear growth slows down or arrests, particularly as weight is maintained or increases, an underlying endocrine cause is likely. The child in this vignette is rapidly gaining weight while his linear growth is decelerating, and has physical examination signs consistent with Cushing syndrome. Other conditions that can follow this pattern include growth hormone (GH) deficiency, hypothyroidism, and genetic disorders, such as Turner syndrome. Children with exogenous obesity will have normal linear growth, and should not undergo evaluation for Cushing syndrome.

Evaluation for Cushing syndrome can be performed by measurement of 24-hour urine free cortisol, overnight dexamethasone suppression, late night salivary cortisol, and in some cases, assessment of diurnal variation in cortisol. Thus, of the choices offered, a 24-hour urine-free cortisol would be appropriate for this patient.

An AM GH level is not recommended for evaluation of growth, even during evaluation for GH deficiency, because GH levels peak in deep sleep and are often very low during the day. This is, in part, the rationale for growth hormone stimulation testing, and why insulin like growth factor 1 (IGF-1) or insulin like growth factor binding protein 3 (IGF-BP3) levels are measured instead. Obtaining a prolactin level can help rule out prolactinoma as the cause of poor growth. Thyroid-stimulating hormone and free thyroxine levels are used when evaluating for hypothyroidism.

Although a child with GH deficiency, hypothyroidism, or a pituitary tumor such as a prolactinoma could have a growth pattern similar to the child in this vignette, the physical examination findings described in this child are most compatible with Cushing syndrome. A 24-hour urine-free cortisol would be the best test to determine the cause of his poor growth.

PREP Pearls
- An endocrine cause of poor growth is likely when linear growth slows down or arrests but weight gain is either normal or increasing.
- Cushing syndrome in children is characterized by linear growth failure, weight gain, pubertal changes from excess androgen production, as well as signs and symptoms similar to adults (easy bruising, facial plethora, myopathy, striae).

ABP Content Specifications(s)
- Identify the most common causes of short stature
- Plan the evaluation of a child with short stature or whose height percentiles have decreased

Suggested Readings
- Nieman LK, Biller BM, Findling JW, et al. The diagnosis of Cushing's syndrome: an Endocrine Society Clinical Practice Guideline. J Clin Endocrinol Metab. 2008;93:1526-1540. DOI: http://dx.doi.org/10.1210/jc.2008-0125.
- Weintraub B. Growth. Pediatr Rev. 2011;32:404-406. DOI: http://dx.doi.org/10.1542/pir.32-9-404.

Question 80

A 14-year-old adolescent is brought to the emergency department (ED) with 2 months of intermittent wet cough. More recently, she has had 2 to 3 episodes of blood-tinged sputum. She has experienced 1 month of increasing fatigue and pallor. She has been increasingly intolerant of physical activity and now experiences shortness of breath when climbing 1 flight of stairs. Today while at school, she experienced a fainting episode and was brought to the ED via emergency medical services. On review of systems, her mother reports a several month history of a rash on the nose and cheeks, as well as decreased range of motion with swelling in her daughter's fingers and wrists. Most recently, her urine has looked like "tea."

On physical examination, the patient has regained consciousness, but is ill appearing with moderate respiratory distress. Her heart rate is 152 beats/min, and her respiratory rate is 40 breaths/min and labored. Her blood pressure is 90/60 mm Hg. Oxygen saturation is 92% in room air. Oropharynx and nares are clear without blood. Her mucous membranes are pale but moist. Auscultation of the lungs reveals diffuse polyphonic wheezing in upper and mid lung fields with decreased breath sounds at both bases. Scattered fine crackles are also audible throughout. The cardiac and abdominal examinations are unremarkable, with the exception of tachycardia. Capillary refill is brisk. Her extremities are warm and well perfused, and there is no digital clubbing. Her hemoglobin is 5.9 g/dL (59 g/L).

You obtain a chest radiograph (Item Q80).

Item Q80. Chest radiograph for the girl described in the vignette. Courtesy of M Nevin.

Of the following, the MOST likely etiology of this patient's hemoptysis is

A.   chronic airway foreign body

B.   cystic fibrosis

C.   pulmonary hypertension

D.   tuberculosis

E.   vasculitis

Question 80   **Preferred Response: E**

The adolescent in this vignette presents with cough and hemoptysis, but also demonstrates a variety of signs and symptoms that are suggestive of multisystem disease. In this patient with rash, arthritis, fatigue, pallor, hematuria (suggested by the tea-colored urine), and syncope, evidence of a pulmonary renal syndrome and autoimmune or vasculitic condition should be sought.

Bleeding from the lungs or airway may occur in a focal or in a diffuse pattern. The most common etiology for focal pulmonary hemorrhage is chronic infection or inflammation; classic examples are tuberculosis and the endobronchial infections that cause bronchiectasis in patients with cystic fibrosis.

The etiologies of diffuse alveolar hemorrhage (DAH) syndromes are multiple and varied, and may be categorized by whether there is pathologic evidence of pulmonary capillaritis. The finding of pulmonary capillaritis denotes inflammation in the pulmonary capillary bed and is a poor prognostic finding in individuals with DAH; those with systemic lupus erythematosus, Goodpasture syndrome, granulomatosis with polyangiitis (previously Wegener granulomatosis), and Henoch-Schönlein purpura, among others, may be affected.

In the absence of capillaritis, the origins of DAH are divided into cardiac and noncardiac causes. Cardiovascular associations include arteriovenous malformations and pulmonary hypertension. Noncardiac etiologies include celiac disease, coagulation disorders, and acute idiopathic pulmonary hemorrhage of infancy. When an exhaustive search for an etiology of diffuse pulmonary hemorrhage is unrevealing, patients may be designated as having idiopathic pulmonary hemosiderosis. Some of these patients will later be diagnosed with an associated condition.

In patients with DAH, the rate of bleeding is typically slow and insidious. Patients may never expectorate blood and instead are likely to present with fatigue, pallor, tachycardia, or exercise intolerance. Initial symptoms may suggest the underlying condition and the diagnosis of DAH may be a secondary one. The classic laboratory finding is a microcytic anemia. Radiographs are often nonspecific, but may demonstrate bilateral alveolar opacities with lower lobe predominance as in the patient in this critique. The diagnosis of DAH is confirmed by the presence of hemosiderin laden macrophages on sputum analysis. Therapy is dependent on underlying condition, but may include systemic steroids and immunosuppressive agents.
The patient in this vignette is not in the age group classically associated with foreign body aspiration. In addition, there is no asymmetry or air trapping on chest radiograph to suggest an inhaled foreign body. Similarly, the radiograph does not reveal nodularity or lymphadenopathy suggestive of tuberculosis. Furthermore, the bleeding in both of these conditions would be expected to be more brisk with notable bright red hemoptysis.

An adolescent may be diagnosed with cystic fibrosis if they have atypical or mild disease. Bleeding from the airways in patients with cystic fibrosis, however, occurs from bronchiectasis, which is a late manifestation of disease. An individual with pulmonary hypertension may present

with syncope, exertional dyspnea, and fatigue. However, the joint, skin, and urinary symptoms found in the patient in this vignette would not be expected.

PREP Pearls
- he most common causes of focal pulmonary hemorrhage are infection and inflammation.
- Diffuse alveolar hemorrhage syndromes may be classified by the presence or absence of pulmonary capillaritis.
- Bleeding in diffuse alveolar hemorrhage is likely to be slow and insidious; patients may present with symptoms of anemia rather than with hemoptysis.

ABP Content Specifications(s)
- Plan the appropriate clinical and diagnostic evaluation of hemoptysis
- Plan the appropriate management of hemoptysis in patients of various ages

Suggested Readings
- Godfrey S. Pulmonary hemorrhage/hemoptysis in children. Pediatr Pulmonol. 2004;37(6):476-484. DOI:http://dx.doi.org/10.1002/ppul.20020.

Question 81

You are evaluating a 2-month-old infant who was born at 28 weeks of gestation and went home from the neonatal intensive care unit 3 days ago. The baby's birth weight was 990 g and her current weight is 1,900 g. The formula-fed infant was discharged on a diet of 22 cal/oz premature formula. The parents have had difficulty finding the formula and ask if the baby could be fed a different type of milk while still maintaining the benefits of premature formula.

Of the following, the MOST accurate statement about feeding options for this infant is

   A.   banked human milk supplies sufficient minerals and calories to support growth and
          prevent metabolic bone disease in low birth weight infants

   B.   compared to formula made for term infants, premature formulas contain higher levels
          of calcium and phosphorus

   C.   formula made for term infants would provide iron in excess of this infant's needs

   D.   the primary cause of decreased bone mineral concentration in premature infants is
          low vitamin D intake

   E.   this infant should receive supplementation with 200 IU/day of vitamin D

Question 81   **Preferred Response: B**

The nutritional needs of premature infants, particularly those less than 2,000 g in weight, differ from term infants' requirements in several areas, including protein and mineral needs. Most mineral accumulation occurs during the third trimester, therefore premature newborns are at risk for developing deficiencies of calcium, phosphorus, iron, copper, and zinc; other mineral deficits (eg, iodine) are possible, but there have been few if any clinical reports of these deficiencies. Calcium and phosphorus are of particular concern because the primary cause of decreased bone mineral density (BMD) in premature newborns is low levels of these minerals as a result of decreased intake or absorption. The current recommendations are that premature newborns consume 150 to 200 mg/kg of calcium and 60 mg/kg to 75 mg/kg of phosphorus each day. Unfortified human milk, even preterm breast milk, and formulas produced for term infants do not provide sufficient calcium and phosphorus to meet these needs. Therefore, preterm babies less than 2,000 g in weight should receive human milk supplemented with fortifier or preterm formula in order to achieve sufficient intake of calcium and phosphorus (Item C81). Banked human milk is primarily term milk and does not provide enough calcium and phosphorus to prevent metabolic bone disease.

Item C81. Approximate Mineral Concentrations in Breast Milk and Formula.

Feeding type	Calcium (mg/L)	Phosphorus (mg/L)	Iron (mg/L)
Term human milk	280	140	0.3
Preterm human milk	250	130	1.2
Preterm human milk and fortifier	1100-1400	640-800	4.6-15
Premature formula	1100-1600*	500-800*	12-18**
Term infant formula	500-600	280-400	12

* Dependent on caloric density of the formula

** must be verified by brand as content may vary markedly

Courtesy of K. Bowen

While BMD is primarily dependent on intake and absorption of calcium and phosphorus, vitamin D intake also plays a role. There are currently no studies of the clinical impact of 25-hydroxyvitamin D concentrations in preterm newborns, so deficiency and sufficiency is based on extrapolation from adult and pediatric populations. Current recommendations are that preterm newborns with a birth weight of more than 1,500 g receive 400 IU of vitamin D daily just as term infants do. For very low birth weight newborns, 200 to 400 IU daily is recommended, with an increase to 400 IU/day once they have achieved 1,500 g in weight. The maximum dose of vitamin D for term infants is 1,000 IU/day and preterm newborns should not exceed this dose.

Iron stores are also laid down during the third trimester, and preterm and low birth weight infants are at risk for iron deficiency that can contribute to neurodevelopmental issues later in the child's life. Repeated blood draws or transfusions also impact the infant's iron homeostasis. Current recommendations are that low birth weight infants receive 2 to 3 mg/kg per day of iron beginning at 1 to 2 months of age. Although the iron concentrations in formula or human milk plus fortifier are quite variable, this route can supply at least part of this iron supplementation. For the infant in the vignette, she may be able to consume enough iron from term or preterm formula.

PREP Pearls
- Most mineral accumulation occurs during the third trimester, therefore premature newborns are at risk for developing deficiencies of calcium, phosphorus, iron, copper, and zinc.
- The primary cause of decreased bone mineral density in premature infants is low levels of calcium and phosphorus caused by either low intake or low absorption.
- Fortified human breast milk is the preferred energy source for preterm infants and supplies adequate minerals for their growth needs. Formula made for preterm infants also supplies adequate calcium and phosphorus.
- Vitamin D supplementation for healthy preterm infants is 200 to 400 IU/day for weight less than 1,500 g and 400 IU/day for weight greater than 1,500 g.

ABP Content Specifications(s)
- Understand the dietary mineral requirements of patients of various ages, including those born prematurely, and the circumstances in which those requirements may change

Suggested Readings
- Bhatia J, Griffin I, Anderson D, Kler N, Domellöf M. Selected Macro/micronutrient needs of the routine preterm infant. J Pediatr. 2013;162(3 Suppl):S48-S55. DOI: http://dx.doi.org/10.1016/j.jpeds.2012.11.053.

Question 82

A 13-year-old adolescent is admitted to the hospital for a tonsillectomy. As part of his pre-operative evaluation, a metabolic panel was ordered that revealed an alkaline phosphatase of 325 U/L (upper limit of normal = 116 U/L). You are asked to evaluate him for this abnormal laboratory result. His past medical history is significant only for tonsillar hypertrophy and related obstructive sleep apnea. His review of systems is negative except for snoring. He does not take any medications. Aside from having grade 3 tonsils, his physical examination is unremarkable. His sexual maturity rating is 3.

Of the following, the MOST likely reason for his abnormal laboratory result is

A.   bone tumor

B.   gallbladder disease

C.   laboratory error

D.   pubertal growth

E.   vitamin C deficiency

Question 82   **Preferred Response: D**

Adolescence is a period of physical, cognitive, and psychosocial development. Linear growth velocity during puberty increases to about 9.5 cm per year in boys and 8.3 cm per year in girls. This bone growth is associated with an elevated serum alkaline phosphatase (ALP).
The development of secondary sexual characteristics is triggered by the increased secretion of pituitary gonadotropins. Sexual maturity rating (SMR) can be used to depict genital development.

Item C82A and Item C82B show the SMR for boys and girls.

Item C82A. Stages of Breast and Pubic Hair Development.
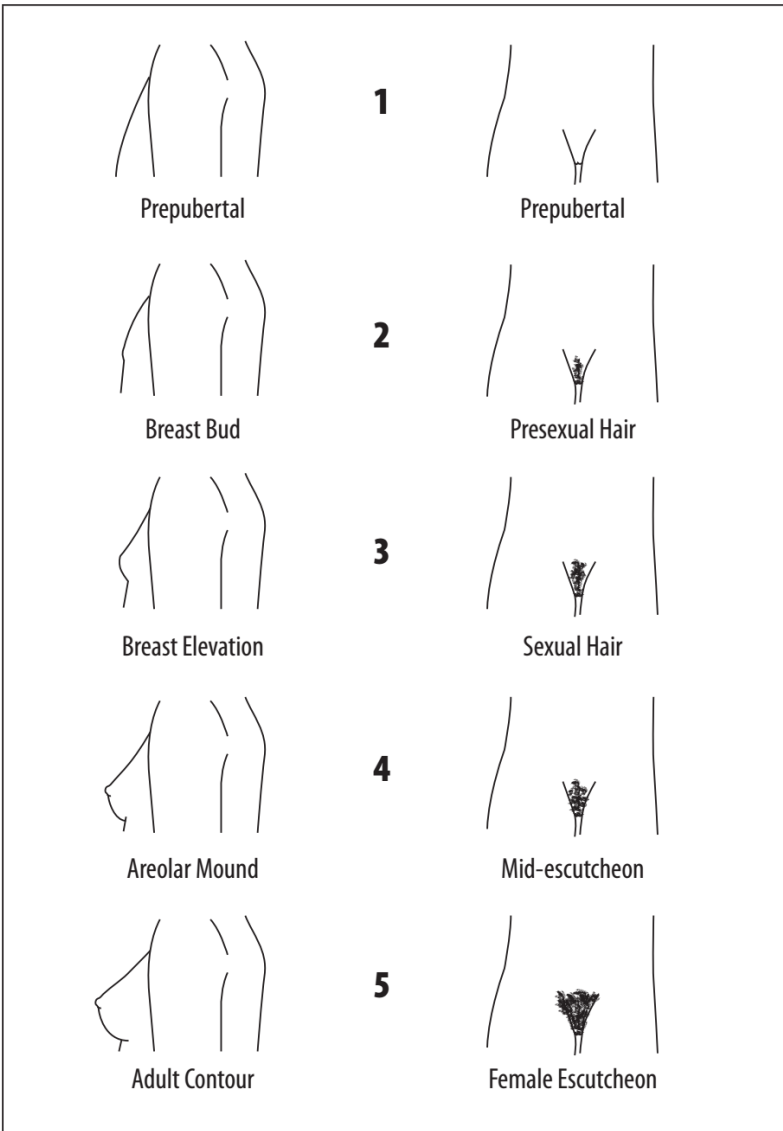
Reprinted with permission from Bordini B, Rosenfield RL. Normal pubertal development: part II: clinical aspects of puberty. *Pediatr Rev*. 2011;32(7):282

Item C82B. Stages of Genital Development in Males.
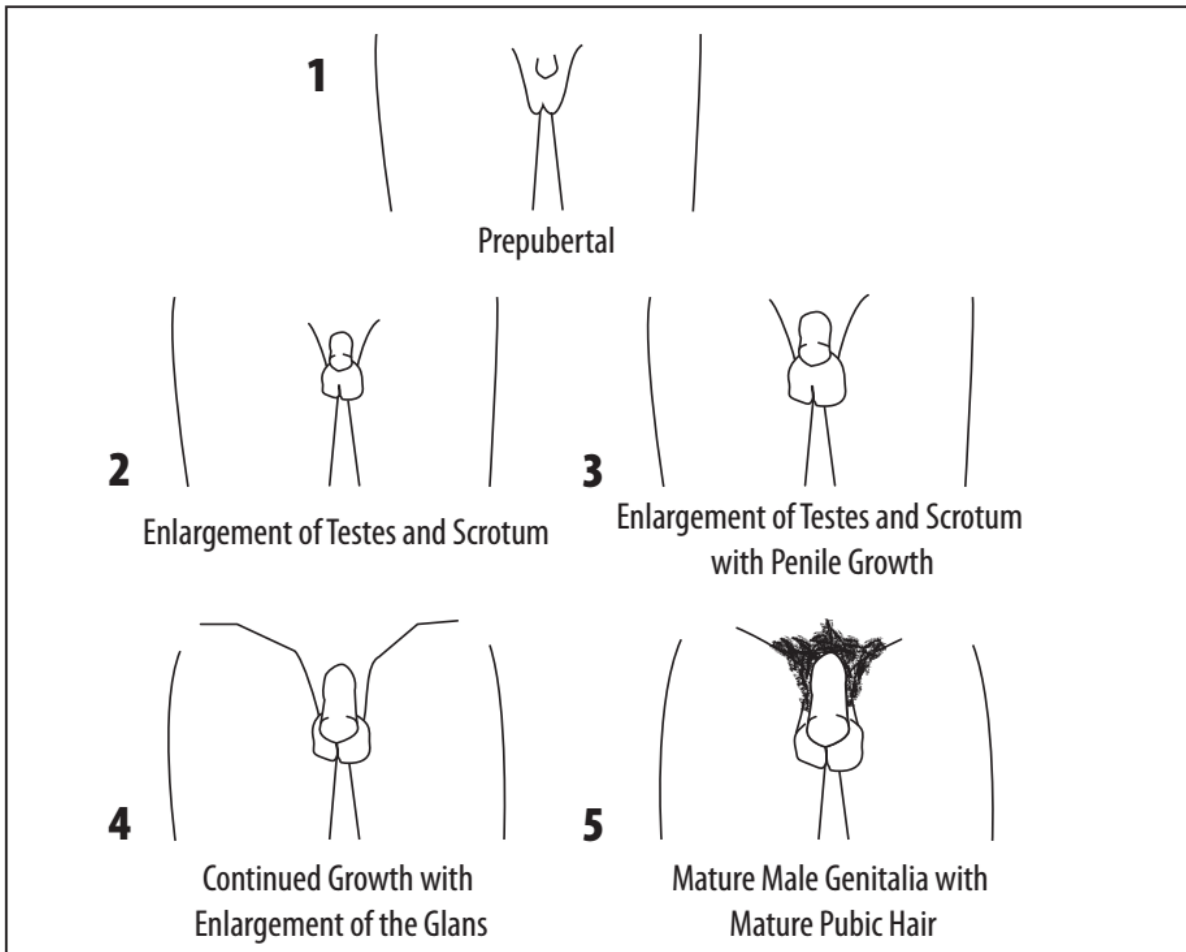
1
Prepubertal

2
Enlargement of Testes and Scrotum

3
Enlargement of Testes and Scrotum with Penile Growth

4
Continued Growth with Enlargement of the Glans

5
Mature Male Genitalia with Mature Pubic Hair

Reprinted with permission from Bordini B, Rosenfield RL. Normal pubertal development: part II: clinical aspects of puberty. *Pediatr Rev.* 2011;32(7):283

The first signs of puberty are breast development (girls) and testicular enlargement (boys). The typical age of the onset of puberty can vary by ethnicity, particularly among girls. A recent study by Susman and colleagues looked at the longitudinal development of secondary sexual development in a multiracial population and found the mean age for each stage of sexual development (see suggested reading 5).

While biliary disease can cause elevated ALP, typically it is 3 to 4 times the upper limit of normal. Bone tumors can also cause elevated ALP, but would be an unlikely diagnosis in the adolescent in this vignette with a review of systems significant only for snoring. Lower than expected ALP values can be seen with vitamin C deficiency. Laboratory error is a possibility; however, it is developmentally appropriate for this patient to have an elevated ALP, making pubertal growth a better explanation for this finding.

Other laboratory changes during puberty include:

1. Increases in luteinizing hormone and follicle-stimulating hormone. Luteinizing hormone increases throughout puberty. Follicle-stimulating hormone increases, but then tends to plateau when SMR 3 is achieved.
2. All of the sex hormones, including estradiol and testosterone, increase during puberty.
3. Insulin secretion also increases during adolescence.

PREP Pearls
- Elevated alkaline phosphatase can be seen during linear growth associated with adolescence.
- Increases in luteinizing hormone and follicle stimulating hormone are seen in puberty. Luteinizing hormone increases throughout puberty. Follicle-stimulating hormone increases, but can plateau when sexual maturity rating 3 is achieved.
- All of the sex hormones, including estradiol and testosterone, increase during puberty.
- Insulin secretion increases during adolescence.

ABP Content Specifications(s)
- Recognize the stages of sexual development and the range of age of onset of each
- Recognize laboratory values that change in girls and boys during puberty

Suggested Readings
- Corathers SD. Focus on diagnosis: the alkaline phosphatase level: nuances of a familiar test. Pediatr Rev. 2006;27(10):382-384. DOI: http://dx.doi.org/10.1542/pir.27-10-382.

Question 83

A 16-year-old adolescent girl presents to your office for evaluation of a 2-month history of hair loss. Her physical examination demonstrates an area of incomplete alopecia at the vertex. Within the affected area are hairs of differing lengths and 2 areas of hemorrhage (Item Q83).

Hair loss as described for the girl in the vignette. Reprinted with permission from Krowchuk DP, Mancini AJ, eds. Pediatric Dermatology. A Quick Reference Guide. Elk Grove Village, IL: American Academy of Pediatrics; 2012.

Of the following, the MOST likely diagnosis is

A.   alopecia areata

B.   friction alopecia

C.   tinea capitis

D.   traction alopecia

E.   trichotillomania

Question 83   **Preferred Response: E**

The adolescent in the vignette has an area of incomplete hair loss where hairs of varying lengths are observed. These physical findings suggest trichotillomania (hair-pulling disorder), a form of traumatic alopecia in which individuals repetitively twist, twirl, or pull hair. The areas of hemorrhage observed in the adolescent in the vignette represent sites from which hairs were pulled (Item C83A).

Item C83A

The girl described in the vignette has an area of hair loss within which hairs of differing lengths may be seen. Two areas hemorrhage (arrows) are present at sites where hairs were pulled. Adapted and reprinted with permission from Krowchuk DP, Mancini AJ, eds. Pediatric Dermatology: A Quick Reference Guide. Elk Grove Village, IL: American Academy of Pediatrics; 2012.

Trichotillomania usually involves the scalp, but any hair-bearing area can be affected (eg, eyebrows, eyelashes). In young children, trichotillomania often represents a habit similar to thumb sucking. For example, children unconsciously twirl or pull hair while watching television. In such cases, parents may be advised to offer a gentle reminder when the behavior is observed. In older children and adolescents, trichotillomania often represents a compulsion, and is considered among the obsessive-compulsive-related disorders. Comorbid depression and anxiety are not uncommon and therapy is more challenging. Cognitive behavioral therapy, especially habit reversal training (HRT), can be highly successful in reducing hair pulling, but treatment is intensive and finding skilled therapists can be difficult. In HRT, patients are taught to monitor

hair-pulling behavior (identifying circumstances or emotions associated with pulling), avoid triggers, and initiate a competing response when the urge to pull hair develops (eg, manipulating a stress ball or rubber band, clenching fists). There are no US Food and Drug Administration-approved pharmacologic agents for the treatment of trichotillomania. Selective serotonin reuptake inhibitors are often employed, but evidence supporting their efficacy is lacking. The tricyclic antidepressant clomipramine has been shown to reduce hair-pulling urges and increase the ability to resist such urges. N-acetylcysteine has been investigated for use in trichotillomania, but data regarding its benefit are conflicting.

Several other disorders may produce circumscribed areas of hair loss without scalp scarring and therefore may mimic trichotillomania. These include:
- Alopecia areata: round to oval patches of complete hair loss; the scalp appears normal (Item C83B)
- Friction alopecia: round to oval patch of hair thinning located at the occiput in young infants who spend much of their time in the supine position (Item C83C)
- Tinea capitis: in the most common form, 1 or more patches of alopecia are present, as well as scale and "black-dot" hairs (the remnants of broken hairs within follicles) (Item C83D)
- Traction alopecia: thinning of hair in areas where the hair is being stretched as the result of braiding or creating ponytails (often seen at temporal-parietal hairline) (Item C83E)
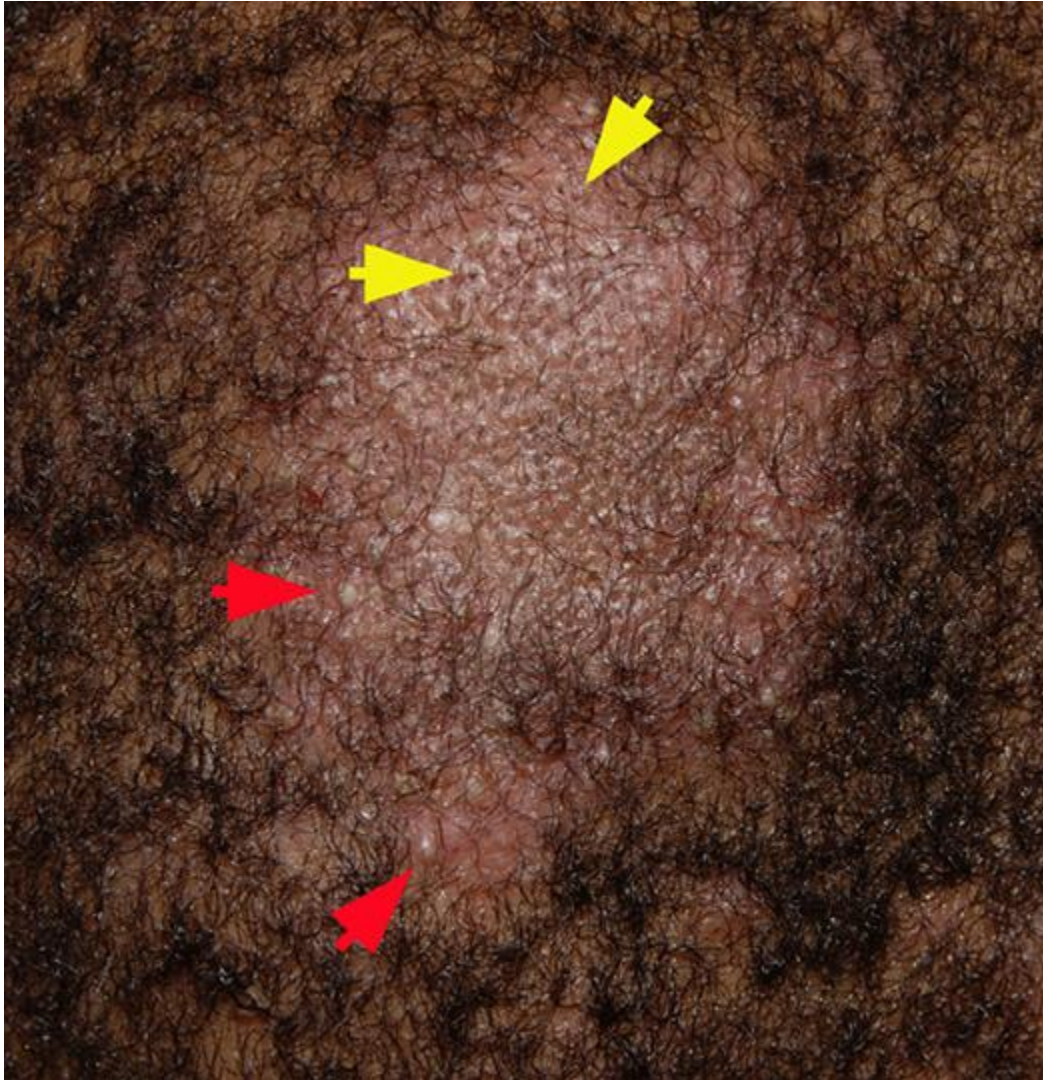
Item C83C

In alopecia areata, round or oval patches of complete hair loss are observed. Courtesy of D. Krowchuk.

Item C83C

Friction alopecia appears as thinning of the hair most often at the occiput in young infants. Courtesy of D. Krowchuk.

Item C83D

In the most common form of tinea capitis, there are one or more patches of hair loss within which one may see scale, "black-dot" hairs (yellow arrows), or pustules (red arrows). Adapted and 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 C83E

Thinning of the hair at sites of tight braiding (often the temporal-occipital region) is characteristic of traction alopecia. Adapted and 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.

PREP Pearls
- When evaluating hair loss, it is useful to consider: (1) is the hair loss localized or generalized, and (2) is there scarring of the scalp? Most hair loss in children is localized and scarring is absent.
- The leading causes of localized hair loss are tinea capitis, traction, friction, alopecia areata, and trichotillomania.
- In trichotillomania, the hair loss is incomplete (unlike in alopecia areata or tinea capitis) and, within the affected area, hairs are of differing lengths.

ABP Content Specifications(s)
- Recognize the clinical findings associated with trichotillomania, and manage appropriately

Suggested Readings
- Bloch MH, Panza KE, Grant JE, Pittenger C, Leckman JF. N-acetylcysteine in the treatment of pediatric trichotillomania: a randomized, double-blind, placebo-controlled add-on trial. J Am Acad Child Adolesc Psychiatry. 2013;52(3):231-240. DOI:http://dx.doi.org/10.1016/j.jaac.2012.12.020.

Question 84

A 10-year-old boy presents to the emergency department after he was hit in the head with a baseball. His vital signs show a temperature of 37°C, blood pressure of 120/80 mm Hg, pulse of 60 beats/min, and respiratory rate of 15 breaths/min. He is awake and alert, complaining of a headache and blurry vision. He denies any other pain, nausea, vomiting, difficulty breathing, or dizziness. There is a boggy, tender area over his right temporal area with no other signs of trauma. Pupils are 3 mm, equal, and reactive. When he looks straight ahead, his right eye deviates medially. Extraocular movements and cranial nerves are otherwise intact. Mental status examination and the remainder of the neurologic examination are unremarkable. A computed tomography of the head confirms your suspected diagnosis.

Of the following, the MOST appropriate initial step in management is

A.   hospital admission for observation

B.   intubation and mechanical ventilation

C.   magnetic resonance imaging of the brain

D.   neurosurgery consultation

E.   ophthalmology consultation

Question 84   **Preferred Response: D**

Based on the mechanism and area of injury, the right-sided lateral rectus palsy, and positive findings on computed tomography, it is likely the child in this vignette has an epidural hematoma (EDH). Because this injury could lead to neurologic deterioration, and the likelihood of spontaneous resolution is low, a neurosurgery consultation should be obtained.

The presence of bleeding between the skull and the dura mater is known as EDH. The most common cause of EDH is blunt trauma, and the most frequent source of bleeding is arterial. A common mechanism of such trauma involves being struck in the temporal region of the skull, causing temporal skull fracture and injury to the middle meningeal artery. Characteristic appearance on computed tomography is a lenticular-shaped density, as shown in Item C84A. The classic progression of EDH is an initial loss of consciousness at the scene, followed by a lucid interval that can last several hours, followed by neurologic deterioration caused by expanding hemorrhage and cerebral edema. This can lead, in some cases, to increased intracranial pressure, herniation syndrome, and death. It is important to note that only approximately 20% of patients with EDH experience the classic initial loss of consciousness. In many cases, the neurologic examination is normal, so a high index of suspicion should be maintained in patients with persistent symptoms of vomiting, headache, and signs of skull fracture. In such patients, computed tomography of the head should be obtained. Although the child in this vignette is lucid, the presence of extraocular motion abnormality is ominous, and could represent uncal herniation. In a unilateral brain hemorrhage, downward pressure on the temporal lobe could cause the uncus to herniate through the tentorium, exerting pressure on the midbrain structures. An early sign of this is a lateral rectus palsy caused by dysfunction of the sixth cranial nerve.
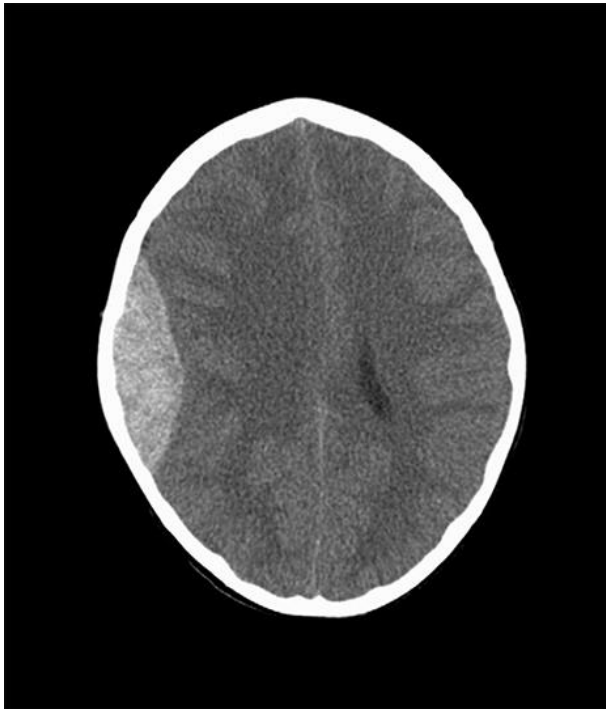
Item C84A

Computed tomography for the boy described in the vignette. The lenticular-shaped density is a classic presentation for an epidural hematoma. Reprinted with permission from Schunk JE, Schutzman SA. Pediatric head injury. Pediatr Rev. 2012;33(9):398-411.

Bleeding between the dura mater and arachnoid mater is known as subdural hematoma (SDH). Similar to EDH, SDH is most commonly caused by blunt trauma, though in contrast, it is not usually associated with an overlying skull fracture. Other than blunt trauma, SDH can be caused by acceleration-deceleration injury, such as occurs in motor vehicle collisions or abusive shaking of an infant, leading to tearing of bridging veins between the dura and arachnoid maters. Classic appearance on head computed tomography is crescentic (Item C84B). SDH is most often associated with underlying brain edema, injury, and ischemia. Thus, patients are likely to present with significant depressed consciousness or signs of elevated intracranial pressure. In previously unrecognized SDH, or in cases of abuse, SDH may be chronic and of varying ages. If SDH is suspected, computed tomography should be obtained, and emergent neurosurgical consultation should be sought.

Item C84B

Computed tomography in a patient with a subdural hematoma. Note the crescentic pattern characteristic of a subdural hematoma. Reprinted with permission from Schunk JE, Schutzman SA. Pediatric head injury. Pediatr Rev. 2012;33(9):398-411.

For any patient with traumatic brain injury, the clinician should frequently assess the patient's degree of neurologic dysfunction and coma. A Glasgow Coma Score (GCS) should be obtained (Item C84C). Emergent treatment for increased intracranial pressure and severe herniation syndrome may be necessary in cases of severe coma (GCS < 8). This may include endotracheal intubation and mechanical ventilation for airway protection, brief hyperventilation, and osmotherapy including hypertonic saline or mannitol.

Item C84C. Glasgow Coma Scale. (Best Score is 15)

Activity	Best Response	Score
Eye Opening	Spontaneous	4
	To verbal stimuli	3
	To pain	2
	None	1
Verbal	Oriented	5
	Confused	4
	Inappropriate words	3
	Nonspecific sounds	2
	None	1
Motor	Normal spontaneous	6
	Localizes pain	5
	Withdraws to pain	4
	Abnormal flexion	3
	Abnormal extension	2
	None	1

Reprinted with permission from Schunk JE, Schutzman SA. Pediatric Head Injury. *Pediatr Rev.* 2012;33;398-411

For the boy in the vignette, observation alone is not the best course of action, because he will likely require evacuation of the hemorrhage. Intubation and mechanical ventilation is not necessary, because he is awake, alert, and protecting his airway. Magnetic resonance imaging is a sensitive and specific modality for intracranial injury. However, the diagnosis of epidural hematoma is already evident from the computed tomography study. Ophthalmology consultation is not necessary because the origin of his extraocular abnormality is neurologic.

PREP Pearls
- Intracranial epidural hematoma (EDH) is usually associated with a skull fracture, is commonly caused by disruption of a meningeal artery, and usually does not include underlying cerebral edema or contusion
- Intracranial subdural hematoma (SDH) is usually not associated with a skull fracture, is commonly caused by disruption of bridging veins, and usually includes underlying edema and contusion.
- An emergent computed tomography of the head and neurosurgical consultation should be sought in cases of suspected or confirmed SDH or EDH.

ABP Content Specifications(s)
- Recognize the clinical findings associated with epidural hematoma, and manage appropriately
- Recognize the clinical findings associated with subdural hematoma with and without skull fracture, and manage appropriately

Suggested Readings
- Atabaki SM. Pediatric head injury. Pediatr Rev. 2007;28(215):215-223. DOI:http://pedsinreview.aappublications.org/content/28/6/215.

Question 85

A 13-year-old adolescent is hospitalized with fever, bloody stools, and weight loss. She has a history of multiple infections. Vital signs show a temperature of 38.3°C, respiratory rate of 24 breaths/min, heart rate of 130 beats/min, blood pressure of 101/70 mm Hg, and a weight of 32.5 kg. On physical examination, she is cachectic, has abdominal tenderness over the left lower quadrant, and hepatosplenomegaly. Laboratory data shows:

- White blood cells, 4,600/μL (4.6 x 109/L)
- Hemoglobin, 8.1 g/dL (81 g/L)
- Platelets, 185 x 103/μL (185 x 109/L)
- Differential, 63% segmented neutrophils, 23% bands, 8% lymphocytes, 4% monocytes, 2% eosinophils
- HIV enzyme-linked immunosorbent assay, positive
- HIV viral load, 312,729 copies/mL

A resident working with you asks about HIV testing in the adolescent.

Of the following, the MOST accurate response is

    A.   adolescents should routinely be tested for HIV according to prevalence

    B.   the US Centers for Disease Control and Prevention mandates pre-test counseling be provided prior to testing for HIV

    C.   disclosure of HIV status is associated with lower self-esteem

    D.   parental permission is needed for disclosure of HIV status to adolescents

    E.   physicians should accept parental requests to withhold the diagnosis

Question 85   **Preferred Response: A**

In 2011, the American Academy of Pediatrics published a policy statement supporting routine HIV testing in adolescents at least once, regardless of risk factors, when the prevalence of HIV in the population is more than 0.1%. When the prevalence is lower than 0.1%, testing based on risk factors is recommended. Any adolescent who is sexually active is at risk. Adolescents are considered to be high risk if they have multiple sexual partners, are men who have sex with men, exchange sex for money, or use intravenous drugs. It is recommended that high risk youth be tested annually.

The US Centers for Disease Control and Prevention (CDC) recommends universal testing for HIV starting at 13 years of age, based on prevalence instead of risk-based testing. While the CDC recommends that patients should be informed that testing is planned, it should be performed unless patients opt out. Separate written informed consent or pre-test counseling are not recommended, as these can be barriers to evaluation. The CDC policy is influenced by an increasing number of HIV cases, missed opportunities for testing, and missed cases when only risk-based testing is performed. For example, in 2006, 5% of the over 1 million HIV-infected individuals living in the United States were adolescents and young adults, and nearly half of these were unaware of their infection. However, individual state laws vary and can inhibit implementing the CDC recommendation. Clinicians can reference the Compendium of State HIV Testing Laws from the National HIV/AIDS Clinician's Consultation Center (http://nccc.ucsf.edu/) to determine how local laws and the CDC recommendations apply in their setting.

While there may be hesitation to disclose status because of the stigma associated with the diagnosis of HIV, withholding the diagnosis can have negative consequences such as preventing access to social support, which is known to buffer psychological distress in those with HIV. In some studies, disclosure of HIV status is related to greater quality of social support, greater self-esteem and lower levels of depression. The American Academy of Pediatrics supports disclosure of information that is developmentally appropriate. Most perinatal infections are disclosed by 10 years of age.

The laws that govern consent and confidentiality for HIV care vary by state. However, public health statutes and legal precedent support medical evaluation and treatment of HIV without parental knowledge or consent. Confidential community-based HIV testing is available if it cannot be achieved in a particular practice for concerns of insurance billing or other reasons. Local testing sites can be identified through the CDC web site (http://gettested.cdc.gov/).

PREP Pearls
- The American Academy of Pediatrics supports routine HIV testing in adolescents at least once when the prevalence of HIV in the population is more than 0.1%.
- Although local laws may differ, the US Centers for Disease Control and Prevention recommends universal HIV screening without separate written informed consent or pre-test counseling.

- Disclosure of status congruent with the patient's developmental stage is supported by the American Academy of Pediatrics.

ABP Content Specifications(s)
- Recognize and apply ethical principles regarding the care of children and adolescents with AIDS/HIV infection

Suggested Readings
- American Academy of Pediatrics. Adolescents and HIV infection: the pediatrician's role in promoting routing testing.Pediatrics. 2011;128(5):1023-1029. DOI: http://dx.doi.org/10.1542/peds.2011-1761.

Question 86

You are evaluating a 10-hour-old term female neonate. The neonate was born to a 34-year-old gravida 2, para 2 woman by normal vaginal delivery. Antenatal history was significant for bilateral hydronephrosis. A third trimester ultrasonography reported the anteroposterior renal pelvic diameter was dilated at 15 mm bilaterally.

The neonate's current vital signs show a weight of 3.0 kg, temperature of 37°C, heart rate of 140 beats/min, respiratory rate of 40 breaths/min, blood pressure of 80/46 mm Hg, and oxygen saturation of 97% by pulse oximetry on room air. Physical examination reveals a comfortable pink newborn, with clear and equal breath sounds bilaterally, no murmur, and no abdominal mass.

Of the following, you are MOST likely to inform the parents that the risk for congenital anomaly of the kidneys and urinary tract is

A.   low because the patient has normal blood pressure

B.   low because there is no antenatal history of oligohydramnios

C.   high because renal pelvic diameter is more than 10 mm

D.   high if the patient currently has elevated creatinine

E.   uncertain until the results of postnatal ultrasound become available

Question 86   **Preferred Response: C**

Hydronephrosis (dilatation of the renal pelvis with or without dilatation of the renal calyces) can be easily diagnosed on antenatal ultrasonographic examination (from 12th to 14th week of gestation). Antenatal hydronephrosis could be transient, secondary to urinary tract obstruction or vesicoureteral reflux (VUR). Urinary tract obstruction or VUR may be associated with abnormal renal development (renal dysplasia) or lead to renal injury.

Transient antenatal hydronephrosis, not associated with clinically significant renal dysplasia or risk for renal injury, is common; therefore, testing of these newborns is not indicated or helpful.

Postnatal management of newborns diagnosed antenatally with hydronephrosis aims to identify those patients with significant congenital anomalies of the kidneys and urinary tract (CAKUT) and minimizing associated long term renal injury and other adverse effects (urinary tract infections, bladder dysfunction). The degree of hydronephrosis and presence of bilateral hydronephrosis are used to identify neonates with increased risk for CAKUT and needing detailed investigations. Hydronephrosis severity is graded based on the anterior posterior renal-pelvic diameter (RPD), measured on renal ultrasonography. Antenatal RPD is affected by gestational age, maternal hydration, and degree of fetal bladder distention. Antenatally, RPD of 4 to 10 mm is categorized as mild pelvic dilatation and greater than 10 mm as severe pelvic dilatation. Postnatally, RPD of:
    1.   Less than 7 mm is categorized as normal
    2.   7 to 8 mm is categorized as mild hydronephrosis
    3.   9 to 15 mm is categorized as moderate hydronephrosis
    4.   Greater than 15 mm is categorized as severe hydronephrosis

Severe hydronephrosis and bilateral hydronephrosis on ante- or postnatal renal ultrasonography are associated with the greatest risk for CAKUT, requiring surgical intervention. Also, fetuses with RPD greater than 15 mm in the third trimester have been reported to have increased risk for CAKUT. Patients with pre- and postnatally diagnosed hydronephrosis should be managed in conjunction with specialists and further evaluation should be done in discussion with them. Further imaging studies in these patients include voiding cystourethrography, diuretic renal scan, and serial ultrasonography.

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. Therefore, elevated creatinine in a 10-hour-old neonate is not a good indicator of underlying kidney function.

Maternal oligohydramnios is indicative of decreased fetal urine and impaired renal function. It is associated with severe renal dysplasia or obstruction of the urinary tract (as in posterior urethral valves). Maternal history of severe oligohydramnios is associated with increased neonatal risk for pulmonary hypoplasia and these patients may present with respiratory problems immediately after birth. The 2 most common congenital anomalies associated with prenatal hydronephrosis;

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ureteropelvic junction (UPJ) obstruction and VUR are not usually associated with maternal oligohydramnios.

In the neonatal period, UPJ obstruction and VUR rarely present with elevated blood pressure. In patients with congenital renal dysplasia, blood pressure is normal at initial diagnosis, as these patients have high urine output caused by tubular injury and increased sodium losses from damaged or dysplastic tubules. Hypertension develops later in association with progressive renal failure.

PREP Pearls
- Antenatal hydronephrosis can be transient, secondary to urinary tract obstruction or vesicoureteral reflux (VUR).
- Transient antenatal hydronephrosis is not associated with clinically significant renal dysplasia or risk for renal injury; therefore, testing of these newborns is not indicated or helpful.
- Hydronephrosis associated with urinary tract obstruction or VUR may be associated with abnormal renal development (renal dysplasia) or increased risk for renal injury.
- Severe hydronephrosis (> 10 mm renal-pelvic diameter) and bilateral hydronephrosis are associated with increased risk for congenital anomalies of the kidneys and urinary tract.

ABP Content Specifications(s)
- Plan the diagnostic evaluation of abnormalities of the kidneys, urinary collecting system, bladder, and urethra

Suggested Readings
- Carmody JB, Carmody RB. Question from the clinician: management of prenatal hydronephrosis. Pediatr Rev. 2011;32(12):e110-e112. DOI: http://dx.doi.org/10.1542/pir.32-12-e110.

Question 87

You are caring for a newborn delivered at a gestational age of 26 weeks with complex congenital cardiac disease. A resident is assisting you in the nursery. On physical examination, you find an intrauterine growth-restricted newborn with a 3/6 systolic ejection murmur. He is on nasal cannula oxygen with a pulmonary examination that is unremarkable. His mother prefers not to breastfeed and you are discussing his nutritional needs with your resident.

Of the following, the BEST choice for protein intake in this newborn is

A.   0.5 g/kg per day

B.   1 g/kg per day

C.   2 g/kg per day

D.   4 g/kg per day

E.   6 g/kg per day

Question 87   **Preferred Response: D**

The newborn described in this vignette has increased protein and caloric needs caused by significant prematurity and congenital cardiac disease. Item C87 has a review of protein requirements for infants and children of different ages.

Item C87. Protein Requirements for Infants and Children of Different Ages.

Age	Protein requirement in g/kg/day
24-30 weeks	3.5-4
30-40 weeks	3
Term infant to 12 months	2-3
Children 1-12 years	1.5-2.5
Children 12-18 years	1-1.5

Courtesy of C. Waasdorp Hurtado

Protein accounts for 10% to 15% of energy intake. The typical Western diet has increased protein intake, with an average of 70 g to 100 g per day. Protein requirement in adults is 0.75 to 1 g/kg per day. Proteins are digested to amino acids, which are then used for protein synthesis and molecular signaling. Premature infants have increased needs because of increased protein synthesis and elevated enteral losses. Many children with chronic medical conditions have increased metabolic needs, including increased requirements of protein. These conditions include cancer, cystic fibrosis, failure to thrive, and other scenarios in which children require additional calories. There are also conditions that require lower protein intake, including chronic renal and liver disease.

PREP Pearls
- Premature infants have increased protein needs.
- Many children with chronic medical issues have increased protein needs.
- A term infant requires 2 to 3 g/kg per day of protein.
- An adult requires 0.75 to 1g/kg per day of protein.

ABP Content Specifications(s)
- Understand the dietary mineral requirements of patients of various ages, including those born prematurely, and the circumstances in which those requirements may change

Suggested Readings
- Kleinman RE, Greer FR. Pediatric Nutrition. 7th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2013: 1431 pp.

Question 88

A 17-month-old boy presents to your clinic with developmental delay, poor growth, hypotonia, and microcephaly. He has very little speech beyond repetitive sounds. The mother reports that he rolled over at 5 months of age, sat at 9 months of age, and began crawling at 16 months of age. He had a full endocrinology workup for his slow growth, which was unremarkable. His birth history is significant for mild intrauterine growth retardation and an unusual high-pitched cry that has persisted. The remainder of his past medical history is notable for gastroesophageal reflux and hypospadias. His height and weight are at the first percentile; his head circumference is at the second percentile. Dysmorphologic examination shows microcephaly, mild hypertelorism, broad nasal bridge, short palpebral fissures, bilateral epicanthal folds, and micrognathia (Item Q88). No cardiac murmur is noted. A high resolution chromosome analysis is ordered and reveals the diagnosis.

Item Q88. Boy described in the vignette.
Courtesy of L. Parsley.

Of the following, the MOST likely diagnosis is

A.   Angelman syndrome

B.   cri-du-chat syndrome

C.   fragile X syndrome

D.   Russell-Silver syndrome

E.   Williams syndrome

Question 88   **Preferred Response: B**

The child in this vignette has a classic history consistent with cri-du-chat syndrome. This is a genetic disorder caused by a chromosomal deletion of varying size of the short arm of chromosome 5. It occurs in 1 out of 15,000 to 1 out of 50,000 births. Classic features include a high pitched or cat-like cry (> 95%), small head, epicanthal folds, micrognathia, broad nasal bridge, hypertelorism, downward-slanting palpebral fissures, and moderate to severe intellectual disability. Other systemic congenital malformations are not common, but can include congenital heart disease, renal anomalies, hypospadias, cryptorchidism, and ear tags. The characteristic cat-like cry is secondary to laryngeal and epiglottic anomalies. Cri-du-chat syndrome can be diagnosed based on clinical presentation, along with a high resolution karyotype, fluorescent in situ hybridization (FISH) for 5p-, or a chromosomal microarray. If highly suspected, a high resolution karyotype would make the diagnosis. The American College of Medical Genetics and the American Academy of Pediatrics recommend all children presenting with developmental delay have a chromosomal microarray, which would not only pick up a gross chromosomal deletion as in the child in the vignette, but also microdeletions that could be easily missed on a high resolution karyotype. An array would also be helpful at clarifying specific breakpoints and genes involved in the deletion. There exists genotype and phenotype correlation dependent on the size of the actual deletion. Early intervention and therapies can improve the prognosis and social communication of patients with cri-du-chat. Most patients have prenatal and postnatal growth retardation. They tend to have sweet and affectionate personalities, along with hyperactivity. Some patients have sensorineural deafness, so a formal audiologic examination is recommended. Survival is high with low morbidity because of the low incidence of major congenital malformations. Most cases are caused by a de novo random event; however, parental analysis should be performed to exclude a balanced translocation in the unaffected parent, which could incur an increased risk for recurrence.

Angelman syndrome manifests with severe developmental delay, significant speech impairment, gait ataxia, microcephaly, seizures, and a happy demeanor with inappropriate laughing and excitability. They lack the classic facial dysmorphology and cat-like cry seen in cri-du-chat syndrome. It is diagnosed with DNA methylation analysis of the 15q11.2-q13 chromosome region (78%) or UBE3A sequence analysis (11%).

Fragile X syndrome is a disorder caused by a FMR1 full gene mutation (> 200 CGG repeats) or other loss-of-function mutations characterized by moderate intellectual disability in affected males and mild intellectual disability in affected females. Males with an FMR1 full gene mutation develop a classic dysmorphologic facial appearance as they age, which includes macrocephaly, long face, prognathism, and large, outwardly-rotated ears. Patients also have joint laxity and macroorchidism after puberty. Behavioral abnormalities are common.

Russell-Silver syndrome is caused by abnormal methylation of the paternal imprinting center of chromosome 11p15.5 or maternal uniparental disomy for chromosome 7 (UPD7). It presents with intrauterine growth retardation, followed by proportionately short postnatal growth deficiency, normal head circumference, fifth finger clinodactyly, triangular facies, and sometimes limb-length asymmetry.

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Williams syndrome is caused by another contiguous gene deletion on chromosome 7 involving the elastin gene (ELN), which can be diagnosed with a high resolution karyotype, FISH analysis, or a chromosomal microarray. Patients with Williams syndrome present with cardiovascular disease (peripheral pulmonary stenosis, supravalvar aortic stenosis, elastin arteriopathy), classic "elfin" facies (broad forehead, wide smile with full lips and wide-spaced teeth, full cheeks), connective tissue pathology (hypotonia and hyperextensibility), mild to moderate intellectual disability, a friendly personality, hypothyroidism, growth problems, and hypercalcemia.

PREP Pearls
- Cri-du-chat syndrome is caused by a deletion of the short arm of chromosome 5 characterized by a high pitched or cat-like cry (> 95%), small head, epicanthal folds, micrognathia, broad nasal bridge, hypertelorism, downward-slanting palpebral fissures, and moderate to severe intellectual disability.
- Cri-du-chat syndrome can be diagnosed based on clinical presentation along with a high resolution karyotype, fluorescent in situ hybridization (FISH) for 5p-, or a chromosomal microarray.
- The American College of Medical Genetics and the American Academy of Pediatrics recommend all children presenting with developmental delays have a chromosomal microarray, which would not only pick up a gross chromosomal deletion, but also microdeletions that could be easily missed on a high resolution karyotype.

ABP Content Specifications(s)
- Understand the significance of a gross chromosomal deletion
- Plan appropriate parental evaluation when an infant is born with a structural chromosomal abnormality

Suggested Readings
- Mainardi CP. Cri-du-Chat syndrome. Orphanet J Rare Dis. 2006;1:33. DOI: http://dx.doi.org/10.1186/1750-1172-1-33.
- Rodríguez-Caballero A, Torres-Lagares D, Rodríguez-Pérez A, Serrera-Figallo MA, Hernández-Guisado JM, Machuca-Portillo G. Cri du chat syndrome: a critical review. Med Oral Patol Oral Cir Bucal. 2010;15(3):e473-e478. DOI:

Question 89

The mother of a 3-year-old boy brings him to the office for evaluation of penile discharge. She became concerned when she noticed some white matter while bathing him. The boy has been otherwise well. He is toilet trained and has not been complaining of pain with urination. On physical examination, you notice a partially retractable foreskin with a small amount of pasty, cheesy material under the foreskin. There is no swelling or erythema of the shaft or glans penis. There are no penile lesions and no discharge from the meatus.

Of the following, the MOST appropriate next step is to

A.   evaluate for possible sexual abuse and refer to child protective services

B.   instruct the mother to retract the foreskin fully by applying increasing force daily

C.   obtain a culture and sensitivity of the material and begin antibiotic treatment

D.   offer reassurance and encourage gentle washing of the area daily

E.   refer to urology for circumcision

Question 89   **Preferred Response: D**

View Peer Results The boy in the vignette has normal physiologic partial phimosis and smegma. Desquamated epithelial cells that are trapped under the foreskin create smegma. This accumulation of epithelial debris is physiologic. His mother should be reassured and counseled on the appropriate care of the uncircumcised foreskin, which includes gentle washing with soap and water with regular bathing. Forcible retraction of the foreskin should be avoided. Although parents may mistakenly believe that the smegma suggests an infection, no culture and sensitivity or treatment with antibiotics is needed. This is also not the type of discharge that would be seen in cases of sexually transmitted diseases, therefore no evaluation for sexual abuse is warranted. Referral to urology for evaluation of phimosis or performance of circumcision is not needed, because this is a physiologic process.

The foreskin typically extends about 1 cm beyond the glans penis and provides protection to the urethral meatus and glans. Phimosis is the inability to retract the foreskin or prepuce to expose the glans penis, and is caused by either a small foreskin opening or adhesions. In young boys, this is normal or physiologic and the ability to fully retract the foreskin increases with age. Physiologic phimosis is present in almost all newborns, because of normal adhesions that exist between the foreskin and glans penis. After birth and during childhood, penile growth and physiologic erections aid in the natural process that loosens the adhesions and leads to foreskin retraction. The degree of normal retractability by age varies a great deal, and reports of norms have differed. There is a high likelihood that physiologic phimosis will resolve spontaneously. Partial phimosis is quite common and may still be present in one-third of 5- to 7-year-old boys. The foreskin can be retracted fully, or enough to visualize part of the glans penis, in most boys by the age of 12 to 13 years.

The uncircumcised penis should be washed with soap and water regularly when bathing. As the foreskin naturally begins to retract, proper hygiene that includes cleaning and drying underneath should be emphasized. Patients or parents can be taught to perform gentle stretching exercises, but forcible retraction should be avoided. A 1- to 2-month course of topical corticosteroids applied to the preputial outlet may be used, in addition to stretching exercises, to speed up the natural process of obtaining full retraction of the foreskin.

Pathologic phimosis is a truly nonretractable foreskin secondary to distal scarring of the prepuce. This may be the result of previous trauma or inflammation. It is important for primary care clinicians to distinguish pathologic cases, because they will need referral to a pediatric urologist or surgeon.

Paraphimosis occurs when the foreskin is retracted and gets trapped behind the coronal sulcus of the glans penis, leading to venous and lymphatic congestion. Painful swelling of the shaft and glans penis distal to the constriction progresses, which further hinders return of the foreskin to its natural position, and promotes more congestion. Eventually, this can impede blood flow to the glans penis with the potential for permanent damage or gangrene. Paraphimosis is a urologic emergency. Treatment requires adequate pain control and timely reduction of the foreskin back to the normal position. If the general practitioner is not able to perform the reduction, or if signs

of arterial compromise are present, immediate urologic consult is indicated. Anticipatory guidance to decrease the risk of paraphimosis should inform the patient and family that the foreskin must always be pulled down to its normal position to cover the glans penis after cleaning or sexual intercourse.

PREP Pearls
- Smegma, desquamated epithelial cells trapped under the foreskin, is a common physiologic finding in uncircumcised males.

Question 90

A 2-year-old boy presents to the emergency department having fallen down a flight of 5 stairs and hitting his head on a ceramic floor. On further history, his mother reports that, since birth, he has had excessive bruising in response to seemingly minor trauma, bruising in unusual places such as the small of the back and the dorsal aspects of the proximal arms, frequent gum bleeding with tooth brushing, and having had excessive bleeding following circumcision. There is no known family history of abnormal bleeding symptoms. Given this history, a head computed tomography is obtained that demonstrates an intracranial bleed. Laboratory evaluations are shown:

Laboratory Test	Results	Normal Range
Prothrombin time	12.2 seconds	(11 to 15 seconds)
Partial thromboplastin time	32 seconds	(30 to 50 seconds)
Platelet count	203 x 10^3/μL(203 x 10^9/L)	(140 to 450 x 10^3/μL)
Laboratory Test	Results	Normal Range
Prothrombin time	12.2 seconds	(11 to 15 seconds)
Partial thromboplastin time	32 seconds	(30 to 50 seconds)
Platelet count	203 x 103/μL(203 x 10^9/L)	(140 to 450 x 10^3/μL)

Of the following, the MOST appropriate next immediate action would be

    A. call child protective services
    B. infuse intravenous immunoglobulin
    C. infuse factor VIII
    D. infuse factor IX
    E. transfuse leukocyte-depleted single donor platelets

Question 90   **Preferred Response: E**

The clinical findings seen in the child in this vignette suggest a heritable bleeding disorder. Of note is that he has had unusual bleeding since birth (bleeding following a circumcision), repeated mucosal bleeding (such as gum bleeding while tooth brushing), and bruising in locations not readily explainable by trauma (such as the dorsal aspect of the upper arms). Given the strong suspicion for a heritable bleeding disorder and the presence of intracranial blood following trauma, immediate action is needed. The normal prothrombin time (PT), partial thromboplastin time (PTT), and platelet number effectively rule out disorders of the coagulation cascade or platelet number. It is likely that the child in this vignette has a disorder of platelet function. In platelet function disorders, platelets are present, but cannot activate to effectively form a clot. The most common heritable platelet function disorders are Bernard-Soulier syndrome (a disorder of platelet adhesion) and Glanzmann thrombasthenia (a disorder of platelet aggregation). The most appropriate management for life-threatening bleeding in a child with a known or suspected platelet function disorder is to transfuse functional platelets.

The formation of a functional clot requires 2 components, fibrin and platelets. Fibrin is the end product of the coagulation cascade. The absence of any of the factors in the coagulation cascade can result in failure to form a clot and is associated with a prolonged PT or PTT. The PT and the PTT are effective measures of the functionality or presence of the components of the coagulation cascade, except for the conversion of fibrinogen to fibrin. While the absence of functional fibrinogen could explain the scenario presented in the vignette, none of the answers provided would correct a disorder of fibrin, which is treated with infusion of cryoprecipitate.

Von Willebrand factor is a linking factor that allows functional platelets to bind to fibrin to form a clot. Von Willebrand disease (VWD) is the result of decreased function or absence of von Willebrand factor. Von Willebrand disease has multiple phenotypes ranging from mild to severe bleeding disorders that mirror the degree of dysfunction or absence of the von Willebrand factor. There are several different types of VWD, including types 1, 2A, 2B, 2M, 2N, and 3. Von Willebrand disease is the most common heritable bleeding disorder. While severe VWD could explain the scenario in the vignette, the most common subtypes have mild to moderate mucosal bleeding and patients typically present later in life. In addition, none of the answers provided would have corrected for VWD, which would have required the infusion of von Willebrand factor concentrates or cryoprecipitate.

As the platelet number was normal in the child in this vignette, this is not idiopathic thrombocytopenic purpura requiring intravenous gamma globulin. The normal PTT rules out types A and B hemophilia, so infusion of factor VIII or IX is not indicated. The severity of the reported bleeding, especially with circumcision and gum bleeding, makes child abuse unlikely in the absence of additional concerning history, therefore child protective services does not need to be contacted.

PREP Pearls
- The formation of a clot requires normal platelet numbers and function, as well as the ability to form fibrin.
- The treatment for bleeding in a patient with a platelet function disorder is the infusion of platelets with normal function.
- Two of the most common heritable platelet function disorders are Glanzmann thrombasthenia and Bernard-Soulier syndrome.

ABP Content Specifications(s)
- Identify the etiology of bruising in a child with a normal or slightly increased platelet count

Suggested Readings
- Carpenter SL, Abshire TC, Anderst JD. Evaluating for suspected child abuse: conditions that predispose to bleeding. Pediatrics. 2013;131(4):e1357-e1373.DOI: http://dx.doi.org/10.1542/peds.2013-0196.
- Journeycake JM, Buchanan GR. Coagulation disorders. Pediatr Rev. 2003;24(3):83-91. DOI: http://dx.doi.org/10.1542/pir.24-3-83.
- Sharathkumar AA, Pipe SW. Bleeding disorders. Pediatr Rev. 2008;29(4):121-130. DOI: http://dx.doi.org/10.1542/pir.29-4-121.

Question 91

You see an 8-year-old boy who has been struggling in school for the past several years. He was recently tested and identified by his school as having significant learning disabilities in both reading and writing. Neither of his parents had any personal experience with this kind of learning difficulty; in fact, they had found school to be relatively easy. They express worries about how their son will cope and adjust to this problem.

Of the following, the MOST preferred strategy to recommend is

    A.   ask the school to adjust his curriculum such that he almost never experiences "failure" on academic assignments

    B.   emphasize that parents should uphold firm or "tough love" academic expectations for him at home

    C.   encourage him to develop a close nurturing relationship with an educator at the school

    D.   encourage him to view that his disability is something to cope with rather than something in his power to control

    E.   minimize attention given to his areas of normal functioning in favor of focusing on disability instruction

Question 91   **Preferred Response: C**

There are many contributions to a child's overall self-esteem, including experienced family and peer relationships, physical and emotional development, and academic accomplishments. A learning disability creates self-esteem challenges because of the child experiencing repeated academic "failures" and social isolation from peers. Poor self-esteem on top of a learning disability creates many more problems for the child.

Promoting resilience and empowerment for a child with a learning disability can improve their self-esteem, reduce social isolation, and improve academic performance. There is no single path for generating resilience given the multiple overall contributions. However, elements for addressing self-esteem should include:

1.   Providing the child with resources and opportunities to become competent and capable (ie, special education instruction)
2.   Encouraging the child to become a self-advocacy expert in their own best path toward accomplishments
3.   Ensuring that the child has a social network that supports stress mediation and increases their sense of competence

Having at least 1 caring adult within the school that the student knows they can turn to and who will be their advocate is thought to be a critical element to academic success. Therefore, encouraging a nurturing relationship with an educator at the school is likely to improve a child's self-esteem and promote resilience.

Removing all possibilities for experiencing "failure" would not be a productive strategy because learning requires being challenged and thus occasionally experiencing failure. Also, developing resilience to adversity is a very important life skill, which cannot be learned without actually managing stress and adversity. Parents enforcing rigid academic expectations at home (such as negative consequences for receiving a poor grade) will be unlikely to promote coping if done without regard to the child's own learning style and abilities. Instead, it is more helpful if parents expect their child to exert an effort in learning, effort in homework completion, and achievement commensurate to their abilities. Viewing a learning disability just as a burden to be coped with is a less adaptive and self-effective strategy than developing self-effectiveness and mastery over it. For instance, it should become natural for the child to tell an educator in a self-assertive fashion, "I learn best when I…." Parents celebrating a child's accomplishments rather than only focusing on their child's struggles or failures can enhance self-esteem. Thus, if parents only talk about and discuss what the child is struggling to do, a cumulative message of inadequacy would be delivered.

PREP Pearls
- A learning disability creates self-esteem challenges because of the child experiencing repeated academic "failures," and some degree of social isolation from peers.
- Having at least 1 caring adult within the school that the student knows they can turn to and who will be their advocate may be critical to academic success for a child with a learning disability.

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ABP Content Specifications(s)
• Understand strategies to improve the self-esteem of children who have learning disabilities

Suggested Readings
• Brooks RB. Children at risk: fostering resilience and hope. Am J Orthopsychiatry. 1994;64(4):545-553.
• Roer-Strier D. University students with learning disabilities advocating for change. Disabil Rehab. 2002;24(17):914-924. DOI: http://dx.doi.org/10.1080/09638280210148611.

Question 92

A 13-year-old adolescent is transferred into your practice and comes for an initial visit and physical examination. He was full term, has never been hospitalized or had surgery, and takes no medications. He has a history of a heart murmur and the family brings you a copy of his last cardiac evaluation done 5 years ago. The report states that he has a bicuspid aortic valve. He plays baseball and soccer without any symptoms. On physical examination today, he has a heart rate of 72 beats/min, respiratory rate of 18 breaths/min, and blood pressure of 115/65 mm Hg in the right arm and 105/65 mm Hg in the right leg. He has no jugular venous distension. His lungs are clear to auscultation. His cardiac examination reveals a regular rhythm. His S1 and S2 are normal. There is a 1/6 systolic murmur at the left mid sternal border, which does not radiate into the neck. There is no rub or gallop. His abdominal examination is normal. His femoral pulses are brisk. He has been invited to participate in a rugged 2-week survival course and his parents want your opinion as to whether it is safe for him to participate and what his long term prognosis is.

Of the following, the MOST accurate advice or assessment for them at this time is

A.   the gradient across his aortic valve is likely 5 to 30 mm Hg

B.   he has a 75% risk of developing aortic stenosis during his lifetime

C.   he will need lifelong endocarditis prophylaxis

D.   his blood pressures suggest an associated coarctation

E.   the presence of a murmur means that he will need exercise restriction

Question 92   **Preferred Response: D**

One to 2% of the population has a bicuspid aortic valve (BAV). There may not be any murmur, especially early in life, or an ejection click may be heard. There is an association of BAV and coarctation of the aorta, and the blood pressures (BP) reported for the adolescent in this vignette are suggestive of coarctation. Blood pressures in the legs should always be higher than the upper extremities because of peripheral amplification of systolic pressure as blood travels down the aorta. If the BP is lower in the legs than the upper extremity, one should suspect a coarctation. Coarctation, depending on the severity, is a significant comorbidity in that it can cause left ventricular hypertrophy, congestive heart failure, aortic dilation, hypertension, and aneurysm if not treated. Bicuspid aortic valve may also be seen in conjunction with Williams syndrome and Loeys-Dietz syndrome. Approximately 30% of people with BAV will develop complications during their lifetime, including aortic regurgitation, aortic stenosis, and dilation of the proximal aorta in adulthood. These problems are rare early in life; only 2% of adolescents with BAV will develop aortic stenosis or regurgitation.

The presence of a murmur does not necessarily mean that the patient will need exercise restriction, but the BP does suggest coarctation of the aorta, which would require restriction. Although the murmur is soft and the gradient across the aortic valve is likely to be mild, this finding is not the most important in this patient because of concern for concomitant coarctation.

This patient will have a 30% risk of developing complications of BAV, rather than 75%. Endocarditis prophylaxis is no longer recommended for patients with BAV. Endocarditis prophylaxis is recommended only for those patients with unrepaired cyanotic congenital heart disease, repaired congenital heart disease which utilize prosthetic material, or those with residual defects or cardiac transplant with valvular disease.

PREP Pearls
•   Bicuspid aortic valves may not be symptomatic in childhood, but frequently become so later in life.
•   Bicuspid aortic valves are associated with coarctation of the aorta.

ABP Content Specifications(s)
•   Understand the natural history of a bicuspid aortic valve

Suggested Readings
•   Beekman RH III. Coarctation of the aorta. Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. 8th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2012: 1044-1060.

Question 93

You are seeing a 4-year-old girl in your office for a sick visit. Her parents are concerned that the girl is having difficulty walking up stairs and has been falling more than usual. She has been complaining of pain in her thighs, especially when walking long distances. The girl had coldlike symptoms 2 weeks ago and developed a rash on her hands and elbows 2 days ago. The parents deny any sick contacts, travel history, or animal exposures. The remainder of the review of systems is unremarkable. On physical examination, you note erythematous, hyperkeratotic, flat papules overlying her metacarpophalangeal joints and elbows bilaterally. Strength testing is 3/5 with hip flexion and 4/5 with knee flexion and extension bilaterally. There is no upper extremity weakness. Her reflexes and sensation are normal.

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

A.   creatinine phosphokinase testing

B.   edrophonium testing

C.   electromyography

D.   genetic testing of the dystrophin gene

E.   magnetic resonance imaging of the spine

Question 93   **Preferred Response: A**

The girl in the vignette has signs and symptoms consistent with juvenile dermatomyositis (JDM). The rash and proximal muscle weakness are characteristic of JDM. The best next step in evaluating this patient is to look for signs of muscle inflammation with muscle enzyme testing.

The clinical presentation of JDM affects several different systems (Item C93). Children often present with constitutional signs including fever, fatigue, malaise, anorexia, and weight loss. Musculoskeletal involvement usually presents as proximal muscle weakness and pain, with weakness occurring predominantly in the lower limb girdle and lower extremities making it difficult to climb stairs or walk. Gowers and Trendelenburg signs are often present. Weakness can affect the anterior neck flexor and back muscles, making holding up the head and overhead arm use difficult. Dysphonia and nasal speech can be an ominous sign, indicating pharyngeal, hypopharyngeal, and palatal muscle weakness that places the patient at risk for aspiration. Children may have arthralgia or even a mild arthritis, and some develop contractures because of myofascial inflammation.

Most patients will develop the pathognomonic Gottron papules and heliotrope discoloration of the eyelids. Gottron papules are erythematous to violaceous, sometimes scaling, flat-topped lesions located over the extensor surfaces of interphalangeal or metacarpophalangeal joints. The heliotrope rash is often accompanied by eyelid swelling.

Some patients present with a malar type rash. There is often dilation of the capillary loops of the nailfold with corresponding cuticle overgrowth. Lipodystrophy may occur. Cutaneous vasculitic ulcerations are commonly seen at the corners of the eyes, in the axillae and at pressure points. Calcinosis may occur in the muscle or subcutaneous layer, and can be debilitating. Some patients experience visceral vasculopathy, which can cause severe abdominal pain, melena, and hematemesis, and can be life threatening. Although serious cardiac involvement is rare, conduction defects have been associated with mortality. Respiratory muscle weakness is also rare, but can cause restrictive pulmonary disease. Osteopenia and osteoporosis are associated with JDM independent of treatment with corticosteroids.

Laboratory evaluation for suspected JDM should include markers of serum sarcoplasmic muscle enzymes including creatine phosphokinase (CPK), aspartate transaminase (AST), alanine aminotransferase (ALT), aldolase, and lactic acid dehydrogenase (LDH) levels. Elevated levels of these enzymes are evidence of cellular breakdown. Because AST and ALT elevations can be caused by muscle or liver disorders, simultaneously obtaining CPK and gamma-glutamyl transferase (GGT) levels can help determine the origin. CPK and AST are usually elevated 20 to 40 times normal early in the disease, though they will occasionally be normal in the acute phase. Generally CPK rises first, and aldolase and LDH are last to increase. LDH level can correlate with disease activity. Additional useful laboratory tests include C-reactive protein (CRP) level and erythrocyte sedimentation rate (ESR), markers for inflammation that can help differentiate an inflammatory myopathy, like JDM, from other myopathies. The presence of factor VIII–related antigen or von Willebrand factor antigen would be evidence of endothelial cell damage

associated with vasculitis. Neopterin, a marker of activated monocytes and macrophages, is increased with inflammation, infection, or malignant disease.

Several autoantibodies are associated with JDM. Rheumatoid factor is usually negative. The evidence regarding antinuclear antibody (ANA) positivity is widely variable, with studies demonstrating between 10% and 85% positivity in JDM. Myositis-specific autoantibodies (MSAs) and myositis-associated antibodies (MAAs) are more commonly positive in adult JDM patients.

Muscle biopsy is the gold standard in diagnosis, and the typical findings in dermatomyositis are necrosis, phagocytosis, perifascicular atrophy, fiber size variation, and perivascular inflammation. However, muscle involvement is patchy and pathology can be missed if a good biopsy site is not identified.

Edrophonium testing is used for diagnosing myasthenia gravis. This diagnosis is unlikely for the girl in the vignette, who has a distinctive rash and does not have progressive weakness or signs of muscle involvement other than her legs.

Electromyography (EMG) can be useful in the diagnosis of JDM, but is invasive and can be difficult in children. Although EMG will help to localize the problem at the nerve or muscle level, it would not be the next step in testing. The dystrophin gene is used to test for muscular dystrophy. The patient in the vignette has an inflammatory muscle disease with rash, weakness, and abnormal inflammatory markers that would not be seen in muscular dystrophy.

Magnetic resonance imaging (MRI) is often used to identify areas of myositis or to determine a biopsy site. MRI of the musculature has largely replaced electromyography, because it is less invasive. MRI of the spine would be appropriate if there were a concern for spinal cord pathology. The girl in the vignette did not have any changes in sensation or sensory level consistent with spinal cord compression, and her family did not report urinary or stool incontinence.

PREP Pearls
- Creatine phosphokinase, aspartate transaminase, alanine aminotransferase, aldolase, and lactic acid dehydrogenase levels are usually elevated in juvenile dermatomyositis.
- The gold standard for diagnosing juvenile dermatomyositis is muscle biopsy.
- Proximal muscle weakness is the pattern of muscle involvement associated with juvenile dermatomyositis.

ABP Content Specifications(s)
- Recognize the clinical findings associated with dermatomyositis
- Plan the appropriate laboratory evaluation for dermatomyositis

Suggested Readings

- Khan S, Christopher-Stine L. Polymyositis, dermatomyositis, and autoimmune necrotizing myopathy: Clinical Features.Rheum Dis Clin North Am. 2011; 37:143- 158. DOI: http://dx.doi.org/10.1016/j.rdc.2011.01.001.

Question 94

A 13-year-old obese adolescent presents to your office for evaluation of left anterior knee pain. The pain began 2 or 3 weeks before this visit and had an insidious onset. You observe that the patient has a slight limp and out-toed gait on the left side. He denies history of trauma, knee swelling, knee instability, and catching or locking of the knee joint.

Of the following, the MOST important element to include in this patient's physical examination is

A.   evaluation of hip rotation

B.   palpation of the medial joint line

C.   palpation of the patellar facets

D.   to assess anterior translation of the tibia relative to the femur with the patient supine and the knee flexed to 30 degrees (Lachman maneuver)

E.   to load the tibia onto the femur and rotate the lower leg with the patient in a prone position and the knee flexed to 90 degrees (Apley grind test)

Question 94   **Preferred Response: A**

Hip pathology can present as pain referred to the knee. Slipped capital femoral epiphysis (SCFE) should be considered in an obese early adolescent boy with insidious-onset knee pain such as the boy in the vignette. Children with SCFE generally hold the affected hip(s) in an externally rotated position and have limited internal rotation. The patient in the vignette is an obese peripubertal child with anterior knee pain and an out-toed gait (caused by obligate external hip rotation). This scenario should prompt evaluation for SCFE.

With SCFE, the femoral head epiphysis "slips" away from the femoral neck. The peak incidence of SCFE is at ages 12 to 15 years in boys and ages 10 to 13 years in girls. Although approximately 70% of affected children are above the 80th percentile for weight, pediatric providers should bear in mind that 30% of patients with SCFE are not overweight. SCFE is bilateral in about 40% of cases. In some cases, patients present with bilateral symptoms whereas in others, SCFE develops in the contralateral hip subsequently. SCFE is classified as acute in patients with symptoms of less than 3 weeks' duration and chronic in patients with symptoms lasting 3 weeks or more. SCFE can be further classified as stable (when patients can bear weight) or unstable. Children with SCFE typically present with a painful limp but some children will not have pain. Because comorbid endocrine abnormalities (eg, hypothyroidism, panhypopituitarism) exist in 5% of patients with SCFE, patients should be asked about systemic symptoms. On physical examination, obesity and decreased hip internal rotation are the most common findings. Hip radiographs typically show widening of the physis on anterior-posterior view. On the lateral view, a "Klein line" drawn along the superior border of the femoral neck should intersect the femoral head, and should be symmetric with comparison of both hips. The absence of these features is diagnostic of SCFE. Magnetic resonance imaging (MRI) should be considered for patients with normal radiographs and a high index of suspicion for SCFE. This may help detect a "preslip" in which the MRI may demonstrate bone marrow edema adjacent to the proximal femoral physis or slight separation at the physis.

Patients with SCFE should not bear weight and should be referred to an orthopedic specialist for urgent surgery. Internal fixation of the femoral head with screws is the most common treatment. Bilateral surgery is generally performed in patients with bilateral symptoms or in the presence of risk factors (eg, body mass index >35, young age) for a subsequent contralateral slip. Patients with SCFE have an increased risk of developing osteoarthritis; the risk increases with a higher degree of slippage.

Although complete physical examination of the knee is important in patients presenting with knee pain, the boy in the vignette has classic signs and symptoms of SCFE, therefore assessing hip rotation would be the most pressing option. This boy has anterior knee pain of insidious onset. Palpation of the medial joint line would be most useful to assess for a medial meniscal tear, which typically results from an acute injury in a young patient. Tenderness noted on patellar facet palpation could indicate patellofemoral pain syndrome (PFPS). Although PFPS would be in the differential diagnosis of anterior knee pain, this syndrome would not result in an out-toed gait; detection of PFPS is less urgent than detection of SCFE. The Apley grind test is used to assess for a meniscal tear and the Lachman test is used for an anterior collateral ligament (ACL)

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injury. Meniscal and ACL tears occur as a result of an acute mechanism of injury in this age range, but the patient in the vignette reports insidious onset of pain.

PREP Pearls
- Hip pathology can present as pain referred to the knee.
- Obligate external hip rotation is a classic finding with slipped capital femoral epiphysis (SCFE) and can present as an out-toed gait.
- Thirty percent of patients with SCFE are not overweight.
- A child with suspected SCFE should not bear weight on the affected extremity and should be referred for urgent evaluation.

ABP Content Specifications(s)
- Recognize the clinical findings associated with slipped capital femoral epiphysis, and plan appropriate management

Question 95

A previously healthy 12-year-old boy complains of worsening vision in the left eye over the last several weeks without other symptoms. His visual acuity is 20/80 in the left eye and 20/20 in the right eye. Fundoscopic examination of the left eye reveals a chorioretinal scar with surrounding retinitis nasal to the disk and mild overlying vitritis (Item Q95). The right eye is normal.

Item Q95
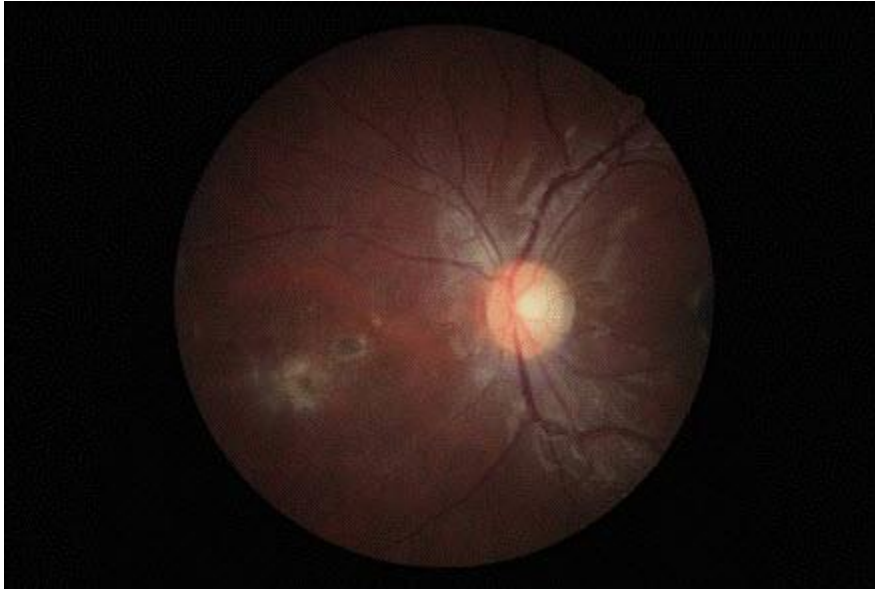
Fundoscopic examination of the left eye for the boy described in the vignette. Courtesy of V. Chen. Reprinted with permission from Pickering LK, et al. Red Book® Online. Elk Grove Village, IL: American Academy of Pediatrics.

Of the following, the test MOST likely to establish the cause of the patient's findings is

A.   blood culture for Treponema pallidum

B.   microscopy of corneal scraping for Acanthamoeba

C.   serum immunoglobulin G for Toxoplasma gondii

D.   serum polymerase chain reaction for cytomegalovirus

E.   swab of ocular mucous membranes for herpes simplex virus culture

Question 95   **Preferred Response: C**

The boy in the vignette has isolated ocular toxoplasmosis as evidenced by the characteristic retinal lesions (chorioretinitis) in an otherwise healthy child. The diagnosis is suggested by detecting immunoglobulin G antibody to T gondii in serum, but is based largely on the characteristic appearance of the chorioretinal lesion, usually described as white focal retinitis with overlying vitreous inflammation that has the appearance of a "headlight in the fog." In addition, an adjacent pigmented retinochoroidal scar also may be seen.

Isolated ocular toxoplasmosis develops in up to 85% of adolescents and young adults after untreated congenital infection. It also can become reactivated years after initial infection in both immunocompetent and immunocompromised individuals. Patients with ocular toxoplasmosis may complain of blurred vision, decreased acuity, epiphora (excessive tear formation), eye pain, floaters, photophobia, or scotoma. Chorioretinitis caused by Toxoplasma infection may lead to vision loss.

In most immunocompetent individuals, Toxoplasma infection either is asymptomatic or results in a self-limited and benign flulike illness. Rare complications include pneumonia, myocarditis, pericarditis, hepatitis, and cutaneous involvement. In patients with immunodeficiency, such as that caused by infection with human immunodeficiency virus, life-threatening manifestations of T gondii infection can include encephalitis, pneumonia, or disseminated disease. Infants with congenital toxoplasmosis are asymptomatic in the majority (70%-90%) of cases, but may develop sequelae such as learning disability, mental retardation, hearing loss, or vision impairment later in life. Infants with symptomatic congenital infection can present with various signs such as lymphadenopathy, rash, hepatosplenomegaly, petechiae, thrombocytopenia, pneumonitis, meningoencephalitis, and chorioretinitis. Although rare, the classic triad of hydrocephalus, cerebral calcifications, and chorioretinitis is highly suggestive of congenital infection caused by T gondii.

Toxoplasma gondii is found worldwide, and the seroprevalence (11% in the United States) of infection varies by geographic area and socioeconomic status. Felines are definitive hosts. Intermediate hosts such as pigs, sheep, and cattle can have tissue cysts in multiple organs. Humans become infected by ingestion of oocysts from soil (or cat feces) or contaminated food or water. Congenital infection results from primary maternal infection during gestation and is estimated to occur in 1 in 1,000 to 1 in 10,000 live births.

Most cases of acquired toxoplasmosis in immunocompetent hosts do not require antimicrobial therapy. Item C95 shows antimicrobial regimens for the treatment of toxoplasmosis, when indicated. Indications for treatment include pregnancy, immunocompromised status, severe symptoms (eg, chorioretinitis or organ damage), or persistent symptoms.

Item C95. Recommended Treatment for Toxoplasmosis.

	Drug	Pediatric Dosage
Drug of choice	Pyrimethamine[†]	2 mg/kg/day × 2 days, then 1 mg/kg/day (maximum 25 mg/day) × 3-6 weeks
	plus Sulfadiazine	100-200 mg/kg/day PO × 3-6 weeks
	or plus Clindamycin	5-7.5 mg/kg/day IV or PO in 3 or 4 divided doses (maximum 600 mg/dose)
	or plus Atovaquone	1,500 mg PO bid
	or plus Azithromycin	10 mg/kg/day
Alternative	Trimethoprim-sulfamethoxazole	15-20 mg/kg/day TMP plus 75-100 mg/kg/day SMX PO or IV in 3 or 4 doses

[†] Plus leucovorin 10-25 mg with each dose of pyrimethamine.

Adapted and reprinted with permission from Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. *Red Book: 2015 Report of the Committee on Infectious Diseases.* 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:954

The duration of therapy for congenital toxoplasmosis is prolonged, usually lasting approximately 1 year. For immunocompromised patients, long-term antibiotic suppression often is indicated after the initial course of therapy to prevent recurrence.

Ocular manifestations of infection with T pallidum (syphilis) can include uveitis (anterior or posterior) that usually is granulomatous. Posterior uveitis is more common and manifests as chorioretinitis that is multifocal, retinal necrosis, or optic neuritis. Patients usually have decreased visual acuity, often in association with syphilitic meningitis. The diagnosis is made serologically and not with blood culture.

Acanthamoeba causes keratitis that is indolent and can resemble keratitis caused by herpes simplex virus (HSV) or bacteria. Patients typically present with severe pain, photophobia, tearing, and the sensation of a foreign body in the eye. Classic findings include a stromal ring infiltrate and radial keratoneuritis. Diagnosis is made with corneal scrapings to identify the organism.

Cytomegalovirus retinitis is rare in immunocompetent individuals and usually involves the anterior chamber of the eye. The diagnosis can be made with polymerase chain reaction analysis of anterior chamber aqueous fluid. HSV can cause keratitis which presents as ocular pain, visual blurring, and discharge. Patients will have conjunctivitis, chemosis, dendritic lesions of the cornea, and decreased corneal sensation. The diagnosis usually is clinical, but viral culture from ocular mucous membranes may isolate the pathogen. HSV also can cause acute retinal necrosis, usually in immunocompromised individuals and pregnant women. Children presenting with decreased vision are reported to have bilateral acute retinal necrosis caused by HSV. Neonates can have chorioretinitis caused by HSV.

PREP Pearls
- In most immunocompetent individuals, Toxoplasma infection either is asymptomatic or results in a self-limited and benign flulike illness.
- In ocular toxoplasmosis, the chorioretinal lesion is described as white focal retinitis with overlying vitreous inflammation that has the appearance of a "headlight in the fog."
- Isolated ocular toxoplasmosis develops in up to 85% of adolescents and young adults after untreated congenital infection.
- Most cases of acquired toxoplasmosis in immunocompetent hosts do not require antimicrobial therapy.
- Pyrimethamine plus sulfadiazine are the drugs of choice for treating toxoplasmosis.

ABP Content Specifications(s)
- Identify the clinical features associated with congenital and acquired Toxoplasma gondii infestation, and manage appropriately
- Understand the epidemiology of Toxoplasma gondii

Suggested Readings
- American Academy of Pediatrics. Amebic meningoencephalitis and keratitis. In: Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:231-234.

Question 96

A 16-year-old high school football player presents to the pediatric emergency department with pain in his chest and left shoulder after an injury that he sustained during a football game, approximately 1 hour ago. While running and holding the football in his left arm, he was tackled from behind. The patient fell forward and landed forcefully on his chest, with the weight of the opposing player on top of him. At that time, he heard a "cracking" sound and immediately felt pain in his chest and left shoulder.

In the emergency department, the boy is alert and fully oriented. He is very uncomfortable and is holding his hand over the left side of his chest. He tells you that it is difficult to breathe. His vital signs include a heart rate of 100 beats/min, respiratory rate of 24 breaths/min, blood pressure of 130/80 mm Hg, temperature of 37°C, and pulse oximetry of 96% on room air. On physical examination, his breath sounds are clear and equal bilaterally. The patient is taking shallow breaths because of pain, but is not in respiratory distress. He is tender to palpation over the left sternoclavicular junction, as well as over his left first rib, and you note bruising over these areas. He has no focal tenderness on examination of his left shoulder, but he refuses to move his left shoulder due to pain. You find no evidence of trauma to his head and his cervical spine is nontender to palpation. The remainder of his physical examination findings, including a full neurologic examination, is unremarkable.

An electrocardiogram reveals sinus tachycardia with no other abnormality. You administer an intravenous analgesic and order plain radiographs of the patient's chest and left shoulder, which reveal a non-displaced fracture of his left first rib. His left shoulder radiograph reveals no fracture or dislocation. He continues to complain of severe pain over his left sternoclavicular joint and subjective dyspnea, and refuses to move his left shoulder due to pain. His vital signs are unchanged.

Of the following, the BEST next step in his evaluation is

A.   bone scan

B.   computed tomography of the chest

C.   echocardiography

D.   no further imaging

E.   plain radiographs of the ribs

Question 96   **Preferred Response: B**

The teenager in the vignette presents with pain in his left chest, subjective dyspnea, and refusal to move his left shoulder after sustaining a significant blunt traumatic force to his anterior chest wall during a football game. Plain radiography reveals that he has a fracture involving his left first rib. Computed tomography (CT) of the chest is the next best step in his evaluation.

Physical examination and/or plain radiography are sufficient to identify many chest wall injuries in children; however, CT of the chest may be needed to evaluate for bony injuries that are not apparent on plain radiography (eg, posterior sternoclavicular fracture or dislocation) and to investigate for associated intrathoracic injuries (eg, injury to the airway or great vessels). In general, stable children with fractures to the first rib and sternum should be evaluated with CT of the chest because of the high risk for associated intrathoracic injury.

It is important for all pediatric providers to know how to appropriately evaluate children who have sustained chest wall trauma. Pediatric chest wall injuries generally arise from blunt trauma to the thorax. The forceful mechanism of the trauma, such as a motor vehicle accident, can cause additional serious injuries. All children with chest wall injuries should undergo a full physical examination, beginning with assessment of their airway, breathing, circulatory, and neurologic status, and then progressing to a head-to-toe secondary survey after any life-threatening conditions have been addressed. It is important for clinicians to keep in mind that, because the rib cages of children are generally more pliable than those of adults, traumatic forces may be transmitted to their intrathoracic organs; thus, significant intrathoracic injuries such as pulmonary contusion may occur even in the absence of injury to the chest wall structures.

Chest wall injuries may include fractures of the ribs, sternum, clavicles, and scapulae, with rib fractures being the most common in children. Multiple rib fractures may result in flail chest, an uncommon but very serious injury that can cause respiratory insufficiency from a compromise to the structural integrity of the chest wall. Fractures of the upper ribs (especially the first and second), sternum, and scapulae should lead clinicians to consider more serious intrathoracic injuries such as pulmonary contusion, injury to the intrathoracic vessels, cervical spine injuries, and injuries to the trachea or esophagus. This concern is due to the large amount of force generally required to fracture these bones, given their protected anatomic positions.

Posterior dislocation of the sternoclavicular joint or posterior displacement of a medial clavicle fracture are chest wall injuries that are relatively rare, but do occur in children. These injuries can present with very subtle physical examination findings and are often not apparent on plain radiography. Furthermore, they carry the risk of associated injuries to intrathoracic structures including the great vessels, esophagus, and trachea. Physical examination findings in children with posterior sternoclavicular joint dislocations may include pain localized to the sternoclavicular joint, a palpable gap at the joint (which can be subtle), and swelling at the medial end of the clavicle. Clinical symptoms may include difficulty swallowing, shortness of breath, hoarseness, and inability to move the shoulder on the affected side. CT of the chest is the imaging study of choice for these injuries.

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A bone scan may identify fractures that are not apparent on plain radiography, particularly stress fractures, but would not be helpful in excluding injury to intrathoracic structures. Echocardiography certainly has a role in the evaluation of children who have sustained chest wall trauma, as a means of excluding cardiac injury. Echocardiography is indicated for those with abnormal electrocardiograms, ectopy, abnormal cardiac enzyme levels, or other evidence of cardiac injury after thoracic trauma. The boy in the vignette does not meet any of these criteria.

Many children with uncomplicated chest wall injuries, including those with simple clavicle fractures and fractures to the middle or lower ribs will not require further evaluation beyond plain radiography. However, the boy in the vignette has a clinical picture suggestive of a posterior sternoclavicular joint dislocation, and CT is indicated to evaluate for this injury. Furthermore, CT of the chest would be indicated in this patient because of the presence of the rib fracture.

Finally, plain radiography of the ribs would not aid in the evaluation for associated intrathoracic injuries or dislocation of the posterior sternoclavicular joint, which is indicated for the boy in the vignette. Standard anterior-posterior and lateral radiographs of the chest are sufficient for detecting the majority of rib fractures, therefore dedicated plain radiographs of the ribs are not needed in the evaluation of most patients.

PREP Pearls
- The thoracic cages of children are more pliable than those of adults, resulting in the transmission of traumatic forces to the intrathoracic organs; significant intrathoracic injuries may occur even in the absence of injury to the chest wall.
- Computed tomography (CT) of the chest may be indicated to evaluate for injuries to chest wall structures that are not apparent on plain radiography (eg, posterior sternoclavicular fracture dislocations) and to investigate for associated intrathoracic injuries (eg, airway or great vessels).
- In general, stable children with fractures to the first rib or sternum should be evaluated with CT because of the high risk for associated intrathoracic injury.

ABP Content Specifications(s)
- Plan the appropriate evaluation of a child who has experienced chest wall trauma

Suggested Readings
- Garg S, Alshameeri ZA, Wallace WA. Posterior sternoclavicular joint dislocation in a child: a case report with review of literature. J Shoulder Elbow Surg. 2012;21:e11-e16. DOI: http://dx.doi.org/10.1016/j.jse.2011.07.007.

Question 97

You are caring for a 600 g, 25-week gestation male in the intensive care unit. The neonate remains on a ventilator for respiratory distress syndrome 2 days after delivery. He is receiving light-emitting diode phototherapy for indirect hyperbilirubinemia (total bilirubin 6 mg/dL [102.6 µmol/L] and direct bilirubin 0.5 mg/dL [8.6 µmol/L]) while on an open warmer. Over the past 24 hours, the intravenous fluids have been increased to 140 mL/kg per day of intravenous dextrose 10% in water and his urine output has been 4 mL/kg per hour. Ultrasonography of the head reveals a right grade III intraventricular hemorrhage. The laboratory value trend over 48 hours is shown in Item Q97.

Item Q97. Laboratory data

	Birth	24 Hours	48 Hours
Sodium (mEq/L)	142	148	154
Potassium (mEq/L)	3.5	4.4	4.8
Chloride (mEq/L)	98	108	116
Bicarbonate (mEq/L)	20	19	15
Blood urea nitrogen (mg/dL)	12	21	42
Creatinine (mg/dL)	0.9	1.1	1.3
Glucose (mg/dL)	61	150	250

Of the following, the BEST next step in care for this neonate is to

A.   change the intravenous fluids to dextrose 5% in water

B.   discontinue phototherapy

C.   increase the intravenous fluid rate to 180 mL/kg per day

D.   initiate desmopressin

E.   place the neonate in a humidified isolette

Question 97   **Preferred Response: E**

The premature infant in the vignette has clinical evidence of excessive free water loss and would benefit from placement in a humidified isolette. The higher body surface–to-mass ratio of an infant compared to an older child contributes to insensible water loss. This is exaggerated before a gestational age of 28 weeks because of immaturity of the skin leading to evaporation and high transepidermal water loss.

Insensible water loss rates up to 200 mL/kg per day may be seen in extremely premature infants. If tape or topical agents do not compromise the integrity of the premature skin further, the skin will mature rapidly over the first week after birth and approach the insensible water loss rate of 20 mL/kg per day seen in a full-term infant. A premature infant on an open radiant warmer may have an increase of up to 50% in evaporative water loss. Placing the infant in a humidified isolette can markedly decrease these losses.

Full-term and late preterm infants typically require 60 to 80 mL/kg of fluid per day to meet their maintenance requirements and replace ongoing losses. The fluid requirement increases to 90 to 120 mL/kg of fluid per day for infants born before 28 weeks' gestational age, primarily because of the increased insensible losses from evaporative water loss. Caution must be used to avoid fluid overload in premature infants, as noted in some studies that demonstrated increased rates of bronchopulmonary dysplasia in infants with greater fluid intake.

The premature infant in the vignette has a complicated medical course. The increasing serum sodium, chloride, blood urea nitrogen, and creatinine all suggest excessive free water loss that would be improved by placement in a humidified isolette. Changing the intravenous fluid to dextrose 5% in water would drop the serum glucose but not provide additional free water. The infant requires treatment for hyperbilirubinemia and the light emitting diode phototherapy does not increase insensible water loss. Increasing the intravenous fluid rate of the dextrose 10% in water would give more free water, but worsen the infant's hyperglycemia and lead to an osmotic diuresis. Because diabetes insipidus is rarely seen with grade III intraventricular hemorrhage in the premature infant and the urine output is not excessive, initiation of desmopressin is not indicated.

PREP Pearls
- The high body surface–to-mass ratio and immaturity of the skin found in infants of less than 28 weeks' gestational age leads to high transepidermal water loss through evaporation.
- Evaporative water loss can be minimized in the premature infant by placement in a humidified isolette.
- Light-emitting diode phototherapy does not increase transepidermal water loss.

ABP Content Specifications(s)
- Recognize the differences in daily fluid requirements per kilogram of body weight in preterm and full-term infants

- Understand how prematurity and the use of radiant warmers affect insensible water loss, especially in preterm infants

Suggested Readings

- Dell KM. Fluids, electrolytes, and acid-base homeostasis. In: Martin RJ, Fanaroff AA, Walsh MC, eds. Fanaroff and Martin's Neonatal-Perinatal Medicine. 9th ed. St. Louis, MO: Elsevier Mosby; 2011:669-677 .

Question 98

A 10-year-old girl is brought to your office for possible seizures. her parents report that several times, as she started to laugh at something, the girl suddenly leaned over and her eyelids sagged for a few seconds. There was no loss or alteration in consciousness and no twitching or limb jerking. She has been falling asleep at school and her parents report that, on the weekends, the girl naps all afternoon. Recently, she chose to take a nap rather than attend a friend's birthday party. The girl appears to get good sleep at night, her hair and skin seem normal, she has not had any recent illnesses, and she does not have headaches. Her parents are not aware of any recent stressors or toxic exposures, and no one else in the family has similar symptoms. On physical examination, the girl weighs 51 kg (8 kg more than at her health supervision visit 3 months previously) her blood pressure is 98/74 mm Ho, heart rate is 92 beats/min, and respiratory rate is 32 breaths/min. Her physical examination including neurological examination is unremarkable

Of the following, the test MOST likely to yield the correct diagnosis is
   A. Depression screening
   B. Electroencephalogram
   C. Magnetic resonance imaging of the brain
   D. Multiple sleep latency test
   E. Thyroid stimulating hormone

Question 98   **Preferred Response: D**

The girl in the vignette has excessive daytime sleepiness and cataplexy. Cataplexy is the sudden loss of muscle tone provoked by strong emotion such as laughter or fright. These 2 symptoms together suggest narcolepsy, which is diagnosed on a multiple sleep latency test. Narcolepsy is characterized by excessive daytime sleepiness, cataplexy and hypnogogic or hypnopompic hallucinations (hallucinations when falling asleep or waking up, respectively). Weight gain is common in children with new-onset narcolepsy. In children and adolescents, the presentation of narcolepsy can be subtle, with only 1 or 2 of these symptoms present.

In narcolepsy, a sleep study is diagnostic. Overnight polysomnography will confirm the absence of sleep apnea or other sleep disorders causing excessive daytime sleepiness, and the multiple sleep latency test conducted the next day as part of the sleep study will show the presence of rapid eye movement (REM) sleep during at least 2 daytime naps. Normally, naps are too short to allow the emergence of REM sleep.

The differential diagnosis for excessive daytime sleepiness includes depression, disorders of attention, absence seizures, hypothyroidism, conversion disorder, Epstein-Barr virus infection, hypothalamic or pituitary tumors, and other more rare disorders. The girl in the vignette does not have signs or symptoms of any of these disorders. In addition, none of these disorders are characterized by cataplexy, which she does have.

Symptoms of cataplexy can be differentiated from atonic seizures ("drop attacks") and absence seizures because cataplexy is provoked by strong emotion such as laughter or sudden fright. Absence seizures can be provoked by prolonged hyperventilation but not by sudden emotion, and atonic seizures are unprovoked.

The presence in a child of excessive daytime sleepiness, weight gain, and cataplexy should prompt the clinician to consider narcolepsy and order polysomnography with a multiple sleep latency test to make the diagnosis.

PREP Pearls
- The presence of excessive daytime sleepiness, weight gain, and cataplexy should prompt the clinician to consider narcolepsy.
- Cataplexy, the sudden loss of muscle tone provoked by strong emotion such as laughter or fright, suggests a diagnosis of narcolepsy.
- Narcolepsy is diagnosed by polysomnography with a multiple sleep latency test.

ABP Content Specifications(s)
- Recognize the clinical features of narcolepsy and manage appropriately

Suggested Readings
- Martin S. Index of suspicion. Pediatr Rev. 2009;30(3):107-113. DOI: http://dx.doi.org/10.1542/pir.30-3-107.

- Rosen GM, Bendel AE, Neglia JP, Moertel CL, Mahowald M. Sleep in children with neoplasms of the central nervous system: case review of 14 children. Pediatrics. 2002;112(1)e46-e54. Available athttp://pediatrics.aappublications.org/content/112/1/e46.full.

Question 99

An 11-year-old boy has been growing at the third percentile in the last 4 years of age (Item Q99). His parents are concerned because both of their heights are at the 25th percentile and they are worried he will not catch up. The boy is otherwise healthy. He denies headaches or abdominal pain. The boy's bone age is 12 months delayed from his chronologic age. His parents request further evaluation to identify the cause of his poor growth.

Item Q99. Growth chart for the patient described in the vignette.
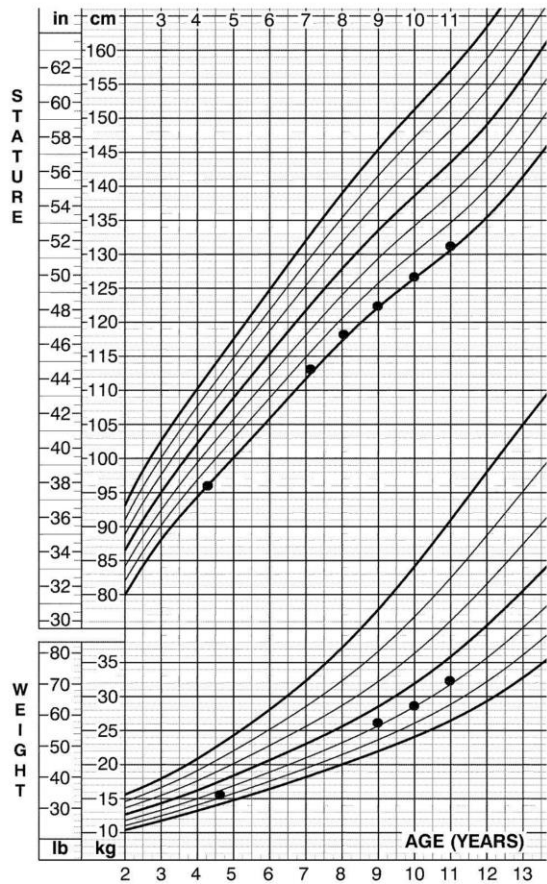
Courtesy of Jordan Pinsker, MD.

Of the following, the BEST next step in the evaluation of this child is to

A.   obtain an insulin like growth factor 1 level

B.   obtain a morning growth hormone level

C.   obtain thyroid-stimulating hormone and free thyroxine levels

D.   order a sella and pituitary magnetic resonance imaging

E.   reassure parents that no further testing is needed

Question 99   **Preferred Response: E**

The child in the vignette is growing at a normal prepubertal growth rate of 5 cm/year (normal 4-7 cm/year) and has a bone age within 2 standard deviations of his chronologic age. Thus for a patient who is tracking at the third percentile, his final height is expected to be at the third percentile (for a man, this is just above 5'4"). Given his normal linear growth velocity and predicted final height, reassurance is appropriate and no further testing is needed. It is important to note that in 2003 the US Food and Drug Administration approved growth hormone (GH) treatment for idiopathic short stature (ISS) defining the condition as the height of an individual greater than or equal to 2.25 standard deviation score (SDS) below the corresponding mean height for a given age, sex, and population group, without indications that catch-up growth would occur. This approximates the 1.2 percentile. The child in the vignette is too tall to meet the criteria defined for treatment of ISS.

In cases where laboratory evaluation for short stature is warranted, measurement of insulin like growth factor 1 (IGF-1) and/or insulin like growth factor binding protein 3 (IGF-BP 3) could be used to assess the GH axis. Random or AM GH levels are generally not helpful, because GH levels peak in deep sleep and are often very low during the day. Thus IGF levels are a better test, and GH stimulation testing can be used if IGF levels are low. Measurement of thyroid-stimulating hormone and free thyroxine may be used to evaluate for hypothyroidism, a common cause of poor linear growth.

If GH deficiency is suspected, or there is concern for another pituitary finding such as a tumor, cranial magnetic resonance imaging (MRI) can contribute significantly to diagnosis. In some studies, more than 25% of children with GH deficiency will have an abnormal finding on MRI, including pituitary hypoplasia, empty sella, hypoplastic anterior pituitary, missing pituitary stalk, and ectopic posterior pituitary. Children with abnormal MRI findings tend to have more severe characteristics of GH deficiency. However, in children such as the boy in the vignette, linear growth is normal with a normal predicted final height, so no further laboratory or radiologic assessment is needed.

PREP Pearls
• An endocrine cause of poor growth is likely when linear growth slows down or arrests while weight gain is either normal or increasing.
• In children with normal growth velocity and normal final height prediction, reassurance is appropriate and no further evaluation is needed.

ABP Content Specifications(s)
• Plan an appropriate diagnostic evaluation to differentiate constitutional growth delay and other conditions causing growth delay

Suggested Readings
• Maghnie M, Lindberg A, Koltowska-Häggström M, Ranke MB. Magnetic resonance imaging of CNS in 15,043 children with GH deficiency in KIGS (Pfizer International

Growth Database). Eur J Endocrinol. 2013;168(2):211-7.
DOI:http://dx.doi.org/10.1530/EJE-12-0801.

Question 100

A 15-year-old adolescent is brought to your office with a complaint of chronic cough. He has had a cough for "as long as he can remember." He has been treated with short courses of antibiotics for bronchitis and with systemic steroids, inhaled corticosteroid, and short acting β-agonist therapy for asthma without reported improvement. His cough is wet and productive of greenish sputum without blood. He is moderately short of breath with exertion. He has had poor weight gain with malodorous stools and has been frustrated by his inability to gain muscle mass with weight lifting.

There is no history of recurrent ear or sinus infections. The patient was born at term without perinatal or neonatal respiratory complications.

On physical examination, you note a thin, alert adolescent in no acute distress. His respiratory rate is 24 breaths/min. Auscultation of lungs reveals bilateral and diffuse coarse crackles without wheezing. A chest radiograph shows a diffuse bilateral increase in interstitial markings with notable peribronchial thickening. There is no focal infiltrate or effusion. Pulmonary function testing is obtained, which confirms moderate obstructive lung disease with a forced expiratory volume in 1 second of 58% predicted.

A computed tomography of the chest is obtained and a representative image is shown (Item Q100).

Item Q100
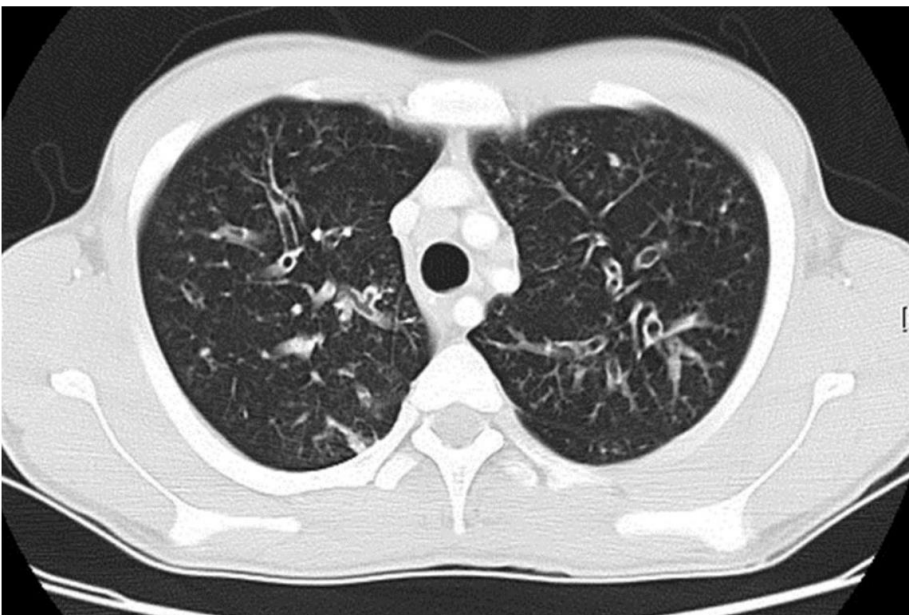
Chest computed tomography scan for the boy described in the vignette. Courtesy of M Nevin

Of the following, the MOST likely etiology of this patient's respiratory complaints is

A.   α-1 antitrypsin disease

B.   cystic fibrosis

C.   hypogammaglobulinemia

D.   Kartagener syndrome

E.   sarcoidosis

Question 100   **Preferred Response: B**

The patient in this vignette has a chronic cough that is productive of purulent sputum and has a computed tomographic (CT) scan showing bronchiectasis. In addition, he demonstrates poor weight gain and stool malodor. These findings are highly suggestive of a diagnosis of cystic fibrosis (CF). Cystic fibrosis is classically diagnosed in infancy, and is now a part of routine newborn screening in all states. It may also be detected in early childhood, but there are many examples of late diagnoses with mild disease mutations and atypical disease.

A chest radiograph may reveal bronchiectasis, but it is insensitive for detection of early disease. In the appropriate clinical setting, high resolution CT is indicated for definitive diagnosis of bronchiectasis. The CT image shown for the patient described in the vignette demonstrates the "signet ring" sign that is pathognomonic for bronchiectasis. Bronchiectasis occurs when there is dilation of the airway and thickening of the airway wall. The radiographic findings are generally representative of permanent airway damage and scarring caused by chronic and severe inflammation.

Focal "traction" bronchiectasis may be found in individuals with pneumonia. Traction bronchiectasis is described when the dilatation of airways occurs within areas of altered lung architecture. This may occur with a consolidative and necrotizing pneumonic infiltrate, but is classically associated with pulmonary fibrosis, where parenchymal scar creates a "pull" on the airway wall and creates distortion. Peripheral bronchi, which lack cartilaginous support, are primarily affected.

Central bronchiectasis may be seen in patients with allergic bronchopulmonary aspergillosis. More diffuse bronchiectasis may be seen in patients with immunodeficiency, chronic aspiration, or in patients with CF or ciliary dyskinesia syndromes (primary ciliary dyskinesia, Kartagener syndrome). Although bronchiectasis is seen in individuals with α-1 antitrypsin disease, the onset of lung disease typically occurs late in the second decade of life and is generally preceded by hepatic involvement. In this patient without recurrent ear, sinus, and skin infections, immunodeficiency is less likely. Although this patient may have ciliary dyskinesia, the presence of the point of maximal cardiac impulses on the left side suggests that he does not have situs inversus totalis, and therefore does not meet the criteria for a diagnosis of Kartagener syndrome. In addition, the patient in this vignette had no significant neonatal history, while the majority of children with ciliary defects will have transient neonatal respiratory distress. Finally, sarcoidosis is unlikely, based on the absence of pulmonary granulomatous disease and lymphadenopathy.

PREP Pearls
- Bronchiectasis generally denotes irreversible airway damage from chronic inflammation or infection.
- Bronchiectasis may occur early in life in children with cystic fibrosis.
- Bronchiectasis causes decrements in lung function and is associated with decreased survival.

ABP Content Specifications(s)
- Plan the appropriate diagnostic evaluation of suspected bronchiectasis
- Formulate a differential diagnosis of bronchiectasis

Suggested Readings
- Redding GJ. Bronchiectasis in children. Pediatr Clin North Am. 2009;56(1):157-171. doi:http://dx.doi.org/10.1016/j.pcl.2008.10.014.
- Sly PD, Gangell CL, Chen L, et al. Risk factors for bronchiectasis in children with cystic fibrosis. N Engl J Med. 2013;368(21):1963-1970. doi: http://dx.doi.org/10.1056/NEJMoa1301725.

Question 101

A 4-day-old newborn presents for his newborn check. His mother, who has not breastfed before, reports that she is having a lot of pain with breastfeeding despite assistance from a lactation consultant. She has heard that tongue-tie can cause breastfeeding problems and wonders whether the baby needs to have his frenulum clipped.

Of the following, the MOST appropriate information to provide this mother is that

   A.   ankyloglossia is common, affecting about one-quarter of all babies

   B.   frenulotomy may be useful in improving maternal nipple pain during breastfeeding

   C.   strong evidence exists that newborn frenulotomy improves the baby's long term speech outcome

   D.   there are reliable tools for determining which infants may benefit from frenulotomy

   E.   there is clear evidence that frenulotomy is not helpful in supporting breastfeeding

Question 101   **Preferred Response: B**

Ankyloglossia, also known as tongue-tie, has long been considered a possible cause of breastfeeding, speech, oral hygiene, and psychological difficulties for either mother or infant. Unfortunately, there is limited objective data as to how often or severe these effects are or whether division of tongue-tie resolves these problems. A literature review concluded that frenulotomy can facilitate breastfeeding, improve milk transfer, and decrease maternal nipple pain for many affected infants and mothers. A recent controlled study demonstrated a statistically significant decrease in maternal nipple pain scores immediately after frenulotomy, compared to mothers of infants who underwent a sham procedure. Interestingly, a commentator recently raised concerns about the ethics of doing a procedure on one patient (the infant) for the benefit of another (the mother experiencing nipple pain). Overall, while much of the evidence in support of frenulotomy is based on subjective data, the majority of studies do demonstrate benefit from frenulotomy for carefully selected infant-mother pairs.

One of the great difficulties in assessing the effects of frenulotomy is in determining which patients have clinically significant issues related to ankyloglossia. Tongue-tie is an anatomic variant that affects approximately 2% to 5% of infants with a male predominance. However, probably no more than half of affected infants have difficulty with breastfeeding, and even fewer will have future problems with speech development. Some lactation consultants and researchers have used the Hazelbaker Assessment Tool for Lingual Frenulum Function to evaluate tongue and frenulum appearance and tongue function with good inter-rater reliability for appearance, but not for all of the function factors. Other researchers have used ultrasonography to assess tongue function pre- and postfrenulotomy. Neither tool is universally available or utilized. The need for intervention is based on more subjective criteria. Most authors recommend a maximal trial of conservative interventions to support breastfeeding before considering frenulotomy, but there are no accepted guidelines as to if or when intervention should occur.

To date, there is not strong evidence to support a relationship between ankyloglossia and later speech problems. In one study, there was no statistically significant difference in speech articulation between controls and children with ankyloglossia who did or did not have a frenulotomy during infancy.

PREP Pearls
- Ankyloglossia is an anatomic variant present in 2% to 5% of newborns with male predominance.
- Frenulotomy can facilitate breastfeeding, improve milk transfer, and decrease maternal nipple pain for many affected infants and mothers.
- There is no widely accepted method to assure detection of those infants with ankyloglossia who will benefit from frenulotomy.

ABP Content Specifications(s)
- Plan the most appropriate management of a short lingual frenulum

Suggested Readings

• Buryk M, Bloom D, Shope T. Efficacy of neonatal release of ankyloglossia: a randomized trial. Pediatrics. 2011;128:280-288. DOI: http://dx.doi.org/10.1542/peds.2011-0077.

Question 102

You are seeing a 15-year-old adolescent for her annual health supervision visit. During the assessment, she reveals that she is sexually active. She reports 2 lifetime sexual partners. Both partners were male. She reports unprotected vaginal and anal sex. Her last menstrual period was 2 weeks ago. Her physical examination is unremarkable. Her urine pregnancy test is negative and screens for sexually transmitted infections are pending.

Of the following, the MOST appropriate counseling would be to

    A.   discuss the importance of dual protection with hormonal contraceptives and barrier methods

    B.   discuss the need for an initial Papanicolaou test to screen for cervical dysplasia

    C.   discuss the need for the patient to have her partners screened for sexually transmitted infections

    D.   explain to the patient that the age of sexual consent is 16 years of age

    E.   explain to the patient that unprotected anal sex does not place her at risk for HIV acquisition

Question 102   **Preferred Response: A**

There are several established guidelines for the provision of preventive services for adolescents. Bright Futures is an initiative of the American Academy of Pediatrics (AAP) that provides guidance and tools for pediatric and adolescent health supervision visits. Bright Futures recommends that sexuality is addressed at each period of adolescence starting at the early adolescent visit. Taking a sexual history allows the clinician to identify individuals at risk for acquisition of sexually transmitted infections (STIs) or unintended pregnancy, as well as sites for anatomic testing and guide risk reduction counseling. The US Centers for Disease Control and Prevention provides a tool to assist clinicians with sexual history taking that emphasizes the 5 Ps: partners, practices, protection from pregnancy, protection from STIs, and past history of STIs.

Sexually active adolescents should receive screening and counseling for sexually transmitted infections and pregnancy prevention. In July 2014, the AAP Committee on Adolescence and the Society for Adolescent Health and Medicine issued the policy statement Screening for Nonviral Sexually Transmitted Infections in Adolescents and Young Adults, which recommends gonorrhea and chlamydia screening for all sexually active female adolescents and young adults, sexually active adolescent and young adult males who have sex with males, and other males from high prevalence areas. The United States Preventive Services Task Force (USPSTF) recommends HIV testing for individuals 15 years of age and older, with subsequent testing based on risk.

Sexually active adolescents should be counseled about contraceptive options. The American College of Obstetricians and Gynecologists and the AAP recommend that adolescents are counseled on the first line use of long acting, reversible contraceptives (eg, intrauterine contraceptives, contraceptive implants) for pregnancy prevention. Additionally, it is imperative to counsel adolescents on the dual use of hormonal contraceptives with barrier methods for the prevention of pregnancy and STIs, and is recommended for the adolescent in this vignette.

The USPSTF recommends cervical cancer screening for women ages 21 to 65 years with cytology (Papanicolaou smear) every 3 years or every 5 years for women ages 30 to 65 years, with a combination of cytology and human papillomavirus testing. The USPSTF specifically recommends against screening for women younger than 21 years of age.

While screening for STIs in sexual partners may be important, it does not negate the need for use of dual protection for the prevention of STIs and unintended pregnancy. Unprotected anal sex is a significant risk factor for HIV acquisition. The age of sexual consent varies by state.

PREP Pearls
•   Sexuality should be addressed during the health supervision visit at each period of adolescence, starting at the early adolescent visit.
•   Taking a sexual history allows the clinician to identify individuals at risk for acquisition of sexually transmitted infections or unintended pregnancy, as well as sites for anatomic testing and guide risk reduction counseling.

American academy of pediatrics   330

- The American Academy of Pediatrics Committee recommends gonorrhea and chlamydia screening for all sexually active female adolescents and young adults, sexually active adolescent and young adult males who have sex with males, and other males from high prevalence areas.
- Adolescents should be counseled on the dual use of hormonal contraceptives with barrier methods for the prevention of pregnancy and sexually transmitted infections.
- The United States Preventive Services Task Force recommends HIV testing for all individuals 15 years of age and older, with subsequent testing based on risk.

ABP Content Specifications(s)
- Provide appropriate counseling with regard to contraception and prevention of sexually transmitted infection for an adolescent engaging in vaginal and/or anal intercourse

Suggested Readings
- American Academy of Pediatrics, Society For Adolescent Health And Medicine. Screening for nonviral sexually transmitted infections in adolescents and young adults. Pediatrics. 2014;134(1):e302-e311. DOI:http://dx.doi.org/10.1542/peds.2014-1024.

Question 103

During the health maintenance examination of a 6-month-old female infant, you note a diaper rash. There is erythema involving the convexities of the thighs and suprapubic area; the skin folds are not involved (Item Q103). Her physical examination is otherwise unremarkable.

Item Q103: Diaper rash for the patient described in the vignette.
Courtesy of D. Krowchuk.

Of the following, the MOST appropriate treatment for this infant's rash is topical

A.   clindamycin

B.   clotrimazole

C.   nystatin

D.   triamcinolone/nystatin

E.   zinc oxide

Question 103   **Preferred Response: E**

The 6-month-old female infant described in the vignette has a diaper rash characterized by erythema and chafing involving the convexities of the thighs and suprapubic area with sparing of the skin folds. This pattern of involvement is characteristic of irritant contact dermatitis (ICD), the most common form of diaper rash. Several factors contribute to the development of the eruption, including excessive moisture (that reduces the skin's ability to withstand frictional forces), frictional forces applied by the diaper, and the irritating effects of fecal enzymes. The most appropriate treatment for ICD is the application, at all diaper changes, of a barrier preparation, such as zinc oxide paste or petrolatum. In addition, parents may be advised to change diapers frequently. If ICD is moderate or severe, a low potency topical corticosteroid (eg, hydrocortisone 1% or 2.5%) may be applied twice daily. The perineal skin is thin and occluded by the diaper, therefore, the potential exists for enhanced absorption of a corticosteroid and the development of adverse effects, such as atrophy or striae. For these reasons, more potent agents should be avoided, including those contained in certain combination products (eg, triamcinolone/nystatin and betamethasone dipropionate/clotrimazole).

Folliculitis represents infection of hair follicles, usually with Staphylococcus aureus. In the diaper area, it is characterized by erythematous papules and pustules centered about follicles. The buttocks are commonly involved (Item C103A). In mild or localized forms of the infection, clindamycin may be applied topically. More extensive cases require an oral antistaphylococcal antibiotic.

Item C103A: Erythematous papules and pustules centered about follicles are characteristic of folliculitis. 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.

Infection of the diaper area with Candida albicans may occur primarily or complicate an existing ICD. The eruption is composed of "beefy-red" erythematous patches that involve the convexities and inguinal creases (Item C103B). Scaling at the periphery of patches, or satellite papules and pustules may be observed. Treatment of diaper candidiasis is with topical nystatin or an imidazole (eg, clotrimazole, ketoconazole, miconazole).
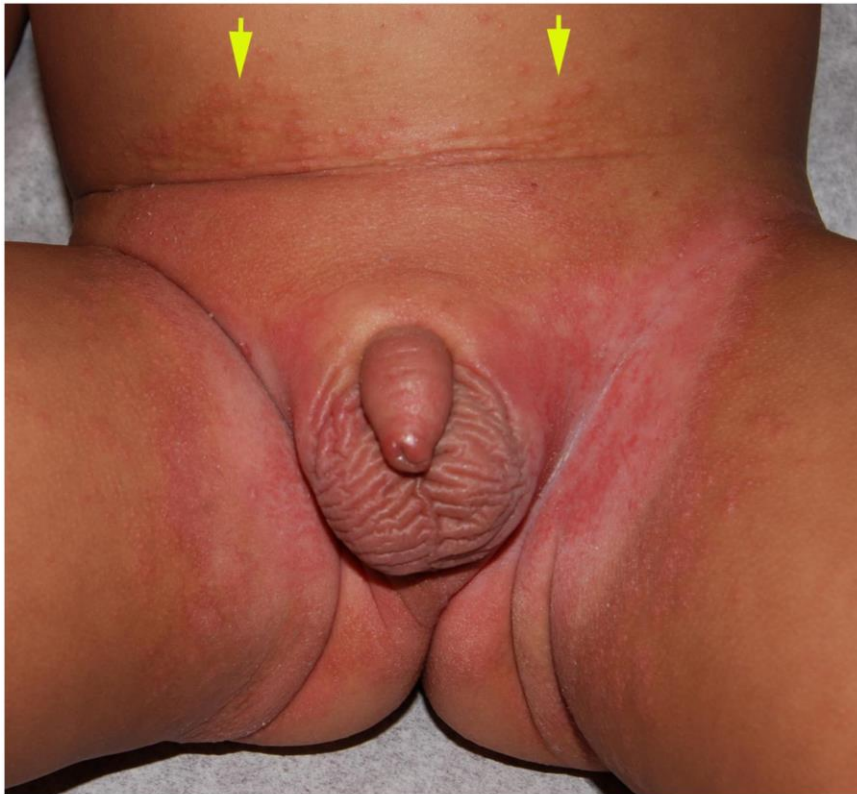
Item C103B. Candidiasis in the diaper area produces "beefy-red" erythematous patches that involve both the convexities and creases. Satellite papules may be observed (arrows). Reprinted with permission from Krowchuk DP. Diaper rash. In: McInerny TK, Adam HM, Campbell DE, Kamat DM, Kelleher KJ, eds. Textbook of Pediatric Care. 2nd ed. Elk Grove Village, IL: American Academy of Pediatrics.

PREP Pearls
- The most common forms of diaper rash are irritant contact dermatitis, candidiasis, and seborrheic dermatitis. Irritant contact dermatitis involves the convexities, but spares the folds, while candidiasis and seborrheic dermatitis involve the convexities and folds. Candidiasis often is bright red, while seborrheic dermatitis appears orange-pink.
- Consider referral to a pediatric dermatologist if a diaper rash does not resolve with appropriate therapy.
- Refer to a pediatric dermatologist infant who have atypical findings (eg, petechiae, papules with hemorrhagic crust), as these may be signs of Langerhans cell histiocytosis.

ABP Content Specifications(s)
- Recognize the etiology of diaper dermatitis, and manage appropriately

Suggested Readings
- Krowchuk DP, Mancini AJ. Diaper dermatitis. Pediatric Dermatology: A Quick Reference Guide. 2nd ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012:501-509.

Question 104

A 12-year-old boy is brought to the emergency department after being involved in a house fire. He is currently awake, alert, and oriented. He is complaining of throat and chest pain. His temperature is 37°C, pulse is 110 beats/min, respiratory rate is 24 breaths/min, and blood pressure is 115/70 mm Hg. His oxygen saturation by pulse oximetry is 100% on room air. Physical examination shows an emotionally upset child in no apparent physical distress. There is no apparent trauma to the head, trunk, or extremities. There is soot around his mouth, nose, and the exposed skin on his neck, hands, and feet. When the soot is washed away, there is no redness or burn to any area of skin. The boy's oropharynx is erythematous. He is breathing comfortably. On auscultation, his lungs are clear with transmitted upper airway sounds, and his heart has a regular rhythm. The boy's extremities are warm and well-perfused. His abdomen is soft, nontender, and non-distended. His pupils are equal, round, and reactive. The remainder of his neurologic examination is unremarkable.

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

A.   dexamethasone, 10 mg intravenously

B.   endotracheal intubation

C.   hyperbaric oxygen therapy

D.   methylene blue, 1 mg/kg intravenously

E.   noninvasive positive pressure ventilation

Question 104   **Preferred Response: B**

The child in this vignette is a victim of a house fire. Although his clinical condition is relatively benign at the time of presentation, there are signs of smoke inhalation. Because airway injury caused by smoke inhalation can be rapidly progressive and life-threatening, endotracheal intubation is the best management option listed.

Inhaled heated air alone rarely causes sufficient physical burn to cause airway compromise. However, smoke has a higher heat capacity because of suspended particulate material, and can thus distribute thermal injury to the upper airway. Significant airway injury can occur without evidence of burns to the skin because the airway is relatively less resistant to thermal injury. Because airway resistance is inversely proportional to the radius of the airway to the fourth power, there is exponentially more airway resistance in younger children or with decreasing inner diameter of the airway as the disease process worsens. Thus, if a child has a history and any sign of smoke exposure, the airway should be evaluated immediately. This holds true even if the child looks well, because if the airway edema worsens while the disease process declares itself, endotracheal intubation becomes more immediately lifesaving but also more technically difficult. Although the child in this vignette looks clinically well, he displays signs of smoke inhalation that could portend progression of airway edema, such as soot around the mouth and nose, transmitted upper airway sounds, and an erythematous oropharynx. Other signs of smoke inhalation not present in this child include singed nasal hairs, soot-stained secretions, and hoarseness.

The best modality for airway evaluation is direct laryngoscopy and bronchoscopy by an airway expert. However, if personnel are not readily available, endotracheal intubation is recommended. Dexamethasone can help reduce airway inflammation and swelling, but is not likely to prevent injury. Although controversial, hyperbaric oxygen therapy can be used to mitigate hypoxic injury in carbon monoxide poisoning from house fires, but is not recommended in a child with a normal neurologic status. Methylene blue can be used to treat methemoglobinemia, which can occur in victims of house fires, but will not be helpful in treating airway injury. Noninvasive positive pressure ventilation can be helpful in cases of upper airway obstruction caused by decreased airway tone, but not from airway swelling.

PREP Pearls
- A child with smoke inhalation may be at a higher risk for airway edema than predicted by clinical appearance.
- The airway of a child with smoke inhalation should be evaluated immediately because airway edema can worsen rapidly and also lead to a technically difficult intubation.
- Signs of smoke inhalation in children include soot around the mouth and nose, pharyngeal erythema, singed nasal hairs, soot-stained secretions, and hoarseness.

ABP Content Specifications(s)
- Recognize the clinical findings associated with airway injury in a patient with an acute burn

Suggested Readings

- Fidowski CW, Fuzaylov G, Sheridan RL, Coté CJ. Inhalation burn injury in children. Pediatr Anaesth. 2009;19(suppl 1):147-154. DOI: http://dx.doi.org/10.1111/j.1460-9592.2008.02884.x

Question 105

A 13-month-old girl is hospitalized with fever and pancytopenia. She has a history of an orthotopic liver transplant for biliary atresia. She currently is on tacrolimus and prednisone for rejection prophylaxis and trimethoprim-sulfamethoxazole for pneumocystis prophylaxis. Vital signs show a temperature of 38.8°C, respiratory rate of 44 breaths/min, heart rate of 130 beats/min, blood pressure of 121/67 mm Hg, and weight of 9.85 kg. On physical examination, she is irritable, mottled and has several erythematous macules over her face. Her abdomen is distended and you note hepatosplenomegaly. Laboratory data shows:

- White blood cells, 1,110/µL (1.1 x 10^9/L)
- Hemoglobin, 7.5 g/dL (75 g/L)
- Platelets, 42 x 10^3/µL (42 x 10^9/L)
- Differential, 1% segmented neutrophils, 18% bands, 78% lymphocytes, 2% monocytes, 1% eosinophils
- Aspartate aminotransferase, 90 U/L
- Alanine aminotransferase, 61 U/L
- Human herpesvirus 6 polymerase chain reaction, 261,000 copies/mL
- Tacrolimus level 11.5 ng/mL

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

A.   human herpesvirus 6

B.   liver transplant

C.   prednisone

D.   tacrolimus

E.   trimethoprim-sulfamethoxazole

Question 105 **Preferred Response: A**

The child in this vignette has had a liver transplant and is on immune suppressive therapy. She has fever, an erythematous macular rash, and hepatomegaly, along with pancytopenia, elevated hepatic transaminases, and a human herpesvirus 6 (HHV-6) polymerase chain reaction with a high copy level, indicating infection with HHV-6. The clinical manifestations of HHV-6 infections can include an acute febrile illness, roseola (exanthem subitum) (Item C105), and neurologic manifestations including febrile seizures and encephalitis. Additional features can include cervical and occipital lymphadenopathy and symptoms attributable to the gastrointestinal and respiratory systems. In transplant recipients, manifestations can include fever, rash, hepatitis, bone marrow suppression, pneumonia, and encephalitis. Additionally, graft rejection can occur.

Item C105: Roseola as seen in human herpesvirus 6 infections.

Courtey of G. Williams. Reprinted with permission from Pickering LK et al. RedBook Online. Elk Grove Villange, IL: American Academy of Pediatrics.

By 2 years of age, nearly all children have been exposed to HHV-6. The seroprevalence in adults is as high as 95%. Human herpesvirus 6 establishes latency and can reactivate in the setting of immune suppression, such as after solid organ transplantation. The incidence of reactivation peaks at 2 to 4 weeks after transplantation. While reactivation of HHV-6 is common, clinical illness ascribable to HHV-6 is not, occurring in only 1% of solid organ transplant recipients.

Prednisone would not be expected to cause pancytopenia. Tacrolimus can cause leukopenia, anemia, and thrombocytopenia. However, the child in this vignette has therapeutic levels and additional symptoms that cannot be explained by receipt of tacrolimus. Similarly, while trimethoprim-sulfamethoxazole can cause bone marrow suppression, it cannot explain all of the findings manifested by the patient in the vignette.

PREP Pearls
- Human herpesvirus 6 (HHV-6) is the causative agent of roseola (exanthema subitum), a classic childhood exanthem characterized by a period of high fevers followed by a generalized maculopapular rash.
- Like other Herpesviridae, HHV-6 establishes latency after primary infection and can reactivate, especially in the setting of immune suppression.
- The manifestations of HHV-6 infections in transplant recipients can include fever, rash, hepatitis, bone marrow suppression, pneumonia, encephalitis, and graft rejection.

ABP Content Specifications(s)
- Recognize the clinical features associated with human herpesvirus type 6 infection

Suggested Readings
- American Academy of Pediatrics. Human herpesvirus 6 (including roseola) and 7. In: Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015: 449-451.

Question 106

You are evaluating a 4-month-old infant in the emergency room for vomiting and decreased oral intake. The baby was born to a 29-year-old primigravida woman at 39 weeks of gestation via normal vaginal delivery. The baby's birth weight was 3.6 kg (50th percentile) and had a length of 50 cm (50th percentile). The mother mentions that his pediatrician has been concerned about his weight gain for the last month. His vital signs show a weight of 5.0 kg (less than the third percentile), length is 55.2 cm (less than the third percentile), temperature of 38.0°C, heart rate of 126 beats/min, respiratory rate of 30 breaths/min, and a blood pressure of 70/60 mm Hg. His physical examination is significant for mild dehydration, excessive watering of the eyes, photophobia, frontal bossing, and wrist widening.

His urinalysis today demonstrates a specific gravity of 1.005, a pH of 7.0, 3+ glucose, and no blood, protein, leukocyte esterase, or nitrites.

Of the following, the MOST likely cause of the patient's findings is

A.   Bartter syndrome

B.   Gitelman syndrome

C.   renal tubular acidosis (type 1)

D.   renal tubular acidosis (type 2)

E.   renal tubular acidosis (type 4)

Question 106   **Preferred Response:D**

The patient described in the vignette was at the 50th percentile for height and weight at birth and is currently less than the third percentile for both weight and height. The decline in weight gain across 2 or more major percentiles is suggestive of failure to thrive in the patient. The history of concerns for the patient's growth, wrist widening, frontal bossing (suggestive of rickets), photophobia, and excessive watering of the eyes also suggests an underlying cause for the patient's failure to thrive. The diagnosis of renal tubular acidosis (RTA) should be considered in a young infant with failure to thrive, recurrent vomiting, rickets, episodes of dehydration, and investigations showing persistent metabolic acidosis, hypokalemia, and nephrolithiasis. Renal tubular acidosis is an inherited or acquired defect in the ability of the kidneys to absorb filtered bicarbonate or excrete ammonia. Renal tubular acidosis is characterized by a normal anion gap metabolic acidosis. Distal (type 1) RTA, proximal (type 2) RTA, mixed (type 3; features of both type 1 and 2) RTA, and hypoaldosteronism (type 4) are the 4 different forms of RTA seen in patients.

The presence of glycosuria, rickets, and photophobia, along with excessive watering of the eyes, is indicative of Fanconi syndrome in a patient with RTA and is most commonly associated with proximal (type 2) RTA, as demonstrated in this infant. Fanconi syndrome, characterized by generalized proximal tubular dysfunction as evidenced by rickets (phosphaturia leading to hypophosphatemic rickets), glycosuria (dipstick positive glycosuria with normal plasma glucose concentration), and aminoaciduria or tubular proteinuria (urine dipstick negative for protein and quantitative urine tests positive for amino acids and protein). Other RTAs (type 1 or type 4) or Bartter and Gitelman syndromes are not associated with Fanconi syndrome.

Renal tubular acidosis occurs from the inability of the kidneys to keep up with the excretion of the daily endogenous acid production arising from dietary proteins and amino acids. The renal response to endogenous acids leads to maintenance of normal plasma HCO_3^-, the extracellular buffer. Urinary ammonia (NH_3) and monohydrogen phosphate (HPO_4^{2-}) are the 2 principal buffers binding the endogenously produced H^+ ions. Of these, increased urinary ammonia excretion is the main adaptive response (can be increased by 10-fold) to increased acid excretion, as urinary phosphate excretion is fixed. Therefore, all forms of RTA are associated with metabolic acidosis (low plasma HCO_3^-) and decreased renal ammonium (NH_4^+) excretion. Ammonia excretion by the kidneys can be explained as a 4-step process, each of which is implicated in different types of RTA. Ammonium (NH_4^+) is produced by the proximal tubule from glutamine and secreted by the proximal tubule into the tubular fluid via Na^+-H^+ exchanger by binding to the H^+ site. This step primarily generates the NH_4^+, which is subsequently processed in the renal tubule to regeneration of new $HCO3^-$ and also contributes to restoration of the bicarbonate pool. The next step is the absorption of NH_4^+ by the thick ascending loop of Henle via the Na-K-2Cl channel by occupying the K site. Ammonium in the cell dissociates into NH_3 and H^+. The apical membrane of thick ascending loop of Henle is impermeable to NH_3, while the basolateral membrane is highly permeable. This leads to increased movement of NH_3 across the basolateral membrane into the medullary interstitium and a gradient of increasing concentration of NH3 in the deeper medulla similar to the countercurrent system. This is the third step in normal ammonium secretion by the kidney. The fourth step involves diffusion of

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NH_3 into the cortical collecting duct, wherein it reacts with H^+ ions secreted by the intercalated A cells. This H^+ secretion (proton donated by H_2CO_3, which dissociates into HCO_3^- and H^+) leads to the addition of a new HCO_3^- molecule to the circulation (Item C106A).
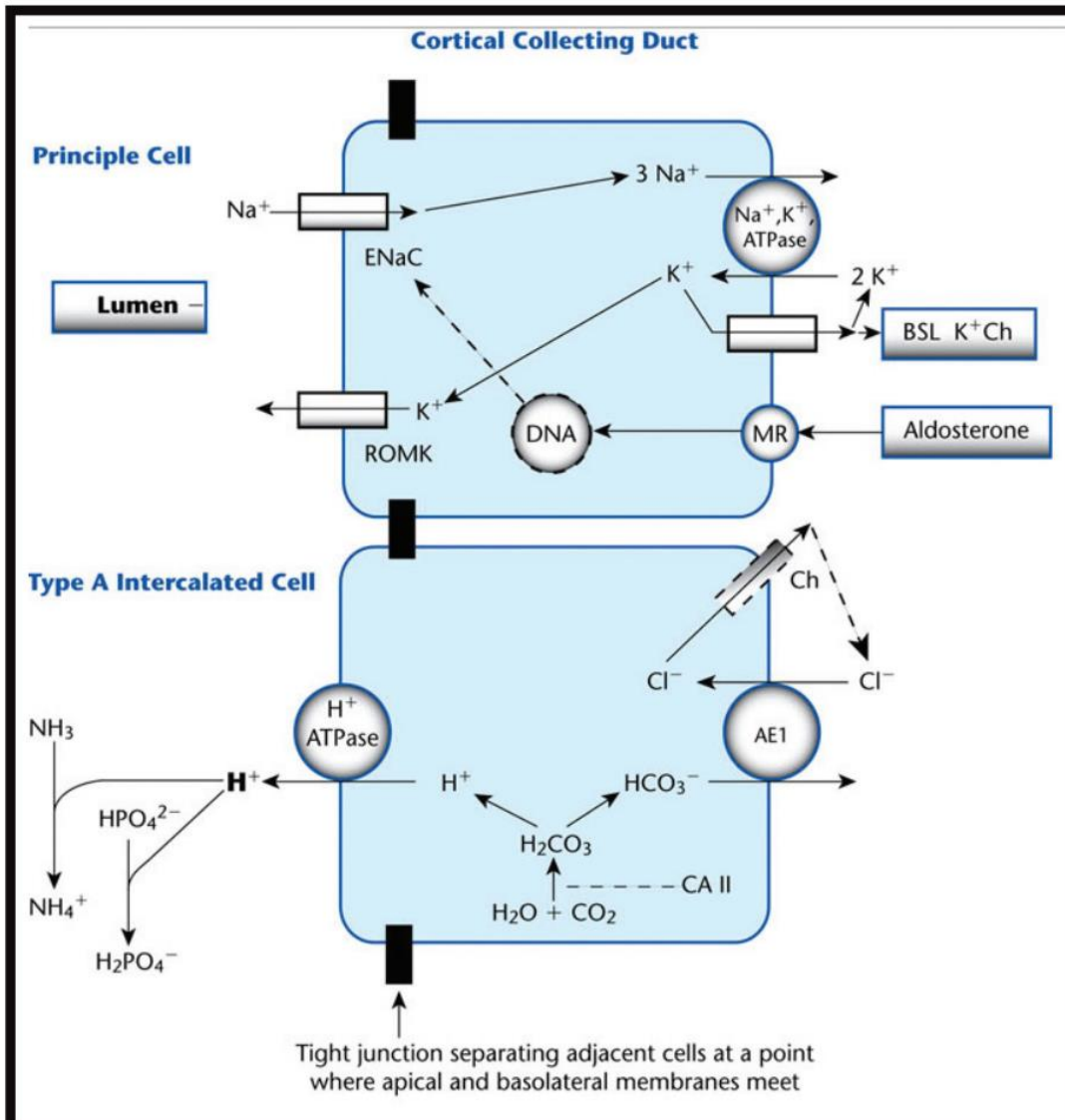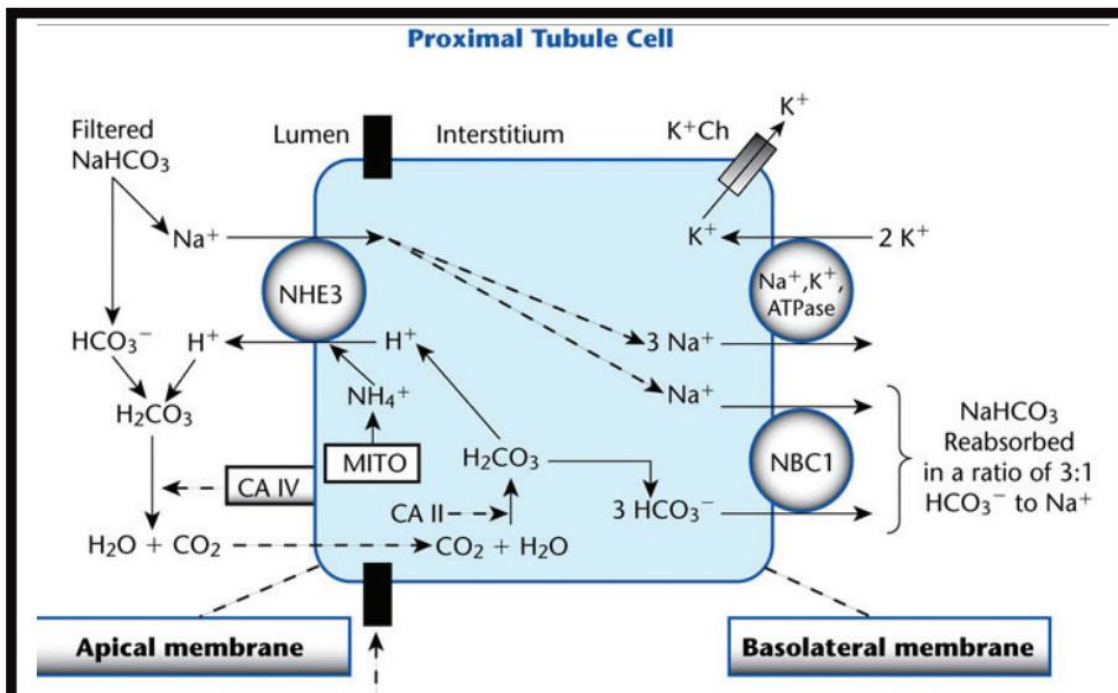

Reprinted with permission from Kallen RJ. Renal tubular acidosis. In: McInerny TK, Adam HM, Campbell DE, Kamat DM, Kelleher KJ, eds. *American Academy of Pediatrics Textbook of Pediatric Care.* Elk Grove Village, IL: American Academy of Pediatrics; 2009:2463

ITEM C106A: Diffusion in the cortical collecting duct.

The kidneys maintain acid base balance in vivo by: 1) reclaiming the filtered preformed HCO_3^- in the tubular fluid 2) regeneration of new HCO_3^- to account for HCO_3^- lost in buffering of the endogenous acids

Eighty percent to 90% of the filtered bicarbonate is reclaimed by the proximal tubule and the remaining by the thick ascending loop of Henle. Reclamation is an appropriate term to describe the processes of the proximal tubule, as the HCO_3^- molecule returning to the circulation is different from the filtered HCO_3^- (Item C106B). Proximal RTA is a defect in the ability of the proximal renal tubules to reclaim filtered HCO_3^- from the tubular fluid. Fanconi syndrome, a form of proximal RTA, has been associated with cystinosis, galactosemia, tyrosinemia, oculocerebrorenal (Lowe) syndrome, and hereditary fructose intolerance. The presence of excessive eye watering and photophobia is indicative of cystinosis as the cause of the patient's symptoms and RTA. A slit lamp examination will reveal the characteristic cysteine crystals and confirm the diagnosis. In proximal RTA, once the serum bicarbonate has dropped to a level (around 12-15 mEq/L [12-15 mmol/L]) wherein the distal renal tubules can absorb the filtered bicarbonate load, the urine can be appropriately acidified and the presenting urine pH may be less than 5.5.

Reprinted with permission from Kallen RJ. Renal tubular acidosis. In: McInerny TK, Adam HM, Campbell DE, Kamat DM, Kelleher KJ, eds. *American Academy of Pediatrics Textbook of Pediatric Care*. Elk Grove Village, IL: American Academy of Pediatrics; 2009:2461

ITEM C106B: Proximal tubule cell.

Impaired regeneration of new HCO_3^- is indicative of a decreased rate of NH_4^+ ammonium (predominant urinary buffer) secretion by the kidney. This may be associated with normal serum potassium (distal type 1 RTA) or elevated serum potassium (hyperkalemic type 4 RTA). Impaired H+ ion secretion (step 4 of ammonium generation) by the cortical collecting duct leads to type 1 RTA. Distal or type 1 RTA is associated with failure to thrive, polyuria, hypokalemia, and medullary nephrocalcinosis (caused by hypercalciuria and hypocitraturia). Hyperkalemic RTA is most frequently seen in patients with chronic renal failure, associated with renal parenchymal injury and scarring. Mineralocorticoid deficiency (decreased production or receptor insensitivity due to genetic or acquired causes) leads to hyperkalemia, which impairs steps 1, 2, and 4 of ammonia generation, leading to type 4 RTA.

Type 3 RTA is rare and associated with marble bone disease. It has findings of both type 1 (impaired regeneration of HCO_3^-) and type 2 (bicarbonate wasting) RTA. Treatment of RTA involves correction of metabolic acidosis via sodium or potassium citrate solutions. Potassium citrate solutions are avoided in hyperkalemic RTA. As the distal tubules are responsible for regulating only 5% to 10% of the acid load, the alkali dose is higher for proximal (10-20 mEq/kg per day), as compared to distal or hyperkalemic (5-8 mEq/kg per day) RTA.

Bartter syndrome and Gitelman syndrome are characterized by hypokalemia and metabolic alkalosis. Bartter syndrome often presents in childhood with growth retardation, hypokalemia, metabolic alkalosis, and polyuria or polydipsia. Bartter syndrome is caused by a primary defect in sodium chloride reabsorption in the medullary thick ascending limb of the loop of Henle, similar to chronic furosemide therapy. Gitelman syndrome generally presents in late childhood or adulthood with muscle cramps (hypokalemia), polyuria, or polydipsia. Mutations in the gene encoding for the thiazide-sensitive Na-Cl transporter in the distal tubule have been identified in patients with Gitelman syndrome. In contrast to Bartter syndrome, patients with Gitelman syndrome have reduced urinary calcium and hypomagnesemia (more common).

PREP Pearls
- The diagnosis of renal tubular acidosis (RTA) should be considered in a young infant with failure to thrive, recurrent vomiting, and episodes of dehydration.
- Proximal RTA is most commonly associated with Fanconi syndrome, characterized by generalized proximal tubular dysfunction, as evidenced by rickets, glycosuria, and aminoaciduria.
- Patients with cystinosis develop cystine deposits in the cornea and the conjunctiva, leading to photophobia, watering, and blepharospasm.
- Bartter syndrome and Gitelman syndrome are characterized by hypokalemia and metabolic alkalosis.

ABP Content Specifications(s)
- Recognize the clinical findings associated with various anomalies of the kidneys, urinary collecting system, and urinary excretion system

Suggested Readings

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- Chan JCM, Scheinman JI, Roth KS. Consultation with the specialist: renal tubular acidosis. Pediatr Rev. 2001;22(8):277-287. DOI: http://dx.doi.org/10.1542/pir.22-8-277.

Question 107

You are caring for a 13-year-old adolescent with a 4-month history of periumbilical pain and intermittent nausea with a normal, soft, nonbloody bowel movement daily. She denies a history of emesis, headaches, joint pain, and weight loss. She has had an extensive evaluation including:

- Normal abdominal radiograph without evidence of constipation
- Normal complete blood cell count, electrolytes, and liver panel
- Stool heme test is negative
- Celiac and Helicobacter pylori serum screens are negative

Of the following, the BEST treatment plan for this patient is

A.   acid blockade

B.   laxatives

C.   no medications

D.   probiotics

E.   tricyclic antidepressant

Question 107   **Preferred Response: C**

The 13-year-old female adolescent in this vignette has functional abdominal pain (FAP) with a negative evaluation for organic etiology. The current literature recommends supportive care with cognitive behavioral therapy. No medications have been shown to have significant benefit in children with functional abdominal pain.

The differential diagnosis for abdominal pain is extensive (Item C107A).

Item C107A. Differential Diagnosis of Abdominal Pain by Organ System.

Gastrointestinal
- Esophagitis (infectious, peptic, eosinophilic)
- Gastritis (infectious, peptic, eosinophilic)
- Ulcers
- Celiac disease
- Structural (malrotation, hernia, duplication, volvulus)
- Inflammatory bowel disease
- Ischemic bowel disease
- Colitis
- Constipation
- Foreign body
- Carbohydrate malabsorption
- Functional gastrointestinal disease (functional abdominal pain, abdominal migraine, irritable bowel syndrome, cyclic vomiting syndrome)

Hepatobiliary and Pancreatic
- Cholecystitis
- Cholelithiasis/biliary colic
- Gallbladder dysfunction (biliary dyskinesia, sphincter of Oddi dysfunction)
- Chronic hepatitis
- Chronic pancreatitis
- Structural etiology (choledochal cyst, annular pancreas)

Genitourinary
- Ureteropelvic junction obstruction, hydronephrosis
- Nephrolithiasis
- Recurrent pyelonephritis or cystitis
- Endometriosis
- Mittelschmerz
- Ovarian torsion
- Pelvic inflammatory disease
- Testicular torsion

Metabolic/Hematologic
- Porphyria
- Hereditary angioedema
- Sickle cell disease
- Collagen vascular disorder

Musculoskeletal
- Trauma
- Tumor
- Infection (Herpes zoster, bacterial)

Respiratory
- Infection

Other
- Mesenteric adenitis
- *Streptococcus* pharyngitis
- Henoch-Schönlein purpura

Modified from Wyllie and Hyams. Chronic abdominal pain of childhood and adolescence. In *Pediatric Gastrointestinal and Liver Disease.* 3rd edition. pages 111-125

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The Rome criteria are useful in establishing the diagnosis of FAP. Childhood functional abdominal pain is defined by the 2006 Rome III criteria as episodic or continuous abdominal pain with some loss of daily function and no or only occasional relationship to psychological stress. There must be insufficient criteria to meet the diagnosis of any other functional gastrointestinal disorder. The evaluation must be negative for evidence of inflammatory, anatomic, metabolic, or neoplastic process to explain the symptoms. The pain must occur at least weekly for 2 months. If the patient demonstrates any of the red flags for pathologic chronic abdominal pain (Item C107B), additional evaluation is required.

Item C107B. Red Flags for Pathologic Chronic Abdominal Pain.

- Weight loss

- Failure to thrive

- Delayed puberty

- Patient younger than 5 years of age

- Significant vomiting or diarrhea

- Gastrointestinal blood loss

- Unexplained fever(s)

- Constitutional symptoms (fever, joint symptoms, decreased energy, weight loss, recurrent oral ulcers)

- Nighttime symptoms awakening child from sleep

- Family history of inflammatory bowel disease, celiac disease, or peptic ulcer disease

- Consistent right upper quadrant or right lower quadrant pain

- Abnormal findings on physical examination

Courtesy of C Waasdorp Hurtado

Based upon the Rome III criteria and current literature, the evaluation for children with chronic abdominal pain should include a thorough history and physical examination looking for possible red flags. If no red flags are identified and the child meets the described diagnostic criteria, the

diagnosis of FAP is made. There is no longer a need to "rule out" other disease processes. If there are red flags or continued concerns for an organic etiology, the evaluation may include complete blood cell count, inflammatory markers, liver and pancreatic enzymes, infectious evaluation, and possible endoscopy with biopsy as clinically indicated.

Management of chronic abdominal pain is multifactorial. The therapy must be rooted in education, reassurance, and ongoing support for the patient and family with a discussion of how the diagnosis is made and why other diseases have been ruled out. It is important for the provider to acknowledge the pain that the child is experiencing is real. The goal is then to focus on function to include school attendance. A symptom diary can be very helpful in identifying triggers. Dietary recommendations focus on avoidance of triggers and limitation of artificial sweeteners that may increase gas. Several studies have demonstrated a reduction in abdominal pain with an increase in dietary fiber, with a goal of age of patient in years plus 5 g (13 + 5 in the case of the adolescent in this vignette).

The role of pharmacotherapy is limited in FAP, with placebo success rates being quite high in functional gastrointestinal disorders. Randomized controlled trials in pediatrics are lacking, therefore limiting recommendations for medications.

Psychological therapies are increasingly recommended for children with chronic abdominal pain and functional abdominal pain. This may include cognitive behavioral therapy, relaxation training, and hypnotherapy. Studies show that children utilizing cognitive behavioral therapy have a higher rate of resolution of pain.

The adolescent in this vignette has chronic periumbilical pain with intermittent nausea and no evidence of constipation. There are no red flags to raise concern for an organic etiology. The literature shows no consistent benefit to treatment with tricyclic antidepressants, probiotics, or acid blockade. With no evidence of constipation, the use of laxatives is not appropriate, however, this should be considered in a child with irritable bowel syndrome – constipation type.

PREP Pearls
- Cognitive behavioral therapy has been shown to be effective in children with functional gastrointestinal diseases to include functional abdominal pain.
- Pharmacotherapy has not been shown to have significant benefit in children with functional abdominal pain, although studies are very limited.
- If a child lacks any red flag symptoms, no significant workup is needed for diagnosis.

ABP Content Specifications(s)
- Plan appropriate evaluation of chronic recurrent abdominal pain
- Formulate an age-appropriate differential diagnosis of recurrent abdominal pain
- Recognize the clinical manifestations of chronic recurrent abdominal pain, and manage appropriately

<u>Suggested Readings</u>

- Gottsegen D. Complementary, holistic, and integrative medicine: recurrent abdominal pain. Pediatr Rev. 2010;31(5):e36-e39. DOI: http://dx.doi.org/10.1542/pir.31-5-e36.

Question 108

A 16-year-old adolescent presents for a health supervision visit. The father reports his son is an endearing, shy teenager with a mild reading disability. The adolescent's height is at the 85th percentile and his weight is at the 35th percentile. The physical examination is remarkable for an adolescent with a thin build, disproportionately long arms and legs, gynecomastia, and small testes.

Of the following, the MOST appropriate next step for his diagnosis is

A.   brain magnetic resonance imaging

B.   karyotype analysis

C.   serum prolactin

D.   serum testosterone

E.   testicular ultrasonography

Question 108   **Preferred Response: B**

The patient in this vignette has Klinefelter syndrome or 47,XXY syndrome, which is the most frequent chromosomal disorder in males, affecting 1 to 2 cases per 1,000 males (0.1%-0.2% of male newborns). Classic presentation includes older adolescents or young males with small testes, gynecomastia (38%), tall stature, long arms and legs, and possibly learning or behavioral difficulties. Only 10% of cases are diagnosed before puberty. It is suspected that only 25% of Klinefelter syndrome cases are actually diagnosed. Most present in adulthood due to infertility, hypergonadotropic hypogonadism, or sexual dysfunction. Almost all affected males have nonobstructive azoospermia (90%+), testicular fibrosis, and decreased fertility in association with their hypoandrogenism. It is diagnosed with a standard karyotype, which is the most appropriate next step in diagnosis of the adolescent male in this vignette. Classic laboratory abnormalities include a significantly elevated follicle-stimulating hormone concentration, increased luteinizing hormone concentration, and low testosterone levels in addition to the abnormal 47,XXY karyotype.

A multidisciplinary approach is best at managing the overall care of a patient with Klinefelter syndrome and could include an endocrinologist, urologist, geneticist, primary care physician, psychologist, psychiatrist, and a fertility specialist. Medication regimens during transition into adulthood focus on testosterone replacement therapy. Patients with Klinefelter syndrome are also prone to thyroid dysfunction, autoimmune diseases, dental caries, metabolic syndrome, varicose veins, thrombosis, and malignancy (breast cancer, lung cancer, lymphoma, and nonseminomatous germ cell tumors). They are not at increased risk for testicular cancer or prostate cancer compared to the general population.

From a behavioral standpoint, many patients with Klinefelter syndrome have a higher incidence of anxiety, depression, attention-deficit/hyperactivity disorder, autism spectrum disorders, substance abuse, or other psychiatric disorders. They tend to be introspective, shy, and sensitive. Infertility can lead to psychological concerns as well. A fertility counselor is quite helpful in discussing the options of testicular sperm extraction, artificial insemination with donor sperm, or adoption. From an educational standpoint, they tend to experience delayed verbal development and learning difficulties (75%).

The best diagnostic test to determine the etiology of a patient presenting with the clinical constellation described in this vignette is a karyotype. A low serum testosterone level and a testicular ultrasound showing testicular fibrosis would be informative, but not diagnostic. A serum prolactin would be helpful in a patient with galactorrhea, not gynecomastia, because of concerns for a secretory pituitary adenoma. Klinefelter patients do not have structural anomalies of the brain, so a magnetic resonance image of the brain would not be helpful.

PREP Pearls
- Klinefelter syndrome, or 47,XXY syndrome, is the most frequent chromosomal disorder in males affecting 1 to 2 patients per 1,000. Although diagnosis is easy, given the gynecomastia and small testes in affected males, Klinefelter syndrome is often missed and untreated until adulthood.
- The best diagnostic test for Klinefelter syndrome is a karyotype.
- Testosterone replacement therapy, as well as identification and management of the comorbid disorders, remain important determinants in positive outcomes for patients with Klinefelter syndrome.

ABP Content Specifications(s)
- Recognize the clinical features associated with Klinefelter syndrome

Suggested Readings
- Aksglaede L, Juul A. Testicular function and fertility in men with Klinefelter syndrome: a review. Eur J Endocrinol. 2013;168(4):R67-R76. doi: http://dx.doi.org/10.1530/EJE-12-0934 .

Question 109

The parents of a 2-day-old, 35-week gestation, male newborn request to speak with the pediatrician about routine circumcision. They are interested in having the procedure performed before discharge. However, they want to discuss whether current methods of analgesia are safe and effective.

Of the following, the MOST accurate statement to include when counseling this family is that

    A.   a dorsal penile nerve block is preferred over a subcutaneous ring block

    B.   oral sucrose is effective as the sole method of pain control

    C.   topical analgesic creams are as effective as a dorsal nerve or subcutaneous ring block

    D.   topical lidocaine cream has a slower onset of action than topical lidocaine-prilocaine cream

    E.   topical lidocaine-prilocaine can cause skin irritation, especially in low birth weight infants

Question 109   **Preferred Response: E**

It is important that pediatric health care providers who perform circumcisions are aware of the risks and benefits of each option for analgesia, as well as the contraindications to circumcision in the newborn period. The adverse effects of topical anesthetic creams are uncommon and are usually mild. However, low-birthweight infants have a higher incidence of skin irritation (erythema, swelling, or blistering) when topical creams are used.

Circumcision is the surgical removal of some, or all, of the foreskin (or prepuce) from the penis. This procedure yields specific health benefits that include prevention of urinary tract infections, decreased acquisition of human immunodeficiency virus, decreased transmission of sexually transmitted diseases, and a lower risk of penile cancer. Analgesia that is safe and effective in reducing the pain associated with newborn circumcision is available and should always be provided.

Both the dorsal penile nerve block and subcutaneous ring block are effective options for analgesia. Onset of the anesthestic effect occurs after approximately 7 minutes for both procedures. Nonpharmacologic techniques are not sufficient when used alone to manage the pain associated with circumcision. Comfortable positioning and oral sucrose may be used as adjunct therapies, but neither should be used as the sole method of analgesia. Topical lidocaine-prilocaine cream does attenuate circumcision pain, but is less effective than either the dorsal penile nerve block or subcutaneous ring block. In addition, there are case reports that suggest a rare risk of methemoglobinemia with lidocaine use. Topical 4% lidocaine has a faster onset of action than lidocaine-prilocaine cream (20-30 versus 60-90 minutes, respectively).

Contraindications to newborn circumcision include significant prematurity, medical instability, blood dyscrasia or family history of a bleeding disorder, and congenital penile abnormalities such as hypospadias or chordee. Complications of circumcision are usually minor and may include bleeding, infection, or a poor cosmetic outcome.

PREP Pearls
- Safe and effective analgesia should always be provided to reduce the pain associated with newborn circumcision.
- When used for circumcision, the adverse effects of topical anesthetic creams are uncommon (though more common in low-birthweight infants) and are usually limited to mild skin irritation.
- Both the dorsal penile nerve block and subcutaneous ring block are effective options for analgesia.
- Neither nonpharmacologic techniques nor topical analgesics are sufficient to manage the pain associated with circumcision.

ABP Content Specifications(s)
- Recognize the conditions in which circumcision is contraindicated
- Plan the appropriate use of analgesia when performing circumcision

Suggested Readings
- American Academy of Pediatrics Task Force on Circumcision. Male circumcision. Pediatrics. 2012;130(3):e756-e785. doi:http://dx.doi.org/10.1542/peds.2012-1990.

Question 110

You are caring for a 4-year-old girl hospitalized for newly diagnosed acute lymphoblastic leukemia. The child has a hemoglobin level of 6 g/dL (60 g/L) and has been tachycardic and fatigued. You order a transfusion of packed red blood cells. One hour into the transfusion, she develops a temperature to 38.5°C.

Of the following, the procedure that is MOST likely to have prevented this reaction is

A.   the administration of diphenhydramine prior to the transfusion

B.   culturing the blood product prior to the transfusion

C.   irradiating the blood product prior to the transfusion

D.   using a leukoreduced (filtered) blood product

E.   using only blood products that have had extended antigen phenotyping

Question 110   **Preferred Response: D**

The transfusion of blood products poses many risks, and children who are immune compromised or undergoing treatment for malignancies are at high risk for adverse transfusion events. The most common types of transfusion reactions include febrile, allergic, or anaphylactic. Blood product transfusions can also transmit infectious diseases, and in immune compromised hosts, can cause transfusion-associated graft-versus-host disease. Acute and delayed hemolytic transfusion reactions and transfusion-associated acute lung injury are rare but serious complications of transfusions.

The infusion of even small numbers of granulocytes can lead to the release of pro-inflammatory cytokines, thereby increasing the risk of a febrile transfusion reaction. In order to reduce this risk, granulocytes are removed from blood products at the time of initial processing or immediately prior to transfusion via filtration, a process called leukodepletion. For the girl in the vignette, leukodepletion of the red blood cells (RBC) prior to transfusion would have had the greatest likelihood of reducing her risk of fever.

Diphenhydramine reduces the risk of an allergic transfusion reaction through the blockade of histamine, but will not influence the risk of fever. While bacterial infections can certainly be transmitted through blood product transfusions and can lead to fevers, culturing the packed cells prior to transfusion is neither practical nor feasible. Once thawed, blood products need to be transfused prior to culture results being available.

Although granulocytes are removed through filtration, many lymphocytes are the size of RBC and are therefore not as effectively removed by filtration. While RBCs are typed by ABO typing, they are not human leukocyte antigen typed or matched to the recipient, and lymphocytes infused with a transfused product may recognize the recipient as foreign. In immune compromised hosts, the host immune system is unable to clear the infused donor lymphocytes, which may then undergo expansion in response to recognition of a foreign antigen and cause a severe, typically fatal, graft-versus-host reaction. In immune competent hosts, this does not occur, as the host immune system recognizes the donor lymphocytes as foreign and successfully eliminates them. Irradiation of blood products prior to transfusion renders donor lymphocytes replication incompetent, eliminating the threat of transfusion-associated graft-versus-host disease.

There is a marked discrepancy in the surface antigens commonly found on the RBCs of different subpopulations or ethnic groups and those commonly found on the RBCs in the general donor pool. These differences can lead to the development of alloantibodies to RBC antigens when transfused with RBCs, with surface antigens not routinely evaluated in routine RBC typing (minor RBC antigens). Patients with alloantibodies to minor antigens are at higher risk of hemolytic transfusion reactions when infused again with RBCs that have had a standard typing for only ABO and Rh antigens. It is important to perform extended spectrum RBC typing, especially in patients receiving chronic transfusions, to obtain the best possible match of RBC surface antigens between donor and recipient in order to reduce the risk of a hemolytic transfusion reaction. This is especially important in children with sickle cell disease.

PREP Pearls
- Leukodepletion removes granulocytes from blood products through filtration, thereby reducing the risk of a febrile transfusion reaction.
- Irradiation of blood products renders donor lymphocyte replication incompetent, thereby eliminating the risk of transfusion-associated graft-versus-host disease.
- Extended spectrum typing can improve the minor surface antigen match on red blood cells between the donor unit and the recipient, thereby reducing the risk of a hemolytic transfusion reaction.

ABP Content Specifications(s)
- Recognize complications associated with transfusion of blood products

Suggested Readings
- Bell MD. Consultation with the specialist: red blood cell transfusions. Pediatr Rev. 2007;28(8):299-304. doi:http://dx.doi.org/10.1542/pir.28-8-299.

Question 111

You are seeing a 1-year-old boy who had an episode earlier today that greatly concerned his parents. While playing with a toy, his older sister came by and took it away, which made him start to cry vigorously. After a couple of crying exhalations, his mother saw him get very quiet and "turn blue." Then his body became limp and he seemed to become "unconscious." The mother said that although it seemed longer at the time, he may have remained this way for about 20 to 30 seconds before he woke up, took a few big breaths, and seemed to "act like his usual self again."

Of the following, the BEST next step in care would be to

A.   arrange for an electroencephalogram to be performed

B.   perform a lumbar puncture

C.   provide reassurance to parents

D.   refer the child to see a cardiologist

E.   refer the child to see a neurologist

Question 111   **Preferred Response: C**

This 1-year-old child most likely has had a breath holding spell, which will happen at one point in time in up to 5% of children. A breath holding spell is a respiratory/autonomic reaction in a young child from the sudden experience of anger, frustration, or pain, typically happening within the first 15 seconds of starting to cry. Breath holding spells most commonly appear between the ages of 6 to 18 months, and when present, should disappear entirely by 5 years of age. During a spell, the child may hold their breath after an exhalation, or hold their breath after an inhalation. About 80% of breath holding spells are cyanotic, in which breath is held in expiration. Cyanotic breath holding spells may result from the sequence of an involuntary Valsalva maneuver during an initial strong cry that slows the venous return to the heart, which then decreases cardiac output to yield cerebral ischemia and unconsciousness. Recovery happens within a minute of falling unconscious by the child suddenly taking a few deep breaths and then looking normal again. The child in the vignette appears to have had a cyanotic breath holding spell (suddenly quiet, turned blue during an exhalation, then recovered quickly).

Less commonly, a breath holding spell will involve a sudden pallid or pale appearance and loss of muscle tone, typically after a painful experience, which looks like fainting. These pallid breath holding spells are thought to come from abnormal vagal responses to a sudden emotion. Occasionally, some tonic-clonic movements may occur, which if they do, may make it more difficult to differentiate from a seizure based on history alone. Breath holding spells are not a choice of the child, as in a child making a threat like "I'm going to hold my breath till I turn blue!", but rather they represent a sudden involuntary reaction of an immature nervous system. The hallmarks of a breath holding spell are that there is no loss of bowel or bladder control, no post-ictal state, and a sequence of events that include crying from a trigger of anger, frustration, or pain right before the incident happens. Normal consciousness and normal breathing are rapidly restored (within 1 minute). It is unusual, but not impossible, to see tonic-clonic movements. Diagnosis is based on the clinical history and reassurance can be provided because of their common and self-resolving nature. If these spells are happening frequently, a parent can be encouraged to video record an episode so that it can be reviewed with their provider to assure that the clinical assessment is correct.

There are no recommended treatments for breath holding spells. There have been occasional associations of breath holding spells with the presence of anemia or iron deficiency without anemia, therefore checking for the presence of anemia and obtaining iron studies is reasonable. Some have suggested that an electrocardiogram (EKG) be performed with all of these children because of the very rare risk of long QT syndrome. However, this is not necessary unless the reported symptom pattern is atypical (ie, no precipitating intense cry or emotional trigger, lasts longer than a minute). An EKG could be ordered for an extra degree of reassurance, however, referral to a cardiologist would only be appropriate if there were any abnormal findings on that study or on the physical examination.

An electroencephalogram (EEG) would be appropriate if the story of the child's symptoms were more consistent with a seizure disorder, such as appearing groggy or sluggish when the episode

ends. With the exception of an acute head injury, an EEG would not be appropriate. Performing a lumbar puncture to evaluate for a central nervous system infection would be appropriate if there was a fever, or persisting symptoms of illness such as listlessness or irritability. Given that the child was immediately "acting like his usual self again," a central nervous system infection is highly unlikely. Referral to a neurologist would also not be necessary, given the lack of any seizure hallmarks such as post-ictal states.

PREP Pearls
- Breath holding spells are nonvolitionally triggered by the child's sudden experience of anger, frustration, or pain
- Nearly 5% of children will experience a breath holding spell.
- Breath holding spells most commonly appear around 1 year of age, and if present, they should disappear entirely by 5 years of age.

ABP Content Specifications(s)
- Plan the appropriate management of breath-holding in toddlers and preschool-age children
- Recognize the clinical features of breath-holding and counsel parents appropriately

Suggested Readings
- Narchi H. The child who passes out. Pediatr Rev. 2000;21(11):384-388.http://pedsinreview.aappublications.org/content/21/11/384.

Question 112

A 7-week-old infant was noted to have a short, soft 2/6 systolic murmur at the left upper sternal border at 2 weeks of age and comes today for a follow-up visit. His mother has noticed that he has been breathing rapidly for the last few days. The baby has been breastfeeding less than usual for the last 2 days, taking feedings every 3 hours, and only for 5 min. When you enter the examination room, you see an infant breathing at 90 breaths/min. The saturation is 68% in room air taken on the right hand. The baby becomes fussy when you do your examination, but the saturations are stable even with crying. The heart rate is 160 beats/min and the blood pressure is 80/60 mm Hg in the left leg. Physical examination shows the chest is clear, the respirations are shallow but there are no retractions, and there is a long 4/6 systolic murmur at the left upper sternal border that begins at S1 and is present for all of systole. You do not appreciate an ejection click or any split to S2. There is no hepatosplenomegaly and the femoral pulses are 2+ bilaterally. You start oxygen in the office and the saturation increases to 75%. You call emergency medical services and warn the emergency department that the baby is on the way. You speak to the cardiologist on call.

Based on the clinical presentation and physical examination, the finding the echocardiogram is MOST likely to show is

A.   atrial septal defect

B.   coarctation of the aorta

C.   pulmonic stenosis

D.   tetralogy of Fallot

E.   ventricular septal defect

Question 112   **Preferred Response: C**

The physical examination in the infant in this vignette is most suggestive of severe pulmonary stenosis (PS). The murmur is very loud, persists throughout systole, and is in the pulmonic position. It starts early in systole and the ejection click that is often appreciated with moderate stenosis is not noted because it has merged with the first heart sound. If this patient had tetralogy of Fallot, the murmur would diminish as the pulmonary flow decreased, especially if this were a hypercyanotic spell. In that case, crying on the part of the infant, with a decrease in systemic vascular resistance, would increase the right to left shunt at the ventricular level and the cyanosis would become successively more profound. If the infant had an atrial septal defect with left to right shunt, one would expect a murmur in the pulmonic position, but not desaturation, as seen in this infant, and you might appreciate a fixed split to the second heart sound. The blood pressure in the lower extremity is normal and the femoral pulses are normal, making the diagnosis of coarctation much less likely. Coarctation could be associated with other left-sided obstructive lesions such as aortic stenosis, but the murmur in that case would be expected in the aortic position (the right upper sternal border). This patient has evidence of decreased pulmonary blood flow as the primary physiologic abnormality. A ventricular septal defect (VSD) would cause a holosystolic murmur, usually at the left mid-sternal border. A small restrictive VSD would cause a loud grade 4/6 murmur, but you would not expect to see desaturation. If the VSD were large and the murmur was caused by excess pulmonary flow, there could be desaturation if there was also pulmonary edema. In that situation, one would expect other signs of congestive heart failure, including hepatomegaly. A chest radiograph would help to differentiate excessive from decreased pulmonary flow.

The natural history of PS varies greatly. Patients may have stable and mild PS and remain asymptomatic during childhood. Pulmonic stenosis severity is differentiated by the gradient across the pulmonary valve on echocardiogram, as well as the estimated right ventricular pressure compared to the pressure in the left ventricle. If there is no left ventricular outflow tract obstruction, the left ventricular systolic pressure is estimated by the systolic blood pressure. Right ventricular pressure can be estimated if there is adequate tricuspid regurgitation to measure the difference between the right atrial and ventricular pressures. The tricuspid regurgitation velocity allows us to calculate the difference in the pressure of the right ventricle and right atrium. Pulmonary stenosis severity is categorized as:
- Mild PS is defined as a gradient up to 30 to 40 mm Hg or right ventricular (RV) pressure less than 50% of systemic or left ventricular (LV) pressure.
- Moderate PS is defined as a gradient of 40 to 70 mm Hg or RV pressure 50% to 75% of the LV pressure.
- Severe PS is a gradient greater than 70 mm Hg or an RV pressure greater than 75% of the LV pressure. The RV pressure correlates with the degree of outflow tract obstruction.

The more severe form of PS can worsen quickly over the first few weeks of life if the newborn does not present immediately after birth. The newborns with this form of PS usually have thick and dysplastic valves and require close monitoring and referral for balloon angioplasty if they begin to show desaturation or respiratory distress. Valve dysplasia is present in 10% to 20% of patients with PS. Balloon angioplasty of critical PS in infancy has a 95% probability of relieving

the obstruction and not requiring re-intervention for up to 5 years. Mild pulmonary insufficiency is common after the procedure.

PREP Pearls
•   The murmur of pulmonic stenosis prolongs as the gradient across the valve increases.
•   Severe pulmonic stenosis may be urgently treated with balloon angioplasty.
•   The murmur in tetralogy of Fallot will decrease during a hypercyanotic spell.

ABP Content Specifications(s)
•   Plan appropriate initial management of severe pulmonary valve stenosis

Suggested Readings
•   Park MK. Obstructive lesions: pulmonary stenosis. Park's Pediatric Cardiology for Practitioners. 6th ed. Philadelphia, PA: Saunders Elsevier; 2014:184-205.

Question 113

A 15-year-old adolescent girl presents to your office for a health supervision visit. She has a history of migraine headaches that have improved with sumatriptan and naproxen as needed, but she continues to have severe headaches that affect her daily activity. The patient is concerned about feeling fatigued when she takes sumatriptan and is exploring more natural treatments for her migraines. She has done research on the internet and has questions about herbal supplements, such as butterbur and biofeedback therapy.

Of the following, your BEST response to her regarding these therapies would be that

    A.   biofeedback therapy is not covered by insurance

    B.   herbal supplements have no serious side effects

    C.   some alternative treatments can be used along with medical therapy

    D.   they are not recommended as they are not approved by the US Food and Drug
           Administration

    E.   you cannot make any recommendations about alternative therapies

Question 113   **Preferred Response: C**

The correct response is that some alternative treatments can be used along with medical therapy. Many patients seek alternative methods of treatment because of medication side effects or poor results with conventional therapy. Biofeedback may or may not be covered by insurance; however, this should not exclude its use by patients who have the resources to try this therapy. It is important to recognize that some herbal supplements can have serious side effects and interact with other medications. Although most herbal supplements are not approved by the US Food and Drug Administration, many are safe and can be recommended as possible treatments. The American Headache Society (AHS) and the American Academy of Neurology (AAN) recommend butterbur as an effective prophylactic treatment for migraine. Physicians may not be familiar with all of the complementary or alternative medicine (CAM) therapies, but it is important for them to be open to discussing these options with families. A nonjudgmental approach to discussing CAM will allow the physician to counsel the patient and family about the risks, benefits, and potential interactions with traditional therapy.

The patient-physician relationship should allow autonomy regarding treatment choices. Practitioners should engage in open and nonjudgmental communication with patients and families regarding CAM. With open communication, a physician can practice nonmaleficence by informing patients of potential harm that some therapies may cause, including potential medication interactions. It may be necessary to monitor specific organ function with prolonged use of some alternative therapies. The physician should also practice beneficence, promoting the best interest of the patient, which includes respecting personal religious, cultural, and medical-related beliefs. By facilitating equal access to all treatments, a physician can demonstrate an ethical approach to CAM.

Many patients with chronic conditions will pursue CAM as an option, especially when a conventional treatment method has significant side effects or is perceived to be ineffective. Patients use CAM for many reasons, and most will use CAM in conjunction with traditional medical treatment. A study on the reasons and associations for CAM use reported that higher education, poorer health status, anxiety, chronic pain, back problems, transformational experiences, or a personal world view were associated with higher rates of CAM use. No association was found between dissatisfaction with conventional therapy and CAM use. In recent studies, up to 70% of patients seen in subspecialty pediatric clinics report CAM use. In neurology clinics, although 60% of patients had tried CAM along with conventional therapy, only 50% of those discussed the use of CAM with their physician. CAM use is widespread and inadequately reported to their physicians, therefore it should be discussed as a routine part of each patient visit.

PREP Pearls
- Up to 70% of families use complementary and alternative medicine (CAM) including nutritional supplements.
- Patients often use CAM along with traditional medical therapy.
- A nonjudgmental approach to the use of CAM can prevent harm resulting from interactions with traditional medical therapy.

ABP Content Specifications(s)
- Recognize and apply ethical principles regarding the use of complementary and alternative medicine

Suggested Readings
- Adams D, Dagenais S, Clifford T, et al. Complementary and alternative medicine use by pediatric specialty outpatients.Pediatrics. 2013;131:225-232. DOI: http://dx.doi.org/10.1542/peds.2012-1220.

Question 114

The parents of an 8-year-old boy with trisomy 21 would like to enroll their child in a soccer league. The child's medical history is significant for ventricular septal defect repair at 6 months of age. The boy does not report any neck pain. His parents deny gait disturbance, bowel or bladder changes, or weakness. On physical examination, the child has low-normal muscle tone and normal reflexes.

Of the following, the MOST appropriate recommendation would be to

   A.   allow the child to participate after discussing his increased risk for atlantoaxial instability with his parents

   B.   allow the child to participate as long as flexion and extension radiographs of the cervical spine are normal

   C.   allow the child to participate as long as magnetic resonance imaging of the cervical spine is normal

   D.   recommend the child not participate, given his history of congenital heart disease

   E.   recommend the child not participate, given his increased risk of atlantoaxial instability

Question 114   **Preferred Response: A**

The parents of the boy in the vignette should be counseled that children with trisomy 21 have an increased risk for atlantoaxial instability. Based on published case reports, children with Down syndrome appear to be at increased risk of developing spinal cord injury with contact sports, but there are no evidence-based studies that quantify the risk.

Atlantoaxial instability refers to excessive motion at the junction of the spinal atlas (C1) and the axis (C2). After age 3 years, the bones of the cervical spine are adequately ossified to be evaluated on plain radiography. The atlantodens interval (ADI), the distance from the atlas to the anterior aspect of the dens on lateral radiograph of the cervical spine, has historically been used to assess for atlantoaxial instability. Approximately 20% of children with trisomy 21 meet the radiographic criteria for atlantoaxial instability; most of these individuals are asymptomatic. In the absence of symptoms, a large ADI has not been associated with injury to the cervical spine. In addition, a child with normal radiographs can subsequently develop atlantoaxial instability. Because ADI measurements on radiography are neither sensitive nor specific for the risk of cervical spine injury, routine screening is not recommended for asymptomatic children. The American Academy of Pediatrics recommends counseling parents that children with trisomy 21 who participate in certain contact or collision sports (eg, soccer, football, and basketball) and sports that require cervical spine flexion/extension (eg, gymnastics) are at increased risk for cervical spine injury. In addition, parents should be advised that trampoline use is not recommended for children with trisomy 21 who are younger than 6 years. Children age 6 years or older should only be allowed to use trampolines with careful supervision.

Children who experience symptoms of spinal cord compression (eg, weakness, neck pain, radicular symptoms, bowel or bladder symptoms) should undergo urgent evaluation of the cervical spine. For these patients, providers should obtain anterior-posterior and lateral cervical spine radiographs with the neck in a neutral position. If findings on these films are normal, flexion and extension films should then be obtained. During health supervision visits, parents should be counseled about the signs and symptoms of cervical myelopathy and the urgent need to seek evaluation and treatment if symptoms develop; physical examination should include a neurologic evaluation to look for weakness and hyperreflexia.

Despite the lack of evidence to support screening, the Special Olympics continues to require cervical spine screening for participation in certain sports. The exact requirements for Special Olympics screening vary by state, therefore providers who care for patients with trisomy 21 should be familiar with their state's requirements.

Screening radiography and magnetic resonance imaging are not indicated for the boy in the vignette because he is asymptomatic. This child had a ventricular septal defect that had been repaired during infancy, which, in the absence of other congenital heart disease and pulmonary hypertension, is not a contraindication to sports participation.

PREP Pearls
- Asymptomatic children with trisomy 21 do not need routine radiographic screening for atlantoaxial instability.
- Children with cervical spine compression symptoms should have prompt radiographic evaluation of the cervical spine, with anterior-posterior and lateral views in the neutral position. Flexion and extension films should only be obtained once films obtained with the cervical spine in neutral position are normal.
- Children with signs or symptoms of cervical spine compression should be urgently referred to a pediatric orthopedic or neurosurgical specialist.

ABP Content Specifications(s)
- Understand the guidelines for sports participation for patients who have Down syndrome

Suggested Readings
- Bull MJ and the Committee on Genetics. Health supervision for children with Down syndrome. Pediatrics. 2011;128(2):393-406. DOI: http://dx.doi.org/10.1542/peds.2011-1605.

Question 115

A 5-year-old girl, visiting the United States from India, is brought to the emergency department with a complaint of fever, abdominal pain, back pain, and lethargy. On physical examination, her temperature is 39.5°C, heart rate is 135 beats/min, respiratory rate is 30 breaths/min, and blood pressure is 95/65 mm Hg. She is thin and ill appearing. Her conjunctivae are pale and mucous membranes are tacky. Capillary refill is 2 seconds. There is an II/VI vibratory systolic ejection murmur. Lungs are clear to auscultation. Her abdomen is diffusely tender to palpation, and the liver and spleen are palpated 4 cm and 5 cm, respectively, below the costal margins. Examination of the spine and paraspinal muscles is normal.

A complete blood cell count shows:
- White blood cell count, 4,000/µL (4.0×10^9/L) with 40% neutrophils, 40% lymphocytes, and 20% monocytes
- Hemoglobin, 8 g/dL (80 g/L)
- Platelet count, 120×10^3/µL (120×10^9/L)

A review of the peripheral smear by pathology is requested (Item Q115).
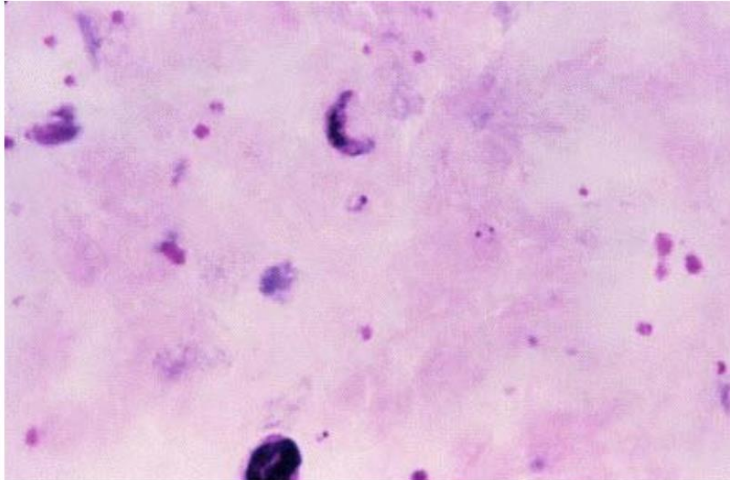
Item Q115. Giemsa-stained peripheral blood smear from the patient described in the vignette. Reprinted with permission from Pickering LK, et al. Red Book® Online. Elk Grove Village, IL: American Academy of Pediatrics.

Of the following, the MOST likely etiology of this child's illness is
A.   Babesia microti
B.   Borrelia species
C.   Ehrlichia species
D.   Plasmodium falciparum
E.   Trypanosoma cruzi

Question 115   **Preferred Response: D**

The patient in the vignette has Plasmodium falciparum (malaria) infection documented in the Giemsa-stained micrograph (Item C115) showing a crescent-shaped P falciparum gametocyte above a ring-form trophozoite (center). This infection is transmitted by the female Anopheles mosquito throughout the tropical areas of the world, most commonly in sub-Saharan Africa, Papua New Guinea, the Solomon Islands, and Vanuatu. Travelers to the Indian subcontinent, like the girl in the vignette, are at moderate risk of infection. Malaria also can be transmitted in more temperate climates, including parts of the United States, where mosquitoes of the Anopheles genus are present.
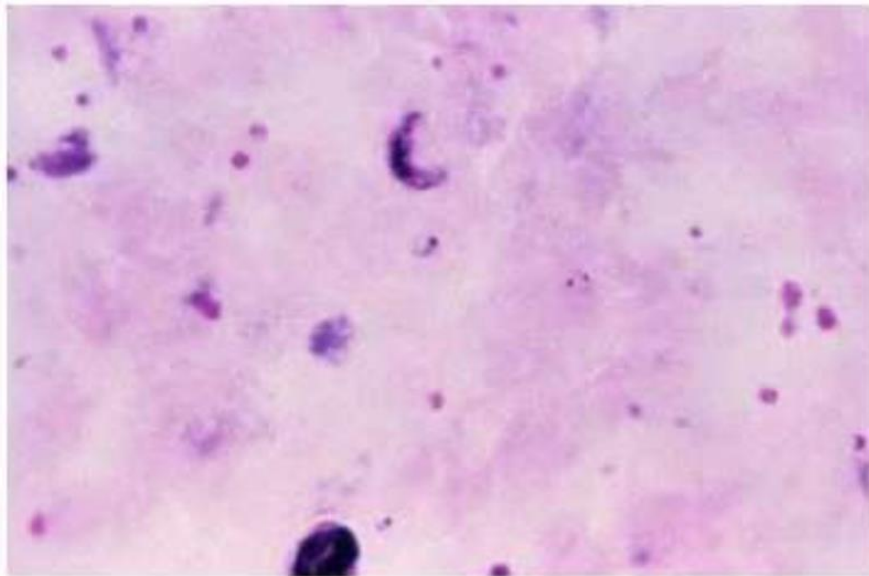
Item C115: *Plasmodium falciparum* as seen on a Giemsa-stained micrograph.

Modified with permission from Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds.

Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed.

Elk Grove Village, IL: American Academy of Pediatrics; 2015:854-855

The most common malaria species are P vivax (most prevalent in the Indian subcontinent and Central America) and P falciparum (worldwide but most prevalent in Africa, Papua New Guinea, Haiti, and the Dominican Republic). Less common are P ovale (most prevalent in West Africa) and P malariae (worldwide distribution). P vivax and P ovale can cause persistent hepatic infection, which may result in reactivation of disease months to years later. Persistent, low-concentration parasitemia caused by P falciparum or P malariae also can result in recurrence of symptoms. Chronic infection caused by P malariae can lead to nephrotic syndrome. P knowlesi,

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found in Southeast Asia, is a primate parasite that can infect humans and cause severe disease due to hyperparasitemia.

Patients with malaria typically present with fever, chills, rigors, and headache, which may be paroxysmal. Nausea, vomiting, diarrhea, abdominal pain, arthralgias, myalgias, and respiratory symptoms can occur. Hepatosplenomegaly is common. Typical laboratory findings include anemia and thrombocytopenia. P falciparum infection can be severe or even fatal, presenting with neurologic manifestations (eg, seizure, confusion, coma), renal failure (acute tubular necrosis), respiratory failure (pulmonary edema), severe anemia (from splenic sequestration or hemolysis), metabolic derangements (hypoglycemia, metabolic acidosis), vascular collapse, and shock. The treatment of malaria depends on the severity of disease, the species, and the likelihood of drug resistance (See suggested reading no. 5 for recommended treatment). Babesia microti causes babesiosis, characterized by mild, nonspecific symptoms such as malaise, anorexia, and flulike symptoms. Clinical signs usually are minimal, unlike those described for the girl in the vignette. Babesiosis is a tick-borne zoonosis most commonly acquired in the Northeastern United States; the primary reservoir is the white-footed mouse, and the primary vector is the tick Ixodes scapularis, which also can transmit Borrelia species. The organism is difficult to distinguish from P falciparum on stained blood smears, but the clinical presentation generally distinguishes one infection from the other.

Infection with Borrelia species can cause relapsing fevers, either tick borne or louse borne (B recurrentis), as well as Lyme disease (B burgdorferi). Relapsing fever is characterized by high fever, shaking chills, myalgias, arthralgias, headache, and nausea. A fleeting rash may occur. Louse-borne disease occurs in Ethiopia, Eritrea, Somalia, and the Sudan. Tick-borne relapsing fever occurs worldwide. The diagnosis is made by observing spirochetes (not ring-form parasites as seen in malaria) on dark field microscopy or stained preparations of peripheral blood smears. The symptoms of Lyme disease typically are indolent and occur in stages (early localized, early disseminated, and late disease). Fever, arthralgias, myalgias, headache, and fatigue are common during the early disseminated stage. The diagnosis of Lyme disease can be made clinically or serologically.

Manifestations of Ehrlichia infection include fever, chills, malaise, headache, and myalgias. Vomiting and diarrhea may also occur. Rash is present in 50% of children. Severe manifestations of disease include disseminated intravascular coagulation, respiratory distress syndrome, renal failure, and encephalopathy. Most cases of ehrlichiosis occur in California, Texas, and in the southeast, northeast, and north central regions of the United States. The diagnosis typically is made serologically or with commercially available polymerase chain reaction assays. Examination of peripheral blood smears to detect morulae (inclusion bodies in the cytoplasm of monocytes) is insensitive.

Most patients infected with the parasite Trypanosoma cruzi (Chagas disease) are asymptomatic. Young children are more likely than adults to be symptomatic. The infection is transmitted by the "kissing bug" (triatomine insects), and occurs primarily in Mexico and Central and South America. A red, indurated nodule may form at the inoculation site on the face or arms. If the site

of entry is the conjunctiva, a Romana sign (unilateral eyelid edema) may occur. Fever, malaise, hepatomegaly, and generalized lymphadenopathy may develop acutely, but resolve without treatment within 3 months. Chagas cardiomyopathy is a late complication that occurs years to decades after the initial infection. The diagnosis is established serologically, but parasites may be observed in blood smears during the acute phase of infection.

PREP Pearls
- The treatment of malaria depends on the severity of disease, the infecting parasite, and the likelihood of drug resistance.
- The most common malaria species in the world are P vivax (most prevalent in the Indian subcontinent and Central America) and P falciparum (worldwide but most prevalent in Africa, Papua New Guinea, Haiti, and the Dominican Republic).
- P vivax and P ovale can cause persistent hepatic infection which may result in reactivation of disease months to years later.

ABP Content Specifications(s)
- Recognize the clinical features of malaria, and manage appropriately
- Understand the epidemiology of malaria

Suggested Readings
- American Academy of Pediatrics. American trypanosomiasis. In: Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:803-805.

Question 116

A 16-year-old adolescent girl is transported to the emergency department by paramedics following a motor vehicle collision. Thirty minutes earlier, she was a passenger in the front seat of a car that struck a tree. She was not wearing a seatbelt and the right side of her head hit the passenger side window. She does not recall any further details about the collision.

The patient was awake when paramedics arrived at the scene, but then had a 1-min generalized seizure, followed by several minutes of decreased responsiveness. Her level of consciousness has improved gradually since the seizure occurred. She is now following commands and answering questions, although she intermittently seems confused. She is maintaining her own airway, has clear and equal breath sounds, and her vital signs are within normal limits. She has had no episodes of vomiting.

On your physical examination, the patient is lying flat with her entire spine immobilized. She is awake, follows your commands, and answers your questions, although she seems confused at times. Her Glasgow Coma Scale score is 14. You detect the scent of alcohol on her breath. She has a 4 x 6 cm boggy hematoma over her right temple, which is very tender to palpation. Her pupils are 4 mm in diameter bilaterally and equally reactive. She has full strength and normal reflexes in all extremities. The remainder of your physical examination is unremarkable.

Of the following, the BEST next step in evaluation of this girl is

A.   computed tomography of the brain

B.   electroencephalogram

C.   plain radiography of the skull

D.   serum ethanol level

E.   urine toxicology screen

Question 116   **Preferred Response: A**

The adolescent girl in the vignette presents with intermittent confusion and a large boggy hematoma over the right temporal region of her scalp, following a motor vehicle collision during which she was an unrestrained passenger. Given her altered mental status, large right temple hematoma, and severe mechanism of injury followed by a seizure with loss of consciousness, computed tomography (CT) of the brain is the best next step in evaluating her for potential intracranial injury.

Among trauma patients, head injury is the leading cause of death and disability. Although most children sustaining head trauma have only minor injuries, a small number will have more serious injuries with the potential for clinical deterioration and significant sequelae. Providers of pediatric care should understand how to appropriately evaluate and initially manage children and adolescents who present with trauma to the central nervous system.

Pediatric head injuries account for more than 600,000 emergency department (ED) visits and an even larger number of visits and calls to primary care providers annually. Most pediatric head trauma results from falls, motor vehicle collisions, automobile-pedestrian accidents, bicycle-related injuries, and sports-related injuries. Most pediatric head injuries are minor, but some can be extremely serious; more than 3,000 deaths related to head trauma occur in US children each year. A challenge for pediatric providers is to differentiate the relatively small number of children at high risk for intracranial complications and clinical deterioration after head trauma from the many who are at very low risk. Clinical symptoms are neither completely sensitive nor specific for significant injury.

Computed tomography of the brain is a rapid and accurate way to identify intracranial injuries in children after head trauma. There is consensus that patients identified as being at high risk for intracranial injury should undergo early noncontrast CT of the brain for evidence of intracranial hemorrhage, midline shift, or increased intracranial pressure. Widespread use of this diagnostic modality has its downsides, however, including exposure of the brains of developing children to ionizing radiation, identification of minor lesions or incidental findings with unclear clinical importance, the need for sedation for younger or uncooperative pediatric patients, and a significant increase in health care costs. The goal should be to identify children with clinically important intracranial injury after head trauma to prevent deterioration and secondary brain injury, while limiting unneeded radiographic imaging in children at very low risk.

Recent studies, including a very large multicenter study conducted through the Pediatric Emergency Care Applied Research Network (PECARN), have yielded validated clinical decision guidelines that provide a useful clinical framework for determining which children are at higher (as well as at very low) risk for clinically important brain injuries after head trauma. Based on the PECARN study, the risk of clinically important traumatic brain injury is estimated at more than 4% for children with any of the following: Glasgow Coma Score of 14 or other signs of altered mental status, a palpable skull fracture (under age 2 years), or signs of basilar skull fracture (≥2 years of age). A CT scan of the brain is thus recommended for these children. The girl in the vignette would fall into this category.

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Brain CT is not recommended for children who are found to be at extremely low risk ($< 0.05\%$) for clinically important traumatic brain injury. This includes children meeting all of the following criteria: normal neurologic examination findings, normal mental status, normal behavior as noted by a caregiver, no loss of consciousness, no vomiting, no severe headache, no evidence of skull fracture (for children younger than 2 years, and no or frontal only scalp hematoma), no signs of basilar skull fracture, no high-risk mechanism of injury, and no concern for inflicted injury.

Children with no alteration of mental status or signs of skull fracture fall into the "intermediate" risk category if they have a history of isolated loss of consciousness, headache, vomiting, or certain scalp hematomas. For these children the PECARN guidelines recommend that the decision about whether brain CT or observation alone is needed should be based on additional clinical factors that include: the presence of multiple intermediate risk factors, age younger than 3 months, worsening symptoms or signs after ED observation, physician experience, and parental preference. Observing a child in the ED when emergent imaging is not indicated provides the opportunity to watch for symptom improvement or worsening. Children who clinically worsen during a period of observation should undergo CT.

Initial management of cases of central nervous system trauma must focus on stabilizing the airway, maintaining adequate ventilation and oxygenation, and maintaining adequate perfusion to the brain and other vital organs, while ensuring continuous cervical spine precautions until a cervical spine injury can be excluded.

Although the patient in the vignette did have a self-limited post impact seizure after sustaining head trauma, she is displaying no indication of ongoing seizure activity in the emergency department, therefore electroencephalography would not be useful at this time.
Plain radiography of the skull may identify a skull fracture in this patient, but this diagnostic study would provide no direct information about the presence of intracranial injury.
The patient's history and clinical presentation suggest that she was likely drinking alcohol before the motor vehicle collision. Although intoxication with ethanol could indeed explain her intermittent confusion, this symptom could also be evidence of a life-threatening intracranial injury. Therefore, a brain CT should not be delayed in obtaining a serum ethanol level in this patient.

Similarly, although the girl may have abused other drugs before the motor vehicle collision, ruling out a serious intracranial injury must take greater priority in her evaluation and management than obtaining a urine toxicology screen.

PREP Pearls
- Initial management of cases of central nervous system trauma must focus on stabilizing the airway, maintaining adequate ventilation and oxygenation, ensuring perfusion to the brain and other vital organs, and continuing cervical spine precautions until cervical spine injury is excluded.
- Computed tomography (CT) of the brain is a rapid and accurate way to identify intracranial injuries in children after head trauma. Risks associated with CT include exposure to ionizing radiation, identification of minor lesions or incidental findings, potential need for sedation, and significant increases in health care costs.
- Recent studies have yielded validated clinical decision guidelines that provide a useful framework for determining which children are at higher risk for clinically important brain injuries after head trauma, while limiting unnecessary imaging in children at low risk.

ABP Content Specifications(s)
- Plan the appropriate initial evaluation and management of acute central nervous system trauma

Suggested Readings
- Kuppermann N, Holmes JF, Dayan PS, et al. Identification of children at very low risk of clinically important brain injuries after head trauma: a prospective cohort study. Lancet. 2009;374(9696):1160-1170. DOI:http://dx.doi.org/10.1016/S0140-6736(09)61558-0.

Question 117

A full-term newborn is delivered by repeat caesarean delivery. The mother requires general anesthesia, with the time from abdominal incision to delivery 10 minutes because of multiple adhesions. Artificial rupture of the membranes occurs at delivery and reveals clear amniotic fluid. The newborn emerges apneic and limp. Upon arrival at the warmer, the newborn is dried and stimulated. Assessment at 30 seconds after delivery reveals an apneic, cyanotic newborn with a heart rate of 30 beats/min.

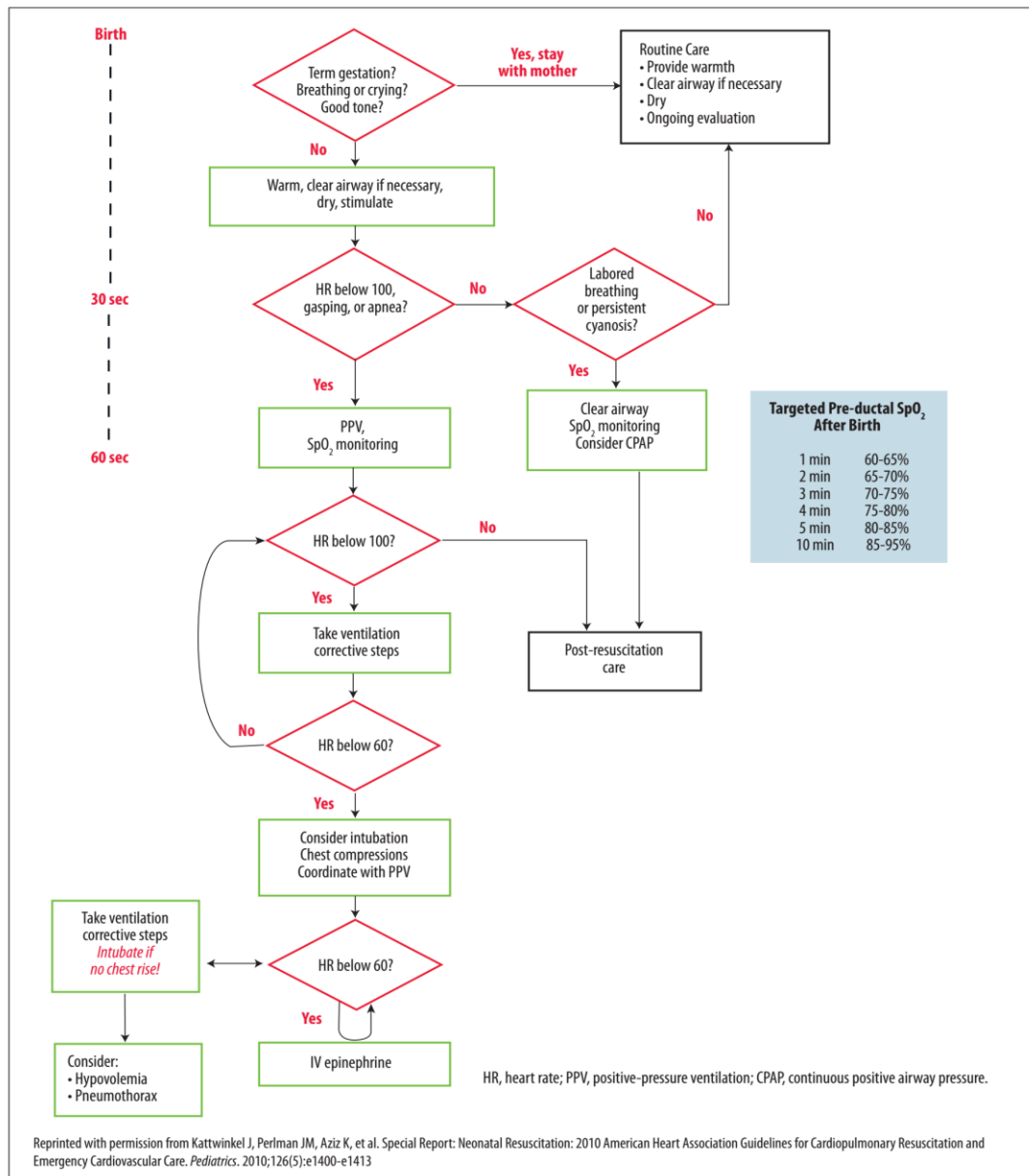Of the following, the BEST next step in management of this newborn is to

A.   initiate bag mask ventilation with 100% oxygen

B.   initiate bag mask ventilation with 100% oxygen and begin compressions

C.   initiate bag mask ventilation with room air

D.   initiate bag mask ventilation with room air and begin compressions

E.   intubate and administer intratracheal epinephrine

**Question 117**   **Preferred Response: C**

The full-term newborn in the vignette who has apnea requires initiation of bag mask ventilation with room air. The sixth edition of the Neonatal Resuscitation Program of the American Academy of Pediatrics and American Heart Association outlines the steps in newborn resuscitation (Item C117A). Newborns should be assessed 30 seconds after delivery, following the initial steps of resuscitation including drying and stimulation. If a newborn has a heart rate less than 100 beats/min, apnea, and/or gasping respirations, positive pressure ventilation should be initiated. Assisted ventilation is the most effective action in the resuscitation of a compromised newborn.

Item C117A. Newborn Resuscitation Process.

Reprinted with permission from Kattwinkel J, Perlman JM, Aziz K, et al. Special Report: Neonatal Resuscitation: 2010 American Heart Association Guidelines for Cardiopulmonary Resuscitation and Emergency Cardiovascular Care. *Pediatrics*. 2010;126(5):e1400-e1413

After 30 seconds of effective assisted ventilation, the newborn should be evaluated again. If the heart rate is below 60 beats/min, compressions should be initiated, coordinated with positive pressure ventilation using a 3:1 ratio, with 90 compressions and 30 breaths occurring in a 1-minute period. Chest compressions may be performed using the thumb technique (Item C117B) or 2-finger technique (Item C117C), compressing to one-third the depth of the anterior-posterior chest. Compressions should be discontinued when the heart rate is greater than 60 beats/min, with the decision to discontinue assisted ventilation being made when the heart rate is greater than 100 beats/min.
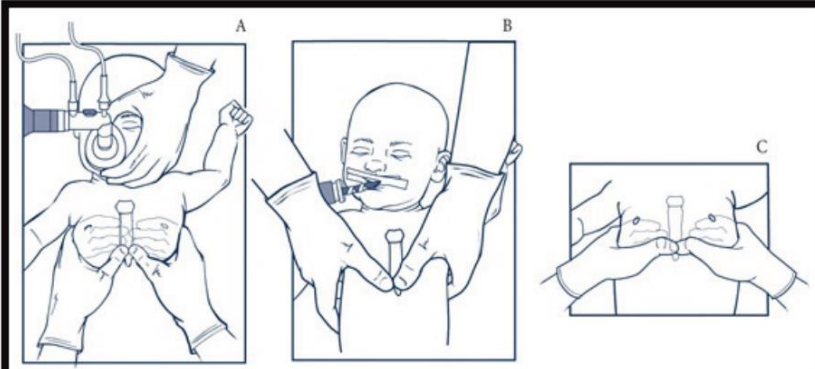
Reprinted with permission from Kattwinkel J, ed. Textbook of Neonatal Resuscitation. 6th ed. Elk Grove Village, IL: American Academy of Pediatrics/ American Heart Association; 2011:139

ITEM C117B: Thumb technique of chest compressions administered from the bottom (A), from the top (B), and for small chests with thumbs overlapped (C).
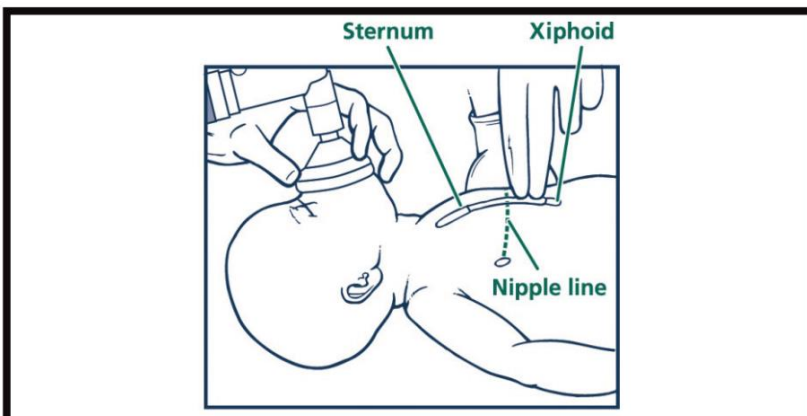
Reprinted with permission from Kattwinkel J, ed. Textbook of Neonatal Resuscitation. 6th ed. Elk Grove Village, IL: American Academy of Pediatrics/ American Heart Association; 2011:139

ITEM C117C: Correct finger position for 2-finger technique.

The 2010 American Heart Association Guidelines for Neonatal Resuscitation recommend that the initial resuscitation of a full-term newborn with positive pressure ventilation be performed using room air. An oximeter probe should be placed on the newborn's right hand (preductal) when assisted ventilation is begun to allow adjustment of oxygen to attain targeted goal saturations during the resuscitation. Although hypoxia may cause multiorgan damage, even brief episodes of hyperoxia may be associated with adverse outcomes.

The full-term newborn in the vignette is found to have apnea 30 seconds after birth and requires bag mask ventilation with room air. Oxygen should not be used until the oxygenation status is determined with a pulse oximeter. Compressions are not indicated until reassessment after 30 seconds of effective assisted ventilation. Intubation and administration of intratracheal epinephrine may be considered if the newborn fails to respond to the initial resuscitation, including compressions and effective assisted ventilation.

PREP Pearls
- Assisted ventilation is the most effective action in the resuscitation of a compromised newborn.
- If the heart rate is below 60 beats/min after 30 seconds of effective positive pressure ventilation, compressions should be initiated.
- Compressions are coordinated with positive pressure ventilation using a 3:1 ratio, with 90 compressions and 30 breaths occurring in a 1-minute period.

ABP Content Specifications(s)
- Recognize the indications for external cardiac massage during resuscitation of a newborn infant, and institute appropriately

Suggested Readings
- Kattwinkel J, ed. Textbook of Neonatal Resuscitation. 6th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2011:328 pp.

Question 118

A 12-year-old boy complains of limb weakness that began 3 months ago. He cannot keep up with peers when playing sports and he has difficulty going up or down more than 1 flight of stairs. The boy can still open jar lids with his hands and his handwriting remains normal. He has no eyelid drooping, double vision, facial weakness, or difficulty swallowing. The limb weakness remains the same throughout the day and does not improve with rest. There is no family history of limb weakness.

On physical examination, the boy's vital signs are normal and his weight is stable from his health supervision visit 9 months previously. He does not have any rashes, skin changes in his hands or knuckles, and there are no striae. He does not have elbow contractures. His neurological examination shows normal cranial nerve function with symmetric weakness of his deltoid, biceps, triceps, quadriceps, and hamstring muscles. His muscle bulk and tone are normal. He has mild tenderness to palpation of his quadriceps muscles and they have a rubbery feel. His reflexes are normal and symmetric. Sensation of light touch is normal in his arms and legs. The boy cannot rise from a chair without using his hands and he uses a Gower maneuver to rise from the floor. An electrocardiogram is normal. You order laboratory tests and the results are shown:

	Result	Normal Range
Alanine aminotransferase	82 U/L	(10 to 40 U/L)
Aldolase	9 U/L	(1 to 1.75 U/L)
Aspartate aminotransferase	98 U/L	(14 to 35 U/L)
Creatine kinase	1,950 U/L	(20 to 180 U/L)
Lactate dehydrogenase	450 IU/L	(127 to 287 U/L)

Of the following, the test MOST likely to identify the correct diagnosis is

A.   acetylcholine receptor antibodies

B.   dystrophin gene sequence

C.   electromyography and nerve conduction study

D.   magnetic resonance imaging of the quadriceps

E.   muscle biopsy

Question 118   **Preferred Response: E**

The subacute onset of proximal muscle weakness with no signs or symptoms of an endocrine or rheumatologic disease suggests an acquired disorder of the muscles, such as polymyositis or dermatomyositis. In the absence of characteristic dermatologic findings of dermatomyositis, muscle biopsy is the best test to determine the diagnosis. In this case, muscle biopsy showed endomysial inflammation and the boy in the vignette was diagnosed with juvenile polymyositis. In this vignette, the history and physical examination findings narrow the differential diagnosis for the boy's weakness to a muscle disorder. Diagnostic testing should focus on determining the kind of muscle disorder he has. His symptoms are not suggestive of myasthenia gravis, which is a disorder of the neuromuscular junction, not of the muscles. So testing for the presence of acetylcholine receptor antibodies is unlikely to yield a diagnosis. Electromyography and nerve conduction study (EMG/NCS) are used to determine whether there is a disorder of the muscle, the neuromuscular junction, or the nerves. In this case, the history and examination findings strongly suggest a muscle disorder, so EMG/NCS will not add additional information. Magnetic resonance imaging of an involved muscle may show edema, but is unlikely to identify a specific diagnosis. The age at onset of 12 years makes Duchenne muscular dystrophy unlikely and a dystrophin gene sequence is not likely to lead to the correct diagnosis.

The differential diagnosis of muscle disorders presenting in childhood can be divided into 2 categories: genetic and acquired. Genetic muscle disorders typically present in the preschool years and have a slowly progressive course. Pediatricians should recognize the more common genetic muscle disorders such Duchenne muscular dystrophy and myotonic dystrophy. Duchenne muscular dystrophy typically presents in boys younger than 6 years with progressive proximal weakness, calf hypertrophy, and creatine kinase of 10,000 U/L or greater. Childhood onset myotonic dystrophy presents with distal weakness (weak ankle dorsiflexion and weak hand muscles), a long, narrow face, and tapering forearms. Acquired muscle disorders include myopathies caused by endocrine or rheumatologic diseases or inflammatory muscle disorders. Acquired muscle disorders have a more abrupt onset than genetic muscle disorders. Signs of an endocrine or rheumatologic disease may be present. Characteristic dermatologic findings of heliotrope rash and Gottron papules suggest a diagnosis of dermatomyositits. If there are no elements to suggest a particular acquired muscle disease, muscle biopsy is the next best diagnostic test.

PREP Pearls
- Typical findings in boys with Duchenne or Becker muscular dystrophy include proximal muscle weakness, calf hypertrophy, and creatine kinase levels of 10,000 U/L or greater.
- Patients with distal muscle weakness involving the hands often notice that they have difficulty opening jar lids.

ABP Content Specifications(s)
- Formulate a differential diagnosis of a muscle disorder of various etiologies

Suggested Readings

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- Andersson PB, Rando TA. Neuromuscular disorders of childhood. Curr Opin Pediatr. 1999;11(6):497-503. Abstract available at http://www.ncbi.nlm.nih.gov/pubmed/10590906.
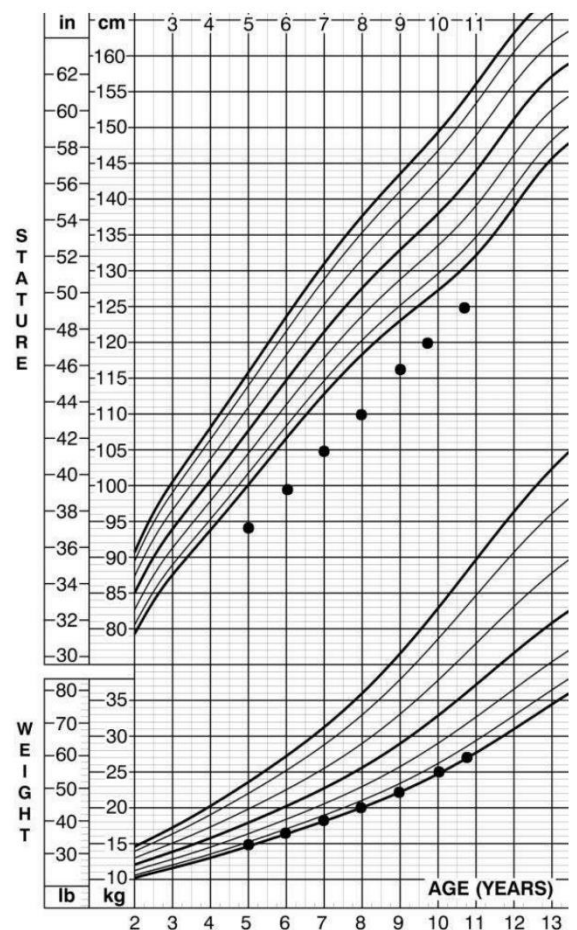
Question 119

A 9-year-old girl presents to your office for evaluation of poor growth. Her height is significantly below the first percentile, but has been at the same percentile for many years. Her weight has always been at the fifth percentile (Item Q119). Midparental height is 152.4 cm (fifth percentile). There is no family history of delayed puberty. You obtain bone age radiographs. Based on her concordant bone age of 9 years, her predicted adult height is 147.3 cm (significantly below the first percentile). The girl's family requests that she be started on growth hormone treatment.

Of the following, the MOST accurate statement regarding this request is that

A.   despite a normal growth velocity, her stature significantly below the first percentile suggests she could benefit from growth hormone treatment for idiopathic short stature

B.   her normal growth velocity suggests that she has idiopathic (or familial) short stature, therefore growth hormone will not be helpful

C.   her significant short stature suggests growth hormone deficiency or another pathological cause and she could benefit significantly from growth hormone treatment

D.   short stature with normal weight gain suggests an underlying syndromic cause of poor growth

E.   short stature with poor weight gain suggests the primary problem is nutritional intake, which must be corrected before growth hormone treatment could be considered

Item Q119. Growth chart for the patient described in the vignette. Courtesy of J. Pinsker

Question 119   **Preferred Response: A**

Growth hormone (GH) was first approved for treatment of acquired growth hormone deficiency (GHD) in 1985. Children with GHD may have an underlying cause of their poor growth, such as a neoplasm, brain cyst, inflammatory condition, infiltrative process, trauma, surgery, or radiation treatment. However, the most common cause of acquired GHD is idiopathic. Children with GHD typically have short stature, abnormal linear growth velocity, and delayed osseous maturation. The child in the vignette has short stature, but has had a normal growth velocity for many years. Thus, it is unlikely she has GHD. This does not mean that she would not benefit from GH treatment, because there are many indications for GH treatment beyond GHD.

In 2003 the US Food and Drug Administration (FDA) approved GH as a treatment for idiopathic short stature (ISS), defining the condition as height of an individual greater than or equal to 2.25 standard deviation scores below the corresponding mean height for a given age, sex, and population group. This approximates the 1.2nd percentile. Predicted height (based on bone age) in ISS is less than 5'3" for males and 4'11" for females, thus this definition does not include children with constitutional delay of growth and maturation who would be expected to have significant catch-up growth. Children with ISS are GH-sufficient, have normal birthweight and -length, and do not have any other clearly identifiable diagnosis predisposing them to have poor growth. Often bone age is not delayed.

Children with GHD can have significant catch-up growth with GH treatment. Although there is less of an impact, the expected mean gain in height for children with ISS averages 3 inches from predicted height. Thus, even with a normal growth velocity, this very short child could benefit from GH treatment. Referral to a pediatric endocrinologist for further evaluation and possible treatment for this FDA-approved indication is appropriate. Specific height velocity charts have been published to show expected changes in height growth under various conditions, including ISS, and can be used to guide treatment.

Evaluation of the growth chart of the child in the vignette shows that she is growing 5 cm/year, a normal prepubertal growth velocity, therefore identification of an underlying cause of poor growth is unlikely. Similarly, there is no indication that she has any underlying syndrome that would make her response to GH treatment above average. With GH treatment, she would be expected to grow approximately 3 inches beyond her predicted height. The child's weight percentiles are above the height percentiles, effectively ruling out a nutritional cause of poor growth.

PREP Pearls
- Children with growth hormone deficiency (GHD) typically have short stature, abnormal linear growth velocity, and delayed osseous maturation.
- Children with idiopathic short stature (ISS) have height at or below the 1.2 percentile, with a predicted height (based on bone age) of less than 5'3" for men and 4'11" for women, and no clearly identifiable diagnosis predisposing them to have poor growth.

- Referral to a pediatric endocrinologist is appropriate for evaluation and management of children with ISS, because this is an US Food and Drug Administration-approved indication for GH treatment.
- Children with ISS, despite a normal growth velocity, respond to GH treatment with a mean height gain of 3 inches beyond predicted final height.

ABP Content Specifications(s)
- Recognize the effects of growth hormone therapy on growth

Suggested Readings
- Allen DB, Cuttler L. Short stature in childhood: challenges and choices. N Engl J Med. 2013;368(13);1220-1228. DOI:http://dx.doi.org/10.1056/NEJMcp1213178.

Question 120

You are called to evaluate a newborn in the delivery room. The newborn was born to a 26-year-old with 1 prior healthy child. No prenatal care or imaging was obtained with the current pregnancy, but the previous pregnancy was notable for maternal group B streptococcal positive cultures. The newborn's Apgar scores are 1 at 1 min and 4 at 5 min. You arrive to find a newborn in acute respiratory distress. Bag-mask ventilation is in progress. The newborn is cyanotic with poor tone and little spontaneous respiratory effort. You place an endotracheal tube and provide positive pressure ventilation. On auscultation, you note asymmetric aeration with primarily right-sided breath sounds. A needle thoracostomy of the left chest returns no air. The abdomen is distended in appearance and tympanitic to percussion. An orogastric tube is placed to decompress the stomach.

Of the following, the MOST likely etiology of this newborn's respiratory distress is

A.   congenital diaphragmatic hernia

B.   hyaline membrane disease

C.   pneumomediastinum

D.   septic shock

E.   total anomalous pulmonary venous return

Question 120   **Preferred Response: A**

The newborn in the vignette suffers from a congenital diaphragmatic hernia (CDH). A newborn with CDH is likely to present immediately after delivery with severe respiratory distress and hemodynamic failure. The affected infant will demonstrate poor air entry into the affected lung and cyanosis; associated findings may include a scaphoid abdomen or a displaced cardiac apex. Although the abdomen is classically scaphoid, it may be distended with air when positive pressure ventilation is used in resuscitation efforts; this is the case for the infant in the vignette. Attempts to prevent abdominal distention are advocated, as abdominal pressure can further limit expansion of functional lung units.

The frequency of CDH has been reported to be 1 in 3,000 to 5,000 live births. However, the true incidence may approximate 1 in 2,000 births when premature fetal terminations and cases of neonatal demise are considered. The most common associated anatomic diaphragmatic defect (95%) is in the foramen of Bochdalek; these defects are posterolateral in location and 80% are found on the left side. Far less frequently, herniation may occur through the foramen of Morgagni; these defects are classically retrosternal in location.

Historically, persistent pulmonary symptomatology and lung hypoplasia have been attributed to compression of the developing ipsilateral lung by abdominal contents displaced into the thoracic cavity during uterine development. However, recent models suggest that lung hypoplasia occurs prior to diaphragm development and that early defects in lung morphogenesis may only be exacerbated by the later presence of intrathoracic compression by abdominal viscera. This data provides some explanation for the fact that pulmonary hypoplasia can be seen bilaterally and not only on the side of the diaphragmatic defect.

Prenatal diagnosis via ultrasonography has advanced in recent decades, but perinatal morbidity and mortality remain high. Right-sided and bilateral defects carry a poor prognosis with mortality rates that approximate 80% and 100%, respectively. Prenatal ultrasonography detects up to 60% of defects, but regional and institutional variability in detection rate exist. Congenital diaphragmatic hernia may be diagnosed by the presence of abdominal organs in the thoracic cavity. Additional associated findings may include displacement of the cardiac axis and mediastinal structures or polyhydramnios. Right-sided defects are difficult to detect on ultrasonography, as the echogenicity of the liver approximates that of the lung. The finding of hydrops carries a poor prognosis.

Congenital diaphragmatic hernia may occur sporadically or as part of a genetic syndrome or association. The autosomal recessive Fryns syndrome is described in up to 10% of infants with CDH; affected patients also suffer from pulmonary hypoplasia, craniofacial anomalies, and distal limb hypoplasia. Other associations include Beckwith-Wiedemann syndrome During delivery of a newborn with CDH, it is advisable to avoid bag and mask ventilation that may overdistend the stomach and result in further compromise of pulmonary mechanics. Placement of a naso- or orogastric tube will allow intermittent decompression, but necessitates monitoring of acid-base balance with concomitant loss of gastric secretions. Mechanical ventilation, when necessary, should be employed, with attention to pulmonary protective strategies. When prenatal diagnosis

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is obtained, delivery should be scheduled at a regional institution with surgical, neonatal intensive care, and extracorporeal membrane oxygenation (ECMO) support. Surgical repair typically occurs early in life; in those infants too unstable to tolerate immediate surgical repair, ECMO may offer a bridging option.

It is important to be mindful of the complications of CDH. Pulmonary hypertension is a well-recognized source of morbidity beyond the neonatal period. All infants with CDH should be screened for pulmonary hypertension with echocardiography in early childhood. When obtained, infant lung function obtained after surgical correction reveals a restrictive pattern with low lung compliance. Obstructive lung disease at 5 years of age is found in 25% of CDH survivors. In adolescent survivors, there is a similar trend toward mild to moderate airway obstruction and bronchodilator responsiveness.

While respiratory distress is commonly encountered in an infant with hyaline membrane disease (HMD), the infant with HMD is more likely to be premature. In addition, while the low compliance of the lung with HMD may result in pneumothorax from positive pressure ventilation and asymmetric aeration, recovery of air on needle thoracostomy would be expected. A severe pneumomediastinum may result in a secondary pneumothorax and related asymmetric aeration, but asymmetry is not expected with a pneumomediastinum alone. Associated findings with pneumomediastinum include subcutaneous air and related crepitus.

The newborn in the vignette may be at risk for sepsis with a history of group B streptococcal infection in the mother and the lack of prenatal care. However, the current presentation is atypical for septic shock or for a congenital cyanotic cardiac defect.

PREP Pearls
- The majority of congenital diaphragmatic hernias (CDH) occur on the left, posterolaterally, and through the foramen of Bochdalek.
- Even with left-sided defects and optimal imaging quality, CDH may not be recognized on prenatal ultrasonography.
- Pulmonary hypertension is a frequent complication for survivors of CDH; infants and children should be screened accordingly.

ABP Content Specifications(s)
- Plan the appropriate management of a child with diaphragmatic hernia.
- Recognize the clinical features of a diaphragmatic hernia

Suggested Readings
- Benachi A, Cordier AG, Cannie M, Jani J. Advances in prenatal diagnosis of congenital diaphragmatic hernia. Semin Fatal Neonatal Med. 2014;19(6):331-337. DOI: http://dx.doi.org/10.1016/j.siny.2014.09.005.

Question 121

A 17-month-old boy presents for his health supervision visit. You last saw him at 12 months of age. The parents have no concerns, but remark that his left eyelid has looked droopy for "awhile" (Item Q121). Reviewing his old chart, you find that he was born at term by repeat cesarean delivery, had an uncomplicated perinatal course, and a normal examination without ptosis at 12 months of age. His growth and development are unremarkable. His physical examination is notable for left-sided ptosis, anisocoria, and a paler pupil on the left than the right. You also palpate a 3 cm left-sided lower cervical lymph node. He has normal movement of his upper and lower extremities.
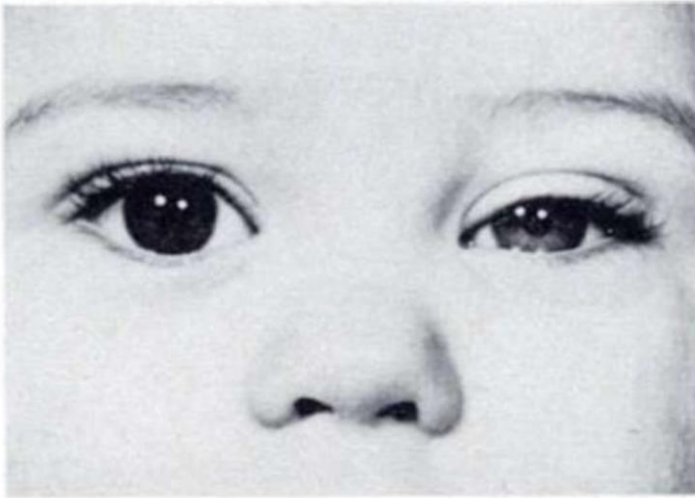
Item Q121. Child described in the vignette. Reprinted with permission from Robinson GC, Dikrainian DA, Roseborough GF. Pediatrics. 1965;35(1):103-107

Of the following, the MOST appropriate management for this child is to

A.   obtain imaging of the head, neck, chest, and abdomen

B.   refer him to neurology for an edrophonium (Tensilon) test

C.   refer him to an ophthalmologist for vision evaluation

D.   refer him to physical therapy for a brachial plexus injury

E.   treat the lymphadenopathy with amoxicillin-clavulanate

Question 121   **Preferred Response: A**

The child in the vignette has Horner syndrome characterized by the presence of ptosis, miosis, anisocoria, facial anhidrosis, and heterochromia (Item C121A). It is most often detected in children younger than 2 years of age. Horner syndrome results from a disruption of the oculosympathetic pathway and can be congenital or acquired. Underlying events may include trauma (birth trauma including brachial plexus injury, unintentional injury, or surgery involving head or neck), neoplasm, infection, or carotid abnormalities. Transient cases may be associated with migraine headaches. Occasionally, there is a family history suggesting a genetic etiology, while in other cases no underlying cause can be found and it is considered idiopathic. The child in the vignette has onset after the first year of life and has concerning physical findings including a large cervical lymph node, therefore further evaluation is necessary. In children, the most common neoplasm associated with Horner syndrome is neuroblastoma. Previously, experts recommended urine catecholamine as an adequate screening test, but a recent study found that imaging was a more effective tool to evaluate Horner syndrome without a known cause. In their study, one-third of children without a known cause for Horner syndrome had tumors detected by magnetic resonance imaging, and of those children, two-thirds had neuroblastoma. Interestingly, none of their patients had abnormal random urine catecholamine screens. For the patient in the vignette, the next appropriate step would be to obtain imaging of the head, neck, and chest. Some experts would add abdominal imaging, but this is controversial unless other physical examination findings suggest the presence of a mass.

Item C121A

Child with heterochromia. Courtesy of M. Rimsza.

Horner syndrome is one form of ptosis, a common eyelid disorder resulting from dysfunction of the muscles that elevate the upper eyelid. The frequency of ptosis in the United States is not defined; a recent study over a 40-year period in 1 county in Minnesota found an incidence of 7.9

per 100,000 people younger than 19 years of age. Of those, nearly 90% were classified as congenital, ie, occurring before 1 year of age, and three-quarters of those with early onset were classified as having simple congenital ptosis (called in some studies myogenic developmental abnormality). In patients who do not have simple congenital ptosis, one must consider genetic, mechanical, mitochondrial, developmental or embryologic, traumatic, neoplastic, and neurological causes (Item C121B).

Item C121B. Causes of Ptosis.

Category	Examples
Developmental/embryonic	• Synkinetic (aberrant innervations of the levator muscle) • Aponeurotic • Superior rectus weakness • Simple congenital ptosis
Genetic	• Blepharophimosis syndrome • Congenital fibrosis of the extraocular muscles • Myotonic dystrophy • Oculopharyngeal muscular dystrophy
Mechanical/obstructive	• Hemangioma • Eyelid tumor • Chalazion • Brow ptosis
Mitochondrial	• Kearns-Sayre syndrome • Chronic progressive external ophthalmoplegia
Neoplastic	• Chest/neck tumors • Neuroblastoma • Neurofibromatosis
Neurological	• Myasthenia gravis • Horner syndrome • Oculomotor nerve palsy • Oculopharyngeal muscular dystrophy
Trauma	• Eyelid trauma • Head, neck, chest surgery • Brachial plexus injury • Other birth trauma

Courtesy of K. Bowen

In addition to addressing the underlying condition, the most important consideration for the pediatrician and ophthalmologist dealing with a child with ptosis is to recognize the risk of developing amblyopia because of obstruction of the visual axis. Treatment of the underlying condition may lead to resolution, and mild cases may not require treatment otherwise. More severely affected patients require corrective surgery because most ptosis will not improve over time.

An edrophonium test would be used to diagnose myasthenia gravis, another rare cause of ptosis. A key characteristic of myasthenia is the worsening of ptosis as the muscle fatigues. The patient in the vignette has persistent ptosis and additional symptoms (lymph node enlargement) that would make myasthenia a less likely diagnosis. While ophthalmology consult may eventually be needed for this patient if the ptosis persists, ruling out a neoplasm is the most immediate concern. Age of onset and additional symptoms make brachial plexus injury unlikely and therefore physical therapy input is not currently required. Given the constellation of symptoms, the noninflamed lymph node is more likely to represent neoplasm than infection, so antibiotic treatment is not warranted.

PREP Pearls
- Ptosis is a common condition resulting from dysfunction of the muscles that elevate the eyelid.
- Congenital ptosis is the most common form and presents during the first year of life.
- Horner syndrome consists of ptosis, miosis, anisocoria, facial anhidrosis, and heterochromia. It can be the result of trauma, infection, neoplasm, migraine, or carotid artery abnormality.
- In children, the most common neoplasm causing Horner syndrome is neuroblastoma.

ABP Content Specifications(s)
- Differentiate the clinical findings associated with congenital ptosis from those of acquired ptosis

Suggested Readings
- Griepentrog GJ, Diehl NN, Mohney BG. Incidence and demographics of childhood ptosis. Ophthalmology. 2011;118(6):1180-1183. DOI: http://dx.doi.org/10.1016/j.ophtha.2010.10.026.

Question 122

The father of a 14-year-old adolescent calls your office because of concerns about his son's behavior. The father says that his son seems withdrawn and paranoid. The adolescent rarely wants to spend time with the family. Whenever the parents plan a family event, the son finds an excuse to not attend. He constantly asks his mother how he looks and spends 30 min selecting his clothes each day. He frequently argues with his 13-year-old sister and insists that she is always looking at him. The father reports that his son has friends at school and that he is an honor roll student.
Of the following, you advise the father that

A.   his son needs an evaluation by a mental health professional for depression

B.   his son's behavior seems appropriate for his developmental age

C.   his son's behavior seems consistent with a narcissistic personality

D.   the parents should be concerned that he will harm his sister

E.   the parents should insist on his participation in family activities

Question 122   **Preferred Response: B**

The primary outcomes of adolescent development include biological and sexual maturation, development of personal identity, formation of sexual relationships, and the development of independence. Adolescent development has 3 main components: physical, psychosocial, and cognitive development. The processes of development can be categorized by early, middle, and late adolescent tasks as described in the second suggested reading.

Adolescents progress from concrete thinking to abstract thinking, which means that the early adolescent has limited capacity to recognize the consequences of risk-taking behaviors. Emotional separation from parents is a characteristic task of adolescent development; therefore, an adolescent's desire to spend less time with their parents should be expected. Additionally, as adolescents adjust to their changing physical appearance and develop a mature self-image, they are often very concerned about their physical appearance and appear very self-absorbed. The behavior of the adolescent in this vignette is appropriate for his developmental age.

The prevalence of depression in adolescents 14 to 18 years of age ranges from 4% to 7%. The American Academy of Pediatrics recommends routine screening of adolescents for depression. It is important to recognize that many adolescents may experience transient depressive symptoms, but an adolescent with clinical depression has depressed or irritable mood, loss of interest or pleasure, and at least 3 of the following symptoms for a 2-week period:
1. Significant weight loss or decrease in appetite (more than 5% of body weight in a month)
2. Insomnia or hypersomnia
3. Psychomotor agitation or retardation
4. Fatigue or lack of energy
5. Feelings of worthlessness or guilt
6. Decreased concentration or indecisiveness
7. Recurrent thoughts of death or suicide

The adolescent described in this vignette is exhibiting normal developmental tasks with his desired separation from family and an interest in self-appearance.

PREP Pearls
• Adolescent development has 3 main components: physical, psychosocial, and cognitive development.
• Adolescents progress from concrete thinking to abstract thinking, which means that the early adolescent has limited capacity to recognize the consequences of risk-taking behaviors.

ABP Content Specifications(s)
- Recognize the sequence of emotional and cognitive development and physical maturation
- Recognize the tasks and features of early, mid, and late adolescent socioemotional development
- Recognize how thought processes in early, middle, and late adolescence influence problem solving and risk taking

Suggested Readings
- American Psychiatric Association. Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition. Washington, DC: American Psychiatric Association; 2013: 991 pp.

Question 123

You are seeing a 10-year-old boy in your office with complaint of joint pain and swelling. He has significant right knee and ankle swelling that started 2 days ago, which is very painful. He also complains of right heel and second toe pain. A review of systems is significant for a scaly rash on the soles of the feet, low-grade fevers (38.3°C), myalgia, painless oral ulcers, and conjunctivitis of 2-day duration. Two weeks ago, the boy had an episode of diarrhea, which self-resolved. While other family members had diarrhea as well, no family member shares his current symptoms. On physical examination, the boy has erythema overlying the right knee and ankle with significant swelling and pain, and a limited range of motion. He is tender to palpation along the right posterior heel and has dactylitis of the second toe. He has 2 shallow ulcers on the buccal mucosa and bilateral conjunctivitis. There are hyperkeratotic skin lesions on his soles bilaterally. The remainder of the physical examination is unremarkable.

Of the following, the boy's MOST likely diagnosis is

A.   Bechet disease

B.   Crohn disease

C.   mononucleosis

D.   psoriatic arthritis

E.   reactive arthritis

Question 123   **Preferred Response: E**

The boy in the vignette has arthritis, conjunctivitis, rash, myalgia, and oral ulcers that were associated with a diarrheal illness 2 weeks ago consistent with reactive arthritis. Reactive arthritis is characterized by arthritis that typically occurs 1 to 3 weeks after an infection of the gastrointestinal or urogenital tract. Microbial cultures are often negative by the time the arthritis presents. Gastrointestinal infections most commonly associated with reactive arthritis are Yersinia, Salmonella, Shigella, and Campylobacter jejuni. Urogenital tract infections commonly associated with reactive arthritis are Chlamydia trachomatis, Neisseria gonorrhoeae, Mycoplasma genitalium, and Ureaplasma urealyticum.

Reactive arthritis most frequently presents as an oligoarticular arthritis of the lower extremity, However, multiple joint and upper extremity involvement can occur in some cases. Other features include dactylitis, enthesitis, and bursitis. Some patients with reactive arthritis can have inflammatory back pain. Extra-articular manifestations of reactive arthritis include conjunctivitis or uveitis. Reactive arthritis associated with urogenital infections can be associated with urethritis and cervicitis. Skin lesions can occur with small hard nodules on the feet or hands called keratoderma blennorrhagicum. Penile lesions such as balanitis circinata can develop. Some patients demonstrate painless ulcers in the mouth. Rarely aortic regurgitation, pericarditis, and conduction abnormalities can occur in prolonged cases.

Although no laboratory studies are diagnostic and there are no specific diagnostic criteria, HLA-B27 positivity is associated with the development of reactive arthritis. Laboratory studies looking for the triggering infections (testing for Chlamydia trachomatis, stool cultures, or evidence of synovial infection by immunohistology) can support the diagnosis. Nonspecific inflammatory markers are frequently elevated.

Treatment of reactive arthritis includes treatment of the preceding infection, when appropriate, and nonsteroidal anti-inflammatory agents. Steroid injections can occasionally be used in monoarticular disease. If the arthritis becomes chronic (lasting >6 months), other treatments such as immunosuppressive agents may be used.

Bechet disease can present with arthritis, gastrointestinal involvement and oral ulcers, however the boy in this vignette has diarrhea that is most likely infectious because the rest of the family had similar findings. In addition, Bechet disease is rare. Crohn disease does present with diarrhea and arthritis, but the diarrhea is noninfectious, chronic and often associated with bloody stools. Mononucleosis can present with arthritis, but in the setting of an infectious diarrhea followed by arthritis, reactive arthritis is most likely. The rash described as hyperkeratotic skin lesions on his soles is consistent with keratoderma blennorrhagicum not psoriasis; therefore, psoriatic arthritis is incorrect.

PREP Pearls
- Reactive arthritis typically occurs 1 to 3 weeks after an infection of the gastrointestinal or urogenital tract.
- Reactive arthritis most frequently presents as an oligoarticular arthritis of the lower extremity.
- Initial treatment of reactive arthritis includes treatment of the preceding infection when appropriate and nonsteroidal anti-inflammatory agents.

ABP Content Specifications(s)
- Recognize the clinical findings associated with reactive arthritis and manage appropriately

Suggested Readings
- Selmi C, Gershwin ME. Diagnosis and classification of reactive arthritis. Autoimmun Rev. 2014;13:546-549. DOI:http://dx.doi.org/10.1016/j.autrev.2014.01.005.

Question 124

A 4-year-old, previously healthy boy presents to the emergency department with a facial laceration that requires 4 simple, interrupted sutures. His vital signs are normal for age, and other than the laceration, his physical examination is unremarkable. The boy has no allergies and there is no family history of adverse reaction to anesthetic or sedative medications. Despite local anesthesia, distraction, and reassurance, the boy is uncooperative and sedation is required to perform the repair. He receives a dose of intranasal midazolam, 0.5 mg/kg. Five minutes later, he is calm and has a normal response to verbal stimuli.

Of the following, while this boy is sedated, it would be MOST appropriate to continuously monitor his

A.   arterial blood pressure

B.   electrocardiogram

C.   exhaled tidal volume

D.   pulse oximetry

E.   pupillary response

Question 124   **Preferred Response: D**

The child in the vignette requires sedation for repair of his facial laceration because he is uncooperative. Sedation is often required for laceration repairs in this age group because anxiety, as well as pain, is often a consideration. Monitoring of pulse oximetry is warranted when a dose of intranasal midazolam is given with the effect of anxiolysis.

In 2002, the American Academy of Pediatrics (AAP) established a continuum of sedation ranging from completely awake to general anesthesia, in which the patient is completely unarousable (Item C124). With increasing levels of sedation there are increased risks to the patient. When choosing appropriate sedation measures, the clinician must consider several patient-specific factors, such as any preexisting medical condition, maturity level, the need for analgesia, noxious nature of the procedure, and capability of facilities and personnel. With anxiolytic sedation, the patient should maintain a normal response to verbal stimuli. Conscious sedation is considered moderate sedation/analgesia, in which the patient has a decreased level of consciousness but responds appropriately to physical stimulation or verbal commands. At this level, the patient still maintains airway protective reflexes. With deep sedation, the patient should exhibit purposeful response to repeated or painful stimuli. Under general anesthesia, the patient is unarousable.

Item C124. Continuum of Sedation.

Awake
↕
Anxiolysis
(Normal response to verbal stimuli)
↕
Moderate Sedation/Analgesia
(Purposeful response to verbal or tactile stimuli)
↕
Deep Sedation
(Purposeful response to repeated or painful stimuli)
↕
General Anesthesia
(Unarousable)
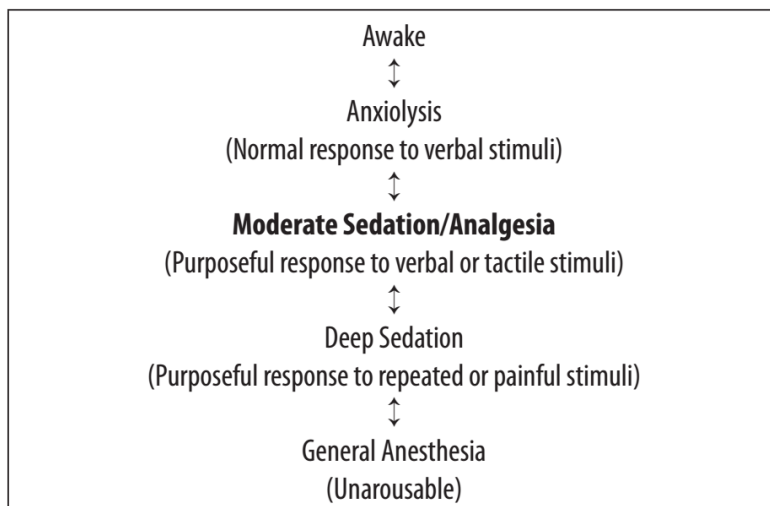
Reprinted with permission from Koh JL, Palermo T. Conscious sedation: reality or myth? *Pediatr Rev.* 2007;28:243-248

A facility in which pharmacologic sedation is used in children should include minimum standards of personnel and equipment. At least 1 individual involved in delivering the sedation should be trained in airway management and pediatric advanced life support. All personnel should have training in basic cardiac life support and education in the sedatives and analgesics used. Size-appropriate resuscitation equipment should be readily available, including ventilation bag and mask, oxygen, and resuscitation medications.

The principal adverse effects of benzodiazepines and narcotics include hypotension, bradycardia, and respiratory depression. Neurologic status and treatment effect can be assessed with verbal and mild tactile stimuli. Monitoring of a patient receiving these medications for pharmacologic anxiolysis should include continuous pulse oximetry, visual assessment of ventilation, and noninvasive blood pressure measurement every 5 minutes. Continuous monitoring of arterial blood pressure is too invasive for this setting. Because these medications generally have minimal effect on cardiac conduction, electrocardiographic monitoring is not required in the absence of significant preexisting cardiovascular disease. Bradycardia can be detected on pulse oximetry. Hypoventilation can be detected visually or on pulse oximetry. Noninvasive capnography by nasal cannula is sometimes used in conscious sedation for procedures involving decreased access for visual monitoring, such as magnetic resonance imaging. Monitoring of exhaled tidal volume would require endotracheal intubation, which is usually not necessary in sedations short of general anesthesia. Monitoring of the pupillary response is not necessary unless an overdose is suspected.

PREP Pearls
- Sedation in children is represented by a continuum ranging from completely awake to anxiolysis, moderate sedation/analgesia, deep sedation, and finally general anesthesia (in which the child is completely unarousable).
- Children undergoing conscious procedural sedation exhibit purposeful reaction to verbal stimulation.
- Children undergoing conscious/moderate procedural sedation require continuous pulse oximetry, visual assessment of breathing, and noninvasive blood pressure monitoring.

ABP Content Specifications(s)
- Plan the appropriate observation and monitoring protocol for a patient who is undergoing procedural sedation
- Understand the differences in procedural sedation, deep sedation, and general anesthesia

Suggested Readings
- American Academy of Pediatrics Committee on Drugs. Guidelines for monitoring and management of pediatric patients during and after sedation for diagnostic and therapeutic procedures: addendum. Pediatrics. 2002;110:836-838. DOI:http://dx.doi.org/10.1542/peds.110.4.836.

Question 125

A 2-year-old boy presents to the emergency department with fever, vomiting, and behavioral changes. He has had increased agitation and repeated episodes of facial twitching. Vital signs show a temperature of 39.5°C, respiratory rate of 38 breaths/min, heart rate of 131 beats/minute, blood pressure of 124/76 mm Hg, and a weight of 12.8 kg. On physical examination, he is irritable, has nuchal rigidity, and several insect bites are noted over his legs. Laboratory data shows:

- White blood cells, 20,200/µL (20.2 x 10^9/L)
- Hemoglobin, 11.3 g/dL (113 g/L)
- Platelets, 341 x 10^3/µL (341 x 10^9/L)
- Differential, 62% segmented neutrophils, 26% lymphocytes, 12% monocytes
- Cerebrospinal fluid (CSF) white blood cells, 145/µL (23% segmented neutrophils, 45% lymphocytes, 32% monocytes)
- CSF red blood cells, 2/µL
- CSF glucose, 49 mg/dL (2.7 mmol/L)
- CSF protein, 15 mg/dL
- CSF West Nile immunoglobulin M, 1:64

The resident working with you asks about the epidemiology of West Nile virus.

Of the following, the BEST response to your resident's question is

A.   avian deaths from West Nile virus have been predictive of human cases

B.   horses serve as a reservoir for avian infections with West Nile virus

C.   a human-mosquito-human cycle of West Nile virus maintains transmission

D.   the mosquito vector for West Nile virus tends to bite during the day

E.   peak case numbers for West Nile virus usually occur in late spring

Question 125   **Preferred Response: A**

Avian species serve as a reservoir for West Nile virus and deaths in the reservoir is predictive of human cases.

The term arbovirus is an abbreviation of arthropod-borne viruses, which alludes to the vectors of transmission (mosquitoes, ticks, sand flies, and midges). Transmission usually occurs between birds or small mammals and the arthropod vectors. Infections in humans and domestic animals do not maintain transmission for most arboviruses. For West Nile virus, birds serve as a reservoir, and avian-mosquito-avian cycles maintain transmission. West Nile virus infections in humans and horses are considered dead end infections. However, there are exceptions to this. In dengue, chikungunya, and yellow fever, arthropods that feed on infected humans can then infect other humans.

Culex mosquitoes are the principal vectors for West Nile virus. Culex mosquitoes preferentially have blood meals at dusk or after dark. Most infections occur in the summer or early fall, which coincides with peak mosquito activity.

West Nile infections are asymptomatic in 80% of cases. Although most infections caused by arboviruses are subclinical, they can manifest as an acute febrile illness or neurologic disease. Manifestations during the acute febrile illness can be nonspecific and include headaches, myalgias, arthralgias, and exanthems. Some arboviruses have a predilection for causing particular symptoms, such as the severe joint pain seen in chikungunya virus infection, and bone pain and retro-orbital headache seen in dengue fever. Neuroinvasive disease occurs in less than 1% of those that are infected with West Nile virus. Manifestations can range from aseptic meningitis to encephalitis and even flaccid paralysis. Although most individuals have asymptomatic infection, La Crosse virus can cause severe neurologic disease, especially in children. In general, severe manifestations of arbovirus infections tend to occur more commonly in adults compared to children. Dengue and yellow fever can cause hemorrhagic disease manifestations.

PREP Pearls
- For West Nile virus, birds serve as a reservoir and avian-mosquito-avian cycles maintain transmission.
- Most West Nile virus infections occur in the summer or early fall, which coincides with peak mosquito activity.
- Although most infections caused by arboviruses are subclinical, they can manifest as an acute febrile illness or neurologic disease, including aseptic meningitis, encephalitis, and flaccid paralysis.

ABP Content Specifications(s)
- Recognize the clinical features associated with arbovirus infection (eg, West Nile, dengue fever)
- Understand the epidemiology of arbovirus, including West Nile virus

<u>**Suggested Readings**</u>

*   American Academy of Pediatrics. Arboviruses. In: Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:240-246.

Question 126

The parents of a 7-year-old girl present to your office for evaluation. The girl was diagnosed with Henoch-Schönlein purpura (HSP) by your associate 2 months ago. Since then, she has had multiple urinalyses showing 2-3+ blood by urine test strips and 10 to 50 red blood cells per high power field (HPF). Her record shows physical examinations on previous encounters remarkable only for palpable purpuric macules on the legs. Further studies include renal ultrasonography and serum chemistry, which were normal when last checked 6 weeks ago.

Her vital signs today show a temperature of 37.9°C, heart rate of 72 beats/min, respiratory rate of 18 breaths/min, and blood pressure of 110/60 mm Hg. Her physical examination is significant for palpable purpuric macules on the legs with occasional confluence. According to the parents, her rash is much improved.

Her urinalysis today demonstrates a specific gravity of 1.035, a pH of 6.0, 3+ blood, 1+ proteinuria, and no leukocyte esterase 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.   children have poorer prognosis than adults with renal involvement because of HSP

B.   her hematuria is unrelated to her history of HSP

C.   microscopic hematuria is uncommon in patients with HSP

D.   nephrotic range proteinuria would increase her risk for progressive renal disease

E.   rash persisting for more than a month indicates severe renal involvement

Question 126   **Preferred Response: D**

Henoch-Schönlein purpura (HSP) is one of the most common causes of vasculitis in children. The clinical manifestations of HSP include nonthrombocytopenic palpable purpura, arthralgia, abdominal pain, and renal involvement. Henoch-Schönlein purpura is self-limiting in the majority of patients, however, renal manifestations secondary to HSP vasculitis may be chronic and associated with renal failure.

Renal involvement is reported in 20% to 50% of the patients with HSP and accounts for the long term morbidity with this vasculitis. Renal manifestations of HSP include transient or persistent (lasting greater than 6 months) hematuria, recurrent gross hematuria (along with recurrent skin rash), proteinuria, nephritic syndrome (hematuria, hypertension, and azotemia), nephrotic syndrome (nephrotic range proteinuria, hypoalbuminemia, edema), or nephritic-nephrotic syndrome. Microscopic hematuria is the most common renal manifestation of HSP vasculitis. Proteinuria (mild and non-nephrotic), with and without hematuria, is more common than nephrotic range proteinuria. Nephrotic syndrome (proteinuria, hypoalbuminemia, and edema) is present in only a minority of the patients. The presence of nephrotic syndrome, renal insufficiency (elevated blood urea nitrogen and serum creatinine), and hypertension (nephritic syndrome) is associated with poorer renal prognosis in patients with HSP vasculitis. Adults with HSP vasculitis have similar clinical manifestations to children, but are at increased risk for progressive renal disease. Risks and severity of renal involvement caused by HSP vasculitis cannot be predicted from the severity of extrarenal involvement such as skin rash, joint, or abdominal pain.

Renal manifestations in patients with HSP appear commonly in the first month after diagnosis (around 80%) and in the majority of patients (95%) by 6 months. Therefore, patients presenting with extrarenal manifestations of HSP should have a urinalysis at presentation and frequently thereafter to look for evidence of renal involvement secondary to HSP vasculitis. The frequency of urinalysis after the initial onset of HSP rash is determined based on the urinary findings at presentation. As the majority of the patients develop renal manifestations in the first month, weekly urinalysis in the first 4 weeks will lead to early identification of renal involvement. It is estimated that 2% to 5% of the patients with renal involvement caused by HSP progress to end stage renal disease. Patients with nephritic or nephrotic syndrome have a higher risk of developing chronic kidney disease (10%-20%), compared to patients with persistent hematuria or proteinuria that does not fall into the nephrotic or nephritic range (around 2%). Patients with renal manifestations associated with increased risk for chronic kidney disease merit frequent urinalysis and should be managed in consultation with a pediatric nephrologist.

PREP Pearls
- Renal involvement is reported in 20% to 50% of the patients with Henoch-Schönlein purpura (HSP) and accounts for the long term morbidity with this vasculitis.
- Microscopic hematuria is the most common renal manifestation of HSP vasculitis.
- The presence of nephrotic syndrome, renal insufficiency (elevated blood urea nitrogen and serum creatinine), and hypertension (nephritic syndrome) is associated with poorer long-term renal prognosis in patients with HSP.

- As the majority of patients develop renal manifestations in the first month, weekly urinalysis in the first 4 weeks will lead to early identification of renal involvement.

ABP Content Specifications(s)
- Recognize the renal findings associated with Henoch-Schönlein purpura
- Understand the prognostic implications when Henoch-Schönlein purpura is associated with nephrotic syndrome

Suggested Readings
- Adrogue HE, Flores FX. Henoch-Schönlein purpura. In: McInerny TK, Adam HM, Campbell DE, Kamat DM, Kelleher KJ, eds.American Academy of Pediatrics Textbook of Pediatric Care. Elk Grove Village, IL: American Academy of Pediatrics; 2009:2112-2115.

Question 127

You are preparing to discharge a 2-month-old preterm infant with a history of duodenal atresia, requiring prolonged total parenteral nutrition exposure that resulted in cholestasis. Recent laboratory work shows:

- Total bilirubin, 5.7 mg/dL (97.5 µmol/L)
- Direct bilirubin, 3.5 mg/dL (59.9 µmol/L)

In reviewing the growth chart with the dietician, the age-adjusted height is at the tenth percentile and the weight is at the third percentile. The infant is currently receiving a partially hydrolyzed formula concentrated to 24 kcal/oz. You would like to initiate a dietary supplement to maximize growth.

Of the following, the supplement that is MOST likely to be well tolerated and result in improved growth is

A.   flaxseed oil

B.   medium chain triglyceride oil

C.   olive oil

D.   omega-3 polyunsaturated fatty acids (fish oil)

E.   soybean oil

Question 127   **Preferred Response: B**

The infant in this vignette has a history of duodenal atresia status post-bowel resection, resulting in short bowel syndrome with intestinal failure. The infant has cholestasis caused by chronic total parenteral nutrition use. Despite both enteral and parenteral nutrition, this patient is failing to thrive. The best way to maximize calories without requiring large increases in volume is to use medium chain triglyceride oil (MCT oil), which is well absorbed in the small bowel. The other listed oils also increase caloric density in a small volume, but are not as easily absorbed as MCT oil.

Fat requirements vary by age, with higher needs in infancy that slowly decrease as children pass 2 years of age (Item C127). Premature infants and infants have fat malabsorption, resulting in increased needs that begin to decrease by 6 months of age. Fats are needed in our diet to provide essential fatty acids and to assist with the absorption of fat soluble vitamins (vitamins A, D, E, and K).

Item C127. Fat Requirements by Age.

Age	American Academy of Pediatrics Recommendations
	Percent of calories from fat
Premature infants	50% of calories
Infants	40% to 50% of calories
1 to 2 years	30% to 40% of calories
2 to 3 years	30% to 35% of calories
4 to 18 years	25% to 35% calories from fat Less than 10% saturated fat

Courtesy of C. Waasdorp Hurtado

Children have increased dietary fat requirements for several reasons, including fat malabsorption, failure to thrive, and increased metabolic demand. Children with complex congenital heart disease must frequently limit their daily volume intake, and additional calories from fats allow for adequate calories while respecting fluid limitations. Fat malabsorption occurs:

1.   Normally in infancy because of immature bile synthesis
2.   In chronic liver diseases due to decreased bile production and secretion
3.   In pancreatic exocrine insufficiency as a result of decreased secretion of enzymes necessary for the breakdown and absorption of fats

PREP Pearls
•   Fat requirements in children decrease with age.
•   Fat requirements in premature infants are higher than in term infants.
•   Increased fat needs are found in children with fat malabsorption, failure to thrive, and increased metabolic demand.
•   Children with complex congenital heart disease may have increased fat needs due to intake restricted by volume limitations.

ABP Content Specifications(s)
•   Understand the dietary fat requirements of patients of various ages, including those born prematurely, and the circumstances in which those requirements may change

Suggested Readings
•   Uauy R, Dangour AD. Fat and fatty acid requirements and recommendations for infants 0-2 years and children of 2-18 years. Ann Nutr Metab. 2009;551(1-3):76-96. DOI: http://dx.doi.org/10.1159/000228997.

Question 128

A 10-year-old boy presents as a new patient to your office for a routine health supervision visit. You note that his right leg and arm are slightly larger than his left. A physical examination reveals macrocephaly, numerous skin tags, 3 to 4 palpable soft nodules on his arms and legs, 2 capillary hemangiomas on his right leg, and unusual freckling to his penis. The mother reports that she has a history of a thyroid goiter, breast cancer, and uterine fibroids.

Of the following, the MOST likely diagnosis is

A.   Beckwith-Wiedemann syndrome

B.   Klippel-Trénaunay syndrome

C.   neurofibromatosis type 1

D.   PTEN hamartoma tumor syndrome

E.   Sotos syndrome

Question 128   **Preferred Response: D**

The patient in this vignette has PTEN hamartoma tumor syndrome (PHTS) caused by a pathogenic variant in the PTEN gene. Patients with PHTS present as Cowden syndrome, Bannayan-Riley-Ruvalcaba syndrome (BRRS), PTEN-related Proteus syndrome, or Proteus-like syndrome. This patient has the BRRS presentation that classically presents with macrocephaly, autism, developmental delay (IQ < 75), intestinal hamartomatous polyposis, lipomas, and pigmented macules of the glans penis.PTEN-related disorders can have a spectrum of presentations. His mother's features are more consistent with Cowden syndrome that predisposes individuals to benign and malignant tumors of the thyroid, breast, and endometrium. They can also have macrocephaly, trichilemmomas (a benign cutaneous neoplasm developing from hair follicles), oral papillomas, and papillomatous papules that present by the second to third decade of life. Vascular features can include arteriovenous malformations or hemangiomas. The lifetime risk for developing specific patterns of cancers is: breast cancer, 85%; epithelial thyroid cancer, 35%; and endometrial cancer, 28%. Other benign tumors commonly seen include lipomas, hamartomatous intestinal polyps, fibromas, and uterine fibroids. It is an autosomal dominant disorder. In this case vignette, it was clearly inherited from his mother.

A suspected diagnosis of PHTS is typically based on clinical signs, but by definition, the diagnosis of PHTS is definitively made only when a PTEN pathogenic mutation is detected. Consensus diagnostic criteria for Cowden syndrome have been developed by the National Comprehensive Cancer Network. There is also a free online risk calculator, produced by the Cleveland Clinical Foundation, for adult and pediatric patients suspected to have PTEN mutations (http://www.lerner.ccf.org/gmi/ccscore/).

This patient has an overgrowth syndrome in the presence of hemihyperplasia. Other disorders that must be considered in this scenario include Beckwith-Wiedemann syndrome, isolated hemihyperplasia, Klippel–Trénaunay syndrome (formerly known as Klippel-Trénaunay-Weber syndrome), and neurofibromatosis type 1.

Beckwith-Wiedemann syndrome is an overgrowth disorder manifested by macrosomia, macroglossia, neonatal hypoglycemia, ear creases and pits, hemihypertrophy, and visceromegaly. Patients also can have embryonal tumors (Wilms tumor, hepatoblastoma, neuroblastoma), umbilical hernia or omphalocele, nephrocalcinosis, medullary sponge kidney disease, cardiomegaly, and nephromegaly. Traditionally, the macrosomia, macroglossia, and hypoglycemia are noted in the neonatal period. Hemihyperplasia is noted in segmental regions of the body or specific organs. Developmental and cognitive outcomes are typically normal. Klippel-Trénaunay syndrome is a condition that impacts the development of blood vessels, soft tissues, and bones with 3 classic features that include a port-wine stain of 1 limb (typically 1 leg), abnormal overgrowth of soft tissues and bones, and venous malformations (varicose veins and a predisposition to deep vein thrombosis).

Neurofibromatosis 1 is an autosomal dominant disorder secondary to an NF1 gene mutation that presents with multiple café-au-lait macules, axillary and inguinal freckling, multiple cutaneous neurofibromas, and iris Lisch nodules. Learning disabilities and macrocephaly are common.

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Hemihyperplasia, if present, is secondary to a plexiform neurofibroma that can be disfiguring and compromise function.

Sotos syndrome is an autosomal dominant disorder caused by NSD1 gene mutations or deletions that presents with cardinal features, which include a typical facial appearance, overgrowth, and learning disability. Facial dysmorphology is characterized by sparse frontotemporal hair, high-bossed forehead, downslanting palpebral fissures, a long narrow face, and a prominent narrow jaw. It does not typically manifest as hemihyperplasia, but a generalized overgrowth of the body.

PREP Pearls
- Within the differential diagnosis of a child with hemihyperplasia, a physician should consider Beckwith–Wiedemann syndrome, Proteus syndrome, PTEN hamartoma tumor syndrome, Klippel–Trénauny syndrome, isolated hemihyperplasia, and neurofibromatosis type 1.
- PTEN hamartoma tumor syndrome caused by a pathogenic variant in the PTEN gene can present as Cowden syndrome, Bannayan-Riley-Ruvalcaba syndrome, PTEN-related Proteus syndrome, or Proteus-like syndrome. This presentation can place patients or other family members at high risk for particular forms of cancer, intellectual disability, and autism.
- Bannayan-Riley-Ruvalcaba syndrome classically presents with macrocephaly, autism, developmental delay (IQ < 75), intestinal hamartomatous polyposis, lipomas, and pigmented macules of the glans penis.
- Cowden syndrome patients can present with macrocephaly, trichilemmomas (a benign cutaneous neoplasm developing from hair follicles), papillomatous papules, uterine fibroids, and early onset cancers of the breast, thyroid, or uterus.

ABP Content Specifications(s)
- Recognize the clinical features and risks associated with overgrowth syndrome

Suggested Readings
- Cleveland Clinic Lerner Research Institute. Risk calculator for estimating a patient's risk for PTEN mutation. Cleveland Clinic Lerner Research Institute website. http://www.lerner.ccf.org/gmi/ccscore/ .

Question 129

During a routine health supervision visit for a healthy 7-month-old male infant, you notice that his right testis is easily palpable in the scrotum, but the left testis is not. The left scrotal sac is smaller than the right and is empty. You are not able to palpate the left testis. The infant was born at full term without complications and has no medical problems. He has been growing appropriately and the remainder of his physical examination is unremarkable. You discuss with his parents the implications of undescended testes.

Of the following, the MOST accurate statement is that

A.   early orchiopexy does not guarantee the preservation of fertility

B.   early orchiopexy will eliminate his increased risk for testicular cancer

C.   he has a 50% risk of a patent processus vaginalis

D.   he is at lower risk for blunt trauma to the testes

E.   he is at lower risk for testicular torsion

Question 129   **Preferred Response: A**

It is important to recognize undescended testes (UDT) on physical examination because of the significant impact of timely management and the potential for serious complications. Early orchiopexy will help to preserve fertility, but it is not a guarantee.

Undescended testis or cryptorchidism is common and is defined by failure of 1 or both testes to descend along the normal pathway into the scrotum. The testis usually begins this abdominal-to-inguinal-to-scrotal descent at 28 weeks of gestation. In infants with UDTs at birth, most testes will descend into the scrotum within the first few postnatal months. Because very few testes descend after 6 months of age, it is important to discuss the implications of UDT with parents, and inform them when it is time to pursue urologic or surgical referral. Referral for examination and possible exploratory surgery with orchiopexy between 6 months and 1 year of age is crucial to outcomes.

It is often difficult to distinguish UDT from retractile testis. Because infertility and malignancy are associated with UDT, it important to make the correct diagnosis. A history of UDT noted at birth, prematurity, and scrotal asymmetry on physical examination are all findings that significantly increase the likelihood that the testis is truly undescended. On physical examination, a retractile testis will be palpable outside the external inguinal ring and can be gently manipulated into the scrotum by overcoming the cremasteric reflex. Ultrasonography does not reliably distinguish an undescended from retractile testis.

The complications of UDT are many. It is estimated that as many as 90% of UDTs have an associated patent processus vaginalis and therefore an inguinal hernia can present at any time. Testes that remain in the inguinal canal are at greater risk for injury from blunt trauma because they can be compressed against the pubic bone. Testicular torsion is more common in UDT than in scrotal testes and the diagnosis is often delayed in cryptorchidism, resulting in a lower rate of salvage of the torsed testis. Impaired spermatogenesis in adults with a history of UDT is common. The contralateral descended testis is also at risk for germ cell loss, infertility, and malignancy. Bilateral involvement and increased duration of suprascrotal location of the testis increase the degree of germ cell dysfunction. After 1 year of age, the histology of the UDT begins to deteriorate. Hence, orchiopexy before age 1 year is preferred, with the ideal time being as soon as possible after 6 months of age. Surgical repositioning of the testis before puberty decreases the risk of testicular cancer, but does not completely eliminate it.

PREP Pearls
- Early orchiopexy for cases of undescended testes is important to help preserve fertility, but it is not a guarantee.
- Undescended testes are associated with an increased risk of infertility, malignancy, blunt trauma (for inguinal testes), testicular torsion, and inguinal hernia.
- Exploratory surgery with orchiopexy between 6 months and 1 year of age yields the best outcome in cases of undescended testis because the histology of an undescended testis begins to deteriorate after 1 year of age.

ABP Content Specifications(s)
•   Recognize complications associated with undescended testes
•   Plan the appropriate management of undescended testes

Suggested Readings
•   Callaghan P. Undescended testis. Pediatr Rev.
          2000;21(11):395.http://pedsinreview.aappublications.org/content/21/11/395.

Question 130

A 15-year-old adolescent boy presents to your office with a 1-month history of progressive fatigue and exercise intolerance. His physical examination is remarkable only for pallor. His weight is 60 kg. He specifically denies any history of hematuria, hematochezia, epistaxis, or unusual bruising. He has had a normal diet. The results of a complete blood cell count are shown in Item 130.

Laboratory test	Result
White blood cell count	11,000/µL (11.0 x 10⁹/L)
Hemoglobin	7.2 g/dL (72 g/L)
Mean corpuscular volume	62 fL
Platelet count	467 x 10³/µL (467 x 10⁹/L)
Reticulocyte count	0.5%

Of the following, the MOST appropriate next step is to advise him to start

A.   325 mg of iron sulfate twice daily on an empty stomach with a glass of orange juice and perform a stool guaiac

B.   325 mg of iron sulfate twice daily on an empty stomach with a glass of orange juice and repeat the blood cell counts in 4 weeks

C.   325 mg of iron sulfate twice daily with meals and perform a stool guaiac

D.   325 mg of iron sulfate twice daily with meals and repeat the blood cell counts in 4 weeks

E.   325 mg of iron sulfate twice daily with milk and perform a stool guaiac

Question 130   **Preferred Response: A**

This patient's presentation with new symptoms of anemia at 15 years of age suggests that his anemia is of new onset and therefore not hereditary. It is most likely that he has an iron deficiency anemia. A complete evaluation for iron deficiency includes a serum iron level, total iron binding capacity, reticulocyte count, and a ferritin level.

The size of red blood cells (RBC), as measured by the mean corpuscular volume (MCV) is in large part determined by the content of hemoglobin within the cell. Any deficiency in the components of hemoglobin will therefore result in a low MCV. The 2 primary components of hemoglobin that can be deficient are iron (deficiency of which causes decreased heme production) or the globin protein. Hemoglobin A, the normal adult hemoglobin variant, consists of 2 ß- and 2 a-globin chains, with the ß-globin gene located on chromosome 11 and the a-globin gene on chromosome 16. Mutations resulting in reduced production of either a- globin or ß-globin result in various thalassemia phenotypes, and present with a microcytic anemia. In order to form hemoglobin, the 4 globin subunits must bind to a molecule of heme, which is dependent on iron. Iron deficiency will therefore also result in a microcytic anemia.

Although the patient in the vignette could theoretically have a variant of thalassemia, his acute findings are more consistent with iron deficiency.

The human body has a tight regulatory system for the absorption of iron, but no mechanism of iron excretion. It would be highly unusual for a male adolescent with a relatively normal diet to develop iron deficiency from dietary restriction. It is therefore very important that any time a male adolescent presents with iron deficiency anemia, a source of iron loss must be sought. Causes of iron loss in this population would include gastrointestinal bleeding, paroxysmal nocturnal hemoglobinuria, and pulmonary hemosiderosis. Paroxysmal nocturnal hemoglobinuria can be evaluated through flow cytometry on a blood sample, and pulmonary hemosiderosis can be initially screened through a chest radiograph. If suspicion of hemosiderosis is strong, sputum analysis for hemosiderin-laden macrophages is recommended. While all 3 should be evaluated in the patient in the vignette, the most common cause would be gastrointestinal bleeding. It would therefore be most appropriate to perform a stool guaiac.

Iron therapy for iron deficiency should include 2 mg to 4 mg of elemental iron per kg of weight daily. Elemental iron is 20% of iron sulfate. For the patient in the vignette, the most appropriate dose of elemental iron would be 120 mg to 240 mg daily, which would be 600 mg to 1,200 mg of iron sulfate daily. Iron is best absorbed on an empty stomach with an acidic or neutral fluid. It is therefore most appropriate to take iron with water or orange juice. The high concentration of casein and whey proteins in milk inhibits iron absorption, and tea contains chelators that will bind the iron and prevent its absorption. Medications that decrease the acidic environment of the upper gastrointestinal tract may also impair absorption of iron.

PREP Pearls
- Iron deficiency in a male adolescent is rarely dietary and needs further evaluation to rule out occult bleeding.
- The nondietary causes of iron deficiency include blood loss (typically gastrointestinal), paroxysmal nocturnal hemoglobinuria, and pulmonary hemosiderosis.
- Iron should be administered on an empty stomach with water or an acidic drink such as orange juice.

ABP Content Specifications(s)
- Plan the appropriate diagnostic evaluation of iron deficiency
- Recognize the clinical findings associated with iron deficiency in patients of various ages

Suggested Readings
- DeLoughery TG. Microcytic anemia. N Engl J Med. 2012;371:1324-1331. DOI:http://dx.doi.org/10.1056/NEJMra1215361.

Question 131

You are seeing a 13-year-old adolescent who has developed significant school avoidance. She has periodically missed school over the past year when she had physical complaints or reported having severe anxious feelings before school. This has worsened recently over the past week with complaints of headache, stomachache, and anxiety before school each morning, causing her to miss school each day. The mother notes that these complaints are relieved when she stays at home by herself or when she goes to work with her mother. The adolescent denies any bullying occurring at her school. She has been a good student, except for missing assignments when she is absent from school. She has a history of being "clingy" with her mother periodically over the years.

Of the following, the management that would MOST likely produce a positive outcome is

A.   arrange for temporary home tutoring while outpatient counseling is initiated

B.   arrange for the parent to remain with the child in the classroom for 2 hours each morning

C.   create a plan for an immediate, unaccompanied return to the classroom

D.   prescribe lorazepam and arrange for her to use it as needed during the school day

E.   set up a plan where the child knows the parent will visit her once a day at school

Question 131   **Preferred Response: C**

School avoidance can happen with or without a psychiatric cause, for instance, avoiding school just because the child prefers their nonschool environment, which is also known as truancy. The adolescent in this vignette might have a separation anxiety disorder (given her history of being "clingy" with mom), or she might have a somatic symptom disorder. Her history of experiencing headaches and stomach aches right before going to school, which are then relieved as she avoids school, is a typical way for anxiety to manifest as physical symptoms. Therefore, her school avoidance is most likely related to anxiety.

Separation anxiety disorder is a developmentally inappropriate and excessive anxiety about separating from home or from an individual with a persistence beyond 4 weeks. While as many as half of early school age children demonstrate some separation anxiety symptoms, only about 4% develop a level of dysfunction consistent with a separation anxiety disorder. There are both genetic and social origins for the development of separation anxiety disorder. There may be an inborn low threshold for experiencing anxiety that enables not just the appearance of separation anxiety disorder, but also other anxiety disorders like generalized anxiety disorder and social phobia. Even in the absence of any particular genetic predisposition for experiencing anxiety, highly anxious parenting may teach children to adopt a fearful view of their world.

School avoidance can be a major problem when it occurs because it typically becomes increasingly difficult to resolve the longer the child remains out of school. One reason why prolonged avoidance is such a problem is that our brains interpret anxiety relief from avoidance as proof that a fear was well founded, and thus future anxious reactions to the same situation deepen. For children avoiding school, this means that their fears about school usually increase the longer their duration of avoidance, and it becomes more and more difficult to get them to return.

The hallmark of an effective school avoidance intervention involves getting the child back into school immediately without their parent sitting next to them. Supports of many forms can be provided as appropriate while the child is at school, such as homework or class work modifications, a plan for how the child will receive support by school staff, schedule modifications, etc. If any persisting anxiety is present, enrollment in psychotherapy would be appropriate. If the trigger for the avoidance was a truly aversive situation such as school bullying, then that will need to be addressed.

Temporary home tutoring is counter productive for anxiety driven school avoidance because it makes it easier for the child and family to avoid a return to school. Arranging for a parent to remain in the classroom is a strategy that parents might request for a young child with separation anxiety, but this is likely to only delay the separation crisis, as it nonverbally communicates to the child that they cannot handle the situation on their own, and it is distracting to child and classroom function while the parent is present. Setting up a plan for the parent to visit the child during the day at scheduled times effectively creates additional separation experiences for the child each day, and a daily opportunity for a child to "build their case" as for why they need to

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leave mid-day. A single separation at the start of school is usually easier on both parents and children in this situation. Pharmacologic intervention is not indicated.

PREP Pearls
- School avoidance is often related to anxiety, which may manifest as headaches and stomach aches before going to school.
- School avoidance becomes more intervention-resistant the longer the child remains out of school. An unaccompanied return to school as soon as possible is needed for these children.

ABP Content Specifications(s)
- Distinguish between separation anxiety and truancy as a cause of school absence
- Understand the relationship between separation anxiety with school phobia/refusal in patients of various ages
- Plan the appropriate management of separation anxiety of various etiologies
- Recognize the family dynamics associated with separation anxiety

Suggested Readings
- Bagnell AL. Anxiety and separation disorders. Pediatr Rev. 2011;32(10):440-446. DOI: http://dx.doi.org/10.1542/pir.32-10-440.

Question 132

A 4-year-old girl is brought to the emergency department with decreased level of consciousness. She is hypotensive and in a wide complex tachycardia on arrival. She is cardioverted with 2 J/kg and regains sinus rhythm. Her electrocardiogram shows low voltages with ST segment elevations (Item Q132). Her mother states that she was ill with a diarrheal illness 2 weeks ago and has been tired since that time, but became acutely ill and drowsy today. On physical examination, you see a lethargic child with pallor. Her heart rate is 180 beats/min, her respiratory rate is 40 breaths/min, and her breathing is shallow. Her blood pressure is 75/40 mm Hg. She is cool in her extremities with a capillary refill time of 4 seconds. Her pulse oximetry is not picking up well.

You notice that her jugular veins are distended. Her chest examination is significant for rales throughout and retractions. Her cardiac examination is significant for a difficult to palpate point of maximal impulse that is displaced to the left and weak. She has a regular rhythm, but it is rapid. You appreciate S1 and S2 and hear a third heart sound in early diastole but no murmur. Her chest radiograph shows pulmonary edema and an enlarged heart. You are planning admission to the intensive care unit and getting consultations arranged. You place the child on 100% oxygen by non-rebreather mask and establish intravenous access.
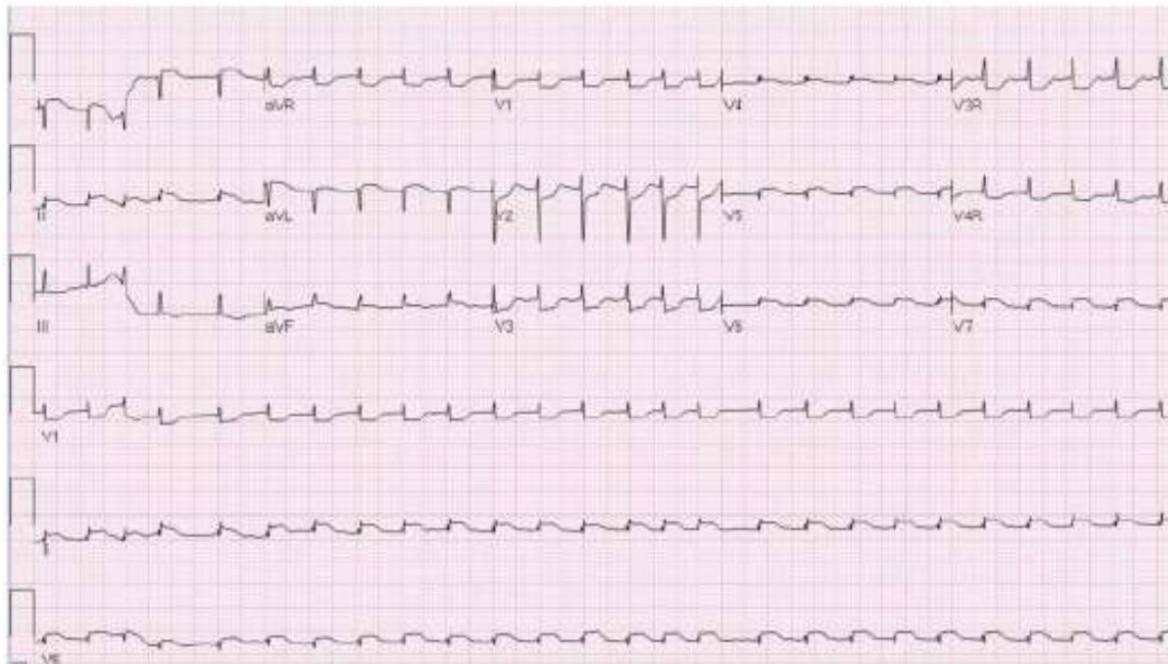
Item Q132. Electrocardiogram for the girl described in the vignette. Courtesy of E.A. Greene

Of the following, the intervention which would be MOST likely to improve the patient's status now is

A.   amiodarone, 5 mg/kg intravenous bolus

B.   dopamine, 10 µg/kg per min

C.   esmolol infusion, 50 µg/kg per min

D.   intravenous immunoglobulin infusion, 1 g/kg

E.   normal saline, 40 mL/kg intravenously

Question 132   **Preferred Response: B**

The girl in this vignette is in cardiogenic shock from viral myocarditis with an electrocardiogram (ECG) consistent with myocardial damage and low voltages. At this point in the patient's management, inotropic support is needed; of the choices given, the BEST next intervention would be dopamine. Her systemic output is very poor and additional support may well be required in this child. The girl presented in ventricular tachycardia (VT), which is a common presenting tachyarrhythmia in viral myocarditis and is associated with a more complicated course.

Intravenous immunoglobulin (IVIG) may be helpful later in the management of this patient, but would deliver too great of a fluid load at this point in the resuscitation, as the patient is already showing symptoms of pulmonary edema. Esmolol is a good treatment for suppression of VT, but would be contraindicated in this patient's management because of its negative inotropic effect. Amiodarone is also a good medication for treatment of VT, but a large bolus of 5 mg/kg, especially if given quickly, will very likely make the patient more hypotensive. A normal saline bolus may make the patient's pulmonary edema worse.

Given the patient's decompensation and presentation with VT, she will likely need advanced mechanical support, extracorporeal membrane oxygenation (ECMO), to support cardiac function and allow for management of her arrhythmias safely. Before ECMO can be initiated, dopamine can be used to help maintain cardiac output and treat cardiogenic shock. Careful monitoring needs to be maintained when dopamine is used because although it may improve function, it may also stimulate atrial or ventricular arrhythmias. Soon after initiation of inotropic support, antiarrhythmics would be indicated, especially if any further ectopy was noted. Lidocaine is a good option for ventricular ectopy in this setting, as there is less risk of hypotension.

Arrhythmias are common in myocarditis. Ventricular arrhythmias are most common, but there is also risk of atrioventricular block. Many viral etiologies are possible, including enteroviruses, adenovirus, parvovirus, cytomegalovirus, influenza A (H1N1), herpes, varicella, mumps, measles, rabies, hepatitis, rubella, rubeola, respiratory syncytial virus, HIV, and Epstein-Barr virus. Diagnostic evaluation is directed towards assessment of the degree of cardiac failure and impending respiratory failure, as well as determining the underlying etiology. A chest radiograph will show cardiomegaly and pulmonary edema. An ECG may show a pattern of myocardial damage, which can mimic ischemia or low voltages throughout the leads if there is not an arrhythmia on presentation. Intensive care will be needed to monitor for development of ventricular arrhythmia or heart block. An echocardiogram will be needed to assess the degree of systolic ventricular dysfunction and chamber size. Blood work, including cardiac enzymes and B-type natriuretic peptide, helps to assess the degree of myocardial damage and heart failure and will also aid in the overall evaluation and follow-up, but will not change the initial management. The initial management will be dictated by the clinical assessment of cardiac output. Viral polymerase chain reaction studies by serology or biopsy help establish the exact viral etiology, but does not impact the initial clinical management. Magnetic resonance imaging is also helpful to confirm the diagnosis of myocarditis.

PREP Pearls
- Support with inotropic agents such as dopamine is useful in the initial management of patients with myocarditis.
- Initial presentation of myocarditis may be a ventricular arrhythmia.
- Cardiogenic shock and arrhythmias may require extracorporeal membrane oxygenation in the initial management of myocarditis.

ABP Content Specifications(s)
- Recognize the clinical findings associated with myocarditis
- Recognize pathogens commonly associated with myocarditis
- Plan an appropriate diagnostic evaluation of myocarditis

Suggested Readings
- Anderson BR, Silver ES, Richmond ME, Liberman L. Usefulness of arrhythmias as predictors of death and resource utilization in children with myocarditis. Am J Cardiol. 2014;114(9):1400-1405. DOI:http://dx.doi.org/10.1016/j.amjcard.2014.07.074.

Question 133

A 2-year-old African American boy presents to your clinic for evaluation of fever. His mother states that he has had fever up to 39°C that started 5 days ago and his eyes and lips are red. The boy had 1 episode of vomiting yesterday and 1 watery stool today. He is eating and drinking less than usual, but urinating normally. The boy has a sibling that is currently well. On physical examination, the boy is irritable. His temperature is 39.2°C, heart rate is 120 beats/min, and his blood pressure is 80/50 mm Hg. He has bilateral, nonpurulent conjunctivitis. His lips and tongue are red with hypertrophied papillae. He has a morbilliform rash on his chest. The dorsum of his hands and feet appear edematous. The remainder of his physical examination is unremarkable. You order laboratory studies. His erythrocyte sedimentation rate is 40 mm/h and his white blood cell count is 15,200/μL (15.2 x 10^9/L). His other laboratory results are normal.

Of the following, the MOST concerning feature of the boy's presentation that warrants further evaluation is his

A.   duration of fever

B.   erythrocyte sedimentation rate

C.   tachycardia

D.   vomiting and diarrhea

E.   white blood cell count

Question 133   **Preferred Response: A**

The boy in the vignette has Kawasaki disease (KD). The most concerning feature that requires further evaluation of his symptoms is the duration of fever. The boy has several features of KD including fever of 5 days' duration, changes in his extremities, polymorphous rash, conjunctivitis, and mucosal findings. He has a slightly elevated erythrocyte sedimentation rate, which is nonspecific and could occur with any type of immune system activation, including a viral infection. The boy's tachycardia is likely caused by fever and not something that would necessarily warrant further workup. He had vomiting and diarrhea only once and urination is normal. The boy's white blood cell count is only slightly elevated and is not concerning in the setting of an acute illness.

The differential diagnosis of a child with this presentation is broad. Early in its course, KD mimics several conditions, including several viral illnesses that can present similarly, such as adenovirus, enterovirus, influenza, roseola infantum, and Epstein-Barr virus. In very young or unimmunized patients the practitioner must consider measles. Bacterial infections that can mimic KD include scarlet fever, cervical lymphadenitis, Rocky Mountain spotted fever, and leptospirosis. The skin peeling associated with KD is also seen in toxin-mediated diseases such as toxic shock syndrome or scalded skin syndrome. Drug hypersensitivity reactions, including Stevens-Johnson syndrome, can mimic KD. More rare conditions such as systemic juvenile idiopathic arthritis, and periodic fever syndromes can present with continuous fever of unknown origin. Mercury poisoning can also present similarly to KD.

The diagnostic criteria for KD include fever for at least 5 days, with at least 4 of the following: bilateral, non-exudative, bulbar conjunctivitis; oropharyngeal changes; cervical lymphadenopathy; polymorphous rash; and peripheral extremity changes. The oropharyngeal changes can include strawberry tongue, erythema of the oropharyngeal mucosa, or erythematous or cracking lips. The extremity changes include erythema or edema of the palms and soles in the acute phase or periungual desquamation of the fingertips in the later subacute phase of KD. The diagnosis can be difficult in cases where the patient has fever for 5 or more days but does not meet criteria for diagnosis. Incomplete KD is the term used for patients with fever for 5 or more days, but with only 2 or 3 of the principal clinical features. In such cases, echocardiography is recommended. KD can be diagnosed in patients when coronary artery disease is detected with 2-dimensional echocardiography. Other laboratory study abnormalities that support a diagnosis of KD are low albumin, elevated alanine aminotransferase, significantly elevated platelets, elevated white blood cell count, or sterile pyuria.

Risk factors for coronary artery aneurysms include male sex, age younger than 1 year, prolonged fever, delayed diagnosis, fever that persists after treatment, low hemoglobin, high white blood cell count, high absolute band count, very high erythrocyte sedimentation (ESR) or C-reactive protein, low platelet count, or low albumin. It should be noted that after intravenous immunoglobulin treatment, ESR is no longer helpful in determining the level of inflammation.

PREP Pearls
- The diagnostic criteria for Kawasaki disease (KD) include fever for at least 5 days and at least 4 of the following: bilateral, nonexudative, bulbar conjunctivitis; oropharyngeal changes; cervical lymphadenopathy; polymorphous rash; and peripheral extremity changes.
- No laboratory study is diagnostic for KD, but elevated erythrocyte sedimentation rate, elevated C-reactive protein, elevated platelets, anemia, low albumin, elevated alanine aminotransferase, elevated white blood cell count, and sterile pyuria can support the diagnosis.
- Echocardiography should be performed in cases of incomplete or suspected KD.

ABP Content Specifications(s)
- Formulate a differential diagnosis of Kawasaki disease

Suggested Readings
- Scuccimarri R. Kawasaki disease. Pediatr Clin North Am. 2012;59:425-445. DOI:http://dx.doi.org/10.1016/j.pcl.2012.03.009.

Question 134

A 12-year-old boy presents to your office for evaluation of right elbow pain. He denies any history of trauma, but describes a 2-month history of medial elbow pain with baseball pitching. He pitches for 4 different teams throughout the year. On physical examination, the boy is lacking 10 degrees of extension of the right elbow. He is tender over the medial epicondyle. Radiographs of the right elbow show slight widening of the medial epicondyle apophysis.

Of the following, the BEST initial step in management is to recommend

A.   cast immobilization of the elbow for 4 to 6 weeks

B.   complete rest from throwing for 4 to 6 weeks

C.   limiting his pitching to only fastballs

D.   referral to orthopedic surgery for surgical management

E.   use of a hinged elbow brace while throwing for 4 to 6 weeks

Question 134   **Preferred Response: B**

The boy in the vignette has Little League®elbow (also called youth baseball elbow), a widening of the distal humerus medial epicondyle epiphysis. He should rest completely from throwing for at least 4 weeks. Little League elbow is an overuse injury that results from repetitive overhead motion, eg, pitching in baseball. Despite the name, Little League elbow is also seen in children who participate in nonthrowing sports that involve overhead motion, eg, tennis. The diagnosis of Little League elbow can be made clinically. Affected individuals are typically between the ages of 10 and 14 years. On physical examination, there is tenderness over the medial epicondyle of the elbow, and many children also have decreased range of motion in the elbow. Radiographs may show widening of the medial epicondyle epiphysis; contralateral radiographs may be helpful for comparison if this finding is subtle. Some children have an avulsion of a piece of the medial epicondyle epiphysis (Item C134).

Item C134. Avulsion of the inferior portion of the medial epicondyle epiphysis. Courtesy of R.L. Carl

The treatment for Little League elbow is rest, followed by an exercise program, ideally under the direction of a physical therapist or athletic trainer familiar with pediatric injuries. For throwing athletes, the exercise program should include a gradual increase in throwing frequency, distance,

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and intensity. For avulsion injuries, with more than 0.5 to 1 cm of separation, surgery may be required.

Limiting repetitive overhead motion will decrease the risk of Little League elbow. Many youth baseball programs (eg, Little League) have set an upper limit for the number of pitches a young athlete can throw during games and practices.

Many overuse injuries in children affect the physes (major growth plates) and apophyses (minor growth areas where tendons attach). In adults, tendons and ligaments tend to be the weakest part of the skeleton. In children, the cartilaginous physes and apophyses are especially vulnerable to injury. With repetitive activity, children may develop irritation, separation, or even avulsion injuries involving the physes and apophyses. These overuse injuries typically occur when repetitive activities apply traction or pressure on a growth area. Osgood-Schlatter syndrome, irritation of the tibial tubercle apophysis, is probably the most well-known form of apophysitis. In Osgood-Schlatter syndrome, the quadriceps muscle pulls indirectly on the patellar tendon, which puts traction on the tibial tubercle. Sever disease refers to calcaneal apophysitis resulting from both traction on the apophysis by the Achilles tendon and repetitive impact to the heel with running and jumping. Little League shoulder is an irritation of the proximal humeral physis commonly seen in young athletes who participate in sports with repetitive overhead activity. There are multiple apophyses of the pelvis. Pelvic apophysitis is most often seen in adolescent athletes, with the anterior superior iliac spine the most commonly irritated location. Avulsion injuries can also occur at the pelvic apophyses.

The child in the vignette should not be treated with immobilization of the elbow, because this can lead to stiffness and should be avoided whenever possible. Research evidence is mixed regarding whether throwing certain pitches (eg, breaking or curveball pitches) increases injury rates. In this child, throwing any type of pitch would not be appropriate given his history and physical examination findings. Surgery is generally indicated in cases with 5 to 10 mm of separation (depending on the type of sport played and the patient's symptoms), a finding not seen in this patient. A hinged elbow brace may relieve some of the force on the medial epicondyle, but is not a substitute for rest.

PREP Pearls
- Little League elbow is an irritation of the medial epicondyle apophysis that occurs in athletes who participate in sports with repetitive overhead motions, most commonly between the ages of 10 and 14 years.
- The cartilage growth centers of the pediatric skeleton are vulnerable to repetitive stress that can cause overuse injuries.

ABP Content Specifications(s)
- Identify the common overuse injuries in athletes
- Plan the appropriate management of an athlete with an overuse injury

<u>Suggested Readings</u>

*   Sarwark JF, LaBella C. Pediatric Orthopaedics and Sports Injuries: A Quick Reference
         Guide. 2nd ed. Elk Grove Village, IL: American Academy of Pediatrics; 2014: 445 pp.

Question 135

A 15-month-old boy presents with sudden onset of fever, left knee swelling, and refusal to bear weight. On physical examination, his temperature is 39.5°C, heart rate is 140 beats/min, respiratory rate is 30 breaths/min., and blood pressure is 95/65 mm Hg. He is ill appearing, but not toxic. His left knee is swollen with mild overlying erythema and is exquisitely painful with any movement. A plain radiograph is normal, but ultrasonography of the knee reveals moderate fluid in the joint space. Fluid obtained by arthrocentesis shows 75,580 white blood cells/μL with 95% neutrophils. The joint fluid culture is growing gram-positive cocci in pairs and chains that are ß-hemolytic on sheep blood agar (Item Q135).

Of the following, the BEST antimicrobial choice for treating this patient's infection is

A.   amoxicillin/clavulanate

B.   cefotaxime

C.   clindamycin

D.   penicillin

E.   vancomycin

Item Q135. Sheep blood agar plate showing growth from the joint fluid aspiration for the boy described in the vignette. Courtesy of R. Jerris. Reprinted with permission from Pickering LK, et al. Red Book® Online. Elk Grove Village, IL: American Academy of Pediatrics.

Question 135   **Preferred Response: D**

The boy in the vignette has pyogenic arthritis of the left knee caused by group A Streptococcus. He should be treated with parenteral penicillin until clinical improvement permits the transition to oral therapy. The incidence of septic arthritis is approximately 5 to 12 per 100,000 children and is more common in boys than girls (2:1). Most cases occur in children younger than 10 years (peak age, 2 to 3 years). Bacterial pathogens typically invade the joint space and synovium by hematogenous spread, but can be introduced through penetrating trauma or operative procedures. Staphylococcus aureus (methicillin-susceptible and methicillin-resistant) is the most common cause of septic arthritis in children outside the neonatal period, followed by group A Streptococcus (C135).

Item C135. Pathogens Associated With Septic Arthritis in Children.

Gram-positive
- *Staphylococcus aureus*
- Group A *Streptococcus*
- *Streptococcus pneumoniae*
- Group B *Streptococcus*
- Coagulase-negative staphylococci

Gram-negative
- *Kingella kingae*
- Enterobacteriaceae (eg, *Escherichia coli, Enterobacter* species)
- *Neisseria gonorrhoeae*
- *Salmonella*
- *Haemophilus influenzae*
- *Pseudomonas aeruginosa*
- Other (eg, *Brucella, Pasteurella*)

Fungal
- *Candida* species
- *Cryptococcus neoformans*
- Endemic mycoses (histoplasmosis, coccidioidomycosis, blastomycosis)

Aytpical pathogens
- Mycobacteria

Courtesy of D. Palazzi

Pyogenic arthritis caused by group B Streptococcus, coagulase-negative staphylococci, Enterobacteriaceae, or Candida species occurs most often in neonates, especially low birthweight infants. Kingella kingae, Streptococcus pneumoniae, and Haemophilus influenzae arthritis typically occur in young children (<2-3 years of age), however H influenzae is now a rare cause because of widespread immunization. Although Salmonella septic arthritis is more common in patients with sickle cell disease, staphylococcal infection is far more prevalent. Neonates and sexually active adolescents may develop infection from Neisseria gonorrhoeae. Unusual causes (eg, Brucella, Pasteurella, Pseudomonas, mycobacterial, fungal) require specific exposure, penetrating injury, or immunocompromised status.

Unlike children with toxic synovitis or arthralgia, children with pyogenic arthritis typically are ill-appearing with fever, significant joint pain and swelling, and pseudoparalysis of the affected area. Inflammatory markers usually are markedly elevated, and there is widening of the joint space with joint effusion on radiographic imaging. Synovial fluid characteristically reveals a white blood cell count of more than 50,000/µL (50 × 109/L) with a predominance of neutrophils. To prevent bony destruction, especially in infants and children with prolonged symptoms, surgical drainage of pyogenic arthritis of the hips and shoulders should occur promptly. Needle aspiration of other infected joints may be sufficient, but depends on the clinical presentation and response to antimicrobial therapy. Most children with septic arthritis require approximately 3 weeks of antimicrobial therapy directed toward the isolated or most likely pathogen. Because antibiotics penetrate readily into joint fluid, most clinicians transition from parenteral to oral therapy as soon as the patient demonstrates some clinical improvement (more joint mobility, less pain, resolving fever, inflammatory markers trending down).

The treatment of choice for pyogenic arthritis caused by group A Streptococcus is penicillin. Vancomycin is appropriate therapy when a gram-positive pathogen resistant to other agents is suspected or documented (eg, methicillin-resistant Staphylococcus aureus, cefotaxime-resistant S pneumoniae). Clindamycin is useful for susceptible staphylococci and S pneumoniae. Typically, pyogenic arthritis caused by susceptible strains of pneumococcus and gram-negative pathogens is treated with a third-generation cephalosporin such as cefotaxime. Amoxicillin-clavulanate is appropriate therapy for septic arthritis caused by Pasteurella, because it also has anaerobic coverage, which often is desired for skin and skin structure infections after animal bites.

PREP Pearls
- Children with septic arthritis typically are ill-appearing with fever, significant joint pain and swelling, and pseudoparalysis of the affected area.
- Most cases of septic arthritis occur in children younger than 10 years (peak age, 2 to 3 years) by hematogenous spread of bacteria to the joint space and synovium.
- Staphylococcus aureus (methicillin-susceptible and methicillin-resistant), followed by group A Streptococcus, is the most common cause of septic arthritis in children outside the neonatal period.
- To prevent bony destruction, especially in infants and children with prolonged symptoms, surgical drainage of pyogenic arthritis of the hips and shoulders should occur promptly.

ABP Content Specifications(s)
- Understand the natural history of pyogenic arthritis
- Differentiate the clinical findings of pyogenic arthritis from those of toxic synovitis and arthralgia
- Identify the etiology of pyogenic and acute arthritis

Suggested Readings
- American Academy of Pediatrics. Group A streptococcal infections. In: Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:732-734.

Question 136

While "wrestling" with an older cousin, a 10-year-old healthy boy fell sideways from standing, landing with his weight on his right shoulder. He reports feeling a snapping sensation when his shoulder hit the floor. Physical examination reveals a well-appearing boy with tenderness to palpation and slight swelling over the middle third of his right clavicle without tenting of the skin. The boy has good passive range of motion in his left shoulder, but he reports pain with abduction of the right shoulder. He has full grip strength and normal radial pulses bilaterally. His breath sounds are clear and equal bilaterally, with no respiratory distress. A plain radiograph of his right clavicle, is shown (Item Q136).

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

A.   obtain computed tomography of his right clavicle

B.   obtain magnetic resonance imaging of his right shoulder

C.   obtain orthopedic consultation for reduction of his fracture

D.   place a posterior long arm splint on his right upper extremity

E.   place his right upper extremity into an arm sling

Item Q136. Radiograph for the boy described in the vignette. Courtesy of D. Krowchuk.

Question 136   **Preferred Response: E**

The boy in the vignette has a minimally displaced right clavicle fracture, with no associated neurovascular compromise. The most appropriate next step in his management is to place his right upper extremity in an arm sling.

Clavicle fractures are the most common fracture type in children. Most clavicle fractures involve the midshaft, whereas fractures of the medial third are the least common type (accounting for only 2%-3% of cases).

Clavicle fractures arise most commonly from direct trauma (typically a fall onto the shoulder), but may also result from indirect forces (such as transmitted force from a fall onto an outstretched hand). Most are greenstick-type fractures involving the clavicular midshaft, with the thick clavicular periosteum preventing significant displacement or angulation in children younger than 10 years.

Children with clavicle fractures typically report pain localized to the clavicle that worsens with movement of the shoulder. Patients will often cradle the arm on the injured side with the opposite one to prevent movement of the arm. Some children may report feeling a "snap" or "crack" at the time of the injury. Physical examination may reveal localized swelling, a visible bulge and/or bruising over the clavicle, or even tenting of the skin over the fracture site. In some cases, the injury can go unnoticed and is later recognized when a lump appears as a bony callus forms. Despite the clavicle's anatomic location close to the brachial plexus and subclavian vessels, neurovascular complications from clavicle fractures rarely occur unless a powerful direct traumatic force results in significant displacement of the fracture fragments. Despite the rare occurrence of neurovascular complications, physical examination should include careful assessment for neurovascular injury, as well as a careful lung examination to assess for associated pulmonary complications such as pneumothorax or hemothorax.

Plain radiography is generally the diagnostic mode of choice for the initial evaluation of isolated clavicular injuries. Several views may be needed to identify more subtle nondisplaced fractures. Computed tomography may occasionally be needed to identify clavicle fractures (particularly those involving the proximal third) that are not apparent on plain radiography and to assess for associated serious intrathoracic injuries. The boy in the vignette has a clavicle fracture with minimal displacement, and he displays no signs or symptoms of associated intrathoracic injuries. Therefore, computed tomography of the clavicle would not be the best next step in his management.

Magnetic resonance imaging of the shoulder would not be indicated in the boy's management, as there are no clinical findings suggestive of an injury involving his right shoulder joint. Orthopedics consultation for closed reduction of this patient's clavicle fracture would not be indicated; the fracture is not significantly displaced, and the boy displays no signs of associated neurovascular compromise. Emergent orthopedic consultation is rarely necessary for children with clavicular fractures. Indications for emergent orthopedic consultation include open fractures (including fractures with significant tenting of the overlying skin), fractures with associated

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neurovascular compromise, midshaft fractures with significant displacement (>2 cm), medial clavicle fractures with posterior or significant anterior displacement, or grossly unstable distal injuries. Patients with more than 1.5 cm of clavicle shortening caused by the fracture should be referred for prompt orthopedic follow-up.

Nonoperative management is the initial treatment of choice for the vast majority of uncomplicated clavicle fractures. Arm immobilization in a sling for comfort for 3 to 4 weeks, followed by 3 weeks of rest from contact sports is recommended in most cases. Healing occurs rapidly, typically with formation of a callus. Parents should be advised that as a part of the normal healing process a lump will likely become apparent as a callus forms, and may persist for a year or longer. Adequate pain control, as well as close follow-up until symptoms have resolved and shoulder function has normalized, is also essential in the management of clavicle fractures. Placing a posterior long arm splint on the right upper extremity would not be useful for the boy in the vignette, because this would not stabilize his injury. Restricting shoulder motion to less than 30 degrees, with either an arm sling or figure-of-8 bandage, best stabilizes clavicle fractures. Because slings are typically more comfortable and more widely available than figure-of-eight bandages, with similar outcomes, they are commonly used as the initial treatment of choice for children with clavicle fractures.

PREP Pearls
- Children presenting with clavicle fractures generally report pain that is localized to the clavicle but worsens with movement of the shoulder. Patients often cradle their arm on the injured side with the opposite one to prevent movement.
- Plain radiography is the diagnostic modality of choice for the initial evaluation of isolated clavicular injuries.
- Emergent orthopedic consultation is rarely necessary for children with clavicular fractures. Indications include fractures that are open, those with significant tenting of the overlying skin, those with associated neurovascular compromise, midshaft fractures with significant displacement (>2 cm), medial clavicle fractures with posterior or significant anterior displacement, or grossly unstable distal injuries.

ABP Content Specifications(s)
- Recognize the clinical and radiographic findings associated with a fracture of the clavicle, and manage appropriately

Suggested Readings
- Bachman D, 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.

Question 137

A mother delivers a neonate precipitously, at 25 weeks' gestation, in the labor and delivery unit of a community hospital. The membranes spontaneously rupture at the time of delivery, revealing clear amniotic fluid. The very-low-birth weight neonate has poor tone, no spontaneous cry, and rare gasping respirations. The newborn is brought over to the warmer.

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

A.　　　dry and stimulate the newborn with warm towels

B.　　　initiate positive pressure ventilation with 100% oxygen

C.　　　insert the newborn up to the neck in a polyethylene bag

D.　　　place the newborn in a head down position

E.　　　put a portable warming pad around the newborn

Question 137   **Preferred Response: C**

The extremely low-birthweight (ELBW) neonate in the vignette should be placed in a polyethylene bag immediately after delivery. The immature skin and underdeveloped subcutaneous fat found in an ELBW newborn contributes to significant heat loss in the immediate neonatal period. The use of a polyethylene barrier has been shown to decrease heat loss and minimize hypothermia in neonates born at less than 28 weeks of gestation.

Multiple strategies have been used to enhance thermoregulation of the ELBW infant in the delivery room, because both hypothermia and hyperthermia are associated with increased morbidity. The ambient temperature of the delivery room should be increased to at least 26°C to decrease radiative heat loss. To minimize conductive heat loss, a radiant warmer should be operational at delivery with an activated heated mattress (always placed under the receiving blanket to prevent thermal contact burns). The ELBW infant should immediately be placed up to the neck in a polyethylene bag or wrap, leaving the head exposed for airway management (Item C137A). The use of a polyethylene barrier, such as a bag or wrap minimizes evaporative and convective heat loss. A hat may be placed on the head to further decrease radiative heat loss. The skin of the ELBW newborn should not be wiped dry or stimulated because the fragile epidermal layer is easily damaged by friction.

Item C137A. Premature newborn placed up to the neck in a polyethylene bag .Courtesy of R. B. Dail

Additional aspects of delivery management in the first minutes after birth can significantly affect the long-term outcome of the ELBW infant. The resuscitation table should be flat, because placement of the infant head down (Trendelenburg) may lead to intraventricular hemorrhage. Excessive positive pressure with assisted ventilation may cause a pneumothorax and can contribute to the development of chronic lung disease. Oxygen therapy should be administered through an oxygen blender guided by the use of a pulse oximeter. Hypoxia and hyperoxia are both associated with increased morbidity in ELBW infants. Although no agreement has been

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reached on the optimum oxygen saturation level for preterm infants, targeted preductal saturations are recommended immediately after delivery based on time after birth (Item C137B), as outlined in the sixth edition of the Neonatal Resuscitation Program of the American Academy of Pediatrics and American Heart Association. Many neonatal providers begin assisted ventilation of a premature newborn with 30% to 40% oxygen rather than room air because of the high likelihood of surfactant deficiency, but the use of 100% oxygen is avoided unless supported by pulse oximetry.

Item C137B. 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

PREP Pearls
- Multiple strategies to enhance the thermoregulation of a premature infant in the delivery room should include increasing the ambient temperature of the delivery room, the use of a radiant warmer, a warming mattress, a polyethylene barrier, and a hat.
- The delivery of oxygen therapy to a premature infant in the delivery room should be guided by the use of a pulse oximeter and administered via an oxygen blender.

ABP Content Specifications(s)
- Plan appropriate initial care for a very-low-birth-weight infant

Suggested Readings
- Kattwinkel J, ed. Textbook of Neonatal Resuscitation. 6th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2011:329 pp.

Question 138

A 14-year-old adolescent is brought to the emergency department by his parents. He has complained of neck pain for 3 days since riding a roller coaster at a theme park. The day before this visit, his right arm started to tingle and became weak, and this morning, his left arm started to tingle. He has not had urinary or fecal incontinence, changes in vision, or headache. He has not been sick recently, although his sister had an upper respiratory infection 3 weeks ago. He does not recall any specific head or neck trauma.

On physical examination, the patient's temperature is 36.8°C, blood pressure is 102/74 mm Hg, and his heart rate is 92 beats/min. He has no rashes and no tenderness to palpation of his vertebral spinous processes. There is a palpable mass in his lower abdomen that you suspect is a distended bladder. His neurologic examination demonstrates flaccid paralysis of his right upper extremity. His left upper extremity has only antigravity strength and he is unable to provide any resistance. Reflexes are diminished in his biceps and triceps, with normal patellar reflexes. His toes are flexor on plantar stimulation. His mental status, cranial nerves, sensation, coordination, and gait are normal.

Of the following, the test MOST likely to yield the correct diagnosis is

A.   computed tomography angiography of the neck

B.   electromyography and nerve conduction study

C.   lumbar puncture

D.   magnetic resonance imaging of the brain

E.   magnetic resonance imaging of the cervical spine

Question 138   **Preferred Response: E**

The boy in the vignette has had rapid onset of bilateral arm weakness, sensory symptoms, and urinary retention, suggesting an acute process involving the cervical spinal cord. Magnetic resonance imaging of the cervical spinal cord is the diagnostic test of choice, as transverse myelitis is the most likely diagnosis. Transverse myelitis is an acute onset, immune-mediated demyelination of the spinal cord, typically involving contiguous vertebral levels in the cervical and thoracic spinal cord. The most common presenting symptoms and signs are pain, weakness, and sensory changes at the level of the lesion. Sometimes sensory symptoms precede sensory deficits. The differential diagnosis of an acute process of the cervical spine includes a compressive mass lesion such as tumor or abscess, an expanding spinal cord syrinx, direct spinal cord bacterial or viral infection, or acute ischemia.

For the boy in the vignette, who does not have signs or symptoms of active infection, the best test to evaluate his symptoms is magnetic resonance imaging (MRI) of the cervical spinal cord. MRI of the brain would not identify a lesion in the cervical spinal cord. Computed tomography angiography of the neck would show abnormalities of the arteries but not the spinal cord itself. Electromyography and nerve conduction studies test the muscle, nerve, and neuromuscular junction, which would not be helpful in this case. Lumbar puncture would likely be performed once the MRI of the cervical spinal cord shows transverse myelitis, but it is not the best test to diagnose an acute cervical spinal cord lesion.

PREP Pearls
- Sudden onset of bilateral arm weakness with sensory loss is suggestive of a cervical spine lesion.
- Magnetic resonance imaging of the spinal cord is the diagnostic test of choice when there is suspicion of an acute, nontraumatic spinal cord lesion.

ABP Content Specifications(s)
- Plan the appropriate diagnostic evaluation of suspected transverse myelitis
- Recognize the clinical manifestations of transverse myelitis

Suggested Readings
- Amador CD. Index of suspicion. Pediatr Rev. 2001;22(9):303-308. DOI: http://dx.doi.org/10.1542/pir.22-9-303.

Question 139

A 23-month-old girl presents to your office for evaluation of breast enlargement. The bilateral breast tissue was noted 4 months after birth and has not significantly changed in size over the last 2 years. Linear growth remains normal. Physical examination reveals sexual maturity rating 3 breast tissue bilaterally, with enhancement of the breast and areola. There is no pubic hair. The child's parents are concerned that the breast enlargement has not regressed and request further evaluation.

Of the following, the MOST likely diagnosis is

A.   central precocious puberty

B.   estrogen-secreting ovarian tumor

C.   premature adrenarche

D.   premature thelarche

E.   virilizing adrenal tumor

Question 139   **Preferred Response: D**

The girl in the vignette has premature thelarche, a normal variant of puberty. In this condition, 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. In many cases there will be regression of breast tissue over time, though in some cases the breast tissue persists. The development of breast tissue before the age of 2 years is common and is almost always benign. Because all children younger than 2 years go through a period of "mini puberty" when gonadotropin-releasing hormone (GnRH) is not inhibited, it is very difficult to distinguish premature thelarche from true central precocious puberty in these young children based on biochemical testing. In general, it is not recommended that testing such as luteinizing hormone (LH), follicle-stimulating hormone (FSH), or estradiol levels be performed in these cases unless there are very clear progressive symptoms.

Other normal variants include early menarche (due to an underlying genitourinary cause) and isolated pubic hair of infancy.

Central precocious puberty in girls is defined as breast budding before 8 years of age, but outside the age group of premature thelarche. In central precocious puberty, early activation of the normal pubertal system is manifested by pulsatile GnRH secretion with subsequent LH and 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 is the development of pubertal signs resulting from other than 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), seen in congenital adrenal hyperplasia (CAH) or adrenocortical cancer (ACC). With adrenal disease, there is usually no breast development because the adrenal glands are secreting androgens only. Children showing signs of significant early androgen exposure, such as pubic hair with acne, clitoromegaly, voice deepening, and rapid height acceleration, should be tested for dehydroepiandrosterone sulfate (DHEA-S) level (elevated in ACC), 17-hydroxyprogesterone level (abnormal in the most common form of CAH), and a total testosterone level (to evaluate for an androgen-secreting ovarian tumor). In the event that all laboratory testing is normal, or the symptoms are so minor that testing is not indicated, the diagnosis of premature adrenarche can be made. With premature adrenarche there is isolated pubic hair development without breast budding or other estrogen effects.

PREP Pearls
- Premature thelarche occurs in girls younger than 2 years of age who show breast development but have no other signs of puberty.
- Girls with premature thelarche will not have rapid height acceleration consistent with a pubertal growth spurt.
- Generally, no workup or treatment is needed for premature thelarche.

ABP Content Specifications(s)
- Understand the significance of premature thelarche

Suggested Readings
- Muir A. Precocious puberty. Pediatr Rev. 2006;27(10):373-381. DOI: http://dx.doi.org/10.1542/pir.27-10-373.

Question 140

You have been caring for an 8-year-old girl who was hospitalized with right lower lobe pneumonia and pleural effusion. She presented to the emergency department 4 days ago with fever to 40.2°C, cough, and moderate respiratory distress. She has continued to require 2 L of oxygen via nasal cannula to maintain oxygen saturations greater than 91%. She has been persistently febrile.

A chest tube was placed on admission and an exudative pleural fluid collection was confirmed by laboratory analysis. Culture of this fluid grew Staphylococcus aureus. The patient has been treated with intravenous vancomycin. Despite confirmed adequate placement of the chest tube, radiographic imaging reveals a persistent dense opacity and fluid collection in the inferior aspect of the right lung. Fibrinolytic therapy is instilled into the pleural space without resolution of the airspace disease. Ultrasonography reveals a complex and loculated fluid collection.

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

A.   aggressive chest physiotherapy with hypertonic saline 4 times daily

B.   computed tomography and computed tomographic-guided lung biopsy

C.   continued observation on current antibiotic therapy

D.   repeat instillation of fibrinolytic therapy via indwelling chest tube

E.   video-assisted thoracoscopic surgery with lung decortication

Question 140   **Preferred Response: E**

The child in the vignette has a parapneumonic effusion as a complication of her community acquired pneumonia. In children with pneumonia, parapneumonic effusion or empyema complicates the clinical course in 28% to 53% of cases.

Pneumonia causes an inflammatory response in the pleura and in the pulmonary parenchyma. A pleural exudative response to this irritation may result in the development of a parapneumonic effusion or an inflammatory fluid collection between the parietal and visceral pleura. Initially, this pleural fluid is likely to be clear with a low number of white blood cells. With persistence and transition to a complicated parapneumonic effusion, there is an influx of inflammatory mediators, development of purulence, and deposition of fibrin. The fluid may become loculated with the presence of fibrous strands. It is important to recognize the complicated parapneumonic effusion prior to the development of fibrotic change with irreversible lung restriction.

As disease progresses, changes occur in the pleural fluid, which include a decrease in glucose and pH, as well as an increase in lactate dehydrogenase (LDH). The Light criteria have been used to characterize pleural fluid as exudative if at least 1 of the following criteria are met:
- o   The ratio of pleural fluid protein to serum protein is greater than 0.5
- o   The ratio of pleural fluid LDH to serum LDH is greater than 0.6
- o   LDH in pleural fluid is greater than two-thirds the normal upper limit value

The sensitivity and specificity of the Light criteria for determination of an exudative pleural effusion have been reported as 98% and 80%, respectively. While the Light criteria are commonly utilized in clinical care, they have not been validated in pediatric patients.

Of note, recent data have shown that serum C-reactive protein levels are significantly higher in children with empyema, as compared to children with uncomplicated pneumonia. In the imaging of pneumonia and parapneumonic effusions, it is often challenging to differentiate between lung consolidation and pleural fluid with the use of plain radiographs. While a lateral decubitus radiograph may demonstrate "layering" with free flowing pleural effusions, the same is not expected with a complex or loculated parapneumonic fluid collection. Ultrasonography involves no exposure to ionizing radiation and is a useful modality in differentiating between a pleural and parenchymal process. It is likely superior to computed tomography (CT) in the demonstration of loculations. The use of CT should be limited to complex cases where parenchymal abscess or bronchopleural fistulae are being considered in the differential.

In general, small and moderate uncomplicated parapneumonic effusions can be treated with antibiotics and do not routinely require chest tube drainage. Drainage with chest tube insertion is warranted when parapneumonic effusions are large in size or when purulence has been demonstrated; drainage is also indicated if the effusion is associated with respiratory compromise or distress. Chest thoracotomy tube drainage is an effective intervention for free flowing effusions. When effusions are complex or loculated, they may respond to instillation of fibrinolytic agents (such as tissue plasminogen activator or urokinase) through the indwelling thoracostomy tube. Video-assisted thoracotomy (VATS) should be performed when there is

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persistence of moderate to large effusions and ongoing respiratory compromise, despite 2 to 3 days of management with chest tube and completion of fibrinolytic therapy.

The patient in the vignette has now failed antibiotic therapy, chest thoracotomy tube drainage, and fibrinolysis. The next most appropriate step in management is a VATS with decortication of the loculations.

Hypertonic saline draws fluid into the airway after nebulized inhalation and may provide symptomatic relief in cystic fibrosis or bronchiolitis by rendering secretions less tenacious. The fluid collection in this case is not in the airway and would not respond to this therapy. Continued observation, repeat instillation of fibrinolytic therapy, and lung biopsy are not indicated. Timely management of the complicated effusion is warranted in order to prevent late complications, including restrictive lung disease.

PREP Pearls
- An elevated C-reactive protein on admission in a patient with radiographic pneumonia is a useful marker of inflammation and may predict the presence of a complicated effusion.
- Computed tomography (CT) is not superior to ultrasonography in the evaluation of complicated parapneumonic effusions and the use of CT should be limited because of the risks of exposure to ionizing radiation.
- Small- and moderate-sized parapneumonic effusions may respond to intravenous antibiotic therapy alone.
- Video-assisted thoracotomy should be performed when there is persistence of moderate to large effusions and ongoing respiratory compromise, despite 2 to 3 days of management with chest tube and completion of fibrinolytic therapy.

ABP Content Specifications(s)
- Understand the etiology of pleural fluid accumulation
- Recognize the normal and abnormal characteristics of pleural fluid
- Plan the appropriate management of an empyema
- Plan the appropriate diagnostic evaluation of pleural disease

Suggested Readings
- Carter E, Waldhausen J, Zhang W, Hoffman L, Redding G. Management of children with empyema: pleural drainage is not always necessary. Pediatr Pulmonol. 2010;45(5):475-480. DOI: http://dx.doi.org/10.1002/ppul.21200.

Question 141

You are seeing first time parents for a prenatal visit. The mother is trying to determine which feeding practices will fit with her lifestyle as a biology graduate student. She has heard that breastfeeding promotes a healthier immune system compared to formula feeding and asks what the mechanism of this protection is.

Of the following, the BEST response to her question is that

F.   formula with added probiotics is immunologically equivalent to human milk

G.   the immune components of human milk prevent transmission of human milk-borne viruses

H.   the immunologically active component of human milk is immunoglobulin G

I.   the intestinal flora of breast- and formula-fed infants are very similar and have little effect on the baby's immune response

J.   most of the immunologically active components of human milk enhance the baby's immune response at the mucosal level

Question 141   **Preferred Response: E**

Among the important benefits of breastfeeding is the support it provides to the infant's immune function. Most of the immune supporting components of breast milk exert their effects within the gastrointestinal tract at the mucosal level. The primary immunoglobulin in breast milk, and the most studied bioactive component, is secretory immunoglobulin A (sIgA); immunoglobulin M and immunoglobulin G occur in much smaller amounts. The sIgA exerts its effects by binding to microbial antigens, inhibiting their adhesion to host cells and preventing their penetration of the mucosal barrier. In addition, sIgA promotes phagocytosis and is active in regulating local immune response. Other bioactive proteins, most notably lactoferrin, lysozyme, α-lactalbumin, and casein, support immunity by several mechanisms, among them: inhibition of bacterial growth; prevention of microbial mucosal adhesion and penetration; support of gastrointestinal (GI) tract development and repair; and enhancement of macrophage activity. In addition, several of these protein components, along with oligosaccharides (a carbohydrate component of breast milk) promote the growth of beneficial bacteria. Lipids present in breast milk are active against viruses and Giardia. An important recent concept in breastfeeding is that the immune components of human milk promote local immune protection in the GI tract, while at the same time limiting a more extensive inflammatory response by inhibiting inflammatory stimulation and limiting production of and promoting breakdown of toxic oxygen radicals, histamine, leukotrienes, and other cytokines.

The American Academy of Pediatrics (AAP) strongly supports breastfeeding both for its nutritional and non-nutritive benefits. The AAP recommends exclusive breastfeeding for the first 6 months of life and continued breastfeeding for at least 1 year with the addition of complementary foods starting at 6 months of age. Breastfed infants experience decreased rates or severity of infectious disease (upper and lower respiratory tract infections, respiratory syncytial virus bronchiolitis, otitis media, nonspecific GI infection, and necrotizing enterocolitis), atopic disease (asthma, atopic dermatitis), sudden infant death syndrome, GI diseases (Celiac disease, inflammatory bowel disease), leukemia, obesity, and diabetes mellitus. While less clear, there is evidence that breastfeeding results in enhanced neurodevelopment.

The bacterial microflora of the gut is also critical in the development of local immunity. Oligosaccharides are important for creating a healthy microflora, and cow milk-based formula contains much lower levels of oligosaccharides than human milk does. The microflora of breast versus formula-fed infants are quite different. In human milk-fed infants, Lactobacillus bifidus and Bifidobacterium are the predominant organisms, while for formula fed-infants, the most common organisms are gram negative. This difference in microflora likely contributes to differences in immune response. Adding probiotics to formula may improve the gastrointestinal microflora, but does not provide the other immune benefits of human milk.

While breastfeeding helps protect the infant against various pathogens, breast milk can be the vehicle to transmit some viruses from mother to infant. Notable among these viruses are HIV and cytomegalovirus.

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PREP Pearls
- The immunoactive components of human milk act primarily within the gastrointestinal tract at the mucosal level.
- Immunologically active proteins in human milk, including secretory immunoglobulin A, support immune function by inhibiting bacterial growth, preventing microbial mucosal adhesion and penetration, supporting gastrointestinal tract development and repair, and enhancing macrophage activity.
- Oligosaccharides in human milk promote growth of the gastrointestinal microflora, which differs between human milk-fed and formula-fed infants.

ABP Content Specifications(s)
- Understand the qualitative and quantitative differences between human milk and various infant formulas
- Recognize the presence and importance of various antibodies (including secretory IgA) in human milk and colostrum

Suggested Readings
- American Academy of Pediatrics. Breastfeeding and the use of human milk. Pediatrics. 2012;129(3):e827-e837. DOI:http://dx.doi.org/10.1542/peds.2011-3552.

Question 142

You are asked to present to the pediatric residents at your local training program on adolescent health. You remind the residents of the top causes of adolescent and young adult (age 15-24 years) mortality and stress the importance of thorough psychosocial risk assessments during adolescent preventive care visits.

Of the following, the top 3 leading causes of death for this age group are unintentional injury,

A.   heart disease, and suicide

B.   HIV, and suicide

C.   homicide, and HIV

D.   homicide, and suicide

E.   malignant neoplasms, and suicide

Question 142   **Preferred Response: D**

According to the Child Health USA 2012 report, the leading causes of adolescent death are unintentional injuries, followed by homicide and suicide (Item C142). Sex and racial disparities exist. The mortality rate of males between 15 to 19 years of age was more than twice that of females in 2010. Additionally, non-Hispanic, American Indian/Alaska Native adolescents had the highest rate of mortality among both males and females. Non-Hispanic black males had the second highest rate; non-Hispanic, Asian/Pacific Islander females had the lowest rate. Unintentional injury was the leading cause of death for adolescents of all racial and ethnic groups, except non-Hispanic black males, for whom homicide was the leading cause of death. The major causes of adolescent mortality are largely preventable, therefore routinely reviewing and counseling on issues such as driving and personal safety with adolescents are important in the provision of adolescent preventive services. Bright Futures discusses provision of anticipatory guidance to adolescents.

Item C142. Mortality Rates Among Adolescents Aged 15-19 Years, by Selected Leading Cause and Sex, 2010
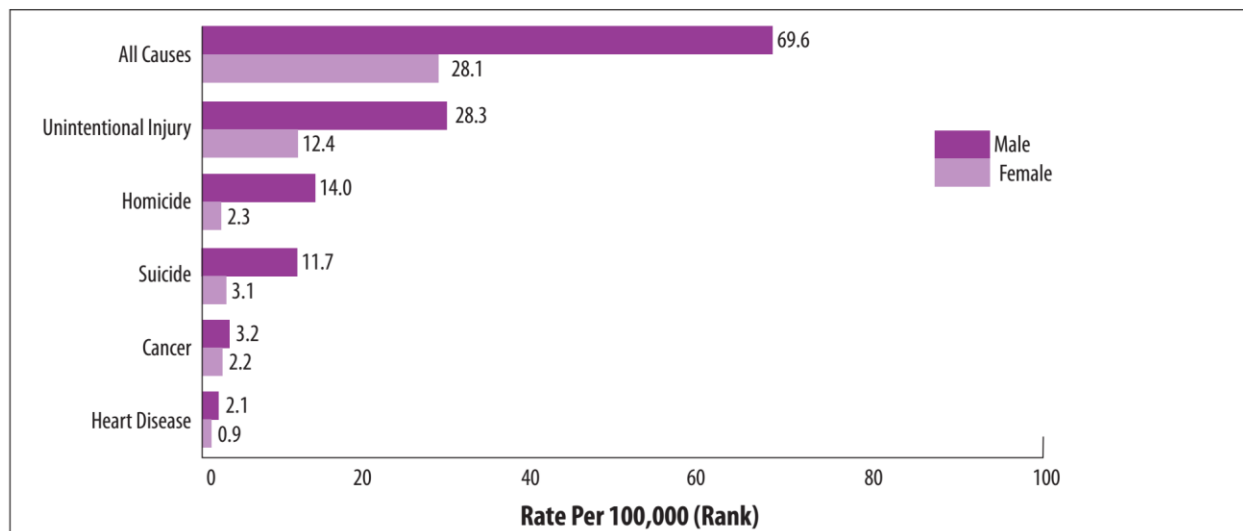
Source: Centers for Disease Control and Prevention, National Center for Health Statistics. Compressed Mortality File 1999-2010. CDC WONDER Online Database, compiled from Compressed Mortality File 1999-2010 Series 20, No. 20, 2012.

PREP Pearls
- The leading causes of adolescent death are unintentional injuries, followed by homicide and suicide.
- Sex and racial disparities exist in the causes of adolescent mortality.
- Non-Hispanic, American Indian/Alaska Native adolescents had the highest rate of mortality among both males and females.
- Bright Futures discusses provision of anticipatory guidance to adolescents.

ABP Content Specifications(s)
- Understand the importance of routinely reviewing behavioral risk factors (eg, school, extracurricular activities, diet, exercise, substance use, sexuality, stress, personal safety, driving, sleep) in adolescents

Suggested Readings

- American Academy of Pediatrics. Bright Futures. Bright Futures website. http://brightfutures.aap.org/.

Question 143

A 15-year-old adolescent presents to your office with right-sided knee pain 10 days after a basketball injury. She was running down the court during a game, came to a sudden stop, and felt "a pop" in her knee. The patient was seen at an urgent care clinic on the day of injury, where radiographs were performed and were unremarkable. The urgent care clinician fitted her with a knee immobilizer and recommended follow-up with her primary care physician. On physical examination, you note a large right knee effusion. The patient is guarding, and therefore you are unable to adequately assess for ligamentous laxity.

Of the following, the MOST appropriate next step would be to

A.   allow full return to sports when swelling subsides

B.   aspirate the knee to relieve swelling

C.   obtain magnetic resonance imaging of the knee

D.   prescribe oral naproxen twice daily for 10 days

E.   provide the adolescent with a patella-stabilizing brace

Question 143   **Preferred Response: C**

The girl in the vignette reports a specific injury mechanism, feeling a "pop" at the time of her injury, and acute onset of swelling. These features point to the presence of an intra-articular injury. An anterior cruciate ligament (ACL) injury is most likely in this case, given the mechanism of injury and rapid onset of swelling. Magnetic resonance imaging (MRI) of the knee is the best test to evaluate the integrity of the ligaments, articular cartilage, and menisci. Another reasonable option would be to recommend protected ambulation with crutches, gentle range-of-motion exercises, application of ice to reduce swelling and a return visit to the clinic for repeat examination in about 2 weeks.

Approximately 75% of ACL tears are noncontact injuries, with female athletes at particularly high risk. Common mechanisms of injury include sudden deceleration, landing from a jump, and twisting or changing direction.

The diagnosis of ACL injury can be made by taking a careful history and demonstrating ligamentous laxity with the Lachman maneuver. To perform a Lachman maneuver, the examiner flexes the patient's knee to 30 degrees, stabilizes the femur with 1 hand, and attempts to pull the tibia anteriorly with the other hand. Increased anterior translation of the tibia relative to the femur compared with the contralateral side is indicative of ACL injury. Although radiographs should be obtained to assess for bony injuries, MRI is the preferred test to evaluate for ligamentous injury, meniscal tear, or cartilage injury. Surgical reconstruction is preferred for athletes who wish to return to sports involving jumping or direction change. The surgical technique for skeletally mature individuals involves drilling through the bone in the area where the physis is located. Therefore, young children should be referred to an orthopedic surgeon experienced in ACL reconstruction using physeal sparing surgical procedures.
Because children and teens with a history of ACL injury are at high risk for early arthritis, prevention of these injuries is extremely important. Exercise programs emphasizing balance, strength, and landing mechanics have been shown to reduce rates of injury. Athletes in sports with high injury rates, for example, soccer and basketball, appear to derive the largest benefits from neuromuscular training programs.

For the athlete in the vignette, a return to sports is inappropriate because her history and physical examination are suggestive of intra-articular injury. Aspiration of the knee would not be helpful in this case, because the fluid would likely reaccumulate and there is a risk of infection. Nonsteroidal anti-inflammatory medications can be used for pain control, but are unlikely to affect the girl's clinical course 10 days after the injury. A patella-stabilizing brace would be an appropriate initial treatment for a patella subluxation or dislocation, without associated osteochondral fracture. Although this injury is in this girl's differential diagnosis, it would be important to rule out an ACL injury before implementing this type of treatment.

PREP Pearls
- Magnetic resonance imaging (MRI) of the knee is the best test to evaluate the integrity of the ligaments, articular cartilage, and menisci.
- Anterior cruciate ligament (ACL) injuries are typically noncontact injuries, and affect female athletes disproportionately.
- An athlete with an acute injury mechanism and joint effusion has a high likelihood of intra-articular injury.

ABP Content Specifications(s)
- Recognize the clinical findings associated with sports-related internal derangement of the knee

Suggested Readings
- Labella C, Carl R. Preventing knee ligament injuries in young athletes. Pediatr Ann. 2010;39(11):714-720. DOI:http://dx.doi.org/10.3928/00904481-20101013-10.

Question 144

A 4-year-old girl is brought to the hospital after falling backwards into a campfire pit. She has been intubated and sedated in anticipation of painful dressing changes. Her weight is 20 kg. Her vital signs include a temperature of 37°C, pulse of 120 beats/min, respiratory rate of 20 breaths/min, and blood pressure of 90/50 mm Hg. The girl has second degree burns involving her neck, posterior trunk, and buttocks, estimated to cover approximately 20% of her total body surface area. Her pupils are 2 mm, equal, and reactive. On auscultation, her heart has a regular rate and rhythm with no murmurs, and her lungs are clear bilaterally. Her abdomen is soft, nontender, and nondistended. Her extremities are warm and well-perfused, with a capillary refill time of 2 seconds. There are no other external signs of trauma.

Of the following, the MOST appropriate fluid management for this child's first 24 hours of treatment is

A.   5% dextrose with 0.2% sodium chloride (NaCl), 800 mL

B.   5% dextrose with 0.2% NaCl, 1,440 mL

C.   5% dextrose with 0.2% NaCl, 1,600 mL

D.   lactated Ringer solution, 800 mL

E.   lactated Ringer solution, 1,600 mL

Question 144   **Preferred Response: E**

The child in the vignette has suffered a burn injury covering 20% of her total body surface area. Fluid management in the first 24 hours should include an intravenous (IV) crystalloid solution at a rate to cover resuscitation plus maintenance fluid requirements according to the Parkland formula. In this case, the correct answer is 1,600 mL of lactated Ringer solution.

Burns can be divided into 4 categories depending on the depth. First-degree burns are limited to the epidermis, second-degree burns extend to part of the dermis, third-degree burns involve the entire dermis, and fourth-degree burns extend to the muscle or bone. Another important descriptor of a burn is the total body surface area (TBSA), which can be assessed using the "Rule of Nines" (Item C144). Patients who have suffered burns have a high fluid requirement because of increased evaporative insensible losses from skin damage and extravasation of fluids from increased capillary permeability due to a systemic inflammatory state. Failure to adequately restore or maintain intravascular volume can adversely affect hemodynamics and lead to lactic acidosis and multiple organ failure.

ITEM C144: The "rules of nines" altered for the anthropomorphic difference

of infancy and childhood."
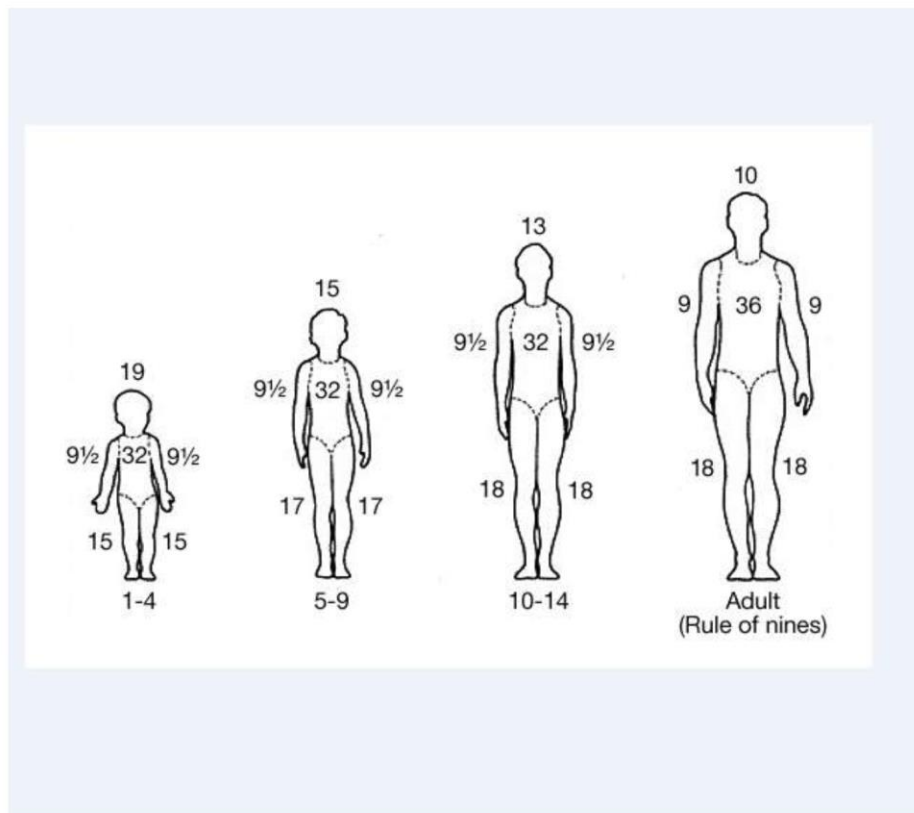
Reprinted with permission from Eichenberger MR. Pediatric Trauma:

Prevention, Acute Care, Rehabilitation. St. Louis, MO: Mosby; 1993:570.

Children with burns—second degree or greater—involving more than 15% TBSA generally require IV fluid replacement. The "Parkland formula" can be used to plan the fluid requirement for the first 24 hours:

Estimated fluid volume (mL) = 4 × body weight (kg) × %TBSA burn

For the child in the vignette, the required fluid volume would equal 4 × 20 kg × 20 = 1,600 mL. Because the purpose of fluid administration is to maintain intravascular volume, the choice should be isotonic, such as normal saline or lactated Ringer solution. For children younger than 2 years, maintenance IV fluids containing dextrose should be added over and above the Parkland formula fluid. Colloids should be avoided unless the serum albumin level is extremely low, because capillary extravasation of albumin may exacerbate tissue edema. If the child is in acute shock, characterized by hypotension or evidence of decreased end-organ perfusion, additional boluses of isotonic fluid may be required. Care must be taken to not overhydrate the patient, because increased intravascular hydrostatic pressure can exacerbate edema, and fluid overload can lead to respiratory and multiple organ failure.

PREP Pearls
- The percentage of total body surface area of a burn can be estimated using the "Rule of Nines."
- The increased fluid requirement for a burn patient is because of increased evaporative insensible losses from skin damage and extravasation of fluids from increased capillary permeability.
- A pediatric burn patient requires isotonic fluid replacement in the first 24 hours, the volume of which can be estimated with the Parkland formula.

ABP Content Specifications(s)
- Plan the appropriate initial management of a burn covering a substantial portion of the body surface area

Suggested Readings
- Hansbrough JF, Hansbrough W. Pediatric burns. Pediatr Rev. 1999;20:117. DOI: http://dx.doi.org/10.1542/pir.20-4-117.

Question 145

A term male newborn is born to a 35-year-old woman known to have hepatitis C. The mother is anxious to know if her newborn has acquired the infection.

Of the following, the test that would BEST identify early infection in the newborn is

A.   polymerase chain reaction at birth

B.   polymerase chain reaction at 4 months of age

C.   polymerase chain reaction at 18 months of age

D.   serology at 4 months of age

E.   serology at 18 months of age

Question 145   **Preferred Response: B**

There are special considerations when testing for vertical transmission of hepatitis C virus (HCV). Polymerase chain reaction (PCR) assays that can detect viremia are preferred over serology for early diagnosis of HCV infection, given that transplacentally acquired maternal anti-HCV antibodies can persist beyond 1 year of life. Unfortunately, even in infected infants, viremia can be low level or episodic. Therefore, a single negative test is not conclusive. A consensus statement by the National Institutes of Health states that infants should be tested for HCV RNA on 2 occasions between 2 and 6 months of age or be tested for antibodies after 15 months of age. A PCR at 18 months of age or at birth is not preferred as a means of early diagnosis of HCV infection.

Immunoglobulin G (IgG) antibody assays and nucleic acid amplification tests are the 2 major means of diagnosing HCV infections. Immunoglobulin M assays for HCV are not available. Neonates born to HCV-infected mothers are expected to be antibody-positive at birth, given transplacental transfer of maternal IgG. Serologic tests prior to 18 months of age cannot distinguish infection in the child from infection in the mother. Therefore, serology performed at 4 months of age, is not a correct option. Serology could be performed at 18 months of age, when maternal IgG would be expected to be undetectable; however, this option would not best identify infection early.

The principal means of transmission of HCV is exposure to contaminated blood or body fluids. Certain risk factors warrant screening for HCV infection in children. These include having a HCV-infected mother, recipients of blood products or transplants prior to 1992, international adoptees and refugees, HIV-infected children, adolescents with any history of illicit intravenous drug use, adolescents with multiple sexual partners, victims of sexual assault, and children with unintentional needle stick injuries.

Vertical transmission accounts for the majority of cases of HCV infection in children. Approximately 5% to 10% of neonates born to mothers with chronic infection will acquire HCV; this incidence is about 750 cases per year in the United States. Young children can spontaneously resolve HCV infection. Twenty-five percent to 75% of HCV-infected infants will not develop chronic infection.

PREP Pearls
- Neonates born to hepatitis C virus (HCV)-infected mothers are expected to be antibody-positive, given transplacental transfer of maternal anti-HCV antibodies.
- Polymerase chain reaction assays that can detect viremia are preferred over serology for early diagnoses of vertically transmitted HCV infection and should be performed between 2 and 6 months of age.
- Vertical transmission accounts for the majority of cases of HCV infection in children.

ABP Content Specifications(s)
- Plan appropriate diagnostic evaluation, including timing of testing, for hepatitis C virus infection
- Identify the risk factors for the acquisition of hepatitis C virus infection

Suggested Readings
- American Academy of Pediatrics. Hepatitis C. In: Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:423-430.

Question 146

You are seeing a 16-year-old adolescent in your office for a health supervision visit. The patient received a renal transplant from his father 2 years ago and is on a steroid-free immune suppressive protocol. According to the parents, he is doing well post-transplantation, with no concerns from the transplant team. His current medications include tacrolimus, mycophenolate mofetil, atenolol, and oral magnesium and phosphorus supplements. He is afebrile with a respiratory rate of 18 breaths/min, heart rate of 78 beats/min, and blood pressure of 119/76 mm Hg. His physical examination shows a well-healed postsurgical abdominal scar.

Of the following the MOST accurate statement regarding this adolescent is

    A.   he is at increased risk for graft failure after renal transplant because of his age

    B.   his graft survival is the same as a patient who receives a deceased donor kidney

    C.   his growth velocity is the same as a patient treated by dialysis

    D.   his infection risk is similar to the general population

    E.   his immune suppressive protocol is associated with poorer growth than those containing steroids

Question 146   **Preferred Response: A**

In patients with deteriorating chronic renal failure, a discussion of treatment options for renal replacement therapy should be initiated once the glomerular filtration rate declines to less than 30 mL/min per 1.73 m2 (onset of stage 4 chronic kidney disease). Treatment options for renal replacement therapy include dialysis (hemodialysis or peritoneal dialysis) and renal transplant. Renal transplant is the treatment of choice for renal replacement therapy in patients with renal failure (adults or children).

According to the United States Scientific Registry of Transplant Recipients, outcomes of renal transplantation in children have improved over the last 25 years. There are various factors affecting the outcome of renal transplantation in children. According to a North American Pediatric Renal Trials and Collaborative Studies report, patients with increased risk for renal allograft failure 10 years post-transplant are likely to be older, nonwhite, have a history of focal segmental glomerulosclerosis, have received a deceased donor kidney, and are less likely to have bladder dysfunction. Similar findings for children with renal transplant were reported from the analysis of Organ Procurement and Transplant Network database from 1995 to 2000. Patients aged 12 years and older, such as the adolescent in this vignette, are at increased risk for allograft failure. This is attributed to nonadherence to immunosuppressive medications and transition to adult healthcare services.

Renal allografts from a living donor are associated with higher survival rates, as compared to those from a deceased donor because of better circumstances for donor organ removal, storage, and engraftment. The potentially injurious effect on deceased donor allograft may lead to increased graft immunogenicity, which may influence the development of chronic allograft dysfunction.

Growth impairment is a common problem in children with chronic renal failure and is attributed to inadequate nutrition, metabolic acidosis, renal osteodystrophy, and growth hormone insensitivity. In the setting of an optimally functioning renal allograft, these factors are reversed, and children show improved growth and growth velocity. To prevent graft rejection, patients with functioning renal allograft are on immunosuppressive medications, which also increase the risk for infection in comparison to the general population.

Daily glucocorticoid therapy following renal transplantation (more than 5 mg/m2 per day) is a contributor to poor growth in children following renal transplantation. Strategies to reduce the cumulative glucocorticoid dose have included alternate day steroids, late or early steroid withdrawal, and immunosuppressive regimens avoiding steroids completely (steroid avoidance transplant protocol). Steroid avoidance protocols have been used only in patients at low risk for allograft rejection. Although studies are limited, improved growth outcomes without increased adverse effects of allograft rejection are reported with steroid avoidance. The beneficial effects of growth are most prominent in children younger than 5 years of age with steroid avoidance.

PREP Pearls
- Patients 12 years of age and older are at increased risk for renal transplant allograft failure.
- Living donor renal allografts are associated with higher survival rates, as compared to those from a deceased donor.
- Improved growth and growth velocity is seen in children post-renal transplantation.
- Immunosuppressive regimens with steroid avoidance have improved growth outcomes without increased adverse effects of allograft rejection.
- Patients receiving immunosuppressive medications are at increased risk for infections.

ABP Content Specifications(s)
- Understand the prognosis for a patient who has undergone renal transplantation

Suggested Readings
- Butani L, Perez RV. Effect of pretransplant dialysis modality and duration on long-term outcomes of children receiving renal transplants. Transplantation. 2011;91(4):447-451.
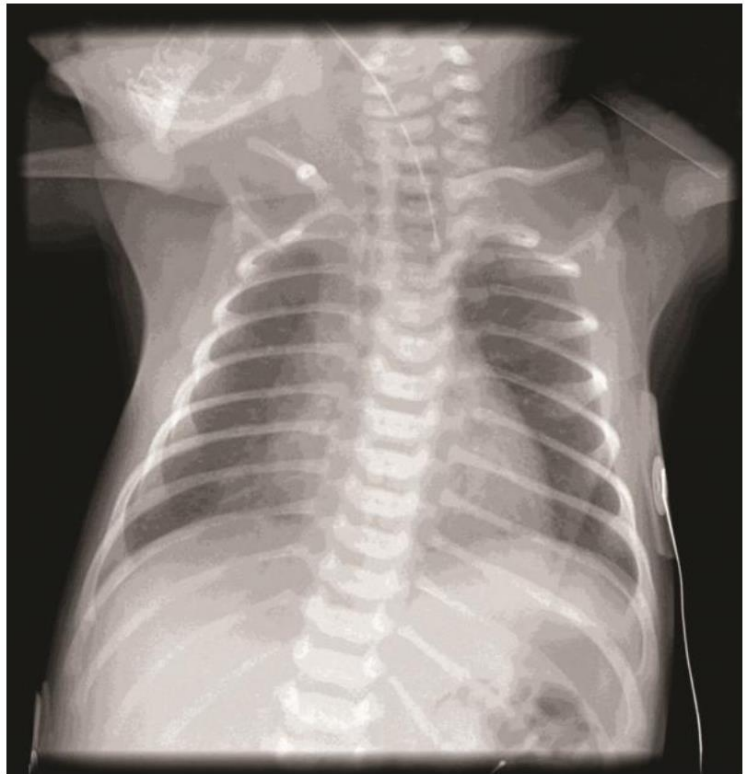
Question 147

You are called to evaluate a 1-hour-old newborn who is experiencing some coughing and choking that improves with suctioning. He was delivered by spontaneous vaginal delivery following a prenatal course complicated by polyhydramnios. Vital signs show a temperature of 37°C, heart rate of 120 beats/min, and a respiratory rate of 25 breaths/min. On physical examination, the newborn is drooling with copious oral secretions. His examination is otherwise unremarkable.

A chest radiograph is taken (Item Q147). Laboratory studies are remarkable for a normal complete blood cell count and electrolytes. A blood culture is pending.

His symptoms worsen significantly following the first attempt to breastfeed.

Of the following, the MOST likely diagnosis is

    A.   congenital diaphragmatic hernia

    B.   congenital esophageal stricture

    C.   esophageal atresia

    D.   esophageal duplication cyst

    E.   pulmonary sling

Item Q147. Chest radiograph for the newborn described in the vignette. Courtesy of C. Waasdorp Hurtado

Question 147   **Preferred Response: C**

There are 5 main types of esophageal atresia (Item C147). Esophageal atresia with a distal tracheoesophageal fistula (TEF) is the most common, accounting for 86%. This is followed by isolated esophageal atresia at 8% and isolated TEF at 4%. The least common are esophageal atresia with a proximal H-type TEF or a double TEF, both accounting for 1% of esophageal atresia patients. Infants with esophageal atresia have a high incidence of prematurity and approximately 50% have other anomalies.

ITEM C147: Variants of esophageal atresia and tracheoesophageal fistula.
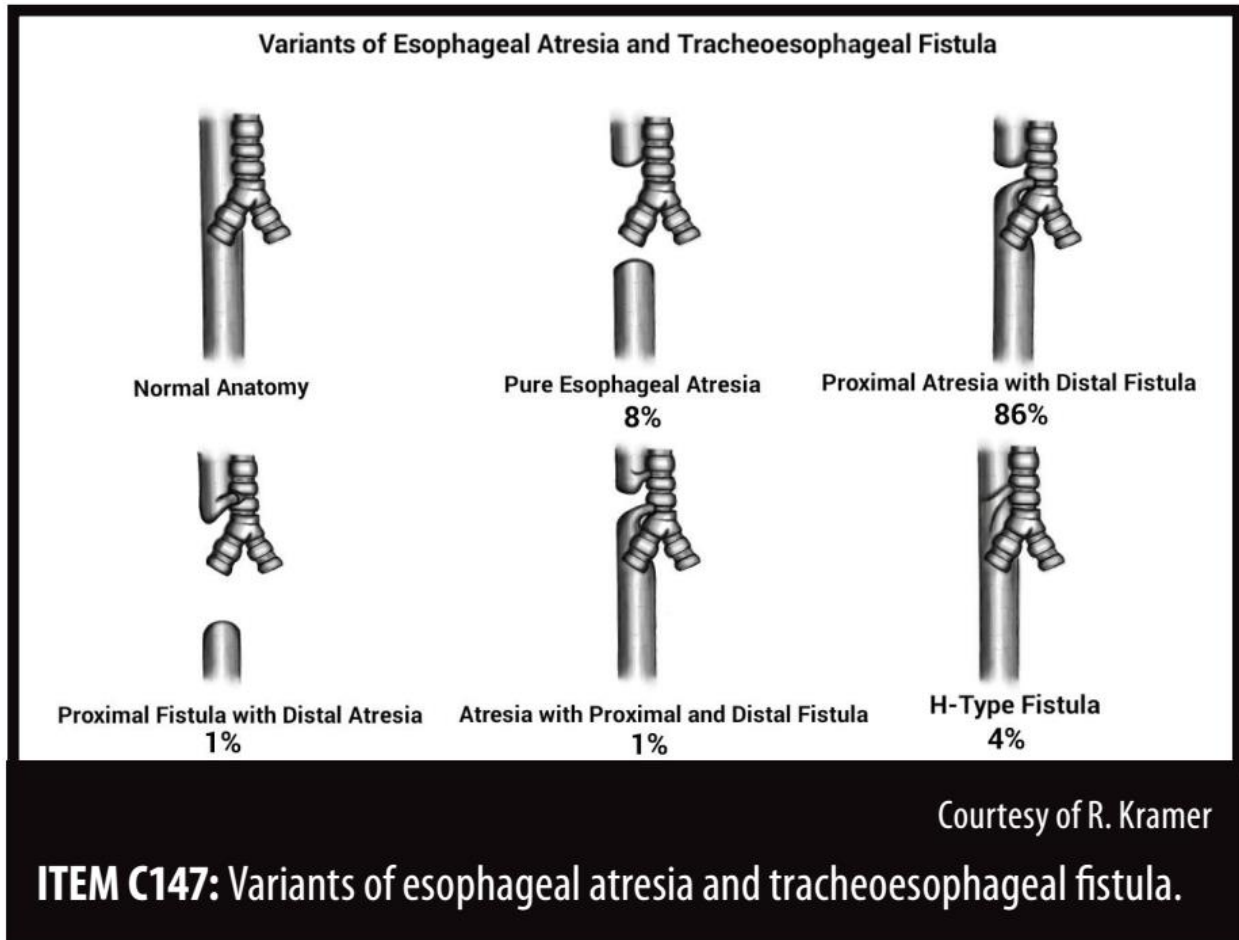
Other esophageal anomalies include congenital esophageal stenosis, presenting with 3 variants: fibromuscular stenosis, cartilaginous tracheobronchial remnants, and esophageal webs. These rare anomalies affect 1 in 25,000 to 50,000 live births and, in 85% of patients, are an isolated anomaly. They often present at the time of pureed food introduction or later with failure to thrive.

Duplication cysts can occur anywhere along the gastrointestinal tract. Duplications occur in 1 out of 4,000 to 8,000 live births, with about 33% affecting the foregut. The cysts usually have a

smooth muscle wall and are lined with either secretory, respiratory, or alimentary tract mucosa. The cysts can increase in size, obstructing the lumen, resulting in intolerance of feeds. Vascular rings are ligamentous and vascular structures that encircle the trachea and esophagus. These are very rare, making up less than 1% of vascular anomalies. Presentation often includes feeding difficulty or respiratory compromise. Vascular rings may be indirectly visualized with radiographs and by barium esophogram, but are best visualized with angiography, computed tomography, and magnetic resonance imaging.

For the newborn in this vignette, congenital diaphragmatic hernia would have a chest radiograph demonstrating intestines above the diaphragm. Esophageal stricture and vascular ring would not have this significant intolerance of secretions and, depending on the severity of the stricture, the orogastric tube should have passed. Esophageal duplication cysts do not commonly obstruct the entire lumen of the esophagus and a cystic structure should be seen on the chest radiograph.

PREP Pearls
- Esophageal atresia with a distal tracheoesophageal fistula is the most common form of esophageal atresia.
- Chest radiograph after orogastric tube placement is used as the initial evaluation for esophageal atresia.
- Esophageal strictures, duplication cysts, and vascular rings can also reduce the lumen of the esophagus and impact swallowing.

ABP Content Specifications(s)
- Recognize the structural anomalies that interfere with normal esophageal function

Suggested Readings
- Achildi O, Grewal H. Congenital anomalies of the esophagus. Otolaryngol Clin North Am. 2007;40(1):219-244. DOI:http://dx.doi.org/10.1016/j.otc.2006.10.010.

Question 148

You are called by the newborn nursery to evaluate a baby that is having difficulty feeding. On physical examination, you find a small, receding mandible, a cleft palate, bifid uvula, midfacial hypoplasia, and an abnormal red eye reflex consistent with a cataract. The tongue is posteriorly positioned. The newborn's father looks similar to the baby and reports a history of severe myopia, mitral valve prolapse, and a cleft palate repaired as a child.

Of the following, the test MOST likely to be abnormal in this newborn is

A.   audiogram

B.   brain magnetic resonance imaging

C.   echocardiogram

D.   liver function tests

E.   skeletal survey

Question 148   **Preferred Response: A**

The baby in the vignette has Stickler syndrome, which is a connective tissue disorder that is associated with midfacial hypoplasia, cleft palate, Pierre Robin sequence (PRS), hearing loss, and abnormalities of the eye, including high grade myopia, cataracts, and increased risk for retinal detachment. Pierre Robin sequence is the clinical constellation

of micrognathia, cleft palate, and glossoptosis in an individual that can lead to life-threatening obstructive apnea and feeding difficulties in the neonatal period. The obvious physical examination findings in this patient include the facial dysmorphology and abnormal red eye reflex secondary to the cataract. Health care providers should be mindful of the high risk of hearing loss as well because it could impact normal cognitive and speech development. Skeletal findings can include femoral head dysplasia, scoliosis, spondylolisthesis, and joint laxity early in life that progresses to early onset osteoarthritis, or kyphosis. It can be autosomal dominant or autosomal recessive in its inheritance pattern and can present with variable phenotypic expression, even within the same family.

Management involves a comprehensive multidisciplinary craniofacial clinic, correction of refractive errors with glasses, ophthalmologic treatment for retinal detachment, hearing aids for sensorineural and conductive hearing loss, and, if the PRS is severe, tracheotomy for respiratory status stabilization. Other methods utilized to stabilize the airway include prone positioning, nasopharyngeal airways, tongue lip adhesion, or a mandibular advancement procedure. Sometimes a new technique called mandibular distraction osteogenesis will take place very early in life in lieu of a tracheostomy. Distraction osteogenesis is a technique by which the lower jaw can be gradually moved forward over several days, utilizing a device that is surgically attached to the jaw. As the jaw moves forward, the tongue also moves forward, and the airway obstruction is relieved. This treatment can be performed as early as the first week of life.

An audiologic examination should be performed early on, as patients can have both conductive and sensorineural hearing loss in the neonatal period or develop it overtime. Therefore, patients with PRS need audiologic evaluations every 6 months through 5 years of age, and annually thereafter.

Pierre Robin sequence (PRS) (Item C148) can present as an isolated finding, in association with additional congenital malformations (PRS-Plus), or in association with a defined genetic disorder (syndromic PRS). The most common genetic disorders associated with PRS are 22q11.2 deletion or Stickler syndrome. In fact, 11% to 18% of babies with PRS will have Stickler syndrome. Clinical testing exists for both of these disorders. It can also be seen with other disorders including Treacher Collins syndrome, Nager syndrome, Miller syndrome, chromosomal abnormalities, teratogenic exposure, and some skeletal dysplasias (Kniest dysplasia, campomelic dysplasia). The respiratory problems with PRS require careful attention, as many patients will require prolonged stays in the neonatal intensive care unit, or surgical interventions such as tracheotomy or a gastrostomy tube. The respiratory and feeding difficulties can persist into childhood. Obstructive sleep apnea, failure to thrive, and long term developmental difficulties are common. A genetics workup is strongly recommended in the newborn period, as it can prove

valuable for accurate diagnosis and an appropriate care plan with specific surveillance guidelines for the child. At the minimum, one should order a high resolution karyotype, microarray, and a detailed ophthalmological assessment at birth and 6 months of age. A targeted family history should be taken for the presence of clinical features suggestive of Stickler syndrome in the family, such as osteoarthritis, myopia, and short stature. If Stickler syndrome, as in this case, is highly suspected, then an audiologic examination should be ordered. Stickler syndrome is not commonly associated with hepatic dysfunction, brain abnormalities, or congenital heart defects, so liver function tests, brain magnetic resonance image, or an echocardiogram would not be indicated in this scenario. A skeletal survey may show abnormalities, but would not take immediate precedence over clinical assessment for hearing loss in the newborn period for the patient in this vignette.

Courtesy of L. Parsley

ITEM C148: Newborn with Pierre Robin sequence.

PREP Pearls
- Stickler syndrome is a connective tissue disorder that is associated with midfacial hypoplasia, cleft palate, Pierre Robin sequence (PRS), hearing loss, and abnormalities of the eye, including high grade myopia, cataracts, and increased risk for retinal detachment.
- Pierre Robin sequence is the clinical constellation of micrognathia, cleft palate, and glossoptosis in an individual that can lead to life-threatening obstructive apnea and feeding difficulties in the neonatal period.
- Pierre Robin sequence can present as an isolated finding, in association with additional congenital malformations (PRS-Plus), or in association with a defined genetic disorder (syndromic PRS). The most common genetic disorders associated with PRS are 22q11.2 deletion or Stickler syndrome.

ABP Content Specifications(s)
• Identify the clinical features associated with Pierre-Robin sequence

Suggested Readings
• Evans KN, Sie KC, Hopper RA, Glass RP, Hing AV, Cunningham ML. Robin sequence: from diagnosis to development of an effective management plan. Pediatrics. 2011;127(5):936-948. DOI: http://dx.doi.org/10.1542/peds.2010-2615.

Question 149

A 2-year-old, African-American boy presents to your office for a new patient health supervision visit. His birth and past medical history are unremarkable. His growth and development are appropriate for his age. On physical examination, you notice an extra digit adjacent to the fifth toe bilaterally (Item Q149). His mother reports that several family members on the paternal side had similar extra digits, but they are otherwise well.

Item Q149. Foot of the child described in the vignette. Courtesy of D. Krowchuk.

Of the following, the MOST common pattern of inheritance for this condition is

A.   autosomal dominant

B.   autosomal recessive

C.   paternal nondisjunctional event

D.   X-linked dominant

E.   X-linked recessive

Question 149   **Preferred Response: A**

Polydactyly, the presence of supernumerary digits, is the most common congenital deformity of the hands and feet. It is usually an isolated condition that is inherited as an autosomal dominant trait. However, it may be a feature of several syndromes, so it is important for the pediatric health care provider to be familiar with the appropriate management and indications for referral. Polydactyly is classified based on the location of the extra digits and the degree of formation. Preaxial polydactyly involves the thumb or great toe; postaxial affects the fifth digit; mesoaxial involves the central 3 digits. Well-formed or rudimentary further describes the shape and form of the extra digit.

Molecular defects seen in patients with polydactyly include chromosomal abnormalities as well as single gene defects. Polydactyly is associated with a number of syndromes including trisomy 13, chondroectodermal dysplasia, Meckel-Gruber, otopalatodigital, Bardet-Biedl, and short rib polydactyly syndrome.

Management of polydactyly depends on the digit location and formation. The rudimentary skin tag form may be simply tied off, but cosmetic outcomes and patient care may improve with excision. Another consideration is functionality, that is, proper fit of shoes. Well-formed digits may have bone and neurovascular structures, therefore, referral to a surgeon is necessary and radiologic evaluation may be helpful to define the anatomy. For patients with signs and symptoms of an underlying genetic disorder, referral to a genetic specialist and evaluation for comorbidities is warranted.

PREP Pearls
- The most common pattern of inheritance for isolated polydactyly is autosomal dominant.
- The location and degree of supernumerary digit formation determines the most appropriate management in cases of polydactyly.
- Because polydactyly may occur as part of a syndrome, providers must be aware of potential comorbidities and the indications for referral to a specialist.

ABP Content Specifications(s)
- Plan the appropriate management of polydactyly and understand when referral is appropriate

Suggested Readings
- Carrigan RS. The upper limb: polydactyly. In: Kliegman RM, Stanton BF, St Geme JW III, Schor NF, Behrman RE, eds.Nelson Textbook of Pediatrics. 20th ed. Philadelphia, PA: Saunders; 2015: 3302-3310.

Question 150

A 15-year-old adolescent girl presents to your office with a 1-month history of progressive fatigue and exercise intolerance. Her medical history is remarkable for autoimmune thyroiditis for which she takes 112 µg of oral levothyroxine daily. She has recently had normal thyroid stimulating hormone, thyroxine (T4), and free T4 levels. Her physical examination is remarkable only for pallor. She specifically denies any history of hematuria, hematochezia, epistaxis, or unusual bruising. She has had a normal diet for age. The results of a complete blood cell count are shown:

- White blood cell count   11,000/µL (11.0 x 10^9/L)
- Hemoglobin   7.2 g/dL (72 g/L)
- Mean corpuscular volume   116 fL
- Platelet count   467 x 10^3/µL (467 x 10^9/L)
- Reticulocyte count   0.5%

Of the following, her fatigue and pallor are MOST likely a result of damage to the

A.   adrenal cortex

B.   adrenal medulla

C.   gastric parietal cells

D.   hepatocytes

E.   pancreatic islet cells

Question 150   **Preferred Response: C**

The differential diagnosis of macrocytic anemia in children includes vitamin B12 deficiency, folate deficiency, and bone marrow failure. Folate deficiency is most often dietary, typically in children whose diets rely heavily on goat milk. When vitamin B12 (also called cobalamin) is consumed, it attaches to haptocorrin and travels to the duodenum, where it is hydrolyzed and released from the haptocorrin. The free vitamin B12 then binds to gastric intrinsic factor and travels to the ileum. It is then absorbed in the ileum, enters the blood stream, and binds to transcobalamin. Vitamin B12 deficiency in pediatrics most often occurs either because of an absence of the terminal ileum (where B12 is absorbed), or because of a deficiency of intrinsic factor. The deficiency of intrinsic factor and subsequent vitamin B12 deficiency is called pernicious anemia. Congenital pernicious anemia occurs when there is a genetic defect resulting in hypofunctional or absent intrinsic factor. Pernicious anemia in adolescents typically results from gastric atrophy and achlorhydria caused by antibodies to the parietal cell and intrinsic factor. Left untreated, vitamin B12 deficiency results in a macrocytic anemia, in addition to neurologic symptoms including paresthesias, ataxia, and gait abnormalities, as a result of posterior and lateral spinal column degeneration. Vitamin B12 deficiency can be treated with the parenteral administration of vitamin B12.

The patient in the vignette has a medical history remarkable for autoimmunity, suggesting that her macrocytic anemia also has an autoimmune origin. Thus, it is likely that her macrocytic anemia is caused by autoimmune damage to the gastric parietal cells.

Autoimmune damage to the adrenal cortex results in Addison disease, a deficiency of cortisol (glucocorticoids) and often aldosterone (mineralocorticoids). It will not cause a macrocytic anemia. The adrenal medulla produces catecholamines. It can be damaged and underproduce catecholamines or have a tumor such as a pheochromocytoma and overproduce catecholamines. Neither will result in a macrocytic anemia. Hepatic injury in the form of alcoholic cirrhosis can coexist with vitamin B12 deficiency, but it is not, by itself, a cause of macrocytic anemia. Injury to the pancreatic islet cells results in an insulin deficiency and diabetes, but not macrocytic anemia.

PREP Pearls
- Vitamin B12 deficiency results in a macrocytic anemia and neurological changes as a result of the degeneration of the posterior and lateral spinal columns.
- Intrinsic factor is produced in the gastric parietal cells and is necessary for the absorption of vitamin B12.
- The most common cause of vitamin B12 deficiency in adolescents is autoimmune gastric atrophy and achlorhydria, resulting in damage to the gastric parietal cells, with a resultant deficiency of intrinsic factor.
- Vitamin B12 is absorbed in the terminal ileum.

ABP Content Specifications(s)
- Recognize the causes of macrocytic anemia

American academy of pediatrics   486

Suggested Readings

- Kaferle J, Strzoda CE. Evaluation of macrocytosis. Am Fam Physician. 2009;79(3):203-208.

Question 151

A 16-year-old adolescent is having problems with his sleep. He says he can never fall asleep before midnight, and usually falls asleep around 1:00 AM or even 2:00 AM. He wakes at 6:00 AM for school, but only after his mother has to struggle to get him out of bed. He says he feels sleepy throughout most of the school day and occasionally falls asleep in class. On most days, he takes a nap right after getting home from school, sleeping for more than 1 hour. He says he has always been an "evening person." Upon questioning, his mother says he does not snore at night. She says at home they all tend to be evening people. They have instructed him to turn off all of his electronics at 11:00 PM, but this has not seemed to help him fall asleep any earlier. On physical examination, his tonsils are 2+ and symmetric.

Of the following, the next MOST appropriate step is

A.   encourage that he catch up on sleep on the weekends by staying in bed past noon

B.   recommend that he discontinue taking naps after school

C.   recommend that he perform a 20-min workout about 1 hour before going to bed

D.   refer him to an ear/nose/throat surgeon to consider a tonsillectomy

E.   make a referral for him to have polysomnography

Question 151   **Preferred Response: B**

The adolescent in the vignette is chronically getting about 4 to 5 hours of sleep each night, and is regularly taking a 1-hour nap every afternoon. He has dysfunction at school because of feeling sleepy and not paying attention in class. His parents appropriately have advised him to turn off all of his electronics at 11:00 PM, but this did not resolve his problem. His family members are all reported to be "evening people," therefore it would be worth asking further if there are family distractions happening for this patient at night.

Adolescents tend to have phase advanced sleep, meaning that there is a natural tendency while progressing into adolescence of falling asleep later in the evening, followed by waking up later in the morning. Some schools now try to match their schedules to that pattern, by scheduling high school classes to start later in the morning than classes for younger children.

The total number of hours a day of sleep that humans need is fairly consistent, so when there is chronically short duration sleep at night, a sleep deficit is created that requires payback to be able to maintain normal level functioning. This experienced sleep deficit is what is most likely leading this adolescent to take a nap every afternoon. A downside of taking naps lasting beyond about 15 min in duration is that "sleep inertia" is created in which it is harder for someone to feel fully awake for the subsequent hours after waking. Even after taking his afternoon nap of approximately 1-hour duration, he is likely to experience sleep inertia rather than feeling truly refreshed upon awakening. More importantly in this situation, the use of afternoon naps decreases the need for sleep later that evening, and impairs the ability to easily fall asleep at night. The most immediate need for the adolescent in this vignette is sleep hygiene management. Both patient and parent will need to commit to not allowing the taking of naps after school, while also providing a reasonable opportunity for him to fall asleep before or up to 11:00 PM (ie, the home activity winds down, no personal electronics, etc). It may be hard for him to make himself stay awake after school at first, given the routine that has been established, but doing so should lead to a rapid resolution of the problem.

Extra catch-up sleep on the weekends can help to restore an accumulated sleep deficit and restore clear cognition. However, a big downside of sleeping in until noon or later every weekend is that this perpetuates a phase-advanced circadian rhythm, which does not help the adolescent's sleep schedule during the following school week. Maintaining a relatively consistent sleep pattern every day of the week is another key way to help maintain an appropriate-for-circumstances biological clock.

Obtaining regular exercise has been shown to help improve sleep habits overall. However, performing significant physical exercise within an hour of the intended bedtime typically impairs being able to settle the mind and body down and be able to fall asleep. Exercise should happen earlier in the day to be able to obtain sleep benefits at night.

Referring the patient to a surgeon to consider a tonsillectomy may be appropriate in the setting of significant problems with snoring, nocturnal enuresis, or gasping respirations. If these symptoms are present, a polysomnogram should be considered before a referral to a surgeon. Tonsils that

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are 2+ sized, as in the patient in this scenario, would not be a very likely cause of sleep apnea difficulties in someone with an otherwise normal oropharynx.

Polysomnography would be the appropriate next step to document apneic pauses if sleep apnea is suspected. In this vignette, the patient's history suggests that he is having sleep hygiene problems and referral for a sleep study is not necessary, presuming an uncomplicated course with sleep hygiene interventions.

Sometimes sleep can be disrupted because of nightmares, particularly for individuals with post-traumatic stress disorder for whom repeated nightmares may decrease sleep quality and cause sleep avoidance behaviors. Nightmares occur during rapid eye movement sleep when muscle tone and movement is inhibited, and generally in the latter part of the morning. Nightmares are different than sleep terrors in that children between 4 and 12 years of age may experience them. Sleep terrors usually occur during the first third of sleep at night, cause abrupt apparent awakening (though typically not full consciousness) with a loud scream, and physical agitation that is unresponsive to parental calming.

PREP Pearls
• Chronic daytime sleepiness is often related to poor sleep habits.
• Sleep hygiene interventions are the first line treatment for most sleep problems in young people.
• For someone with chronic insomnia, taking naps during the day is likely to interfere with their ability to obtain sleep at night.

ABP Content Specifications(s)
• Plan the appropriate management of bedtime refusal/frequent awakening
• Distinguish between nightmares and night terrors
• Recognize symptoms that reflect poor sleep quality and plan appropriate evaluation

Suggested Readings
• Cespedes EM, Gillman MW, Kleinman K, Rifas-Shiman SL, Redline S, Taveras EM. Television viewing, bedroom television, and sleep duration from infancy to mid-childhood. Pediatrics. 2014;133(5):e1163-e1171. DOI:http://dx.doi.org/10.1542/peds.2013-3998.
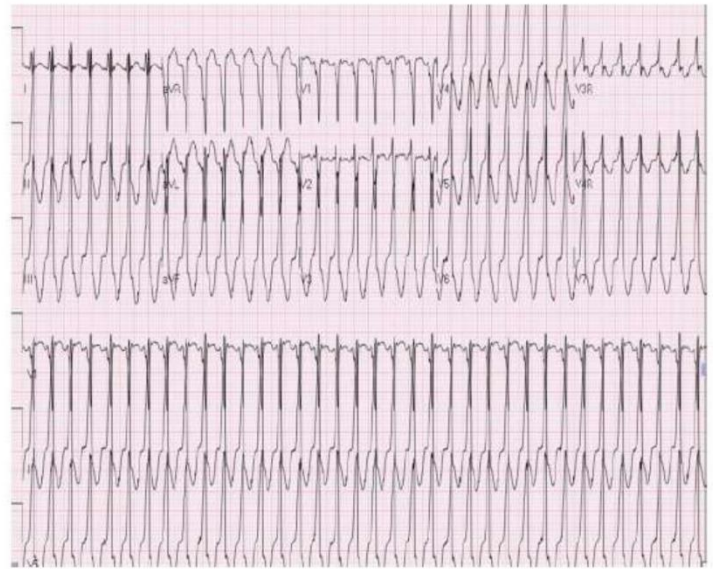
Question 152

An 11-year-old girl faints as she walks off the soccer field at sleep away camp. The temperature at camp has been 95°F (35°C) all day and she has been outside and active. Emergency medical services (EMS) was called and she was taken to the small emergency department near the camp. By history, she was unresponsive on arrival with a heart rate of 190 beats/min, blood pressure of 65/45 mm Hg, and respiratory rate was 15 breaths/min. While she was being evaluated, she regained consciousness, her heart rate dropped to 90 beats/min, and her blood pressure normalized to 105/70 mm Hg. She was observed for several hours and discharged home. She comes to see you in the office next week. The family has requested a copy of the EMS rhythm strip, but does not have it with them. She is feeling well today and has an unremarkable physical examination. She is anxious to get back to playing competitive soccer.

The patient faints again in your office and you call 911. You obtain an electrocardiogram in the office (Item Q152).

Of the following, the BEST next step is

A.   adenosine, 200 µg/kg intravenously

B.   cardioversion with 2 joules/kg

C.   digoxin, 5 µg/kg slow intravenously

D.   normal saline, 20 mL/kg intravenously

E.   propranolol, 40 mg orally

Item Q152. Electrocardiogram for the girl described in the vignette. Courtesy of E.A. Greene

Question 152   **Preferred Response: B**

This patient is unconscious, unstable, and in ventricular tachycardia. According to the American Heart Association Pediatric Advanced Life Support (PALS) recommendations, the preferred treatment is electrical cardioversion. Adenosine is incorporated into the PALS recommendations, but only in the treatment of stable wide complex tachycardia in order to differentiate between ventricular tachycardia (VT) and supraventricular tachycardia (SVT). There is an uncommon form of VT that is adenosine-sensitive as well. In the case of the child in this vignette who is unconscious, one would not want to delay electrical cardioversion. Digoxin is not the treatment for VT; it is used for atrial arrhythmias and for some patients with SVT. A normal saline bolus would help to raise the patient's blood pressure, but would not convert the abnormal cardiac rhythm. Propranolol is a β-blocker, a class of medication useful in long term treatment of VT. The oral form would not work quickly enough for an unstable patient.

When faced with a patient in wide complex tachycardia, especially if unstable, it is safest to assume that the rhythm is VT. Even if the rhythm is an SVT with bundle branch aberrancy, if the patient is unstable, then synchronized cardioversion is the treatment of choice. A starting dose of 2 J/kg is recommended. If the patient were stable, there would be more time to differentiate SVT from VT. Ventricular tachycardia will more classically show atrioventricular (AV) dissociation. This feature is the most helpful in differentiating SVT with aberrancy from VT. Inspection of the electrocardiogram for a regular P to P interval and a regular R to R interval will eventually show these to stop matching up one to one if there is AV dissociation as would be expected in VT. Adenosine may convert SVT by causing transient AV block, but can also cause at least a momentary pause in VT if it involves the posterior fascicle of the left bundle. In a more stable patient, who does not require immediate cardioversion, amiodarone may be used to treat VT. Lidocaine and procainamide are other medication choices to consider.

There are several subtypes of VT. The most common type in the pediatric population arises from the right ventricular outflow tract (RVOT) and may be benign. This form of ventricular tachycardia will have an inferior axis (negative in V1) because it comes from the superior aspect of the heart. There will also be left bundle branch morphology because the activation arrives in the left ventricle after the right ventricle (Item C152A). This type of VT or ventricular ectopy may be benign if it suppresses with the sinus tachycardia that occurs with exercise, is always monomorphic, and the heart is structurally normal. Monomorphic VT is regular (all the QRS complexes are the same) and is consistent with the VT originating from one area of the ventricle and not changing its axis in the midst of the arrhythmia. It often is seen on stress tests during the recovery phase when the heart rate is decreasing. This type of VT may cause hypotension if rapid or prolonged, but may be well tolerated, especially if it is not sustained.
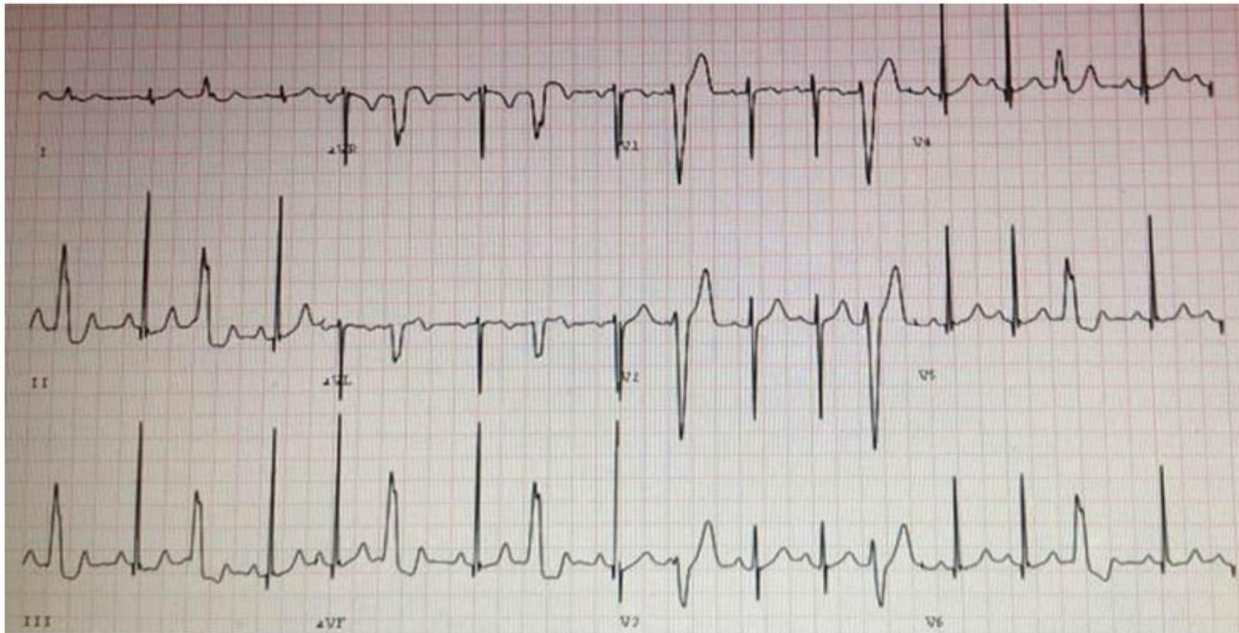
Item C152A. Example of Premature ventricular contractions (PVCs) from the right ventricular outflow tract. The PVCs have an inferior axis (negative in V1) and a left bundle branch morphology (wide with a RsR' in V6) that demonstrate the origin of the complexes. Courtesy EA Greene

Ventricular tachycardia that is not from the RVOT, worsens with exercise or increases with catecholamines, is polymorphic, is associated with congenital heart disease, and is more likely to be dangerous. Polymorphic refers to the morphology of the VT; the QRS will differ beat to beat and the VT may change axis during the arrhythmia. This suggests a more disorganized VT not originating from 1 focus. This form of VT is of higher risk. Arrhythmogenic right ventricular cardiomyopathy or dysplasia (ARVD), catecholamine-sensitive polymorphic VT (CPVT), and long QT (LQT) are inheritable arrhythmia syndromes capable of causing polymorphic ventricular tachycardia. Torsades de pointes is a specific form of polymorphic VT seen with LQT syndrome (Item C152B). In each of these syndromes, the first presentation may be collapse or sudden cardiac arrest.
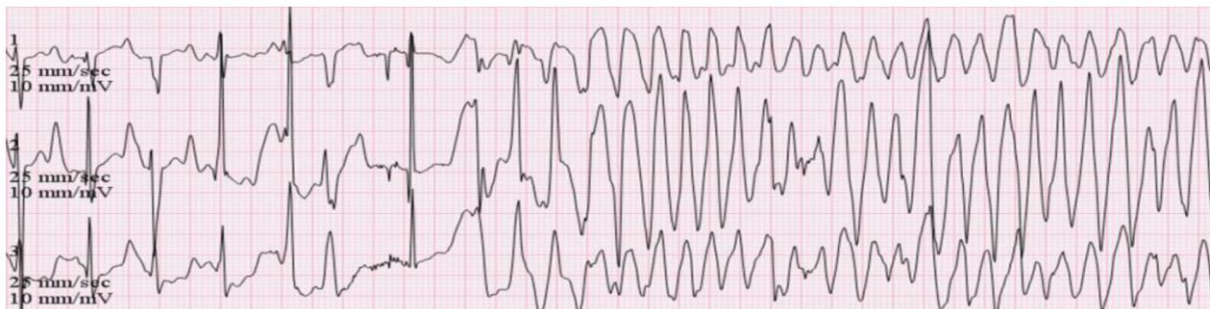
Item C152B. Example of Torsades de Pointe (TdP) in patient with Long QT syndrome. There is T wave alternans prior to initiation and premature ventricular complexes.   Courtesy EA Greene

Long term management of VT depends on the type of VT, any underlying genetic abnormality, and cardiac anatomy. If the ectopy suppresses with exercise, this is reassuring. Polymorphic VT associated with congenital heart disease, CPVT, LQT syndrome, or ARVD is much higher risk. In patients with one of the inheritable arrhythmia syndromes, a combination of medication and protection with an implantable cardiac defibrillator will be needed if VT has developed.

PREP Pearls
•   Unstable wide complex tachycardia requires cardioversion.
•   Monomorphic (premature ventricular contractions with the same morphology and axis throughout) ectopy from the right ventricular outflow tract in a patient with a structurally normal heart, which suppresses with exercise, is a lower risk type of ectopy.
•   Several hereditary arrhythmia syndromes are associated with a more malignant form of tachycardia.

ABP Content Specifications(s)
•   Recognize the role of hyperthyroidism in persistent sinus tachycardia

Suggested Readings
•   Cannon BC, Snyder CS. Disorders of the cardiac rhythm and conduction. In: Allen HD, Driscoll DJ, Shaddy RE, Feltes TF, eds. Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. 8th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2012: 441-472.

Question 153

You are seeing a 14-year-old adolescent girl in your office after her yearly ophthalmology appointment, where she was found to have a cataract in her left eye. During your review of systems, she describes bilateral wrist pain that started 5 months ago. She also reports pain in her hands that interferes with writing and school work, which started at the same time as her wrist pain. She has noticed bilateral knee swelling and pain over the last month. You are concerned that the patient may have juvenile idiopathic (rheumatoid) arthritis.

Of the following, the MOST specific sign or symptom for this diagnosis is

A.   fatigue

B.   frequent diarrhea

C.   nighttime back pain

D.   pain with joint movement

E.   rash localized to the face

Question 153   **Preferred Response: D**

Among the response choices, the symptom most specific for juvenile idiopathic arthritis (JIA) is pain with joint movement. The signs and symptoms associated with arthritis are decreased range of motion with either passive or active movement, joint swelling, and pain with range of motion. Although joints may be erythematous and warm, these signs are not always present and can be difficult to assess if the examiner has warm or cold hands. Fatigue can occur with any autoimmune condition, but is not specific to this diagnosis. Frequent diarrhea occurring in a patient with arthritis should raise a concern for inflammatory bowel disease (IBD). Patients with IBD may have arthritis as an early or presenting symptom, therefore, any patient with arthritis should be screened for gastrointestinal issues. Nighttime back pain in a child should raise the concern for leukemia. Patients with leukemia will often present with bone pain that is severe, and occasionally arthralgia or even arthritis. Patients with JIA have pain that is worse in the morning, whereas patients with leukemia typically have significant nighttime pain. Arthritis or arthralgia associated with a facial rash should raise concerns for infection or a systemic autoimmune condition. Several common infections, such as mononucleosis or parvovirus, can present with arthritis and a rash. Arthritis and rash can also be the initial presentation of systemic lupus erythematosus.

The differential diagnosis when evaluating a child for JIA is broad. Arthritis may be a presenting symptom for reactive processes such as poststreptococcal arthritis, rheumatic fever, serum sickness, and postinfectious arthritis (these illnesses tend to be self-limiting or have other symptoms); infections that can present with symptoms of arthritis include septic arthritis, discitis, or osteomyelitis; mononucleosis, parvovirus, and Lyme disease. Other autoimmune conditions with findings similar to JIA include lupus, mixed connective tissue disease, Sjögren syndrome, sarcoidosis, and inflammatory bowel disease. Joint enlargement caused by conditions other than arthritis can include trauma; benign tumors such as osteoid osteoma or osteoblastoma; malignancies such as leukemia, neuroblastoma, osteosarcoma, Ewing sarcoma, and rhabdomyosarcoma. Because there is no diagnostic laboratory study for JIA, it is often a diagnosis of exclusion. Laboratory findings that support the diagnosis of JIA, are anemia, leukocytosis, elevated acute phase reactants, and hypergammaglobulinemia.

Juvenile idiopathic arthritis consists of 6 types of arthritis with different presenting features (Item C153A).

Item C153A. Classification of Juvenile Idiopathic Arthritis.

Type	Presenting Features
Systemic	• Arthritis in ≥1 joint(s) / preceded by fever for at least 2 weeks, which occurs daily for at least 3 days. • Fever usually accompanied by an evanescent rash, lymphadenopathy, hepatomegaly or splenomegaly, and/or serositis.
Oligoarticular	1-4 joint(s) affected with arthritis during the first 6 months of disease.
Polyarticular	• Arthritis affects ≥5 joints. • Can be rheumatoid factor positive or negative (only 10%-15% of patients with JIA are rheumatoid factor positive).
Psoriatic	Arthritis and at least 2 of the following: dactylitis, nail pitting, or a family history of arthritis.
Enthesitis-related	Arthritis or enthesitis with at least 2 of the following: sacroiliac tenderness or lumbosacral pain, presence of HLA-B27, onset of arthritis in a male greater than 6 years old, anterior uveitis, and/or family history of HLA-B27 associated disease. *HLA-B27 alone is not diagnostic, and in high-risk individuals with a family history of enthesitis-related arthritis, about 20% developed this disease. In a European population study, ankylosing spondylitis was found in 1.3% of HLA-B27 positive individuals in the population at large and in 21% of HLA-B27 positive relatives of B27 positive patients with spondylitis.*
Undifferentiated	Arthritis that fulfills criteria in no other category, or in two or more juvenile idiopathic arthritis categories.

Uveitis, also known as iridocyclitis, is a serious complication of JIA resulting from chronic nongranulomatous inflammation of the anterior eye chamber affecting the iris and ciliary body. The uveitis associated with JIA is usually clinically silent with an insidious onset. Risk factors for uveitis in patients with JIA include JIA subtype, age at onset of disease, and antinuclear antibody (ANA) status. The highest risk group is female patients with pauciarticular JIA who are ANA positive and diagnosed before the age of 4 years. Screening guidelines have been developed for uveitis based on risk (Item C153B). The severity of uveitis does not correlate well with arthritis activity; therefore the status of joint disease should not affect the frequency of screening. While ANA is often positive in patients with JIA who have uveitis, it is important to remember that ANA is positive about 57% of the time in all patients with JIA.

Item C153B. Uveitis Screening Guidelines for Patients With Juvenile Idiopathic Arthritis.

Juvenile Idiopathic Arthritis Onset type	ANA Status	Disease Onset <7 years	Disease Onset >7 years
Pauciarticular or polyarticular	Positive	Every 3-4 months 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	Positive or Negative	Yearly	Yearly

Courtesy of A. Brown

Cataracts are among the late complications of JIA, resulting from chronic uveitis or chronic ophthalmic steroid use and are not associated with specific laboratory findings. Patients with JIA can develop bone erosions, and these patients are more likely to be rheumatoid factor (RF) positive. Osteoporosis may result from chronic steroid use, methotrexate use, or the JIA itself. Vitamin D and calcium should be supplemented in patients with JIA to help prevent osteoporosis. Leg length discrepancy, resulting from increased inflammation in affected joints that increases blood flow and growth factors, occurs in patients with JIA with prolonged poor disease control. Some of these patients may have increased acute phase reactants, such as erythrocyte sedimentation rate, C-reactive protein, or thrombocytosis.

Current treatment recommendations for JIA with low disease activity include an initial nonsteroidal anti-inflammatory drug (NSAID) trial (Item C153C). 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 to NSAID treatment, or if more than 4 joints are involved, then methotrexate with or without systemic or intra-articular glucocorticoids is recommended. If the patient fails methotrexate therapy, then a tumor necrosis factor-α (TNF-α) inhibitor, usually adalimumab or etanercept, is added to therapy. If the patient fails these regimens, then abatacept (a T-cell modulator) is recommended. In patients with 5 or more active joints, NSAIDs should not be used as monotherapy. In patients with systemic JIA (fevers, rash, and arthritis) the recommendations are different. The first-line drugs are NSAIDs, but systemic corticosteroids are used if there is inadequate response. If there is still failure to control the arthritis then methotrexate is used in patients without fever and rash. If fever and rash are present then abatacept, anakinra (an IL-1 blocker), or toculizumab (an IL-6 blocker) is recommended.

Item C153C. Medication Dosages for Juvenile Idiopathic Arthritis Treatment.

Nonsteroidal anti-inflammatory Drugs	Anti-inflammatory Dose*	Dosage Forms
Ibuprofen	10 mg/kg/dose 4 times a day OR 13 mg/kg/dose 3 times a day (maximum dose 2400 mg/day)	Pill or liquid
Naproxen	10 mg/kg/dose twice daily (maximum dose 1000 mg/day)	Pill or liquid
Meloxicam	0.25 mg/kg/dose once daily (maximum dose 15 mg/day)	Pill or liquid
Celecoxib	50 mg twice daily for children 2-17 years of age and weighing 10-25 kg 100 mg twice daily for patients weighing >25 kg (maximum dose 200 mg/day)	Capsules (may be opened, sprinkled onto apple sauce, and taken immediately with water)

*The nonsteroidal anti-inflammatory drug doses listed are anti-inflammatory doses. Smaller doses are used for fever and pain relief.

Courtesy of A. Brown

PREP Pearls
- Juvenile idiopathic arthritis is a clinical diagnosis. Emergent conditions such as leukemia and septic arthritis must be excluded.
- Juvenile idiopathic arthritis can be managed initially with nonsteroidal anti-inflammatory drugs.
- A serious and preventable complication of juvenile idiopathic arthritis is damage caused by uveitis.

ABP Content Specifications(s)
- Recognize the long-term complications associated with juvenile rheumatoid (idiopathic) arthritis
- Plan the appropriate management of juvenile rheumatoid (idiopathic) arthritis, while recognizing side effects of some therapies
- Recognize the clinical findings associated with the various types of juvenile rheumatoid (idiopathic) arthritis
- Formulate a differential diagnosis for juvenile rheumatoid (idiopathic) arthritis

Suggested Readings
- Bernard R. Approach to the child with joint inflammation. Pediatr Clin North Am. 2012;59:245-262. DOI:http://dx.doi.org/10.1016/j.pcl.2012.03.003.

Question 154

You are volunteering as an event physician for a regional high school softball tournament. A 14-year-old adolescent presents to the medical tent for evaluation of her left eye after being hit by a pitch during a game. On physical examination, you note blood in the anterior chamber of the left eye. The patient has normal pupillary reflexes and extraocular eye movements. She has no periorbital tenderness and normal sensation around the eye. Examination of the cornea with a cobalt blue light following fluorescein staining does not reveal any defects. You discuss this injury with the adolescent and her parents.

Of the following, the MOST appropriate statement to include in the discussion is that this type of injury

A.   carries a risk of secondary hemorrhage that is highest 7 to 14 days after the injury

B.   does not require restriction from sports participation

C.   is less common in the pediatric population than in adults

D.   is likely to result in some permanent vision loss

E.   merits urgent evaluation by an ophthalmologist

Question 154   **Preferred Response: E**

The girl in the vignette has a hyphema, a collection of blood in the anterior chamber of the eye that results from disruption of the blood vessels of the iris. An athlete who sustains a hyphema should be evaluated urgently by an ophthalmologist. Hyphema carries the risk of additional bleeding, and a large collection of blood can result in staining of the cornea or glaucoma, conditions that can affect visual acuity.

Hyphema usually results from direct trauma to the eye. Affected individuals typically report pain and blurry vision. Physical examination reveals a collection of blood in the anterior chamber. Treatment includes rest and cycloplegic medication. Nonsteroidal anti-inflammatory drugs should be avoided because they may increase the risk of bleeding. Secondary hemorrhage occurs in up to one-third of patients with hyphema, with the risk being highest 2 to 7 days after injury. Evidence suggesting that rest prevents rebleeding is limited, but most ophthalmologists recommend restricting physical activity until the hyphema resolves and the risk of rebleeding has passed. Hyphema is more common in children than adults, with the highest incidence seen between 10 and 20 years of age. Surgery may be indicated for large hyphemas that could potentially cause optic nerve damage, but vision loss after hyphema is rare. There are no published return-to-play guidelines following eye injuries; an ophthalmologist should provide clearance before the child returns to sports.

Severe pain, lack of normal extraocular motion, disruption of the sclera or cornea, and decreased visual acuity are signs and symptoms of globe rupture. Globe rupture is an emergency; these patients should have an eye shield placed and be referred to the emergency department for ophthalmologic evaluation.

Sports and recreational activities account for about one-quarter of the eye injuries seen in the emergency department. Basketball, baseball, softball, and football are the sports with the highest risk of eye injury. Common sports-related eye injuries include corneal abrasions and corneal foreign body. Approximately 80% of eye injuries occur in individuals not wearing eye protection; appropriate sports eyewear can reduce the risk of eye injury. The American Academy of Pediatrics (AAP) and the American Academy of Ophthalmology (AAO) issued a joint statement in which they "strongly recommend" appropriate protective eyewear for athletes in moderate or high-risk sports. Safety glasses should have shatter-resistant polycarbonate lenses.

PREP Pearls
•   The presence of a hyphema merits urgent referral to ophthalmology.
•   Athletes who participate in sports with moderate or high risk of eye injury should wear appropriate protective eyewear to reduce their risk of injury.

ABP Content Specifications(s)
•   Understand the criteria for return to play in sports after an eye injury

Suggested Readings

*   American Academy of Pediatrics Committee on Sports Medicine and Fitness. Protective eyewear for young athletes.Pediatrics. 2004;113(3 Pt 1):619-622. http://pediatrics.aappublications.org/content/113/3/619.long.

Question 155

A mother brings her 4-month-old and 7-year-old daughters in for evaluation of rash. The 7-year-old has had recurrent itching of the scalp and physical examination findings shown in Item Q155. The 7-year-old has been previously treated for this finding with permethrin.
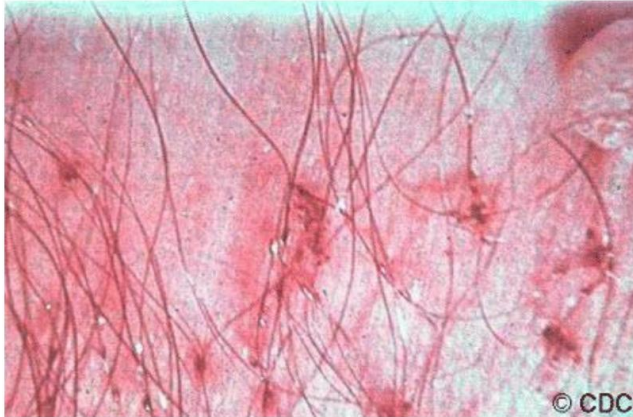
Item Q155. Scalp findings for the 7-year-old girl described in the vignette. Courtesy of Centers for Disease Control and Prevention.

Of the following, the BEST treatment for the 4-month-old girl is

A.   benzyl alcohol lotion

B.   ivermectin lotion

C.   lindane shampoo

D.   malathion lotion

E.   permethrin lotion

Question 155   **Preferred Response: E**

The children described in the vignette have head lice (pediculosis capitis) with nits and scalp excoriations visible on physical examination of the 7-year-old girl. Permethrin, a topical insecticide, is the treatment of choice for the 4-month old infant in the vignette. Permethrin 1% lotion is available without a prescription; it is applied to the scalp and hair for 10 minutes, and then washed out. A repeat application is recommended in 9 to 10 days to kill newly hatched lice, because the medication does not affect unhatched eggs. Permethrin is not recommended for children younger than 2 months.

The female louse lives approximately 1 month and lays up to 10 eggs (nits) each day at the base of a host's hair shaft. After approximately 8 days, the egg capsules hatch nymphs that mature over the next 8 days into adult lice. The adults have mouth parts used to suck blood and grasp host hairs. They move about by crawling and are transmitted by close person-to-person contact. Pubic lice usually are spread through sexual contact.

Clinical manifestations of head, body (pediculosis corporis), and pubic (pediculosis pubis) lice include intense itching and small, erythematous maculopapular lesions with excoriations at the site of bites. Body lice can survive away from a blood source for 5 to 7 days. Pubic lice typically survive for up to 36 hours away from a host, but may live for 10 days under ideal conditions. For the 4-month-old infant in the vignette, the best option for treating head lice is over-the-counter permethrin because none of the other topical agents are recommended for young infants (Item C155). Lindane shampoo no longer is recommended for treating children because of neurologic adverse effects and widespread resistance. Pediculicides used to treat pediculosis capitis and corporis can also be used to treat pediculosis pubis. After each treatment, the hair and body should be checked for nits and lice with a nit comb. Bedding and clothing should be washed in hot water, and close contacts should be monitored for lice and treated if infested.

Item C155. Topical Agents for the Treatment of Pediculosis.

Drug	Target	Reapplication	Adverse Effects	Age Limits
Over-the-counter				
Pyrethrins with piperonyl butoxide	Live lice	9 – 10 d	Patients sensitive to ragweed or chrysanthemum can have allergic reactions	≥2 y
Permethrin lotion, 1%	Live lice	9 d	Skin irritation	≥2 mo
Prescription				
Benzyl alcohol lotion, 5%	Live lice	7 d	Skin irritation	6 mo – 60 y
Ivermectin lotion, 0.5%	Live lice	Not without physician advice	Skin irritation	≥6 mo
Malathion lotion, 0.5%	Live lice, some eggs	7 – 9 days if live lice are visible	Skin irritation, flammable	≥6 y
Spinosad, 0.9% topical suspension	Live lice and eggs	7 days if live lice are visible	Skin irritation	≥4 y

Courtesy of D. Palazzi

PREP Pearls
- Topical pediculicides are the treatment of choice for pediculosis.
- Most topical pediculicides kill only live lice, so reapplication in 7 to 10 days is often necessary to treat infestation.
- The only pediculicide approved for use in children younger than 6 months is 1% permethrin lotion.
- The most common adverse effect of topical pediculicides is skin irritation.

ABP Content Specifications(s)
•   Understand the life cycle of human lice
•   Plan the appropriate management for a patient with pediculosis capitis (head lice)
•   Plan the appropriate management for a patient with pediculosis pubis
•   Recognize the clinical findings associated with pediculosis capitis or pediculosis pubis

Suggested Readings
•   American Academy of Pediatrics. Pediculosis capitis. In: Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. Red Book:2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:597-601.

Question 156

A tearful mother brings her 2-year-old son to your office 30 minutes after he pulled on the handle of a mug full of hot coffee left on the edge of the kitchen counter. The coffee spilled onto the boy and burned his right upper arm and chest. He is crying loudly, without stridor or respiratory difficulty. Physical examination reveals a 3 × 3–cm "ruptured" blister on the child's upper chest with an erythematous, tender base. There is also a 3 × 4–cm area of superficial erythema without blister formation on the child's right upper arm. The child's mother states that he has been very healthy and takes no medications. His immunizations are up to date. After administration of an oral analgesic, you clean the boy's burns with soap and water.

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

A.   application of topical lidocaine gel to the burned areas

B.   debridement of the ruptured blister on the upper chest

C.   prescription of a course of oral cephalexin

D.   referral to a burn center due to the upper chest burn

E.   reporting of the injury to child protective services

Question 156   **Preferred Response: B**

The boy in the vignette presents for care of a superficial burn to his right upper arm and a small partial thickness burn wound with an associated ruptured blister on his chest. After administration of an analgesic and cleansing of these burns, debridement of the ruptured blister on his upper chest is the next best step in management.

Burns are a relatively common type of injury in children and adolescents. Pediatric providers should be familiar with the initial assessment and management of burns. Burns may arise from contact with hot objects or liquids, sun exposure, radioactivity, electricity, chemical exposure, or friction. Thermal injuries that occur commonly in children include scald and contact burns sustained from cooking or spilling hot foods or liquids. Scald burns related to bathing are a particular risk in infancy and the toddler years. Contact burns from hot objects such as space heaters, grills, stoves, ovens, irons, hair appliances, campfires, and fireworks are also seen fairly frequently. Fortunately, only a few burns in children require hospitalization; most can be managed on an outpatient basis.

Burn injuries are classified into 3 main types, based on the depth of tissue injury:
- First-degree (ie, superficial) burns: These burns involve injury to the epidermis only, without involvement of the dermis. Clinical features include erythema and mild inflammation of the epidermis, without blister formation. Uncomplicated sunburn is a commonly occurring example. These burns may be painful, but generally resolve within a few days without scarring. Although the injured epidermis often peels within a few days of a superficial burn injury, new epidermal cells will be regenerated. Superficial burn injuries are not generally included in calculations of total body surface area (TBSA) involvement in patients presenting with burns.
- Second-degree (ie, partial thickness) burns: These burns involve injury to both the epidermal and dermal layers of skin. Blister formation typically occurs, as well as tissue edema arising from increased capillary permeability. These burns are generally painful because of exposure of intact sensory nerve receptors in the injured dermis. Superficial partial thickness burns typically heal within 2 weeks without scarring. Deeper partial thickness burns may involve damage to most of the dermis and may have a paler, drier appearance than more superficial partial thickness burns. Healing may take several weeks, and scarring may occur. Deeper partial thickness burns may be difficult to differentiate from full thickness burns.
- Third-degree (ie, full thickness) burns: These burns involve destruction of the epidermis and entire dermal layer of skin. Full thickness burns may appear pale and "waxy" or charred, and often have a leathery appearance. Skin affected by full thickness burns is nontender because of destruction of the cutaneous nerves, though surrounding partial thickness burns may be very painful. Full thickness burns cannot re-epithelialize because of destruction of the entire dermal layer, and skin grafting is often required.

Children presenting for care of superficial burns, or superficial partial thickness burns involving a small percentage (<5%) of TBSA, can generally be treated on an outpatient basis. First-degree

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(superficial) burns generally require only supportive therapy with emollients and oral analgesics such as acetaminophen or ibuprofen for discomfort.

Minor partial thickness burns can be cleansed with mild soap and water, diluted povidone-iodine solution, chlorhexidine gluconate, or saline. Devitalized tissue should be debrided, often by wiping gently with moist gauze. The exposed underlying skin will generally be erythematous, moist, and painful. The burn wound should be dressed promptly to help reduce pain associated with convection of air across the wound. Minor partial thickness burns should be dressed with a topical antimicrobial agent such as bacitracin ointment or silver sulfadiazine and covered with a sterile gauze dressing. Caregivers should be advised to gently cleanse minor burn wounds daily with a clean cloth or gauze in the shower or bathtub, and then to redress the wounds. Minor partial thickness burn injuries generally heal within 1 to 2 weeks. Providing adequate analgesia is an essential component in the outpatient care of minor burns. Pediatricians should administer appropriate analgesia before performing the initial burn wound assessment and care, and anxiolytic agents may even be required in some children.

Regarding the management of blisters associated with minor partial thickness burns, those that are intact provide protection for the underlying tissue and should be left intact, as long as they do not cross joints or otherwise limit activity. Topical antimicrobial ointment does not need to be applied to intact blisters. Once blisters rupture, they should be debrided, because devitalized tissue could serve as a nidus for infection.

Although providing adequate analgesia is an essential component in caring for children presenting with burns, application of a topical lidocaine gel to burned areas is not recommended for children with burns of any degree. There is a scarcity of evidence supporting the effectiveness and safety of this practice. In addition, the use of topical lidocaine preparations on burned skin places children at risk for systemic lidocaine toxicity, which could result in serious complications, including methemoglobinemia, central nervous system toxicity, and cardiotoxicity.

A course of oral cephalexin would not be the best next step in management for the boy in the vignette, who displays no signs of systemic infection. There is no role for the empiric administration of systemic antibiotics after burn injuries. Systemic antibiotics should only be administered to children with clear evidence of infection on physical examination or on culture of the burn wound.

Referral to a burn center is not indicated for the boy in the vignette. The burned area on his right arm is a superficial burn that requires supportive care only, and the burn on his upper chest is a superficial partial thickness burn that is small in size (<5% TBSA). His burns can certainly be managed on an outpatient basis. Children with partial thickness burns involving more than 5% to 10% TBSA should be considered for admission to a hospital. Indications for referral of children presenting with burn injuries to a regional burn center include full thickness burns; partial thickness burns exceeding 10% TBSA; electrical or chemical burns; burns associated with inhalational injury; associated traumatic injuries or comorbid conditions; burns involving the

face, hands, feet, genitalia, or joints; burns suspected to arise from intentional injury; and burns exceeding local specialist or institutional capacity.

Reporting of the injuries sustained by the boy in the vignette to child protective services is not warranted, given that the reported mechanism of injury is consistent with his developmental stage, and that his pattern of burns is not suspicious for inflicted injury. Pediatric burn injuries associated with a delay in seeking care or isolated scald or contact burns to the hands, feet, genitalia, or buttocks without a plausible mechanism should raise suspicion for inflicted injury. Burns to the hands and feet with a "stocking and glove" pattern (clearly demarcated borders without surrounding splash burns) can arise from intentional immersion of the hands or feet in scalding liquid, and should raise suspicion for child abuse. Scald burns to the buttocks and thighs in toddlers can result from forced submersion in a tub of hot water, often following a toilet-training mishap. Suspicious patterns of contact burns with hot objects (such as burning cigarettes, hot irons, cooking pans, hair appliances, or heaters) that do not seem to correlate with the reported history should prompt a thorough evaluation for abuse. If concern for an inflicted burn injury exists, reporting to child protective services and referral of the injured child to a pediatric burn center are indicated.

PREP Pearls
- Children presenting for care of superficial burns or superficial partial thickness burns involving a small percentage (<5%) of total body surface area can generally be managed on an outpatient basis.
- First-degree (superficial) burns generally require only supportive therapy with emollients and oral analgesics, such as acetaminophen or ibuprofen, for discomfort.
- Minor partial thickness burns should be cleansed and devitalized tissue should be debrided. A topical antimicrobial agent such as bacitracin ointment should be applied, and the wound should be covered with a gauze dressing. Intact blisters should be left intact.
- Pediatricians should administer appropriate analgesia before performing the initial burn wound assessment and care, and anxiolytic agents may be required in some children.

ABP Content Specifications(s)
- Plan the appropriate outpatient management of minor burns

Suggested Readings
- Jamshidi R, Sato TT. Initial assessment and management of thermal burn injuries in children. Pediatr Rev. 2013;34:395. DOI: http://dx.doi.org/10.1542/pir.34-9-395.

Question 157

During rounds in the newborn nursery, a mother tells you she is worried because her daughter has had no wet diapers since birth, 18 hours ago. The neonate was born at 41 weeks' gestation by normal vaginal delivery. The pregnancy was unremarkable, including a normal anatomy scan at 18 weeks of gestation. The mother presented in labor, with artificial rupture of the membranes 3 hours before delivery, revealing clear amniotic fluid. The newborn emerged vigorous and has been exclusively breastfeeding since delivery. Physical examination reveals a pink, well-perfused newborn with a normal cardiac and pulmonary examination, an intact spine, and a soft, nondistended abdomen without palpable masses.

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

A.   continue to monitor for urine output

B.   order renal ultrasonography

C.   order serum electrolyte levels

D.   order a voiding cystourethrogram

E.   perform a bladder catheterization

Question 157   **Preferred Response: A**

The newborn in the vignette, with no urine output since delivery 18 hours earlier, should continue to be monitored. Ninety-two percent to 97% of newborns will urinate within 24 hours of birth and nearly all within 48 hours. Urine output can be difficult to appreciate during this period because of both the use of superabsorbent diapers and frequent meconium stooling. A urine bag or cotton ball may be used to document urine output if concerns arise.

A well-appearing newborn may be monitored up to 24 hours after birth for urine output. If anuria persists beyond that point, the history and physical examination should be carefully reviewed for evidence of a pathologic cause (Item C157). Urinary tract abnormalities often are detectable on prenatal ultrasonography. Because amniotic fluid reflects urine production in the late second and third trimester, oligohydramnios is an additional clue to a urinary tract abnormality. On physical examination, a distended bladder should prompt concerns of bladder outflow obstruction whereas a lower back abnormality such as a dimple or hair tuft may suggest an underlying spinal anomaly affecting micturition.

Item 157. Pathologic Causes of Anuria in an Otherwise Well Newborn

Renal	Obstructive	Functional
Autosomal recessive polycystic kidney disease	Anterior and posterior urethral valves	Detrusor-sphincter dysfunction
Dysplastic kidney(s)	Hydrometrocolpos	
Hypoplastic kidney(s)	Ureteropelvic junction obstruction	
	Ureterovesical junction obstruction (ureteroceles)	

Bladder catheterization and bladder/renal ultrasonography should be considered if anuria persists beyond 24 hours after birth in spite of adequate hydration. Renal function should be evaluated with serum electrolytes, blood urea nitrogen, and creatinine levels. Urologic consultation is recommended if there is difficulty performing the bladder catheterization, for issues with urination that persist after catheterization, or for abnormal physical examination findings. Further studies may be indicated, including voiding cystourethrography (VCUG). At this point in time, the newborn in the vignette should continue to be monitored for urine output. Her normal prenatal ultrasound, amniotic fluid volume, and physical examination findings are reassuring. Bladder catheterization, renal ultrasonography, and serum electrolyte levels may be required if she continues to have no urine output. VCUG is not recommended before a review of the initial evaluation results with a urologic consultant.

PREP Pearls
- A well-appearing newborn may be monitored for urine output up to 24 hours after birth. Ninety-two percent to 97% of newborns will urinate within 24 hours of birth and nearly all within 48 hours.
- Prenatal ultrasound screening, amniotic fluid volume, and the physical examination findings should be reviewed when evaluating an anuric newborn.
- The initial evaluation of a newborn with no urine output in the first 24 hours after birth should include bladder catheterization, bladder/renal ultrasonography, and serum electrolyte, blood urea nitrogen, and creatinine levels.

ABP Content Specifications(s)
- Plan the appropriate evaluation of delayed urination in a newborn infant

Suggested Readings
- Abdelhamid A. Index of suspicion in the nursery: unusual cause of feeding intolerance in a term infant. NeoReviews. 2012;13:e732-e735. DOI: http://dx.doi.org/10.1542/neo.13-12-e732.

Question 158

A 6-month-old infant is brought to your office by her parents with the complaint of decreased use of her left arm. She was born at full term to a 38-year-old woman who took fluoxetine during pregnancy and did not take prenatal vitamins. There were no complications during the pregnancy, labor, or delivery. The infant had torticollis noted at 2 weeks of age that resolved with physical therapy. Her parents report that over the past month, the infant's left arm seems to hang by her side, and she is holding her left hand in a fist with increasing frequency. Her physical examination demonstrates a head circumference of 42 cm (10th percentile), length of 67 cm (50th percentile), weight of 8.2 kg (60th percentile), blood pressure of 82/68 mm Hg, and a heart rate of 102 beats/minute. Her anterior fontanel is open and flat. She has no dysmorphic features. There is no torticollis. Her neurological examination demonstrates symmetric facial movements, increased tone in the left upper and lower extremities, and decreased spontaneous movement of the left arm. Deep tendon reflexes are brisker on the left side compared to the right. You order magnetic resonance imaging of the brain (Item Q158).

Of the following, the risk factor MOST associated with this infant's presentation is

A.   advanced maternal age

B.   female sex

C.   fetal selective serotonin reuptake inhibitor exposure

D.   low maternal folate level

E.   neonatal torticollis

Courtesy of D. Morita

ITEM Q158: Magnetic resonance imaging results from the infant described in the vignette.
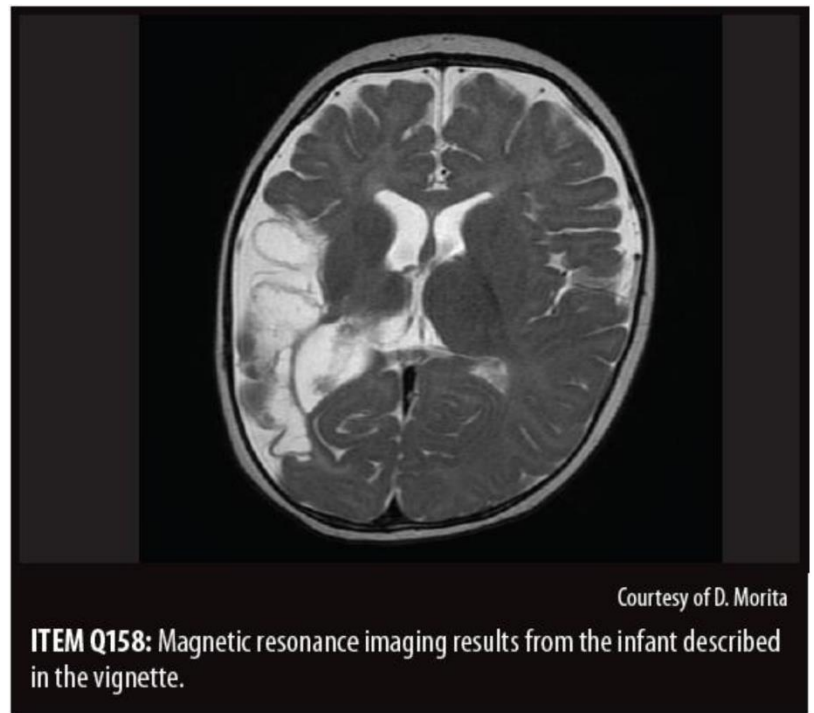
Question 158   **Preferred Response: A**

The girl in the vignette has spastic, hemiparetic cerebral palsy (CP). Of the response choices listed, only advanced maternal age is a risk factor for CP in a full-term infant. Magnetic resonance imaging shows cystic encephalomalacia in the right side of the brain, in a distribution consistent with a focal arterial infarction. This is the cause of her left hemiparesis.

Cerebral palsy is defined as motor impairment caused by a nonprogressive brain injury that occurred during fetal or infantile brain development and the associated disability and impairments that may develop over time. Even though the injury may have occurred prenatally or in the perinatal period, the first symptoms may not appear until 4 to 6 months of age, as described in the girl in the vignette. CP is subdivided into categories based on the limbs that are involved: spastic, hemiparetic CP affects either the right or left side of the body, spastic diplegic or diparetic CP affects both legs, and spastic quadriplegic or quadriparetic CP affects all four limbs. Less commonly seen CP subtypes are athetotic and dystonic. These subtypes are now thought to have different underlying causes along with a different natural history and prognosis. Traditionally, the formal diagnosis of CP has been delayed until age 3 or 4 years, when metabolic or genetic causes of motor disturbance can be clinically ruled out. However, as seen in the girl in the vignette, when a structural abnormality is seen on brain imaging, the diagnosis of CP can be made earlier.

On neuroimaging, focal arterial infarction and brain malformation are the 2 most commonly identified causes of CP in full-term infants. Other causes include periventricular leukomalacia and generalized atrophy. In up to one third of cases, neuroimaging will not show a cause. Risk factors for CP in full-term infants include advanced maternal age, black race, and intrauterine growth retardation. Infant sex and multiple gestation are not risk factors for CP in full-term infants. Fetal selective serotonin reuptake inhibitor exposure can cause excessive jitteriness in the perinatal period, but not CP. Prenatal folate supplementation can prevent neural tube defects such as spina bifida, but does not prevent CP. Neither neonatal torticollis nor its treatment causes CP.

PREP Pearls
- Risk factors for cerebral palsy in full-term infants include advanced maternal age, black race, and intrauterine growth retardation.
- Symptoms of spastic cerebral palsy may not appear until 4 to 6 months of age.
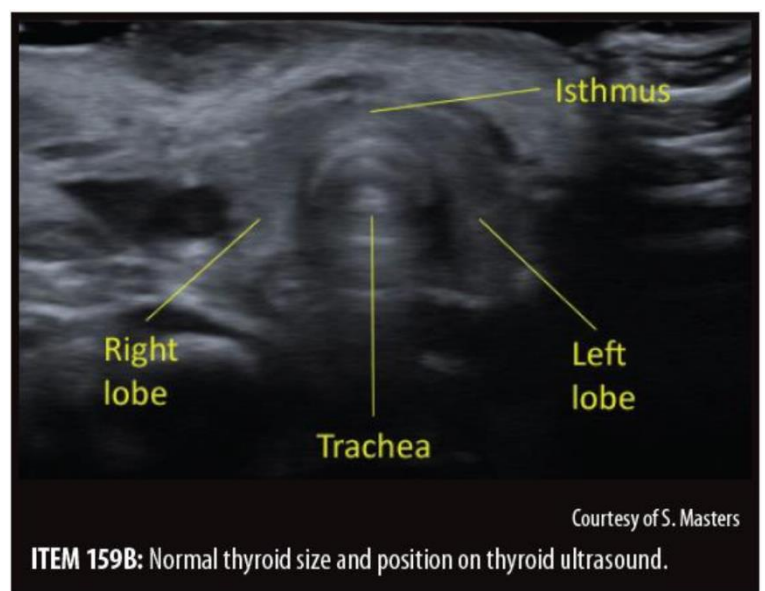
ABP Content Specifications(s)
- Understand the prenatal risk factors associated with cerebral palsy
- Recognize the clinical features associated with cerebral palsy

Suggested Readings
- Novak I, Hines M, Goldsmith S, Barclay R. Clinical prognostic messages from a systematic review on cerebral palsy.Pediatrics. 2012;130:e1285. DOI: http://dx.doi.org/10.1542/peds.2012-0924.

Question 159

You receive a call from your state newborn screening laboratory regarding an abnormal thyroid screening test on a now 6-day-old newborn. In your office, the newborn is asymptomatic and has an unremarkable physical examination. Maternal history is significant for long-term therapy with levothyroxine and previous thyroid surgery. Review of the newborn screening test results show a low thyroxine level and a thyroid stimulating hormone over 100 mIU/L. Repeat laboratory testing confirms these findings. You order imaging studies to further evaluate the newborn's thyroid. Technetium 99m pertechnetate uptake scan shows absence of any uptake over the thyroid, suggesting athyrosis (Item Q159A). Thyroid ultrasonography reveals a normally sized and positioned thyroid gland (Item Q159B).

Courtesy of S. Masters

ITEM Q159A: Absence of uptake over the thyroid gland (red circle) on technetium 99m pertechnetate uptake scan.

Courtesy of S. Masters

ITEM 159B: Normal thyroid size and position on thyroid ultrasound.

Of the following, the MOST likely cause of these findings is

A.   DiGeorge syndrome
B.   levothyroxine use during pregnancy
C.   maternal history of Graves disease
D.   maternal history of Hashimoto disease
E.   neonatal lupus with antiphospholipid antibodies

Question 159   **Preferred Response: C**

The infant in the vignette has congenital hypothyroidism (CH), with a thyroid-stimulating hormone (TSH) level higher than 100 mIU/L. Given this finding, treatment with levothyroxine should be initiated immediately. Optional diagnostic studies that are often performed include thyroid ultrasonography and/or a thyroid uptake scan. A thyroid uptake scan uses either iodine 123 or sodium technetium 99m pertechnetate (99mTc) to identify functional thyroid tissue.

Imaging studies used in the evaluation of congenital hypothyroidism can aid in identifying a cause, which can have important prognostic implications for the child. Imaging study results may include:
*   An ectopic thyroid gland (the most common anatomic cause of hypothyroidism) which establishes a permanent form of thyroid disease.
*   The absence of thyroid gland uptake, which is most often associated with thyroid aplasia or hypoplasia, another permanent cause. However, when radioiodine uptake is absent but ultrasonographic examination reveals a normal gland, as shown in the infant in this vignette, a TSH-receptor defect, iodine-transport defect, or maternal transfer of thyroid antibodies may be present.
*   A normal scan may indicate a transient form of hypothyroidism. These infants should undergo a careful follow-up evaluation after 3 years of age, when it is safe to discontinue levothyroxine treatment with certain precautions.

The infant in this vignette has a normal thyroid ultrasound but negative uptake on 99mTc uptake imaging. A common reason for this discrepancy in imaging findings is maternal transfer of thyroid-blocking antibodies, which block the TSH receptor. Mothers with Graves' disease (either active disease or treated with surgery or radioactive iodine) can have thyroid antibodies present. Antibodies to the TSH receptor (stimulating or blocking) freely cross the placenta and can act in the fetal thyroid gland during the second half of pregnancy and after delivery, causing either neonatal Graves' disease or congenital hypothyroidism. This child had very high thyroid-binding inhibitory immunoglobulins that blocked the functionality of the TSH receptor, resulting in CH. Thus, even though the gland is present, it is functionally turned off while the blocking antibodies are present. This situation can go on for months, and treatment with levothyroxine is required, as in the infant in this vignette, until the antibodies have disappeared.

Ascertaining the cause of a mother's hypothyroidism is important. Mothers who have a history of autoimmune hypothyroidism (Hashimoto thyroiditis), rarely if ever, have antibodies that cross the placenta. Therefore, autoimmune hypothyroidism is unlikely to cause an issue with the infant as long as the mother is taking her thyroid medication and is euthyroid. On the other hand, as mentioned before, a mother with active or a history of Graves' disease may pass antibodies through the placenta to the infant (stimulating or blocking), which could cause either neonatal Graves' disease or CH. Thus infants of mothers with Graves' disease, especially those who have high antibody titers in the third trimester, deserve closer follow-up. This can include assessment of the infant's thyroid function and antibody titers.

Levothyroxine use during pregnancy is safe and will not affect the infant. In fact, maternal thyroxine (or levothyroxine medication) crosses the placenta in small amounts during the first half of pregnancy, when fetal thyroid hormone concentrations are low. This is necessary to keep the fetus euthyroid.

Of the other response choices listed, antiphospholipid antibodies associated with lupus can cause neonatal heart block, but would not affect thyroid function. DiGeorge syndrome, commonly associated with hypoparathyroidism, can also be associated with hypothyroidism. However DiGeorge syndrome is not associated with maternal transfer of thyroid antibodies.

Other possible reasons for hypothyroidism include transient causes, such as iodine overload or maternal anti-thyroid medications.

It is very important for the general pediatrician to recognize that if there is any suspicion of CH, treatment should be initiated with levothyroxine as early as possible. Treatment can be weaned later if a transient cause of hypothyroidism is suspected. For infants with CH , 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 who were treated early with levothyroxine compared with control groups of classmates and siblings. However, even with early treatment visuospatial processing, selective memory, and sensorimotor deficits can occur. In contrast, the prognosis for normal mental and neurologic performance is less certain for infants with CH who are not treated as early as possible, ideally by age 2 weeks. 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.

PREP Pearls
- Imaging studies performed during the evaluation of congenital hypothyroidism can help to determine whether there is a permanent versus transient cause of disease.
- A mother who is hypothyroid after surgery or radioactive iodine ablation for Graves' disease and now taking levothyroxine, or who has active Graves' disease during pregnancy, may pass antibodies to the infant (stimulating or blocking) that could cause either neonatal Graves' disease or congenital hypothyroidism.
- Treatment for congenital hypothyroidism should never be delayed to obtain additional imaging studies or further diagnostic workup. Treatment with levothyroxine should always begin immediately to afford the best developmental outcome.

ABP Content Specifications(s)
- Understand the prognosis for a patient with congenital or acquired hypothyroidism, including neonates whose hypothyroidism is not treated
- Identify the causes of congenital and acquired hypothyroidism

Suggested Readings

- 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: http://dx.doi.org/10.1542/peds.2006-0915.

Question 160

A 14-year-old adolescent is brought to the emergency department via emergency medical services with complaints of shortness of breath, and moderately severe sharp chest and shoulder pain after he was hit by a teammate during football practice.

The patient's medical history is significant for mild persistent asthma and seasonal allergic rhinitis, which are well controlled on leukotriene receptor antagonist monotherapy. The patient did have symptoms of an upper respiratory infection and bronchitis during the preceding 3 days and was completing treatment with azithromycin. There is no antecedent history of vomiting or respiratory distress.

On physical examination, you find a well-developed, well-nourished adolescent in mild respiratory distress. His respiratory effort appears splinted because of chest pain and he is anxious in appearance. His respiratory rate is 30 breaths/min. Oxygen saturation is 94% in room air. Cardiac examination is remarkable for mild tachycardia, but no murmur, rub, or gallop. Auscultation of the lungs reveals moderate aeration. The lungs are clear to auscultation without wheezing, crackles, or asymmetry. Subcutaneous crepitus is demonstrated at the shoulders, neck, and jaw line. The abdomen is soft, nontender, and nondistended. His extremities are well-perfused with symmetric pulses.

A chest radiograph is obtained (Item Q160).

Courtesy of E. Anthony

ITEM Q160: Chest radiograph for the adolescent described in the vignette.
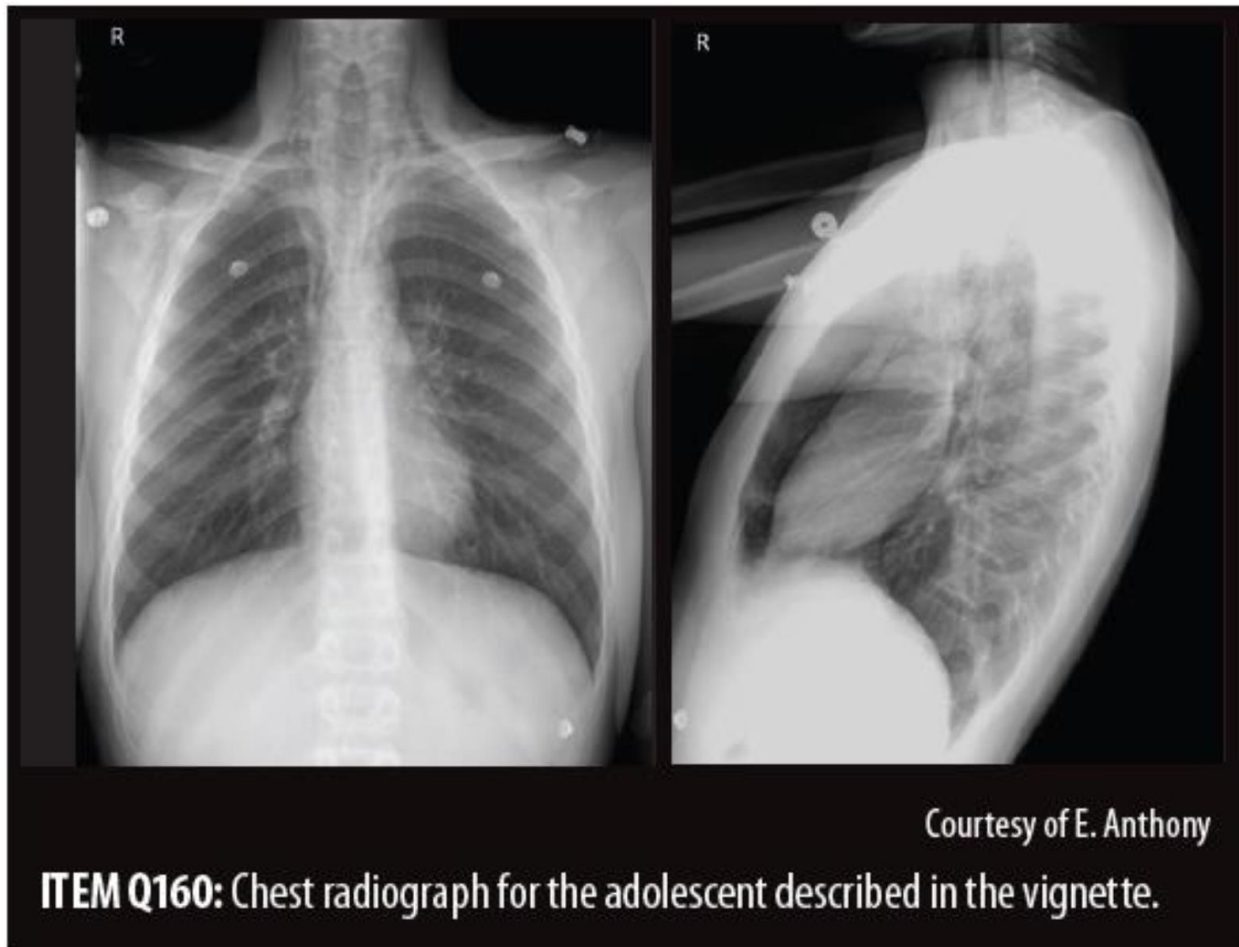
Of the following, the MOST likely explanation for this patient's clinical presentation is

A.   commotio cordis
B.   pneumomediastinum
C.   pneumothorax
D.   splenic rupture
E.   status asthmaticus

Question 160   **Preferred Response: B**

The patient in the vignette has symptoms and signs of a pneumomediastinum. The mechanism of pneumomediastinum involves the tracking of free air from ruptured alveoli along peribronchial vascular sheaths toward the hilum and mediastinum. The most commonly encountered predisposing conditions include asthma exacerbation and infection. Pneumomediastinum has also been described after episodes of choking, with vigorous physical exertion and with physical impact or trauma, with or without associated Valsalva maneuvering. It has also been described with forceful vomiting (including in association with bulimia and Boerhaave syndrome [esophageal rupture from forceful vomiting]), with foreign body inhalation, and in inhalational or intravenous drug abuse. Rarely, pneumomediastinum has been found as a presenting symptom in diabetic ketoacidosis.

Signs and symptoms of pneumomediastinum include chest and neck pain, dyspnea, and sore throat. Dysphonia may be seen. Subcutaneous emphysema and tactile subcutaneous crepitus is frequently encountered. Subcutaneous air collections are often felt at the neck, but may also involve the face and scalp. Hamman crunch describes precordial or substernal crepitance that is typically synchronous with the patient's heart beat. Hamman crunch is nearly pathognomonic for subcutaneous emphysema.

Spontaneous pneumomediastinum demonstrates an incidence that is bimodal; children younger than 7 years of age and adolescents 13 to 17 years of age are disproportionately affected. Pneumomediastinum is usually benign and generally resolves in 1 to 2 weeks with supportive care. Rest, prevention of forced exhalation maneuvers, analgesia, and treatment of comorbid conditions are warranted. Complications are rarely seen with pneumomediastinum, but may include pneumopericardium, pneumothorax, or tension pneumomediastinum. In tension pneumomediastinum, a large collection of mediastinal air may result in decreased cardiac output through direct compression or via decreases in venous return.

Commotio cordis is a rare, but frequently fatal arrhythmia that occurs after the precordial chest is struck or traumatized. This may occur in a motor vehicle crash or in a sport such as baseball or football. Similar blunt trauma may be implicated in patients with pneumomediastinum without commotio cordis.

While a pneumothorax is expected to present similarly with acute onset of chest pain and dyspnea, symmetric lung aeration would not be expected nor is pneumothorax associated with subcutaneous crepitus.

Spontaneous pneumothorax is more common in thin, tall male adolescents. A history of recurrent pneumothorax warrants an evaluation for predisposing factors such as collagen vascular disease. There is no cough, wheezing, prolongation of the expiratory phase of respiration, or silent chest to suggest status asthmaticus in this patient. However, this patient's mild asthma and mild associated air trapping may contribute to the pathogenesis of pneumomediastinum.

Finally, splenic rupture may be the result of traumatic impact and may be associated with sharp shoulder pain (Kehr sign). Other associated symptoms, however, include pain in the abdomen, epigastrium, or left flank. In severe injuries to the spleen, the patient may present in hypovolemic shock caused by massive blood loss.

PREP Pearls
- Subcutaneous emphysema in pneumomediastinum represents the tracking of free air from ruptured alveoli along perivascular sheaths into the hilum and mediastinum.
- The most common causes of pneumomediastinum are infection and asthma.

ABP Content Specifications(s)
- Recognize complications associated with pneumothorax/pneumomediastinum

Suggested Readings
- Bullaro F, Bartoletti S. Spontaneous pneumomediastinum in children: a literature review. Pediatr Emerg Care. 2007;23(1):28-30 .

Question 161

A 6-month-old infant has a large ventricular septal defect complicated by congestive heart failure. His corrective surgery has been delayed because of 2 hospitalizations for bronchiolitis during which he lost weight. He is currently feeding 24 kcal/oz formula, but has not shown any weight gain, and his weight is now below the third percentile for his age. The baby has a good suck, but he takes no more than 60 to 75 mL every 4 hours.

Of the following, the BEST next step to increase this baby's energy intake is to

A.   add microlipid to the current formula

B.   add protein powder to the current formula

C.   change to an amino acid-based formula

D.   increase the caloric density of the current formula to 35 kcal/oz

E.   start total parenteral nutrition

Question 161   **Preferred Response: A**

Like many children with chronic medical conditions, the infant in the vignette has developed growth failure due at least in part to an inability to take in adequate volume and therefore calories to support growth. Multiple other factors contribute to inadequate energy intake in children with hemodynamically significant heart disease, including being in a hypermetabolic state, swallowing dysfunction, gastrointestinal dysfunction, particularly protein-losing enteropathy, and the presence of other genetic anomalies. Children with cardiac conditions that cause hypoxemia, congestive heart failure, or pulmonary hypertension are at particular risk for growth failure, and these children usually require at least 140 kcal/kg per day to meet their energy requirements. Standard infant formula can be concentrated up to 30 calories/oz to increase calories, but an excessive concentration produces a high osmolar load that can impair gastric emptying and increase renal solute load above the kidneys' capacity to handle. Therefore, the primary milk source (human milk or formula) often requires supplementation with glucose polymers or fats such as microlipid emulsion to provide sufficient calories despite relatively small volume intake. Protein powder would not be advisable because it too would increase the osmolar load too much. Even with supplementation, some children will not be able to consume adequate volume and therefore calories; these children may require 24-hour continuous enteral feeds to meet their daily energy requirements. Enteral nutrition would be the preferred route for feeding compared to parental nutrition.

Standard infant formulas provide adequate nutrients to support growth of healthy term infants. However, infants and children with complex medical needs may not receive appropriate nutrition using standard formulas and may require feedings that differ by protein source, carbohydrate, fat ratios, caloric density, and mineral and micronutrient content. For example, standard soy formula does not provide enough calcium and phosphorus to prevent osteopenia in preterm infants, and the increased aluminum content in soy formula may exacerbate this problem. Hydrolyzed and amino acid-based formulas have been developed for children with cow's milk allergy or at high risk for atopic disease, and extensively hydrolyzed formulas have been used for short gut syndrome, hepatobiliary and pancreatic disease, and autoimmune and immunodeficiency diseases. Neither of these formula types would be required for the infant in the vignette whose issues relate to inadequate intake rather than inadequate absorption or immunologic conditions. The primary problems with these formulas include high cost and poor acceptance by infants. Infant formulas have also been modified to manage gastroesophageal reflux by thickening, but data to support this approach are limited.Children beyond infancy may require formula either as the sole source of nutrition or as supplementary oral intake. Standard pediatric formulas provide 30 kcal/oz (1 kcal/mL) and provide sufficient vitamin and mineral content to prevent vitamin D, calcium, phosphorus, and iron deficiency. Adult formulas have a higher calorie-to-nutrient ratio than pediatric formulas. Adult formulas should not be used for low energy children (eg, nonambulatory) because they may gain excessive weight despite being nutrient deficient.

PREP Pearls

•   Children with chronic medical conditions such as congenital heart disease often require increased caloric intake in order to meet energy requirements for normal growth and development.

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- Standard formulas can be concentrated to increase caloric intake, but excessive concentration produces a high osmolar load that can impair gastric emptying and increase renal solute load above the kidneys' capacity to handle.
- Glucose polymers or fats (eg, microlipid) increase the caloric density of formula without increasing the osmolar load.
- Specialized formulas are available for special medical conditions such as amino acid formulas for infants with allergic conditions or extensively hydrolyzed formulas for children with impaired gastrointestinal absorption.
- Soy formula is not appropriate for preterm infants because it provides inadequate calcium and phosphorus and excessive aluminum, which may lead to metabolic bone disease.

ABP Content Specifications(s)
- Know the content of various infant formulas and milk sources, the indications for their use, and possible side effects
- Understand the differences among categories of formula used for special nutritional support and the indications for their use

Suggested Readings
- Kleinman RE, Greer FR. Cardiac disease. Pediatric Nutrition. 7th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2014:1077-1094.

Question 162

A 16-year-old adolescent presents to your office because she is concerned that, unlike her friends, she hasn't started menstruating. Her mother reports that she and the patient's sister had menarche around 13 years of age. The patient's past medical history is remarkable for surgery for strabismus. She denies sexual activity. On physical examination, her height is less than the fifth percentile. Her weight is at the 80th percentile. Her blood pressure and heart rate are normal. She has a soft systolic murmur. The rest of her examination is unremarkable. She has a sexual maturity rating of 2 for pubic hair and breast. Her urine pregnancy test is negative.
Of the following, the MOST appropriate next step in her evaluation is

A.   careful assessment for psychological stressors

B.   a karyotype analysis

C.   to order a prolactin level

D.   to provide reassurance

E.   to screen for nonclassical adrenal hyperplasia

Question 162   **Preferred Response: B**

Primary amenorrhea is defined as the absence of menses by 15 years of age or within 5 years of breast development if that occurs before 10 years of age. Primary amenorrhea is most often caused by an anatomic or genetic abnormality. Gonadal dysgenesis resulting from chromosomal abnormalities accounts for approximately 50% of cases of primary amenorrhea. Other common causes include hypothalamic hypogonadism and congenital anatomical abnormalities of the uterus, cervix, or vagina. Careful history and physical examination are important in the evaluation of the etiology for primary amenorrhea. Pubertal assessment, evaluation of reproductive anatomy, and the presence of dysmorphic features can guide the workup of primary amenorrhea.

Turner syndrome (45 X gonadal dysgenesis) often presents during adolescence as primary amenorrhea. Other features of Turner syndrome include short stature, aortic coarctation, widely spaced nipples, webbed neck, cubitus valgus, strabismus, and congenital lymphedema. The Turner Syndrome Consensus Study Group has published guidelines for the management of girls and women with Turner syndrome. Management strategies address growth-promoting therapies and the induction of puberty. The physical findings in the adolescent in this vignette suggest a diagnosis of Turner syndrome and a karyotype analysis is the most appropriate next step in her evaluation.

While psychological stress can be a reason for amenorrhea, in this scenario, there are other findings that should alert the clinician to the possibility of other reasons for the patient's amenorrhea. Prolactinomas in young female adolescents can present as delayed menarche. However, the other findings in this scenario make a prolactinoma less likely as the etiology of this patient's amenorrhea. Similarly, nonclassical adrenal hyperplasia (NCAH) can present with menstrual irregularities like amenorrhea, but individuals with NCAH usually also have evidence of hyperandrogenism with hirsutism and acne, as well as premature pubarche and tall stature during adolescence.

PREP Pearls
- Primary amenorrhea is defined as the absence of menses by 15 years of age or within 5 years of breast development if that occurs before 10 years of age.
- Primary amenorrhea is most often caused by an anatomic or genetic abnormality.
- Gonadal dysgenesis resulting from chromosomal abnormalities accounts for approximately 50% of cases of primary amenorrhea.
- Karyotype analysis should be done in the case of primary amenorrhea.

ABP Content Specifications(s)
- Recognize the clinical findings associated with primary amenorrhea of various etiologies, and manage appropriately

Suggested Readings
- Bondy CA. Care of girls and women with Turner syndrome: a guideline of the Turner Syndrome Study Group. J Clin Endocrinol Metab. 2007;92(1):10-25.

Question 163

A 5-year-old boy with trisomy 21 and acute lymphoblastic leukemia, diagnosed 1 year ago, is admitted to the hospital with increased work of breathing and dry cough that developed over the last several days. The family reports travel to several cities in the United States over the last month, but there were no known ill contacts or unusual exposures. On physical examination, the boy's temperature is 38°C, heart rate is 120 beats/min, respiratory rate is 60 breaths/min, blood pressure is 95/65 mm Hg, and oxygen saturation is 85% in room air. He has subcostal and intercostal retractions with clear breath sounds on auscultation of the lungs. With the exception of alopecia and mild pallor, his physical examination is otherwise unremarkable.
Laboratory findings were as follows:
- Electrolytes, calcium, serum urea nitrogen, creatinine, serum bicarbonate, and glucose levels were normal
- White blood cell count, 1,600/µL (1.6×10^9/L) with 30% neutrophils, 40% lymphocytes, and 30% monocytes
- Hemoglobin, 9 g/dL (90 g/L)
- Platelet count, 130×10^3/µL (130×10^9/L)
- Lactate dehydrogenase, 1,100 U/L

A chest radiograph is obtained (Item Q163).

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

A.   adenovirus

B.   Cryptococcus neoformans

C.   influenza virus

D.   Pneumocystis jirovecii

E.   Streptococcus pneumoniae

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ITEM Q163: Chest radiograph for the boy described in the vignette

Question 163   **Preferred Response: D**

The boy in the vignette has immune suppression due to leukemia. He has the characteristic findings of Pneumocystis jirovecii (formerly carinii) infection including a nonproductive cough, shortness of breath, significant hypoxia, fever, bilateral perihilar infiltrates on chest radiograph, and an elevated lactated dehydrogenase. The onset of disease can be indolent or fulminant.

Pneumonia caused by adenovirus or influenza may also present with nonproductive cough, tachypnea, and fever, but breath sounds are usually clear and there is a lesser degree of hypoxia than is typical in Pneumocystis jirovecii pneumonia (PCP). Pneumonia caused by Cryptococcus is characterized by cough, chest pain, and fever; chest radiography may show a solitary nodule or focal or diffuse infiltrates. Patients with pneumococcal (Streptococcus pneumoniae) pneumonia typically have productive cough, focal findings on chest auscultation (eg, bronchial breath sounds, crackles, rales over affected area), and focal infiltrates on chest radiography.

Pneumocystis species are ubiquitous, and most immunocompetent children acquire asymptomatic infection and subsequent anti-Pneumocystis antibodies by 20 months of age. PCP occurs almost exclusively in children with immunodeficiency, such as the boy in the vignette who has leukemia treated with chemotherapy and pancytopenia. PCP is a serious, opportunistic infection commonly found in children with human immunodeficiency virus (HIV) infection who are not receiving effective antiretroviral therapy or PCP prophylaxis.

The diagnosis of PCP is confirmed on microscopic identification of fungal organisms in respiratory tract sections or lung tissue. Polymerase chain reaction assays are also sensitive for detecting the organism in clinical specimens. First-line treatment for severe PCP is intravenous (IV) trimethoprim-sulfamethoxazole (TMP-SMX) and corticosteroids for 21 days. Pentamidine IV is recommended for children who cannot tolerate TMP-SMX, or who have severe disease that has not responded to 5 to 7 days of TMP-SMX. For mild to moderate PCP, oral atovaquone may be used. Experience with other antimicrobial combinations used to treat adults is limited in children.

Pneumocystis jirovecii pneumonia prophylaxis is recommended for all children with severe immunosuppression (eg, HIV-infected children with low CD4+ T-lymphocyte counts, primary immunodeficiency, other acquired cell-mediated immunodeficiency such as that following organ transplantation, and patients receiving immunosuppressive chemotherapy). Oral TMP-SMX (daily or three times per week) is the prophylactic drug of choice. If TMP-SMX is not tolerated or cannot be used, alternative agents include oral dapsone (dosed daily or weekly), aerosolized pentamidine (monthly), or oral atovaquone (daily).

PREP Pearls
• Pneumocystis jirovecii pneumonia (PCP) occurs almost exclusively in immunocompromised hosts.
• Nonproductive cough, fever, and increased work of breathing accompanied by significant hypoxia are the most common presenting features of PCP.

- Oral trimethoprim-sulfamethoxazole is the drug of choice for PCP prophylaxis in patients with severe immunosuppression.

ABP Content Specifications(s)
- Plan appropriate prophylaxis for Pneumocystis jiroveci infection in a child who is receiving chemotherapy
- Recognize the clinical features associated with Pneumocystis jiroveci infection, and manage appropriately
- Understand the epidemiology of Pneumocystis jiroveci

Suggested Readings
- American Academy of Pediatrics. Cryptococcus neoformans infections. In: Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:309-311 .

Question 164

A 6-year-old boy has suffered a severe traumatic brain injury and bilateral pulmonary contusions after being struck by a car while walking. Computed tomography of the head shows extensive subdural and subarachnoid hemorrhage, diffuse cerebral edema, ischemia, and herniation of the cerebellar tonsils through the foramen magnum. The neurosurgeon does not consider him an operative candidate because of the nonsurvivable nature of the injury. He has fixed and dilated pupils; absent corneal, gag, and oculocephalic reflexes; no eye movement on cold caloric stimulation; and no response to painful stimuli. He does not breathe over the ventilator. When he is disconnected from the ventilator, his oxygen saturations decrease to the 70% range within seconds. The apnea test could not be performed because of his respiratory instability. His parents accept the inevitability of his death and express a strong desire to donate his organs.

Of the following, the MOST appropriate next course of action is to

A.   inform the family he is not a candidate for organ donation

B.   order a cerebral blood flow scan

C.   perform organ donation after cardiac death

D.   recover all organs for donation in the operating room under anesthesia

E.   recover only the corneas and skin for donation

Question 164   **Preferred Response: B**

The child in the vignette has suffered a severe nonsurvivable traumatic brain injury. It is possible that brain death has occurred because there are no signs of brain activity. Brain death cannot be clinically diagnosed for the boy in this vignette because the apnea test cannot be safely performed. The next step in making the diagnosis is to order an ancillary study such as a cerebral blood flow scan.

Throughout most of human existence, death was defined by the cessation of breathing and circulation. In 1981, the US President's Commission released the Uniform Determination of Death Act which defines death as either the irreversible cessation of circulatory or respiratory functions or irreversible cessation of all functions of the entire brain, including the brainstem. In short, the 2 paths to death include circulatory criteria and neurologic criteria, with the provision that it be irreversible. Since the widespread adoption into clinical practice of brain death criteria in the late 1960s, the majority of donated organs have been recovered from brain-dead donors. An ethical corollary to the 1981 statement is that death should not be caused by the removal of organs. Thus, objective and rigorous criteria must be applied and death should be declared before organ removal for transplantation.

In 1987, and updated in 2011, the American Academy of Pediatrics published guidelines for determining brain death in children and neonates. Prerequisites to testing include the absence of neurologic function, with a known, irreversible cause of coma and the absence of potentially reversible contributors such as abnormal vital signs, medications, or toxins. For example, testing should not proceed if the patient exhibits even the slightest sign of neurologic function, such as pupillary reactivity, spontaneous breathing, or response to stimulus. The clinical examination for brain death consists of 2 full cranial, motor, and sensory neurologic examinations and apnea testing, separated by age-dependent observation periods. The observation period should be 24 hours for full-term newborns (37 weeks' gestational age to 30 days of age), and 12 hours for infants and children (> 30 days to 18 years).

Apnea testing consists of disconnecting the ventilator, allowing the arterial carbon dioxide to rise above 60 mm Hg, and observing for signs of breathing. If the child becomes unstable during that period, as in the child in this vignette, testing should be stopped and the child cannot be declared dead. In such an instance, an ancillary test such as a cerebral blood flow study or an electroencephalography should be performed.

The local organ procurement organization (OPO) should be informed of any patient with a severe neurologic injury, especially when brain death is suspected. Although discussions regarding organ donation may occur at any time when initiated by the family, the family should not be approached by anyone about organ donation before the declaration of death. Until such a time, the child is still alive, and the family may perceive a conflict of interest between their child and potential organ recipients even if the clinicians and OPO representatives have the best of intentions. However, a clinician must pursue a diagnosis of brain death without delay if suspected, much in the way he/she would auscultate the chest to diagnose death based on circulatory criteria. Once brain death is diagnosed, the family should be approached by the OPO

representative regarding consent for organ donation. If consent is declined, support of breathing and circulation should be stopped and the standard bereavement pathway should be followed. There is no requirement for consent or agreement for withdrawal of support in this situation because the diagnosis of death is valid from medical, ethical, and legal standpoints. If brain death is suspected and a family chooses to withdraw life-sustaining therapies before the declaration of death, the possibility of organ donation could be lost. However, prolonging futile care for the patient approaches an ethical gray area. One approach is to inform the family that the usual practice in such cases of devastating neurologic injury is to proceed along the brain death pathway.

Informing the family that the boy is not a candidate for organ donation is not appropriate in this case, because it is still possible he may be declared brain dead with the performance of an ancillary study. Organ donation after cardiac death involves recovery of organs after death is declared based on circulatory criteria in cases of failed resuscitation or after withdrawal of support. In addition to some ethical considerations, organ donation after cardiac death is less than ideal because organs become ischemic after circulation stops. Recovery of all organs for donation in the operating room under anesthesia is not appropriate because organ donation before declaring death is not ethical. Recovery of the corneas and skin only is not the most appropriate next course of action, because brain death has not been diagnosed.

PREP Pearls
- The local organ procurement organization should be notified of any patient with a severe neurologic injury, and should initiate discussions with families regarding organ transplantation after the diagnosis of brain death has been made.
- The diagnosis of brain death should be pursued as soon as it is suspected.
- Death can be declared based on either circulatory (cardiac death) or neurologic criteria (brain death).

ABP Content Specifications(s)
- Recognize and apply ethical principles involved in decisions regarding organ transplantation and donation

Suggested Readings
- Nakagawa TA, Ashwal S, Mathur M, Mysore M; Society of Critical Care Medicine, Section on Critical Care and Section on Neurology of the American Academy of Pediatrics, and the Child Neurology Society. Guidelines for the determination of brain death in infants and children: an update of the 1987 Task Force Recommendations. Pediatrics. 2011;128:e720. DOI: http://dx.doi.org/10.1542/peds.2011-1511.

Question 165

An 18-year-old young man, hospitalized for perforated appendicitis, develops a rash. He underwent laparoscopic appendectomy and has been receiving piperacillin-tazobactam for peritonitis caused by Bacteroides fragilis. Vital signs show a temperature of 38.3°C, respiratory rate of 20 breaths/min, blood pressure of 120/75 mm Hg, and a weight of 65 kg. On physical examination, he has generalized abdominal tenderness and a diffuse morbilliform exanthem. You suspect a drug reaction and choose to change the antibiotic regimen.

Of the following, the antibiotic expected to provide the BEST coverage for this patient's infection is

A.   aztreonam

B.   cefoxitin

C.   clindamycin

D.   imipenem

E.   levofloxacin

Question 165   **Preferred Response: D**

The carbapenem class of antibiotics, which includes imipenem, meropenem, and ertapenem, provide excellent coverage against anaerobic pathogens and, of the choices given, imipenem would be the best therapeutic option for the boy in this vignette. Susceptibility of Bacteroides fragilis to imipenem, metronidazole, and combinations of a penicillin with a β-lactamase inhibitor has generally been preserved. However, resistance to penicillins, cephalosporins, and clindamycin has increased. While carbapenems also provide excellent coverage of gram-negative infections, ertapenem specifically is not active against Acinetobacter species and Pseudomonas aeruginosa.

Monobactams, such as aztreonam, have poor activity against anaerobic bacteria and are not considered adequate as single therapy for polymicrobial intra-abdominal infections. Cefoxitin is the most effective cephalosporin against B fragilis. However, 5% to 15% of isolates can be resistant. Similarly, it is estimated that 5% to 10% of B fragilis will be resistant to clindamycin. In national guidelines that address treatment of intra-abdominal infections in adults and children, clindamycin is not recommended given increasing resistance in the B fragilis group. Additionally, clindamycin does not treat gram-negative bacteria.

Older generation fluoroquinolones also have poor activity against anaerobes. While anaerobic activity is improved in newer generation fluoroquinolones (eg, levofloxacin), national guidelines recommend metronidazole be added to adequately cover anaerobic bacteria when fluoroquinolones are used for the treatment of intra-abdominal infections. In the gastrointestinal tract, anaerobes outnumber aerobic bacteria many times over. Therefore, any infection that involves dissemination of gastrointestinal flora into a normally sterile site (eg, peritonitis resulting from perforated appendicitis) must be considered polymicrobial with anaerobic bacteria playing an important role. Selected antimicrobials must target all potentially involved pathogens. Piperacillin-tazobactam can be an appropriate first line choice, as it targets gram-negative and anaerobic bacteria in addition to treating susceptible gram-positive pathogens such as Enterococcus species.

Anaerobic infections are common in children. Other clinical scenarios that may warrant antimicrobial therapy directed against anaerobes include brain abscesses, oral and dental infections, deep neck infections, pelvic infections, and necrotizing soft tissue infections. In general, anaerobes should be considered potential pathogens in certain infections associated with abscesses and in the setting of tissue destruction with associated gas formation.

PREP Pearls
- Susceptibility of anaerobic bacteria to carbapenems, metronidazole, and combinations of a penicillin with a β-lactamase inhibitor has generally been preserved.
- Clindamycin is not recommended for anaerobic coverage of intra-abdominal infections given increasing resistance in the Bacteroides fragilis group.
- Clinical scenarios that warrant antimicrobial therapy directed against anaerobes include brain abscesses, oral and dental infections, deep neck infections, intra-abdominal infections, pelvic infections, and necrotizing soft tissue infections.

American academy of pediatrics   537

ABP Content Specifications(s)
•   Recognize the common clinical features associated with anaerobic infections

Suggested Readings
•   Brook I. Anaerobic infections in children. Adv Exp Med Biol. 2011;697:117-152. DOI: http://dx.doi.org/10.1007/978-1-4419-7185-2_10.

Question 166

A 14-year-old adolescent presents to your office for a routine health supervision visit. He has no complaints or concerns today and is not taking any medications. Vital signs show a temperature of 37.8°C, heart rate of 60 beats/min, respiratory rate of 16 breaths/min, and blood pressure (BP) of 162/90 mm Hg. His weight is 80.1 kg, height is 155 cm, and his body mass index is 33.3. His physical examination is otherwise unremarkable. His repeat BP after 15 min of relaxation with an appropriate size cuff is 162/94 mm Hg and 165/89 mm Hg.

Of the following, the MOST appropriate categorization of the patient's elevated blood pressures is

A.   hypertensive emergency

B.   malignant hypertension

C.   masked hypertension

D.   stage 1 hypertension

E.   stage 2 hypertension

Question 166   **Preferred Response: E**

Hypertension in children is defined based upon the 2004 National High Blood Pressure Education Program Working Group guidelines. According to the guidelines, high blood pressure (BP) in children is identified using the BP percentiles tables based upon gender, age, and height.

These guidelines categorize BP readings in children as:
1. Normal: systolic and diastolic BP less than the 90th percentile
2. Prehypertension: systolic or diastolic BP greater than or equal to 90th percentile, but less than 95th percentile or more than 120/80 mm Hg (even if < 90th percentile for age, gender, and height)
3. Stage 1 hypertension: systolic or diastolic BP between the 95th percentile and 5 mm Hg above the 99th percentile
4. Stage 2 hypertension: systolic or diastolic BP greater than or equal to 99th percentile plus 5 mm Hg

In the presence of a difference between systolic and diastolic BP readings categorization, the higher value determines the BP category.

The recommendations of the 2004 National High Blood Pressure Education Program Working Group guidelines are similar to the Joint National Committee (JNC 7) recommendations for categorizing high BP in adults. These guidelines categorize BP readings in adults as:
1. Normal: systolic BP less than 120 mm Hg and diastolic BP less than 80 mm Hg
2. Prehypertension: systolic BP 120 to 139 mm Hg or diastolic BP 80 to 89 mm Hg
3. Stage 1 hypertension: systolic BP 140 to 159 mm Hg or diastolic BP 90 to 99 mm Hg
4. Stage 2 hypertension: systolic BP greater than or equal to 160 mm Hg or diastolic BP greater than or equal to 100 mm Hg

For the 14-year-old male adolescent in this vignette with a height near the tenth percentile (155 cm), prehypertension is BP greater than 121/76 mm Hg and less than 125/80 mm Hg, stage 1 hypertension is BP greater than or equal to 125/80 mm Hg and less than 137/93 mm Hg, and stage 2 hypertension is BP greater than or equal to 137/93 mm Hg (Item C166A).

Item 166A. Blood Pressure Percentiles for a 14-year-old Male.

Age, y	BP Percentile	SBP, mm Hg							DBP, mm Hg						
		Percentile of Height							Percentile of Height						
		5th	10th	25th	50th	75th	90th	95th	5th	10th	25th	50th	75th	90th	95th
14	50th	106	107	109	111	113	114	115	60	61	62	63	64	65	65
	90th	120	121	123	125	126	128	128	75	76	77	78	79	79	80
	95th	124	125	127	128	130	132	132	80	80	81	82	83	84	84
	99th	131	132	134	136	138	139	140	87	88	89	90	91	92	92

SBP, Systolic Blood Pressure; DBP, Diastolic Blood Pressure

Reprinted with permission from the 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

Masked hypertension is defined as BP readings that do not meet the criteria for hypertension based upon office readings, but are consistently elevated by out-of-office measurements. Masked hypertension is the opposite of white coat hypertension wherein BP readings are consistently elevated by office readings, but do not meet criteria for hypertension diagnosis based on out-of-office readings. Increased use of 24-hour ambulatory BP monitoring in evaluation of elevated BP readings in out-of-office settings has helped in better characterization of masked and white coat hypertension. Increased long term risk of sustained hypertension and cardiovascular morbidity has been reported with both white coat and masked hypertension.

Hypertensive emergency is defined as severely elevated BP readings, along with clinical features of acute onset end-organ damage (encephalopathy; headache, seizures, mental status changes, focal neurologic symptoms, visual disturbances, and heart failure; chest pain, palpitations, shortness of breath). Patients with hypertensive emergency are at increased immediate risk and require rapid reduction of blood pressure (less than 25% to 30% over the first several hours).

Hypertensive urgency is severely elevated BP readings in asymptomatic patients. Patients with hypertensive urgency may be managed with a slower reduction of BP (normalization in children and below 140/90 in adults) with either intravenous or oral antihypertensives, depending on the patient's symptoms. Malignant hypertension is rapidly rising blood pressure that leads to end organ damage, a finding not seen in the adolescent described in the vignette.

It is important to recheck and confirm systolic and diastolic BP greater than or equal to 95th percentile on 3 or more separate occasions using the techniques detailed in the fourth task force report. A detailed history and physical examination is indicated for all patients with elevated BP. Physical examination should focus on clinical findings suggestive of secondary hypertension Item C166B.

	Finding*	Possible Etiology
Vital signs	Tachycardia	Hyperthyroidism, pheochromocytoma, neuroblastoma
	Decreased lower extremity pulses; drop in blood pressure from upper to lower extremities	Coarctation of the aorta
Eyes	Retinal changes	Severe hypertension, more likely to be associated with secondary hypertension
Ear, nose, and throat	Adenotonsillar hypertrophy	Suggests association with sleep-disordered breathing (sleep apnea), snoring
Height/weight	Growth retardation	Chronic renal failure
	Obesity (high body-mass index)	Primary hypertension
	Truncal obesity	Cushing syndrome, insulin resistance syndrome
Head and neck	Moon facies	Cushing syndrome
	Elfin facies	Williams syndrome
	Webbed neck	Turner syndrome
	Thyromegaly	Hyperthyroidism
Skin	Pallor, flushing, diaphoresis	Pheochromocytoma
	Acne, hirsutism, striae	Cushing syndrome, anabolic steroid abuse
	Café-au-lait spots	Neurofibromatosis
	Adenoma sebaceum	Tuberous sclerosis
	Malar rash	Systemic lupus erythematosus
	Acanthrosis nigricans	Type 2 diabetes
Chest	Widely spaced nipples	Turner syndrome
	Heart murmur	Coarctation of the aorta
	Friction rub	Systemic lupus erythematosus (pericarditis), collagen-vascular disease, end-stage renal disease with uremia
Abdomen	Apical heave	Left ventricular hypetrophy/chronic hypertension
	Mass	Wilms tumor, neuroblastoma, pheochromocytoma
	Epigastric/flank bruit	Renal artery stenosis
	Palpable kidneys	Polycystic kidney disease, hydronephrosis, multicystic-dysplastic kidney, mass (see above)
Genitalia extremities	Ambiguous/virilization	Adrenal hyperplasia
	Joint swelling	Systemic lupus erythematosus, collagen vascular disease
	Muscle weakness	Hyperaldosteronism, Liddle syndrome

*Findings listed are examples of physical findings and do not represent all possible physical findings.

Adapted from Flynn JT. *Prog Pediatr Cardiol.* 2001;12:177-188

Item C166B: Physical examination findings suggestive of secondary hypertension.

In all patients with hypertension (BP ≥ 95th percentile), further evaluation of blood urea nitrogen, creatinine, electrolytes, urinalysis, complete blood cell count, fasting lipid profile, fasting glucose, renal ultrasonography, and echocardiogram (also in patients with prehypertension) is recommended. Fasting lipid profile and glucose is also recommended in overweight and prehypertensive patients, as well as patients with diabetes or chronic kidney disease and family history of hypertension or cardiovascular disease.

Detailed evaluation for secondary causes of hypertension such as plasma renin activity, renovascular imaging (renal scan, duplex Doppler renal ultrasonography, arteriography), plasma

and urine steroids or catecholamines are indicated for young children with stage 1 hypertension or adolescents with stage 2 hypertension.

PREP Pearls
- Children with high blood pressure (BP) readings should be categorized as stage 1 or stage 2 hypertension using the BP percentiles tables (age, gender, and height-based)
- Hypertensive emergency is defined as elevated BP readings, along with clinical features of acute onset end-organ damage.
- In all pediatric patients with hypertension (BP ≥ 95th percentile), blood urea nitrogen, creatinine, electrolytes, urinalysis, complete blood cell count, fasting lipid profile, fasting glucose, renal ultrasonography, and echocardiogram is recommended.
- Detailed evaluation for secondary causes of hypertension is indicated for young children with stage 1 hypertension or adolescents with stage 2 hypertension.

ABP Content Specifications(s)
- Plan the initial clinical and diagnostic evaluation of hypertension
- Formulate a differential diagnosis of hypertension in patients of various ages

Suggested Readings
- Brady TM. Hypertension. Pediatr Rev. 2012;33(12):541-552. DOI: http://dx.doi.org/10.1542/pir.33-12-541.

Question 167

A 4-month-old term male infant is brought to the office for evaluation of vomiting. The vomiting began in the first weeks after birth and has been getting worse. The vomiting is nonbloody and follows most feeds of cow's milk-based formula. The infant's mother reports intermittent bilious emesis and feels they are increasingly projectile. His growth chart shows that his weight has declined from the 50th to 25th percentile over the past 3 months. He is otherwise well, with normal vital signs and no evidence of infection on examination. There are no associated respiratory symptoms or color changes. The parents are mixing the formula correctly.

Of the following, the BEST next step in evaluation is

A.   abdominal radiograph

B.   gastric scintigraphy

C.   pH impedence study

D.   upper endoscopy

E.   upper gastrointestinal series

Question 167   **Preferred Response: E**

The 4-month-old infant in this vignette has gastroesophageal reflux (GER). The differential diagnosis for his reflux includes cow's milk and other protein intolerance, structural etiologies (malrotation, pyloric stenosis, and gastric outlet obstruction), gastritis, gastroparesis, and physiologic reflux. The persistent symptoms and intermittent bilious nature suggest structural issues and dysmotility, however, cow's milk intolerance is the most common cause of worsening GER.

Gastroesophageal reflux occurs in all ages. Gastroesophageal reflux is the physiologic passage of gastric contents into the esophagus. Gastroesophageal reflux disease (GERD) is the passage of gastric contents into the esophagus associated with symptoms or problems. Gastroesophageal reflux in infants is very common, occurring in two-thirds of all infants. The incidence is higher in Western societies. There is a clear hereditary portion to GER and the associated complications (ie, erosive esophagitis and Barrett esophagus, a condition where the lining of the esophagus is damaged by gastric acid). The literature demonstrates an increased risk in certain populations, including preterm or neurologically impaired infants, children with a history of structural or dysmotility issues (esophageal atresia, hiatal hernia, and achalasia), and children with chronic respiratory issues or with a history of lung transplant.

The evaluation of GER is complex because of the lack of a gold standard diagnostic test. Instead, the current testing begins with a complete history and physical examination to document acid in the esophagus, the correlation of acid to symptoms, and evaluation for complications of GER, such as esophagitis. The tests most commonly used include an initial upper gastrointestinal series to evaluate the anatomy when there are concerns for an esophageal web, hiatal hernia, malrotation, or other structural anomaly. Pyloric ultrasonography should be used to evaluate forceful vomiting to rule out pyloric stenosis in young infants. Esophageal pH monitoring can be used to quantify the frequency and duration of GER and provides information on the temporal association between GER and symptoms. Gastric scintigraphy evaluates for postprandial reflux and aspiration, although this test is limited by a lack of consensus on standard technique or normalized values. An abdominal radiograph will not be helpful in the diagnosis of GER. Finally, endoscopy with biopsy provides direct visualization of the esophagus and tissue to evaluate for esophagitis and other conditions, such as eosinophilic gastrointestinal disease, that can clinically mimic GERD.

Gastroesophageal reflux disease is defined as GER with associated esophagitis, poor weight gain, dysphagia, or abdominal and retrosternal pain. Extra-intestinal complications include respiratory symptoms such as chronic cough, wheezing, laryngitis with or without hoarse voice, dental erosions, and recurrent otitis media. Development of Barrett esophagitis or peptic strictures is uncommon in children. The literature suggests that the contribution of GERD on asthma is less than previously believed. Symptoms differ by age (Item C167A).

Item C167A. Symptoms of Gastroesophageal Reflux by Age.

Infant	Older Child/Adolescent
Recurrent Vomiting	Recurrent Vomiting
Feeding Refusal	Abdominal Pain
Poor Weight Gain	Heartburn
Fussiness	Dysphagia
Irritability	Asthma
Sleep disturbance	Respiratory symptoms
Respiratory symptoms	Upper airways symptoms (ie, hoarse voice)

Modified and reprinted with permission from Lightdale JR, Gremse DA.

Gastroesophageal reflux: management guidance for the pediatrician. *Pediatrics*.

2013;131(5):e1684-e1695. doi:10.1542/peds.2013-0421.

Gastroesophageal reflux disease is managed with both lifestyle changes and medications (Item C167B) and can be handled by general pediatricians in most cases. Pediatric gastroenterology should be consulted when primary treatment fails, if medication weaning fails, when children are failing to thrive, or for other complications including significant family history, as noted in Item C167B.

Item C167B. Management of Gastroesophageal Reflux Disease.

Age	Infant	1-year-old and older
Lifestyle Modifications		
Diet	Maternal elimination diet (cow's milk, soy, egg at a minimum)	Avoid triggers, acidic and spicy foods, caffeine and alcohol
	Formula change (soy, protein hydro lysate, amino acid)	Consider postprandial chewing of sugarless gum
	Decrease volume, Increase frequency	Positioning
	Thickening with cereal	Weight loss for obese
	Positioning	
Medications		
Histamine receptor antagonist proton pump inhibitor	Ranitidine 5 to 10 mg/kg/day divided 2 to 3 times per day	Ranitidine 75 to 150 mg twice daily
	Nizatadine 10mg/kg/day divided twice per day	Nizatadine 150 mg twice per day
	Omeprazole 1 to 4 mg/kg/day	Omeprazole 20 to 40 mg/day
	Lansoprazole 1 to 4 mg/kg/day	Lansoprazole 15 to 30 mg/day
Refer to Gastroenterology	Not responding to diet changes	Not responding to diet changes
	Not responding to conservative medical therapy	Not responding to conservative medical therapy
	Unable to wean off medications	Unable to wean off medications
	Family history of ulcerations of Barrett esophagitis	Family history of ulcerations of Barrett esophagitis
	Failure to thrive	

Courtesy of C. Waasdorp Hurtado

Item C167B. Management of Gastroesophageal Reflux Disease.

Age	Infant	1-year-old and older
Lifestyle Modifications		
Diet	Maternal elimination diet (cow's milk, soy, egg at a minimum) Formula change (soy, protein hydro lysate, amino acid) Decrease volume, Increase frequency Thickening with cereal Positioning	Avoid triggers, acidic and spicy foods, caffeine and alcohol Consider postprandial chewing of sugarless gum Positioning Weight loss for obese
Medications Histamine receptor antagonist proton pump inhibitor	Ranitidine 5 to 10 mg/kg/day divided 2 to 3 times per day Nizatadine 10mg/kg/day divided twice per day Omeprazole 1 to 4 mg/kg/day Lansoprazole 1 to 4 mg/kg/day	Ranitidine 75 to 150 mg twice daily Nizatadine 150 mg twice per day Omeprazole 20 to 40 mg/day Lansoprazole 15 to 30 mg/day
Refer to Gastroenterology	Not responding to diet changes Not responding to conservative medical therapy Unable to wean off medications Family history of ulcerations of Barrett esophagitis Failure to thrive	Not responding to diet changes Not responding to conservative medical therapy Unable to wean off medications Family history of ulcerations of Barrett esophagitis

Courtesy of C. Waasdorp Hurtado

PREP Pearls
- Gastroesophageal reflux is the physiologic reflux of gastric contents into the esophagus.
- Gastroesophageal reflux disease (GERD) is the passage of gastric contents into the esophagus associated with symptoms or problems.
- The differential diagnosis for GERD includes cow's milk and other protein intolerance, structural etiologies (malrotation, pyloric stenosis, and gastric outlet obstruction), gastritis, gastroparesis, and physiologic reflux.
- Initial evaluation includes a complete history and physical examination, followed by an upper gastrointestinal series.
- Evaluation of GERD includes an extensive history, physical examination, and family history, followed by appropriate testing and/or referral to pediatric gastroenterology.

ABP Content Specifications(s)
- Recognize the complications associated with gastroesophageal reflux
- Plan the appropriate evaluation of gastroesophageal reflux, and manage appropriately

Suggested Readings
- Craig WR, Hanlon-Dearman A, Sinclair C, Taback SP, Moffatt M. Metoclopramide, thickened feedings, and positioning for gastro-oesophageal reflux in children under two years. Cochrane Database Syst Rev. 2004;(4):CD003502. doi:http://dx.doi.org/10.1002/14651858.CD003502.pub2.

Question 168

You are called to evaluate a baby in the newborn nursery with dysmorphic features. The nurse tells you that the newborn has severe micrognathia and retrognathia with glossoptosis and is having difficulty breathing. Physical examination of the newborn shows small, malformed ears with extreme narrowing of the external auditory canals, a cleft palate, absent lashes and notching of the lower eyelids, hypoplastic facial bones with a prominent nose, and downward-slanting palpebral fissures. The mother has downward slanting palpebral fissures, mild hearing loss, and hypoplasia of the zygomatic complex.

After immediately addressing the respiratory compromise, you explain to the family that the baby is MOST likely to have

A.   congenital heart disease

B.   craniosynostosis

C.   normal intelligence

D.   renal anomalies

E.   vertebral defects

Question 168   **Preferred Response: C**

The baby in the vignette has Treacher Collins syndrome (TCS), which is an autosomal dominant disorder caused by gene mutations in TCOF1 (78%-93%) and POLR1C or POLR1D (8%). Sixty percent are de novo gene mutations. An individual with TCS is at 50% risk of passing it on to their children. Patients have a classic facial dysmorphology that is characterized by hypoplasia of the zygomatic bones and mandible, down-slanting palpebral fissures, prominent nose, micrognathia, retrognathia, external ear abnormalities, coloboma of the lower eyelid, absence of the lower eyelashes, and anterior hair displacement onto the lateral cheekbones (Item C168A). Conductive hearing loss is present in 40% to 50% of patients, mostly secondary to malformation of the ossicles and middle ear cavity hypoplasia. Inner ear structures are typically normal. Less commonly, patients will have a cleft palate or choanal stenosis and atresia. Airway abnormalities are common secondary to the choanal atresia, shortening of the mandible, glossoptosis, and micrognathia. Ophthalmologic abnormalities include coloboma of the lower eyelid, ocular hypertelorism, vision loss, amblyopia, refractive errors, and strabismus. There is significant inter- and intrafamilial clinical variability among affected family members.

Courtesy of L. Parsley

Item C168: Baby with Treacher Collins syndrome.
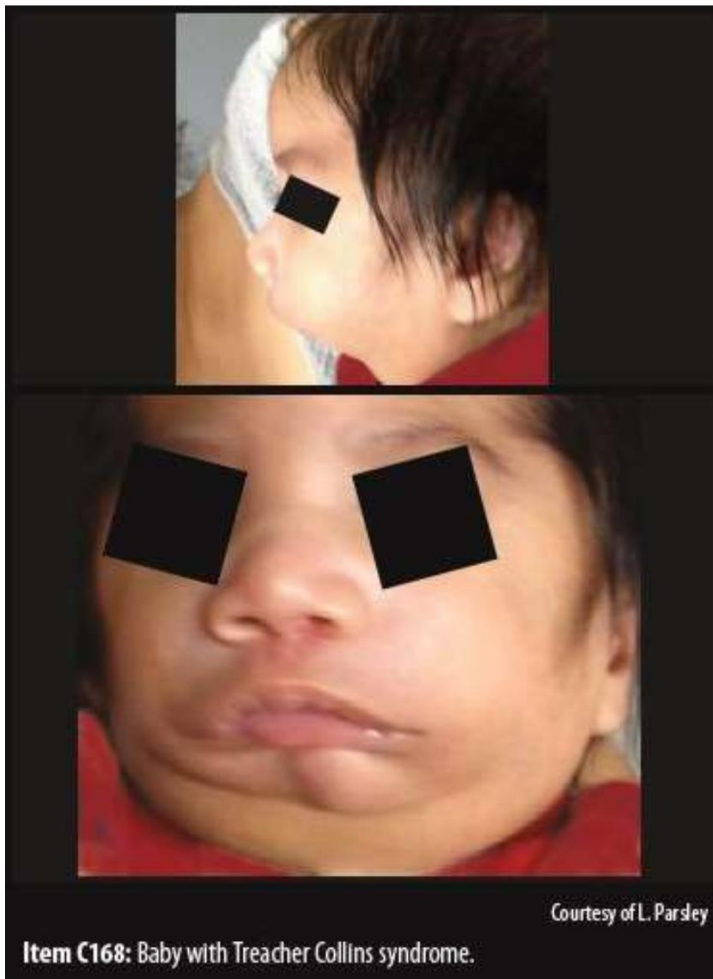
Intelligence is usually normal, which is important for the family and care team to know from an educational and therapeutic standpoint. Fertility is normal. Craniosynostosis is not a feature of TCS, though patients usually have brachycephaly with bitemporal narrowing. Congenital heart disease, renal anomalies, and vertebral defects are not commonly seen. Features of TCS are also seen in Nager syndrome, Miller syndrome, Goldenhar syndrome, Pierre Robin sequence, and nonsyndromic mandibular hypoplasia. With Nager syndrome and Miller syndrome, patients will have the TCS facial dysmorphology, known as mandibular dysostosis, along with additional limb anomalies. Nager syndrome patients have preaxial limb anomalies. Miller syndrome patients have postaxial limb anomalies.

Major management concerns can be stratified by specific age groups (Item C168B).

AGE	INTERVENTION
Birth to 2 years of age	Hearing loss assessment with bone conduction amplification if needed Repair of cleft palate if present Feeding and airway management
3 to 12 years of age	Zygomatic and orbital reconstruction when the cranio-orbitozygomatic bony development is complete (5-7 years of age) External ear reconstruction Speech therapy and educational interventions
13 to 18 years of age	Maxillomandibular reconstruction Orthognathic procedures

PREP Pearls
- Treacher Collins syndrome is an autosomal dominant disorder characterized by a classic facial dysmorphology that is characterized by hypoplasia of the zygomatic bones and mandible, downslanting palpebral fissures, prominent nose, micrognathia and retrognathia, external ear abnormalities, coloboma of the lower eyelid, absence of the lower eyelashes, and anterior hair displacement onto the lateral cheekbones.
- Intelligence and fertility is normal with Treacher Collins syndrome.
- Careful attention to airway management and feeding difficulties is important in infancy and early childhood.

ABP Content Specifications(s)
- Recognize the genetic and clinical features associated with various types of dysostosis, including Treacher Collins syndrome

Suggested Readings
- Katsanis SH, Jabs EW. Treacher Collins syndrome. GeneReviews. http://www.ncbi.nlm.nih.gov/books/NBK1532.

Question 169

The 26-year-old mother of a patient seeks advice about the human papillomavirus (HPV) vaccine. She wonders if she should pursue vaccination for herself. She will be turning 27 years of age in 4 months and she tested positive for HPV on her most recent screening examination. She has received conflicting information about the risks and benefits of this vaccine in cases such as hers, and asks for information and a recommendation.

Of the following, the BEST recommendation for this woman is that she

A.   is already HPV positive, so vaccination is not indicated

B.   is too old to complete the vaccine series, so vaccination is not indicated

C.   should receive only the first 2 doses of HPV vaccine with a 4-week interval

D.   should receive 3 doses of HPV vaccine at 0, 1, and 4 months

E.   should receive 3 doses of HPV vaccine at 0, 1, and 6 months

Question 169   **Preferred Response: E**

All health care providers should know the recommendations, limitations, and schedule for the human papillomavirus (HPV) vaccine. HPV vaccine is recommended for all women through age 26 years. The 3-dose series should be initiated and completed at the recommended minimal intervals to the young mother in this vignette, even though she will be 27 years of age when the third dose is given.

The HPV vaccine is recommended for all males and females beginning at age 11 years, with the option to initiate the series as early as age 9 years. The vaccine series should be offered to female and male patients through age 26 and 21 years, respectively. In addition, unimmunized men ages 22 to 26 years who have sex with men or are immunocompromised should initiate the series. Ideally, HPV vaccination should be administered before the initiation of sexual contact and potential exposure to HPV. However, the vaccine should not be withheld from people who are already sexually active. The HPV vaccine is recommended regardless of sexual orientation. Three formulations of the HPV vaccine are licensed for use in the United States by the US Food and Drug Administration (FDA) and currently recommended by the Centers for Disease Control and Prevention (CDC). Bivalent (HPV2) and Quadrivalent (HPV4) were initially licensed for use. Both vaccines protect against HPV types 16 and 18, which are responsible for 70% of cases of cervical, 87% of anal, 60% of oropharyngeal, and 31% of penile cancers. In addition, HPV4 protects against HPV types 6 and 11, which are responsible for 90% of anogenital warts and almost all cases of juvenile recurrent respiratory papillomatosis. In order to provide greater coverage, a 9-valent vaccine, HPV9, was approved by the FDA on December 10, 2014, for use in girls ages 9 through 26 years and boys ages 9 through 15 years. HPV9 protects against 5 additional HPV types (31, 33, 45, 52, and 58) which cause approximately 15-20% of vulvar, vaginal, and cervical cancers. All formulations may be used in females. HPV2 should not be used in males because of the limited efficacy in prevention of genital warts.

All HPV vaccines should be administered in a 3-dose schedule. The second dose is administered 1 to 2 months after the first dose and the third dose is given 6 months after the first dose. Four weeks is the minimal interval between the first and second doses; 12 weeks between the second and third doses; and 24 weeks between the first and the third doses. There is no accelerated schedule for completing the series, so although the woman in the vignette will be older than 26 years at the time of series completion, the minimal intervals must be adhered to.

No clinical trials indicate a therapeutic effect on existing HPV infections or genital warts, but patients infected with 1 or more HPV types still benefit from vaccination for protection against the remaining HPV types in the vaccine. Hence, the mother in the vignette should be encouraged to complete the HPV vaccine series despite being HPV positive. Testing for previous exposure to HPV is not indicated before immunization. HPV vaccine may be administered to women with an abnormal or equivocal Papanicolaou test result as well as to patients with a history of anogenital warts.

The HPV vaccine is not recommended during pregnancy. The practitioner should inquire about last menstrual period and potential for pregnancy in sexually active female patients; however, a negative pregnancy test is not required before administering the vaccine.

PREP Pearls
- The human papillomavirus (HPV) vaccine is recommended beginning at age 11 years, and may be initiated as early as age 9 years.
- The 3-dose HPV vaccine series should be offered to all female and male patients through ages 26 and 21 years, respectively, and men ages 22 to 26 years who have sex with men or are immunocompromised.
- The recommendations for minimal intervals between the 3 doses must be followed. Four weeks is the minimal interval between the first and second doses; 12 weeks between the second and third doses; and 24 weeks between the first and the third doses.

ABP Content Specifications(s)
- Know the recommendations, limitations, and schedule for the human papillomavirus vaccine

Suggested Readings
- AAP Committee on Infectious Diseases. HPV vaccine recommendations. Pediatrics. 2012;129(3):602-605. DOI:http://dx.doi.org/10.1542/peds.2011-3865.

Question 170

A 4-month-old infant presents to the emergency room for evaluation because her parents feel that she has been "moving her legs less" over the past 2 weeks. On physical examination, she is a happy infant in no distress who smiles at her mother and grabs her hair. The only abnormal finding is flaccidity in her legs bilaterally. A complete blood cell count, complete metabolic panel, creatine kinase, and coagulation profile are all unremarkable.

Of the following, the MOST appropriate course of action is to

A.   admit her to the hospital for overnight observation and a neurology consult

B.   arrange an outpatient appointment with a neurologist in the morning

C.   instruct her to follow-up with her pediatrician in 1 week

D.   recommend a physical therapy evaluation

E.   obtain an immediate magnetic resonance image of the thoracic and lumbar spine

Question 170   **Preferred Response: E**

It can be difficult for parents to identify lower extremity weakness in infants who are not yet mobile and it is imperative that examining physicians identify motor abnormalities quickly. The etiology of the weakness must be also quickly ascertained. If an infant or child is seen in an office setting and found to have new onset weakness, they should be referred to the emergency department. Once there, imaging of the spine at the level of the suspected defect must be obtained rapidly. Injury to the spinal cord may be caused by damage intrinsic to the cord (eg, myelitis) or extrinsic to the cord in the form of spinal cord compression from a hematoma, abscess, or a tumor. Spinal cord compression is a true medical emergency and requires immediate action. The longer there is compression and nerve dysfunction, the greater the likelihood that nerve damage will be permanent. As the infant in the vignette has weakness in her lower extremities but not her upper extremities, cross-sectional imaging of the thoracic and lumbar cord is required. Magnetic resonance imaging (MRI) is the imaging modality of choice when available on an immediate basis. When MRI is not immediately available, computed tomography should be performed.

While many types of childhood cancer can present with spinal cord compression in early childhood, the most common include neuroblastoma and tumors of the central nervous system. Neuroblastoma is an embryonal tumor of the peripheral nervous system and can arise in the adrenal gland or in any of the sympathetic ganglia. It commonly arises in a paraspinal ganglion and tends to track into the spinal canal through the neural foramina. This results in the "dumbbell" sign (Item C170). While neuroblastoma rarely invades the spinal cord, it can cause severe compression (Item C170), where the cord is not visible at all in the thoracic canal.
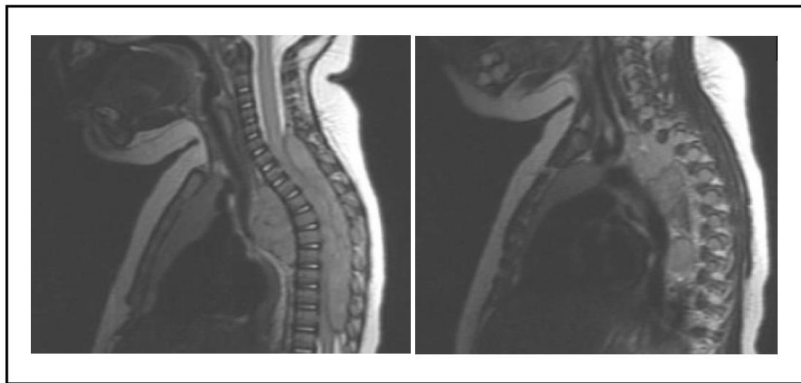
Item C170. Paraspinal neuroblastoma with invasion through the neural foramina into the spinal canal. Courtesy of J Fish.

Once spinal cord compression has been identified, decompression must occur quickly. Depending on the etiology of the compression, decompression can occur by surgical laminectomy or emergent chemotherapy. If the spinal cord compression is caused by a tumor, dexamethasone should be initiated immediately, as a reduction in inflammation can help reduce

the pressure on the spinal cord. If a tumor is noted, a pediatric oncologist should be emergently consulted to determine the most appropriate method for cord decompression.

Admission for observation and a neurological evaluation are appropriate in this circumstance, but only after imaging has been performed and spinal cord compression has been ruled out. Discharge from the emergency room without imaging would not be the most appropriate management in this scenario. While a physical therapy evaluation and program would be appropriate to regain strength in the legs, it should come only after the diagnosis and management plan have been initiated.

PREP Pearls
- New onset weakness is an emergency that requires immediate cross-sectional imaging of the spinal cord from the suspected level of injury down.
- If cord compression is found to be caused by a tumor, dexamethasone should be initiated and an emergent pediatric oncology consult should be obtained.
- Spinal cord decompression can be achieved surgically with a laminectomy, or medically with chemotherapy or radiation, depending on the suspected cause of the compression.

ABP Content Specifications(s)
- Recognize the clinical findings associated with spinal cord compression (eg, from a tumor, from myelopathy), and the need for prompt evaluation

Suggested Readings
- De Bernardi B, Balwierz W, Bejent J, et al. Epidural compression in neuroblastoma: diagnostic and therapeutic aspects.Cancer Lett. 2005;228(1-2):283-299. DOI: http://dx.doi.org/10.1016/j.canlet.2005.01.053

Question 171

You are seeing a 5-year-old boy and his adoptive parent because of educational concerns arising in kindergarten. He was adopted 1.5 years ago, following a brief time in foster care. He has a history of chronic and severe neglect by his biological parents. His adoptive parent says she noted fairly quickly that he was developmentally behind, but had been told that with parental attention and stimulation he was likely to catch up. However, that has not turned out to be the case. Hearing and vision are normal for his age, but communication, as assessed by his speech therapist, has not progressed beyond using occasional 2-word phrases. He is very active and is "quite a handful" according to his mother. His attention and focus is poor, and although he does make eye contact with his parent, he avoids eye contact with others. You think it is likely that this patient has an intellectual disability and have recommended that he be seen by a psychologist for IQ testing.

Of the following, the additional information needed to confirm your suspected diagnosis is

A.   adaptive behavior assessment

B.   attention-deficit/hyperactivity disorder rating scale

C.   autism symptom rating scale

D.   brain magnetic resonance imaging

E.   electroencephalogram

Question 171   **Preferred Response: A**

This vignette describes a 5-year-old child who has a very significant developmental delay (ie, he will only occasionally use a 2-word phrase). He is clearly in need of further evaluation and intervention. Intelligence testing would be appropriate in the process of defining if he has an intellectual disability (ID) and the particular type of difficulty that he has. Establishing a correct diagnosis will help to structure the best school and outpatient care plans for him.

In order to establish the diagnosis of an ID, one cannot rely fully on IQ testing. A significant deficit in adaptive functioning must be found before an ID diagnosis would be valid. According to the current view of ID, someone who has an IQ more than 2 standard deviations below the mean, which means a score of less than 70 but with normal range basic life skills for their age, should not be diagnosed with ID. Adaptive life skills involve self care (such as feeding and dressing yourself) and basic life planning (ie, getting yourself to where you need to go). The ideal way to perform this adaptive life skill assessment is by using a standardized instrument normed for age, which should go along with the IQ testing being performed.

An attention-deficit/hyperactivity disorder (ADHD) rating scale would be a good way to gather inattentive and hyperactive behavior observations from multiple observers. He is said to have a poor attention span and is behaviorally "quite a handful." However, ADHD should not be diagnosed if another diagnosis, in this case ID, provides an explanation for his behaviors. This patient may be developmentally at the mental age of a 2-year-old because of ID, in which case an ADHD diagnosis would not make sense.

While an autism spectrum disorder should be on this child's current differential diagnosis (the reported poor eye contact raises this possibility), utilizing an autism symptom rating scale is not the preferred option for a few reasons. The first is that the vignette specifically stated the diagnosis being evaluated is an ID, for which both IQ and adaptive behavior assessments are the preferred steps rather than having the parent fill out an autism symptom rating scale. The second reason is that an autism symptom rating scale will not diagnose autism, but rather will yield a score indicating the overall likelihood of autism and, if present, the extent of those symptoms. When there is already sufficient reason to be suspicious of autism, as in this case, a clinical assessment of the diagnostic characteristics of autism in the Diagnostic and Statiscal Manual of Mental Disorders, Fifth Edition would be indicated. This child could have either ID, autism, or both.

A brain magnetic resonance image would be indicated for any circumstance in which a child's intellectual or other brain functioning is linked to specific neurological findings, such as spastic hemiparesis, or has functioning that appears to be degenerative or progressive in nature. Magnetic resonance imaging assessments for ID in general are not indicated. An electroencephalogram (EEG) would be reasonable if there was any history of tonic-clonic movements, sudden episodes of altered consciousness with loss of bowel or bladder control, or possible post-ictal states. A child with recurrent seizures may appear more intellectually impaired than they would be if their seizures were under control, so this test is occasionally

pertinent during the evaluation of an ID. For the child in this vignette, there is no reason for an EEG at this time.

While it is true that a very young child with a history of being raised in a neglectful household tends to increase their developmental gains after being placed in a more stimulating and responsive environment, a reassurance-only approach would not be appropriate if the degree of developmental delay is significant.

PREP Pearls
- Children with an IQ of less than 70 will not be diagnosed with an intellectual disability if their adaptive behavior assessment is in the normal range.
- The diagnosis of intellectual disability and autism are separate from each other; a child can have one or the other or both of these disorders.

ABP Content Specifications(s)
- Understand the utility of an adaptive behavioral assessment

Suggested Readings
- American Psychiatric Association. Diagnostic and Statistical Manual of Psychiatric Disorders, Fifth Edition. Washington DC: American Psychiatric Publishing; 2014: 991 pp.
- Shea SE. Intellectual disability (mental retardation). Pediatr Rev. 2012;33(3):110-121. DOI:http://dx.doi.org/10.1542/pir.33-3-110.

Question 172

A 2-day-old newborn is being evaluated for seizures by neurology and has had a computed tomography of the brain that shows findings consistent with tuberous sclerosis. The neonatal intensive care unit nurse tells you that there have been frequent wide complex ectopic beats, but none have been captured on an electrocardiogram. On physical examination, you find a male newborn with weight of 3.8 kg in no distress, heart rate of 130 beats/min, respiratory rate of 34 breaths/min, and blood pressure of 82/55 mm Hg. The heart rhythm is regular. The cardiac examination is unremarkable: there is a normal S1and S2 without any murmurs, rubs, thrills, or gallops. The point of maximal impulse is not displaced. There is no hepatosplenomegaly and the femoral pulses are normal. An echocardiogram is ordered.

Of the following, the MOST likely finding on echocardiogram in this patient is

A.   atrial septal defect

B.   atrioventricular canal defect

C.   dilated cardiomyopathy

D.   rhabdomyomas

E.   tetralogy of Fallot

Question 172   **Preferred Response: D**

The newborn in this vignette has tuberous sclerosis (TS) and is at risk for cardiac rhabdomyomas.

Rhabdomyomas are the most common cardiac tumor in young children and account for 80% of those seen in infants younger than 1 month of age. More than 50% of children with rhabdomyomas have TS complex. The other congenital heart lesions in the response choices, including tetralogy of Fallot, atrial septal defect, and atrioventricular canal, may occur, but are not linked to TS. Dilated cardiomyopathy is not likely. The rhabdomyomas may cause the ventricular septum to enlarge because of the mass effect of the tumor.

Infants with TS and cardiac rhabdomyomas may present with a wide range of clinical symptoms. They may be asymptomatic from a cardiac standpoint, or have symptoms of outflow obstruction with abnormal atrioventricular (AV) valve function. If there is a mass on one of the AV valves, there may be a regurgitant murmur in systole, or a murmur to suggest stenosis in diastole. If there is outflow tract obstruction, there may also be systolic murmurs and, in the case of the right ventricular outflow tract, there may be cyanosis. Patients have been reported with intractable arrhythmias. The newborn described in the vignette has had a report to suggest premature ventricular contractions. This may remain a minor issue or develop into a more serious ventricular arrhythmia. The diagnosis of cardiac rhabdomyoma is usually made with echocardiography and may be confirmed with magnetic resonance imaging. Patients commonly have multiple lesions that range from millimeters to centimeters. They may cause little or no hemodynamic consequence or require multiple medications such as amiodarone and propranolol to control ventricular tachycardia. If there is evidence of obstruction, then surgical resection may be recommended. Everolimus, an inhibitor of the mTOR pathway, can be used to treat subependymal giant cell astrocytomas associated with TS and has been shown to be effective in shrinking the cardiac rhabdomyomas in selected patients. The pathologic structure of the rhabdomyomas may include Purkinje cells, and this may be a mechanism for pre-excitation seen in some patients with this type of cardiac tumor. Patients with this finding may develop supraventricular tachycardia.

Fortunately, however, the most likely course for cardiac rhabdomyomas diagnosed in infancy is that of spontaneous regression. Surgical resection can be avoided unless there is obstruction or arrhythmias that are unable to be controlled medically.

PREP Pearls
- All children with cardiac rhabdomyomas need evaluation for tuberous sclerosis.
- Cardiac rhabdomyomas found in infancy often regress spontaneously in the first year of life.

ABP Content Specifications(s)
- Recognize cardiac conditions associated with tuberous sclerosis

Suggested Readings

• Benyounes N, Fohlen M, Devys JM, et al. Cardiac rhabdomyomas in tuberous sclerosis patients: a case report and review of the literature. Arch Cardiovasc Dis. 2012;105(8-9):442-445. DOI: http://dx.doi.org/10.1016/j.acvd.2012.01.009

Question 173
You are seeing a 12-year-old boy in your office with a complaint of right knee pain and swelling of 4 days' duration. He has pain with movement, making it difficult to walk. He has no fever, rash, conjunctivitis, or diarrhea. There is no history of travel, injury, or tick exposure. The boy was seen 2 weeks ago for fever and cough, at which time a throat culture was negative and he was treated for suspected community-acquired pneumonia. He was then well until the knee swelling occurred. On physical examination, you find an afebrile, well-appearing boy with swelling, erythema, and decreased range of motion of the right knee. He winces when you passively flex his knee. The remainder of the physical examination is unremarkable. The boy was seen in the emergency department 2 days earlier, where an evaluation for septic arthritis was negative.
Laboratory results from the emergency department show:
- White blood cells, 11,300/μL (11.3 x 10^9/L)
- Red blood cells, 4.5 x 10^6/μL (4.5 x 10^{12}/L)
- Hemoglobin, 11.2 g/dL (112 g/L)
- Hematocrit, 37.1%
- Platelet count, 300 x 10^3/μL (300 x 10^9/L)
- Erythrocyte sedimentation rate, 25 mm/h
- C-reactive protein, 2.0 mg/L
- Synovial fluid analysis:
  - Yellow and opaque
  - White blood cell count, 20,000 cells
  - 50% polymorphonuclear leukocytes
  - Low viscosity
  - Gram-stain negative
  - Culture no growth for 48 hours

Of the following, the BEST next step in management of this boy's symptoms is

A.   azithromycin

B.   doxycycline

C.   naproxen

D.   penicillin

E.   prednisone

Question 173   **Preferred Response: C**

The boy in the vignette is well appearing and afebrile, with normal blood counts. His inflammatory markers are only slightly elevated and the joint fluid does not appear to be infectious. The child most likely has reactive arthritis. Naproxen is a nonsteroidal anti-inflammatory drug that is recommended as the first line for treatment of reactive arthritis. The drug, with both pain relieving and mild anti-inflammatory properties, 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. Prednisone is not recommended in this case as this treatment can mask the symptoms of chronic arthritis.

Reactive arthritis is associated with an infection outside the affected joint. Reactive arthritis is a clinical diagnosis based on the presence of oligoarticular arthritis, usually of the lower extremities, and exclusion of other types of arthritis, such as septic arthritis, Lyme arthritis, acute rheumatic fever, trauma, neoplasm and osteomyelitis. Reactive 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 diseases such asChlamydia and gonorrhea. All patients with a history of sexual activity and arthritis should be screened for these diseases. Reactive arthritis is also associated with other genitourinary, gastrointestinal, and upper respiratory infections. The arthritis can appear within days or up to 6 weeks after the infection. After 6 weeks, 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 C173) and with a conservative approach, such as rest and cold therapy. Activity should be limited secondary to pain and can resume as pain improves.

Item C173. Medications to Treat Reactive Arthritis.

Nonsteroidal Anti-inflammatory Drugs	Anti-inflammatory Dose*	Dosage forms
Ibuprofen	10 mg/kg/dose 4 times a day OR 13 mg/kg/dose 3 times a day (maximum dose 2400 mg/day)	Pill or liquid
Naproxen	10 mg/kg/dose twice a day (maximum dose 1000 mg/d)	Pill or liquid
Meloxicam	0.25 mg/kg/dose once daily (maximum dose 15 mg/day)	Pill or liquid (very small pill)
Celecoxib	50 mg twice a day for 2-17 years of age and weighing 10-25 kg; 100 mg twice a day for patients weighing > 25 kg (maximum dose 200 mg/day)	Capsules may be opened and sprinkled onto apple sauce and taken immediately with water

*The doses listed are anti-inflammatory doses for the nonsteroidal anti-inflammatory drugs. Smaller doses are used for fever and pain relief.

Courtesy of A. Brown

PREP Pearls
- Sexually active patients with arthritis should be screened for Chlamydia and gonorrhea.
- Reactive or postinfectious arthritis is associated with genitourinary, gastrointestinal, and upper respiratory infections.
- Postinfectious arthritis should be managed initially with nonsteroidal anti-inflammatory medications.

ABP Content Specifications(s)
- Recognize the clinical findings associated with postinfectious arthritis

Suggested Readings
- Bernard R. Approach to the child with joint inflammation. Pediatr Clin North Am. 2012;59:245-262. DOI:http://dx.doi.org/10.10

Question 174

A 9-year-old girl presents to your office for evaluation of right wrist pain 2 days after falling onto an outstretched arm during a soccer game. On physical examination, the girl is tender over the right ulnar mid shaft with limited pronation and supination. Neurovascular examination of the hand and wrist is unremarkable. Radiographs of the wrist demonstrate anterior bowing of the shaft of the ulna.

Of the following, the MOST appropriate next step would be to

   A.   allow return to activities without immobilization once she regains pronation and supination

   B.   obtain dedicated radiographs of the elbow

   C.   obtain magnetic resonance imaging of the forearm

   D.   recommend cast immobilization of the wrist and forearm

   E.   refer to orthopedic surgery for operative fixation of the ulna

Question 174   **Preferred Response: B**

The girl in the vignette has restricted pronation and supination, which should prompt evaluation for an associated elbow injury. Examination of the elbow after an acute injury should include inspection for deformity, swelling, bruising, and evaluation of motion. Range of motion is often restricted with both acute and chronic injuries.

The medial border of the ulna should be perfectly straight on lateral radiography; curvature suggests plastic deformity, bowing of the bone on radiographs without evidence of cortical dysfunction. Similar to greenstick fractures, plastic deformity is only seen in pediatric patients because of the increased flexibility of young bones. The presence of an ulnar deformity with limited pronation and supination raises concern for a Monteggia lesion—fracture or deformation of the ulna associated with a radiocapitellar dislocation. Although Monteggia lesions are rare, failure to diagnose this condition can lead to subsequent disability. Therefore, any patient with a midshaft or proximal ulnar injury, even patients with plastic deformity or mild greenstick fracture, should undergo dedicated elbow radiography to examine the radiocapitellar joint. Item C174 shows a greenstick fracture of the ulna with an associated radiocapitellar dislocation.

Item C174. Greenstick fracture of the ulna with an associated radiocapitellar dislocation. Courtesy of M Sagan.

Monteggia lesions were classified by Bado into 4 types. The most common is a type 1 injury, with fracture of the proximal or midshaft of the ulna and associated anterior dislocation of the radial head. There are also Monteggia lesion variants, ulnar injuries with radial head subluxation that were not included in Bado's classification system. Monteggia injuries represent fewer than 1% of pediatric fractures and typically occur in children younger than 12 years. A Monteggia lesion identified within 3 weeks of injury often can be treated with closed reduction. Monteggia lesions with delayed diagnosis and fracture healing, or those unsuccessfully treated by closed reduction, require open reduction with ulnar osteotomy to restore normal radiocapitellar joint anatomy.

Elbow dislocations at the ulnar-trochlear joint are rare in children, with the incidence rising during adolescence. Dislocations most often occur in the posterolateral direction. When an elbow dislocation is suspected, radiographs should always be obtained before performing a reduction maneuver, both to confirm the diagnosis and to look for associated fractures.
For the girl in the vignette, a return to sports without imaging to evaluate the elbow and immobilization for the ulnar deformity would not be appropriate. Magnetic resonance imaging is not indicated because a Monteggia lesion, if present, should be visible on plain radiography. Cast immobilization of the wrist and forearm would be appropriate if no radiocapitellar disruption is seen on elbow radiography. The patient should have close follow-up because of the risk for radiocapitellar dislocation in the first few weeks after injury. If a Monteggia lesion is seen on radiography, referral to orthopedic surgery would be indicated. Because the girl in the vignette was injured only 2 days before presentation, the appropriate first step would be to attempt closed reduction under sedation.

PREP Pearls
- For children with any type of acute ulnar deformity or fracture, dedicated elbow radiographs should be obtained to assess for possible Monteggia lesion (radiocapitellar joint dislocation).
- Elbow dislocation at the ulna/trochlear joint is uncommon in children.

ABP Content Specifications(s)
- Recognize the clinical findings associated with sports-related dislocation of the elbow, including associated complications, and manage appropriately
- Recognize the clinical findings associated with sports-related elbow pain, and manage appropriately

Suggested Readings
- Weinstein SL, Flynn HM. Lovell & Winters Pediatric Orthopaedics. 7th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2013.

Question 175

A 14-year-old patient requires treatment for a severe bacterial infection and meets inclusion criteria to participate in a study examining the safety and appropriate dosing in children of a new antibacterial drug. In addition to obtaining informed consent from the patient's parents, appropriate information to share with this patient includes:

- What the study is about
- Why the patient qualifies for the study
- That the study is voluntary
- What procedures are included in the study
- The potential benefits of participating in the study
- The risks of participating in the study
- That the patient will be treated the same whether enrolled in the study or not
- That the patient can withdraw from the study at any time
- An opportunity for the patient to ask questions

Of the following, the process outlined for the patient is BEST known as the

A.   assent process

B.   consent process

C.   dissent process

D.   permission process

E.   regulatory process

Question 175   **Preferred Response: A**

The assent process, as described in the vignette, requires that a reasonable effort be made to enable a child to understand what his/her participation in research would entail and obtain the child's affirmative agreement to participate in the research. Informed assent considers the age and maturity of the child, his/her psychological state, and the nature of the proposed research. Informed dissent occurs when the child decides not to participate in research after having the research protocol and processes explained. In cases of informed dissent, the child's decision should prevail even if the parent or guardian has consented to the research. Institutional review boards (IRBs) can waive informed assent if the capabilities of the child are so limited that they cannot be consulted, the study offers important benefits otherwise unavailable, or if the study already qualifies for a waiver of informed consent.

Informed consent is the process by which full disclosure of research information (Item C175) is discussed with the parent or guardian in nonscientific language, with the opportunity for questions and clarification. This process should be conducted in a noncoercive setting and in a manner such that the parent can comfortably make an informed decision about enrolling the child in the research protocol. If a parent gives permission for a child to participate in a research protocol without having received full disclosure of information regarding the research, that does not constitute informed consent. The concepts of informed assent, dissent, and consent arise from the ethical principles of patient autonomy and basic human rights. These principles are essential for a good relationship between a physician and patient.

• The patient's diagnosis
• The nature and purpose of the proposed research/treatment
• The potential risks and benefits of participation in the research/treatment
• Alternatives to the research/treatment and their risks and benefits
• The risks and benefits of not participating in the research or receiving the treatment
• The expected outcome of participation in the research/treatment

Item C175. Information to be Disclosed as Part of the Informed Consent Process Courtesy of D. Palazzi.

The research regulatory process covers vastly more than the consent or assent processes alone, and includes issues such as IRB registration rules, financial disclosures of investigators, good laboratory practice, investigational drug regulations, electronic record documentation, signature rules, and more.

PREP Pearls
• The assent process requires that a reasonable effort be made to enable a child to understand what his/her participation in research would entail and obtain the child's affirmative agreement to participate.
• Informed consent is the process by which full disclosure of research information is discussed with the parent or guardian in nonscientific language, with the opportunity for questions and clarification.

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- The concepts of informed assent, dissent, and consent arise from the ethical principles of patient autonomy and basic human rights. These principles are essential for a good relationship between physician and patient.

ABP Content Specifications(s)
- Understand the difference between informed consent and assent
- Recognize and apply ethical principles involved in the patient-parent-pediatrician relationship regarding issues of informed consent/dissent/assent

Suggested Readings
- US Food and Drug Administration. Code of Federal Regulations Title 21. 21CFR50.55. Available athttp://www.accessdata.fda.gov/scripts/cdrh/cfdocs/cfcfr/cfrsearch.cfm?fr=50.55.
- US Food and Drug Administration. Guidance for institutional review boards and clinical investigators. Available atwww.fda.gov/oc/ohrt/irbs/default.htm.

Question 176

You are following up on telephone messages at the end of a busy day in your pediatric practice with the assistance of a medical student. One message is from a local attorney, asking if you would be willing to serve as an expert witness in a medical malpractice case involving a pediatric patient. The medical student asks if it is ethical for pediatricians to do this.

Of the following, the MOST accurate statement is that

    A.   it is generally not ethical for physicians to receive compensation for serving as expert witnesses in medical malpractice cases

    B.   it is generally not ethical for physicians to testify as expert witnesses in medical malpractice litigation unless mandated to do so by a court order

    C.   physicians serving as expert witnesses should be aware that transcripts may be submitted as courtroom testimony for peer review

    D.   physicians should not serve as expert witnesses within the state(s) in which they hold medical licensure, to avoid conflicts of interest

    E.   physicians should only agree to testify in cases that are unrelated to their specific medical specialties, to ensure objectivity

Question 176   **Preferred Response: C**

All pediatricians should recognize and apply ethical principles regarding medical testimony and being an expert witness. Transcripts of courtroom testimony in medical malpractice cases may be submitted for peer review, and physicians who serve as expert witnesses should be aware of this. In fact, some medical specialty organizations have established programs in which the content of expert witness testimony is reviewed and critiqued by a panel of peers. Expert witness testimony, along with peer analyses, may even be published in scientific journals.

The American Academy of Pediatrics (AAP) Committee on Medical Liability and Risk Management has issued a policy statement that provides guidance for pediatricians regarding expert witness participation. The guidelines emphasize that the public interest and interests of the medical and legal professions "are best served when scientifically sound and unbiased expert witness testimony is readily available in civil and criminal proceedings." Pediatricians have an ethical and professional duty to "assist in the administration of justice" as "members of the medical community, patient advocates, and private citizens."

Regarding qualifications for physicians who serve as expert witnesses, the AAP recommends that physicians should only contribute as medical experts to cases in which they possess true expertise and related experience. Physicians serving as expert witnesses should hold current, unrestricted medical licenses in their states of practice and be certified by the American Board of Medical Specialties, American Osteopathic Association, or a board with equivalent standards. In addition, expert witnesses must have been actively engaged in clinical practice in the area of medicine about which they testify. The majority of their working time should not be spent doing expert witness work unless they have retired from clinical practice.

Pediatricians serving as expert witnesses should take all steps needed to ensure that the medical testimony they provide in legal proceedings is complete, accurate, unbiased, and based on an excellent understanding of current medical evidence and standard practice related to the case for which they testify.

It is acceptable for physicians to receive reasonable compensation for serving as an expert witness in a medical malpractice case, as long as the compensation is commensurate with the actual time and effort involved and prevailing market value. Compensation for expert witness testimony should never be contingent on the outcome of the case, and contracts between medical expert witnesses and attorneys should be constructed in a manner that is conducive to fairness, accuracy, completeness, and objectivity.

It is ethical for physicians to testify as expert witnesses in medical malpractice cases, even when they are not mandated by a court order to do so, as long as they adhere to ethical standards. The AAP recognizes that pediatricians "have the professional, ethical, and legal duty to assist in the legal process when medical issues are involved."

The assertion that physicians "should not serve as expert witnesses within the states in which they hold medical licensure" is incorrect. In fact, some state courts have "locality rules,"

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requiring that expert witnesses be familiar with the general practices of physicians in the same community, or at least in similar communities. In some cases, state courts have not allowed testimony from expert witnesses who have not practiced within that state.

Physicians should not agree to testify in cases unrelated to their specific medical specialties. The AAP recommends that pediatricians should limit their participation as medical experts to cases in which they possess genuine expertise.

PREP Pearls
- It is ethical for physicians to testify as expert witnesses, provided they take all steps needed to ensure that the testimony they provide is complete, accurate, unbiased, and based on a thorough understanding of current medical evidence and standard practice related to the case.
- The AAP recommends that physicians should only contribute as medical experts to cases in which they possess true expertise and related experience.
- Transcripts of courtroom testimony in medical malpractice cases may be submitted for peer review.

ABP Content Specifications(s)
- Recognize and apply ethical principles regarding medical testimony and being an expert witness

Suggested Readings
- American Academy of Pediatrics Committee on Medical Liability and Risk Management. Expert witness participation in civil and criminal proceedings. Pediatrics. 2009;124:428. DOI: http://dx.doi.org/10.1542/peds.2009-1132.

Question 177

A newborn who was delivered at home is brought to your office 30 hours after birth for the first newborn health supervision visit. She was born at 40 weeks' gestation to a 42-year-old primigravida mother by uncomplicated spontaneous vaginal delivery. The maternal prenatal screening was normal, including negative group B streptococcal screening. The maternal history was remarkable only for chronic hypertension. The newborn has been exclusively breastfeeding since birth. Vital signs include an axillary temperature of 37°C, heart rate of 140 beats/min, and respiratory rate of 50 breaths/min. Physical examination reveals a quiet, slightly ruddy newborn weighing 2,400 g (<3%). There is jaundice of the face and chest, periodic breathing with rare 20 second pauses, slightly decreased tone, and listlessness. The mother describes her newborn as being very quiet for the past 12 hours.

Of the following, the test MOST likely to establish the newborn's diagnosis is a(n)

A.   fractionated bilirubin

B.   ionized calcium

C.   serum glucose

D.   urine cytomegalovirus culture

E.   venous hematocrit

Question 177   **Preferred Response: C**

The small-for–gestational age (SGA) infant in the vignette, who presents with periodic breathing with apnea, decreased tone, and listlessness, a serum glucose test will most likely demonstrate hypoglycemia. SGA infants are at increased risk for hypoglycemia because of insufficient hepatic and skeletal muscle glycogen stores. Some affected infants have superimposed hyperinsulinemia. Hypoglycemia is typically seen during the first 3 days after delivery, and may persist up to 1 week. Symptoms may include jitteriness, apnea, tachypnea, weak cry, lethargy, poor feeding, and seizures. The infant in the vignette may not be getting sufficient breast milk to maintain an adequate plasma blood glucose concentration, leading to symptomatic hypoglycemia.

Small-for-gestational age infants with a history of intrauterine growth restriction (IUGR) have increased perinatal morbidity and mortality. Their intrauterine environment may cause chronic fetal hypoxia and in utero demise. Diminished blood flow during uterine contractions superimposed on a chronically stressed IUGR fetus may lead to intolerance of labor, meconium aspiration, and birth asphyxia. After birth, the infant's decreased subcutaneous fat and increased surface area contribute to excessive heat loss and issues with thermoregulation. Chronic fetal hypoxia may also stimulate erythropoiesis, leading to polycythemia, which may further exacerbate hypoglycemia. The American Academy of Pediatrics Committee on the Fetus and Newborn has recommended monitoring all SGA infants for hypoglycemia for the first 24 hours after birth.

For the infant in the vignette, who has jaundice noted in only the face and chest, the fractionated bilirubin is unlikely to be high enough to cause apnea and decreased tone. The infant is slightly ruddy, suggesting a mildly elevated hematocrit, but it is unlikely to be the primary cause of the infant's clinical findings. Hypocalcemia may present with the findings noted in the vignette, but is not associated with SGA infants unless asphyxia is present. Congenital cytomegalovirus infection is associated with IUGR/SGA and may present with significant central nervous system involvement. However, the underlying cause of the small size of the infant in the vignette is more likely the maternal chronic hypertension.

PREP Pearls
- Small-for-gestational age (SGA) infants are at increased risk for hypoglycemia because of insufficient hepatic and skeletal muscle glycogen stores. Some affected infants have superimposed hyperinsulinemia.
- SGA infants with concurrent intrauterine growth restriction have increased perinatal morbidity and mortality.
- SGA infants should be monitored for hypoglycemia for the first 24 hours after birth.

ABP Content Specifications(s)
- Understand the physiologic and physical abnormalities that may be present in a small-for-gestational-age infant
- Understand the mortality rate in small-for-gestational age infants

Suggested Readings

- American Academy of Pediatrics Committee on Fetus and Newborn. Postnatal glucose homeostasis in late-preterm and term infants. Pediatrics. 2011;127:575-579. DOI: http://dx.doi.org/10.1542/peds.2010-3851.

Question 178

A 10-year-old girl has a 2-year history of poor attention. At school, she stares out the window, often unresponsive to her teacher's directions, occasionally wanders around the playground, and fails to come in on time at the end of recess. Her parents have noticed episodes of similar behavior at home. One time, they noticed that the girl's eyes were "beating" side-to-side for about 10 min and she wouldn't respond to them. The girl reports that she was awake during that episode and could hear her parents, but didn't feel like answering "stupid" questions. Her physical examination is unremarkable.

Of the following, the BEST next step to establish the diagnosis is

A.   administration of a Vanderbilt Assessment scale

B.   begin a trial of methylphenidate

C.   obtain an electroencephalogram

D.   referral to an ophthalmologist

E.   referral to a psychologist

Question 178   **Preferred Response: C**

The girl in the vignette is having focal seizures (previously called complex partial seizures), characterized by altered consciousness with lateral nystagmus. Of the response choices listed, electroencephalography is the best test to evaluate for a seizure disorder. Clinically, it can be difficult to differentiate attentional disorder from focal seizures. Lateral nystagmus during an episode of altered consciousness or inattention is very suggestive of a seizure. Other symptoms that can suggest a seizure include facial twitching, drooling, or stereotyped automatisms or utterances during an episode.

A Vanderbilt Assessment Scale would be helpful in establishing a diagnosis of attentional disorder. However, the 10-year-old girl in the vignette has only had symptoms for 2 years and has associated nystagmus, so attentional disorder is not a likely diagnosis, and a trial of methylphenidate would not be helpful. Ophthalmology referral would be helpful to diagnose nystagmus, but not as helpful as electroencephalography to determine the cause of nystagmus. A psychology referral would be helpful if these episodes were behavioral in origin, but not in making a diagnosis of seizure.

PREP Pearls
- Episodes of altered consciousness with lateral nystagmus are highly suggestive of a focal-onset seizure.
- New-onset nystagmus requires prompt evaluation.

ABP Content Specifications(s)
- Recognize the clinical findings associated with nystagmus and the significance of those findings

Suggested Readings
- Russ SA, Larson K, Halfon N. A national profile of childhood epilepsy and seizure disorder. Pediatrics. 2012;129(2):256-264. DOI: http://dx.doi.org/10.1542/peds.2010-1371.

Question 179

A 14-year-old adolescent presents with emesis this morning, following 3 days of worsening abdominal pain and decreased energy. The patient has a history of pre–B cell acute lymphoblastic leukemia diagnosed 7 years ago, which was treated with chemotherapy for 3 years. His disease recurred and he underwent bone marrow transplant earlier this year. He subsequently developed graft versus host disease and was on a tapering course of prednisone over the last 2 months. He was most recently taking 5 mg orally once daily until his prednisone was discontinued 1 week ago. You order laboratory tests as a part of your evaluation.

Of the following, the MOST likely laboratory abnormality will be

A.   hypochloremia

B.   hyperglycemia

C.   hypokalemia

D.   hyponatremia

E.   metabolic alkalosis

Question 179   **Preferred Response: D**

The boy in the vignette has recently been weaned off glucocorticoid treatment and has developed adrenal insufficiency. Even when slowly weaned off steroid treatment, some patients may experience hypothalamic-pituitary-adrenal (HPA) axis dysfunction and subsequently develop signs of adrenal insufficiency (AI). Symptoms of AI can include weakness, fatigue, anorexia, nausea, and abdominal pain. Acute symptoms can also include muscle and joint pain and hypotension.

The causes of AI can be considered under 2 general headings: primary AI caused by destruction of the adrenal glands themselves, and secondary AI caused by disordered HPA axis function. In primary AI, hyponatremia and hyperkalemia occur because of mineralocorticoid deficiency. This is seen in conditions such as congenital adrenal hyperplasia, Addison disease, or other conditions leading to primary disease.

Secondary AI can occur after exogenous steroid treatment, after cure of Cushing syndrome, and from hypothalamic or pituitary lesions. As occurred with the boy in the vignette, in some cases the HPA axis remains suppressed despite prolonged tapering of exogenous glucocorticoids. The resulting decreased adrenocorticotropic hormone secretion leads to isolated glucocorticoid deficiency, which causes retention of free water and subsequent hyponatremia. Because the renin-angiotensin system is not affected, potassium concentrations are generally normal. However, if the adrenal glands have atrophied from prolonged suppression from exogenous steroid use, then it is possible to have abnormalities in both sodium and potassium.
Other biochemical abnormalities that can occur in both forms of AI include metabolic acidosis and hypoglycemia. Hypochloremia is not consistently associated with AI.

PREP Pearls
- Suppression of the hypothalamic-pituitary-adrenal axis after steroid withdrawal can lead to secondary adrenal insufficiency (AI).
- In secondary AI, isolated hyponatremia without potassium abnormalities is a common finding. This is in contrast to primary AI, where hyponatremia and hyperkalemia are frequently observed.

ABP Content Specifications(s)
- Recognize the clinical features associated with adrenal insufficiency after exogenous corticosteroid therapy has been discontinued, and the complications associated with sudden withdrawal

Suggested Readings
- Antal Z, Zhou P. Addison disease. Pediatr Rev. 2009;30(12):491-493. DOI: http://dx.doi.org/10.1542/pir.30-12-491.
- Loriaux DL. Adrenocortical insufficiency. In: Becker KL, ed. Principles and Practice of Endocrinology and Metabolism. 3rd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2001: 739-742.

Question 180
You have been caring for a 7-year-old girl with severe persistent asthma. She is being treated twice daily with a combined high dose inhaled corticosteroid and long acting β-agonist, and once daily with a leukotriene receptor antagonist. She demonstrates chronic rhinitis for which she is treated daily with a nasal steroid and a nonsedating antihistamine. Her father and paternal grandmother suffer from hay fever. She has never experienced an anaphylactic reaction to an insect sting or food.
Despite her treatment and reported excellent adherence, the patient has an asthma control test score of 17. She continues to require short acting β-agonist administration 3 to 4 times per week for exercise intolerance, cough, or wheezing, and has required 3 courses of systemic steroids in the last 4 months during weather changes or viral illnesses.
The family has a cat, 2 dogs, and a fish. Both parents smoke cigarettes outside of the home and car. Prior radioallergosorbent tests for cat, dog, peanut, and egg allergies were negative.
On physical examination, she has a comfortable respiratory pattern. The lungs are well aerated, but you note a prolongation of the expiratory phase of respiration with a scattered end expiratory wheeze. Forced expiratory volume in 1 second (FEV1) is greater than 80% predicted, but the FEV1/forced vital capacity ratio and maximal midexpiratory flow are mildly decreased.
The parents inquire about the utility of skin testing for allergies in their child.
Of the following, the BEST response to the parents' question is

    A.   further skin testing is not cost effective and relocation of the family's dog and cat is recommended

    B.   further skin testing is reasonable for avoidance measures, but allergy therapy has already been maximized

    C.   further skin testing is unnecessary, as prior radioallergosorbent testing was confirmed as negative

    D.   further skin testing is warranted, as allergies are a likely contributor to their daughter's poorly controlled asthma

    E.   no further testing or therapies are needed, as forced expiratory volume in 1 second is within normal range

Question 180   **Preferred Response: D**

According to National Heart, Lung, and Blood Institute criteria, the patient in the vignette has poorly controlled severe persistent asthma, as evidenced by high asthma-related impairment (exercise intolerance and requirement for "rescue" ß-agonist for symptoms > 2 times per week), elevated risk (3 courses of oral steroids with exacerbation in last 4 months, mildly abnormal lung function), and a low asthma control test score (< 20). In the optimal care of an asthmatic patient, asthma-related control is assessed at each visit to the health care provider. When poor control in the asthmatic patient is encountered, assessments should be made regarding adherence to therapies; additional attention should be given to the potential for comorbid conditions such as allergic rhinitis.

The patient in the vignette demonstrates chronic rhinitis and family members exhibit symptoms that are consistent with atopy. Furthermore, there are multiple potential or proven aero-allergens in the home, including pets and exposure to cigarette smoke.

The prevalence of asthma and allergic rhinitis have increased in parallel in recent decades. Asthma and allergic rhinitis frequently coexist in the same patient. As many as 60% of asthmatic individuals experience rhinitis and there is a corresponding 20% prevalence of asthma in patients with allergic rhinitis.

The complexity found in the overlapping clinical spectra of asthma and allergic rhinitis is exacerbated by the fact that asthma is not a single disease, but rather a variable and multifactorial disease process that may be modified by genetic, epigenetic, and environmental factors. The spectrum of atopic disease is similarly complex and different "endotypes" of atopic disease exist that are unique with regard to their association with asthma. Furthermore, both serum specific (immunoglobulin E [IgE]) and skin allergy tests are prone to variability in interpretation. Therefore, it has been recommended that both skin testing and specific IgE testing be quantified rather than simply reported as positive or negative.

The presence of sensitization to aero-allergens is a recognized risk factor for the development of asthma. In children younger than 3 years of age with recurrent wheezing, evidence of sensitization to 1 or more aero-allergens is considered one of the major criteria that predicts wheezing at school age (asthma predictive index). Moreover, asthma is often preceded by the presence of allergic rhinitis. Patients with allergic rhinitis without a history of asthma may demonstrate asthma-like airway hyperreactivity when exposed to allergens to which they have been sensitized. In addition, the inhalation of allergens in individuals with seasonal rhinitis has been shown to induce bronchial inflammation. The duration of rhinitis may be an independent risk factor for the development of asthma; in patients with moderate to severe persistent allergic rhinitis without asthma, rhinitis duration of greater than 5 years has been found to be a significant risk factor for severe airway hyperreactivity. Extensive research therefore suggests that the upper (nose) and lower (lung) airways should be regarded as a pathophysiological continuum rather than as distinct entities.

Compared to skin allergy testing, serum-specific IgE techniques are less time efficient and are more expensive. Several studies have also found serum testing to be less sensitive than skin testing for the detection of clinically relevant allergies. Serum testing allows evaluation of patients with moderate to severe atopic dermatitis, and patients do not need to stop routine antihistamine therapies prior to testing.

In this vignette, relocation of the family dog and cat is not recommended without evidence to suggest aero-allergen sensitization. Cost effectiveness of allergen testing is likely to be favorable in this patient with severe persistent asthma and the need for multiple physician visits. If significant allergic disease is confirmed, additional therapies, including immunotherapy, may be considered.

Finally, the vast majority of pediatric patients with moderate to severe persistent asthma have normal forced expiratory volume in 1 second. The criteria for asthma-related severity and control are clinically based and spirometric indices are only a portion of the appropriate assessments.

PREP Pearls
- The majority of pediatric patients with moderate-to-severe persistent asthma will demonstrate a normal forced expiratory volume in 1 second with spirometric testing.
- Allergic disease and asthma frequently coexist in the same patient. Allergic rhinitis is a recognized exacerbating factor in patients with asthma. Furthermore, allergic rhinitis may precede the development of asthma and contribute independently to airway hyperresponsiveness.
- Serum immunoglobulin E allergy testing is less sensitive than skin testing for the detection of clinically relevant allergies.
- When allergies are suspected in patients with asthma, skin testing is warranted to help direct therapy.

ABP Content Specifications(s)
- Recognize the frequency of positive immediate-type allergic skin tests in school-age children who have asthma

Suggested Readings
- Boulay ME, Boulet LP. The relationships between atopy, rhinitis and asthma: pathophysiological considerations. Curr Opin Allergy Clin Immunol. 2003;3(1):51-55.
- Chinoy B, Yee E, Bahna SL. Skin testing versus radioallergosorbent testing for indoor allergens. Clin Mol Allergy. 2005;3(1):4. DOI: http://dx.doi.org/10.1186/1476-7961-3-4.

Question 181

During his 9-month health supervision visit, you note that an infant has 2 lower central incisors. The mother reports that she has not begun cleaning the infant's teeth.

Of the following, the MOST appropriate guidance to provide this parent about her infant's dental health is:

   A.   dietary habits play little role in caries development

   B.   fluoride toothpaste should not be used by children younger than 5 years of age because of a high risk of toxicity

   C.   the initial dental examination should occur within 6 months of first dental eruption and no later than 12 months of age

   D.   only professionally applied fluoride treatments are effective in preventing caries

   E.   the parents' dental hygiene has little impact on the infant's oral health

Question 181   **Preferred Response: C**

Dental caries is the most common chronic condition in children, affecting as many as 41% of US children 2 to 11 years of age. An important part of prevention and intervention for this common disease is the establishment of a dental home. The American Academy of Pediatrics, American Academy of Pediatric Dentistry (AAPD), American Dental Association, and Association of Public Health Dentistry recommend that the first dental examination occur within 6 months of the eruption of the first primary tooth and no later than 12 months of age. An established dental home is important for the assessment of the individual child's risk for oral disease and development of a comprehensive plan to address those risk factors. In addition, the dental practitioner is key in providing oral health anticipatory guidance and providing dental care according to established periodicity schedules.

Pediatricians also play a crucial role in supporting dental health, especially in the areas of primary and secondary prevention. Primary prevention for dental caries is focused on parental dental health because the child's oral bacterial flora is determined by that of the mother, and if the mother has heavy colonization with Streptococcus mutans and other cariogenic bacteria, the child is at greater risk for early childhood caries. Secondary prevention focuses on managing risk factors for early childhood caries once the bacterial milieu is established. The 3 components of this approach are dietary counseling, promotion of oral hygiene, and use of fluoride. A diet high in sugary carbohydrates provides the substrate that cariogenic bacteria ferment, leading to lowered pH (which promotes further growth of cariogenic bacteria) and demineralization of dental enamel. Therefore, dietary counseling to promote breastfeeding, to limit sugar, especially in liquids, and to eliminate taking a bottle to bed are aspects of anticipatory guidance that promote dental health.

Current recommendations for oral hygiene are that twice daily tooth brushing should start as soon as the first tooth erupts and flossing should begin as soon as teeth contact each other. If the child is at increased risk for early caries, a small amount of toothpaste can be used beginning in infancy. The AAPD recommends a "smear" of fluoridated toothpaste for children younger than 2 years of age and a "pea-sized" amount for children aged 2 years through 5 years.

Fluoride use has led to a substantial decrease in dental caries. It is effective both topically and orally, and home use, as well as professional application, has demonstrated benefit. Community water fluoridation, when natural water fluoride levels are low, is the most cost effective way to provide fluoride's preventive benefits. Home-administered daily oral fluoride supplements are beneficial in communities with low water fluoride content (Item C181). When that is not available, and as a supplement, fluoridated toothpaste has proven efficacy. Professionally applied topical fluorides in the form of gel, rinse, and varnish (most common) are safe and effective and should be applied twice yearly.

Item C181. American Academy of Pediatric Dentistry Dietary Fluoride Supplementation Schedule.

Age	Community Water Fluoride Concentration		
	< 0.3 ppm F	0.3 to 0.6 ppm F	>0.6 ppm F
Birth to 6 months	0	0	0
6 months to 3 years	0.25 mg	0	0
3 to 6 years	0.5 mg	0.25 mg	0
6 to at least 16 years	1 mg	0.5 mg	0

Courtesy of the American Academy of Pediatric Dentistry

PREP Pearls
- The first dental visit should occur within 6 months of first tooth eruption, but no later than 12 months of age.
- Pediatricians should provide dental anticipatory guidance to discuss diet, dental hygiene, and fluoride to promote good dental health.
- Topical and oral fluoride supplementation are effective in reducing dental caries.

ABP Content Specifications(s)
- Provide appropriate counseling with regard to fluoride use
- Provide appropriate counseling to parents with regard to dental care for their children

Suggested Readings
- American Academy of Pediatric Dentistry. Guideline on fluoride therapy. American Academy of Pediatric Dentistry website. http://www.aapd.org/media/Policies_Guidelines/G_fluoridetherapy.pdf.
- American Academy of Pediatric Dentistry. Guideline on periodicity of examination, preventive dental services, anticipatory guidance/counseling, and oral treatment for infants, children, and adolescents. American Academy of Pediatric Dentistry website. http://www.aapd.org/media/Policies_Guidelines/G_Periodicity.pdf.

Question 182

The local campaign to prevent teenage pregnancy is hosting its annual conference. The primary focus of this year's conference will be risks associated with teenage pregnancy and childbearing.

Of the following, the GREATEST risk associated with pregnancy in this age group is increased

A.   lifetime risk of breast cancer

B.   risk for large for gestational age infants

C.   risk of chromosomal abnormalities

D.   risk of gestational diabetes

E.   risk of placental previa

Question 182   **Preferred Response: E**

While adolescent birth rates have declined for the last few years, the United States continues to have higher adolescent pregnancy and birth rates than other similarly industrialized countries. Adolescent childbearing has been associated with negative consequences for both the adolescents and their children.

Pregnant adolescents have a higher risk of complications, including placenta previa, pregnancy-induced hypertension, and premature delivery. Additionally, adolescent mothers are less likely to graduate from school and are more likely to live in poverty. Children born to adolescent parents are more likely to have poorer health outcomes such as low birth weights and higher rates of infant mortality, as well as lower educational achievement compared with children born to nonadolescent parents.

Early and adequate prenatal care is important for the pregnant adolescent to try to ensure a healthy pregnancy and birth outcomes. Pregnant adolescents should receive resources to assist with smoking, alcohol, and drug cessation, if needed.

Adolescent childbearing has not been associated with an increased lifetime risk of breast cancer, gestational diabetes, or chromosomal abnormalities. In fact, younger age at the time of pregnancy has been associated with lower rates of these conditions.

PREP Pearls
- While adolescent birth rates have declined for the last few years, the United States continues to have higher adolescent pregnancy and birth rates than other similarly industrialized countries.
- Adolescent childbearing has been associated with negative consequences for both the adolescents and their children.
- Early and adequate prenatal care is important for the pregnant adolescent to try to ensure a healthy pregnancy and birth outcomes.
- Pregnant adolescents have a higher risk of complications including placenta previa, pregnancy-induced hypertension, premature delivery, and higher rates of infant mortality.

ABP Content Specifications(s)
- Recognize the age-related risks of pregnancy complications, including associated mortality, in adolescents
- Plan ways to improve the health and outcome of pregnant adolescents
- Understand the socioeconomic and educational problems associated with pregnancy during adolescence

Suggested Readings
- Elfenbein DS, Felice ME. Adolescent pregnancy. In: Kliegman RM, Stanton BF, St Geme JW III, Schor, NE, Behrman RE, eds. Nelson Textbook of Pediatrics. 20th ed. Philadelphia, PA: Saunders Elsevier; 2015:978-982.

Question 183

A 12-year-old boy with no significant medical history presents to your office with perineal pain, difficulty urinating, and hematuria. His symptoms began 14 hours earlier after he slipped and straddled his bicycle seat forcefully while attempting a jump off a retaining wall. At the time of this injury, the boy immediately felt pain in his perineum, but did not seek medical attention. His mother scheduled an appointment with you this morning after the boy reported he was having difficulty urinating and had seen blood in his urine.

The boy is in no acute distress, but appears uncomfortable. His vital signs include a temperature of 37.2°C, heart rate of 100 beats/min, respiratory rate of 14 breaths/min, and a blood pressure of 110/70 mm Hg. His physical examination reveals dried blood and a small amount of fresh blood at the urethral meatus, along with perineal bruising. There are no lacerations visible on your external genitourinary examination. Both of his testes are palpable and nontender, and his abdomen is soft and nontender. There is no tenderness or instability with palpation over his pelvic bones.

Based on your clinical findings, the MOST likely diagnosis for this boy is

A.   anterior urethral injury

B.   bladder laceration

C.   penile shaft fracture

D.   rhabdomyolysis

E.   testicular contusion

Question 183   **Preferred Response: A**

The boy in the vignette presents with perineal pain, difficulty urinating, and hematuria several hours after sustaining a straddle injury. Blood at the urethral meatus and perineal bruising are noted on his physical examination. Based on these clinical findings, the most likely diagnosis is an anterior urethral injury.

Pediatric providers should recognize the clinical findings associated with urethral trauma. The presence of blood at the urethral meatus, as noted in the boy in the vignette, is highly suggestive of a urethral injury. This finding has been reported in up to 90% of patients with anterior urethral injuries. Other clinical indicators of urethral injury may include voiding difficulty, scrotal hematoma or perineal ecchymoses, and a high-riding prostate on rectal examination in males. Urethral injuries may be associated with pelvic fractures. Posterior urethral injuries are typically seen in patients with multisystem trauma and pelvic fractures, whereas anterior urethral injuries typically result from a straddle impact and often occur as isolated injuries. Blunt trauma arising from motor vehicle accidents, direct falls onto the perineum, and straddle injuries are the causes of most pediatric urethral injuries. Penetrating injuries and injuries related to instrumentation of the urethra occur much less commonly.

In patients with suspected urethral injuries, a urinary catheter should never be inserted until retrograde urethrography is performed to assess whether the urethra is intact. Retrograde urethography is the gold standard study for the diagnosis of urethral injury.

Injuries to the bladder may result from blunt or penetrating trauma, with motor vehicle accidents being the most common mechanism in children. Most bladder injuries (>80%) are associated with pelvic fractures and penetration of the bladder by a bony fragment. Clinical findings associated with bladder laceration may include hematuria, dysuria, and pain in the lower abdomen/pelvis. In cases of complete bladder rupture, patients may develop peritonitis and a palpable fluid wave from leakage of urine into the peritoneal cavity. Although the boy in the vignette presents with hematuria, an anterior urethral injury would be much more likely to result from his mechanism of injury than a bladder laceration. In addition, he has no abdominal pain and no findings suggestive of a pelvic fracture on physical examination.

Penile shaft fractures arise from traumatic rupture of the corpus cavernosum. These injuries typically result from the traumatic impact of an erect penis against a hard surface. Patients with penile shaft fractures generally report hearing a cracking sound at the time of injury and present with pain, swelling, and deformity of the penile shaft. None of these findings is present in the boy in the vignette.

Classic signs and symptoms for patients with rhabdomyolysis include weakness, myalgias, red or brown discolored urine, and in some cases, difficulty with urination. Rhabdomyolysis may result from traumatic causes such as crush injuries and compartment syndrome, as well as nontraumatic causes such as excessive exertion, prolonged seizure activity, hypokalemia, metabolic myopathies, illicit and prescription drugs, and infections. Anterior urethral injury is a much more

plausible explanation for the hematuria affecting the boy in the vignette given his history and physical examination findings.

Scrotal trauma may occur as a result of straddle injuries, and injuries can range from minor scrotal/testicular contusions to complete testicular rupture (a surgical emergency). Testicular contusion may present with tenderness to palpation over the scrotum, scrotal edema, and ecchymosis. The boy in the vignette has no tenderness on palpation of his testicles and has blood at the urethral meatus that would not be explained by a testicular contusion.

PREP Pearls
- The presence of blood at the urethral meatus is highly suggestive of a urethral injury. Other clinical indicators may include voiding difficulty, scrotal hematoma or perineal ecchymoses, and displacement of the prostate on rectal examination in males.
- Retrograde urethrography is the gold standard study for the diagnosis of urethral injury.
- A urinary catheter should never be inserted in patients with suspected urethral injuries until retrograde urethrography is performed to assess for an intact urethra.

ABP Content Specifications(s)
- Recognize the clinical findings associated with urethral trauma

Suggested Readings
- American College of Surgeons Committee on Trauma. Abdominal and pelvic trauma. Advanced Trauma Life Support Student Course Manual. 9th ed. Chicago, IL: American College of Surgeons; 2012.

Question 184

You are caring for a 16-year-old adolescent boy with advanced progressive muscular dystrophy. He is currently intubated and sedated because of respiratory failure from pneumonia. This is his fourth episode over the past year. Prior to this event, given the progressive nature of his illness, his parents and primary care physician discussed end-of-life care with him. The patient indicated his desire for withdrawal of life-sustaining treatment if he were to develop respiratory failure again. Per his wishes, the endotracheal tube is removed. After 1 minute, he develops tachypnea and gasping respirations.

Of the following, the MOST appropriate next step is to

A.   administer morphine, 0.1 mg/kg intravenously

B.   administer morphine, 5 mg/kg intravenously

C.   administer vecuronium, 0.1 mg/kg intravenously

D.   provide nonpharmacologic comfort care

E.   start noninvasive positive pressure ventilation

Question 184   **Preferred Response: A**

The child in this vignette has a chronic, progressive neuromuscular disease and has undergone a terminal extubation according to his and his family's wishes. In this setting, therapies should be given with the purpose of alleviating symptoms. The most appropriate medication to treat the symptom of breathing difficulty is intravenous morphine, 0.1 mg/kg.

Palliative care is the practice of integrating medical, spiritual, and psychosocial aspects of care when making medical decisions based on quality of life for a patient in a family-centered manner. Palliative care should begin for all children with progressive medical conditions early enough in the course of disease so that rational decisions can be made with as little stress as possible. As the practice of palliative care expands, primary care physicians will likely play a more integral role. An important aspect of palliative care is defining the goals of end-of-life care for a terminally ill child. Because of repeated respiratory infections from his progressive neuromuscular condition, the child in the vignette prepared an advance directive indicating his desire for withdrawal of life-sustaining therapies in the case of a subsequent episode of respiratory failure. The principle of autonomy, or respect for persons, provides that an appropriately mature 16-year-old child should ideally determine important decisions such as end-of-life care. Although the decision-making power legally lies with his parents, every effort should be made to incorporate the child's wishes into such decisions.

Presumably, for the child in the vignette, open discussions occurred and decisions were appropriately made in a relatively low-stress setting. A "do-not-resuscitate" order may be defined in the outpatient setting, and family members can direct caregivers accordingly. It is not uncommon for families to initially choose mechanical ventilation for a child like the boy in the vignette, in case the cause of acute respiratory failure is quickly reversible. However, a chronically ill or weak child often does not then wean from mechanical support easily. Terminal extubation, which involves removal of an endotracheal tube from a ventilator-dependent patient, is a reasonable option in such cases. For the boy in the vignette, because terminal extubation has been chosen and the condition is not likely reversible, noninvasive positive pressure ventilation would not be appropriate.

In terminal extubation, the patient and family should be informed that it is uncertain how long it will take for the child to die. Opioids and/or benzodiazepines should be readily available, at doses appropriate for the intent to treat symptoms of distress, as opposed to intentionally causing or hastening death. Although these medications may in fact hasten death, the "doctrine of double effect" provides that if a medication has 2 effects, 1 positive (treating symptoms) and the other negative (hastening death), the intent of the positive effect can be honored. This distinction should also be made clear to the family. Although it is practiced in some countries, euthanasia is not widely accepted in the medical community.

For the boy in the vignette, administration of 5 mg/kg of intravenous morphine would be an intentional overdose and would represent euthanasia. Administration of vecuronium without adequate sedation is inhumane in any setting, and in this setting would also constitute euthanasia.

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Comfort care should also be provided, but pharmacologic measures to relieve gasping are more likely to be effective.

PREP Pearls
- It is ethically acceptable to treat symptoms of respiratory distress in a dying patient even if it may hasten death because of the "doctrine of double effect."
- Narcotics and benzodiazepines at standard doses are effective medications to treat the discomfort of respiratory distress in dying patients.
- Palliative care should be incorporated early in the care of children with chronic or progressive medical conditions, and ideally under minimally stressful conditions.

ABP Content Specifications(s)
- Recognize and apply ethical principles with regard to limitations on medical intervention
- Recognize and apply ethical principles involving cardiopulmonary resuscitation and "do not resuscitate" (DNR) orders
- Recognize and apply ethical principles when involved in decisions to withdraw/withhold artificial hydration/nutrition
- Recognize and apply ethical principles when involved in end-of-life care

Suggested Readings
- Kang TI, Munson D, Hwang J, Feudtner C. Integration of palliative care into the care of children with serious illness.Pediatr Rev. 2014;35:318. DOI: http://dx.doi.org/10.1542/pir.35-8-318.

Question 185

A 16-year-old adolescent presents to the office for evaluation of a sore throat after the start of school. She has been febrile and has discomfort with swallowing. Vital signs show a temperature of 37.8°C, respiratory rate of 16 breaths/min, heart rate of 88 beats/min, blood pressure of 117/65 mm Hg, and a weight of 54 kg. On physical examination, she has erythema, edema, and exudates of both tonsillar pillars, tender bilateral anterior cervical lymphadenopathy, and a scarlatiniform rash. Laboratory data shows:

- White blood cells, 12,500/μL (12.5 x 10^9/L)
- Hemoglobin, 11.5 g/dL (115 g/L)
- Platelets, 320 x 10^3/μL (320 x 10^9/L)
- Differential, 1% segmented neutrophils, 18% bands, 78% lymphocytes, 2% monocytes, 1% eosinophils
- Rapid "strep" test, negative

In this patient, the BEST means of distinguishing pharyngitis caused by Arcanobacterium haemolyticum from Streptococcus pyogenes is

A.   age of patient

B.   growth in routine culture

C.   pharyngeal exudates

D.   scarlatiniform rash

E.   time of year

Question 185   **Preferred Response: B**

While Arcanobacterium haemolyticum can grow on sheep blood agar, colonies are small, have narrow hemolysis, and can be missed by laboratory personnel. In contrast, growth of Streptococcus pyogenes is easily detected on routine culture media. Detection of A haemolyticum can be enhanced if rabbit or human blood agar is used. Therefore, the differential growth of S pyogenes compared to A haemolyticum using routine culture media is the best way to distinguish these 2 causes of pharyngitis.

It is not possible to distinguish pharyngitis caused by S pyogenes from that caused by A haemolyticum on clinical grounds. Fever, pharyngeal exudates, lymphadenopathy, and a scarlatiniform rash can be seen in both infections. A haemolyticum, like S pyogenes, can cause invasive infections, although invasive A haemolyticum infections are quite rare and tend to occur in immunocompromised patients. While A haemolyticum infections occur primarily in adolescents and young adults, individuals in this age group also can develop streptococcal pharyngitis, therefore, age alone cannot distinguish these infections. A haemolyticum infections can occur during any season of the year. While the treatment of choice for streptococcal pharyngitis is penicillin, erythromycin is the drug of choice for treating A haemolyticum pharyngitis.

PREP Pearls
- It is not possible to distinguish pharyngitis caused by Streptococcus pyogenes from that caused by Arcanobacterium haemolyticum on clinical grounds.
- While A haemolyticum can grow on the commonly used sheep blood agar, colonies are small, have narrow hemolysis, and can be missed by laboratory personnel.
- Erythromycin is the drug of choice for treating A haemolyticum pharyngitis.

ABP Content Specifications(s)
- Recognize the clinical features associated with Arcanobacterium haemolyticum infection

Suggested Readings
- American Academy of Pediatrics. Arcanobacterium haemolyticum infections. In: Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:246-247.

Question 186

You are evaluating a 15-year-old adolescent in the emergency department (ED) for severe headache. He has been complaining of headaches for the last year. You note 3 ED visits in the last 6 months for headaches. His evaluation includes normal blood urea nitrogen, creatinine, electrolytes, thyroid function tests, urinalysis, and head computed tomography. His vitals show a temperature of 37°C, heart rate of 120 beats/min, respiratory rate of 18 breaths/min, and a blood pressure of 154/96 mm Hg. You make note that on his previous ED visits, his heart rate has ranged 120 to 130 beats/min, and blood pressure has ranged from 149/84 to 166/92 mm Hg. His physical examination is only significant for sweaty palms. You discuss the diagnostic evaluation for this patient's signs and symptoms with your residents.

Of the following, the MOST appropriate next test for evaluating this patient is

A.   abdominal ultrasonography

B.   plasma fractionated metanephrines

C.   metaiodobenzylguanidine (MIBG) scan

D.   urinary total metanephrines

E.   urinary vanillylmandelic acid

Question 186   **Preferred Response: B**

Pheochromocytomas and paragangliomas are catecholamine-secreting neuroendocrine tumors. They arise from the chromaffin cells of the adrenal medulla (pheochromocytoma) or the sympathetic ganglia (paraganglioma or extra-adrenal pheochromocytoma). Pheochromocytomas are often diagnosed in patients with classical paroxysmal symptoms of headache, sweating, flushing, diarrhea, and palpitations, a positive history of familial disease, or as an incidental adrenal mass in patients getting abdominal imaging for other reasons. Pheochromocytomas are also reported in association with multiple endocrine neoplasia type 2, von Hippel-Lindau syndrome, neurofibromatosis type 1, and paraganglioma syndromes. Patients with clinical features and family history of these syndromes are periodically screened for pheochromocytomas and are usually asymptomatic at the time of diagnosis.

The patient in the vignette has hypertension, tachycardia, and sweating, which is suggestive of pheochromocytoma. His prior histories of recurrent emergency room visits are indicative of the paroxysmal nature of the symptoms. Subsequent laboratory evaluation in this patient should focus on identifying excess catecholamine secretion (plasma-fractionated metanephrines) as the underlying cause of the patient's symptoms.

The diagnostic approach to pheochromocytoma includes biochemical testing for excess catecholamine secretion and subsequent demonstration of tumor by imaging. In patients with pheochromocytomas, the excess catecholamines are converted to metanephrines and normetanephrine metabolites within the tumor by the chromaffin cells. Therefore, excess catecholamine can be measured as increased total or fractionated catecholamines (dopamine, norepinephrine, and epinephrine) or total and fractionated metanephrines (metanephrine and normetanephrine). Measurement of fractionated metanephrines in urine or plasma is considered the most sensitive biochemical test for evaluating patients suspected of having pheochromocytoma. Most laboratories measure fractionated catecholamines and metanephrines by high performance liquid chromatography or tandem mass spectroscopy to overcome the problems with false-positive or false-negative results from fluorometric analysis.

Plasma fractionated metanephrines have a high sensitivity (> 95%) and low specificity (85%-89%) for diagnosing pheochromocytoma. In patients with high pretest probability of pheochromocytoma in the presence of clinical symptoms or strong family history, plasma fractionated metanephrines is the initial diagnostic test. A 24-hour urine collection for fractionated catecholamines and metanephrines is reported to have the highest sensitivity (98%) and specificity (98%) for diagnosing pheochromocytoma. Urinary fractionated catecholamines and metanephrines should be the first test in patients with low pretest probability for pheochromocytoma (incidental adrenal mass without imaging characteristics consistent with pheochromocytoma). In younger children in whom obtaining a complete 24-hour urine collection is difficult, plasma fractionated metanephrines is the preferred initial test.

Total urinary metanephrines and vanillylmandelic acid on a spot sample have poor diagnostic sensitivity and specificity, compared with fractionated plasma metanephrines, and are not indicated as the next step for this patient.

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Positive biochemical confirmation of the catecholamine excess is subsequently followed by radiological localization of the tumor. The majority of the tumors (around 95%) are within the abdomen and pelvis. In symptomatic patients, abdominal and pelvic ultrasonography can be the initial imaging. Computed tomography (CT) and magnetic resonance imaging (MRI) with contrast are superior to ultrasonography for identifying and localizing the tumors. Iodinated contrast media can precipitate hypertensive crisis, and it is advisable to avoid using iodinated contrast without α-adrenergic blockade in patients with suspected pheochromocytoma. Recent studies have reported a low risk for hypertensive crisis with the currently used low-osmolar nonionic contrast for CT. Metaiodobenzylguanidine (MIBG) is a radioiodine-labeled compound analogous to norepinephrine, and taken up by the adrenal medullary tissue and frequently by pheochromocytomas and paragangliomas. The MIBG scan is indicated in patients with increased risk for multiple tumors (children with paragangliomas, familial endocrine syndromes) or if malignant lesions are suspected (large adrenal pheochromocytoma on CT scan or MRI).

PREP Pearls
- The classical symptoms associated with pheochromocytoma include paroxysmal headache, sweating, and tachycardia, associated with elevated blood pressures.
- Measurement of fractionated metanephrines in urine or plasma is considered the most sensitive biochemical test for evaluating patients suspected of having pheochromocytoma.
- A 24-hour urine collection for fractionated catecholamines and metanephrines has the highest sensitivity and specificity for diagnosing pheochromocytoma.
- Computed tomography and magnetic resonance imaging with contrast are preferred to ultrasonography for identifying and localizing pheochromocytomas.

ABP Content Specifications(s)
- Recognize the clinical findings associated with pheochromocytoma
- Recognize disorders commonly associated with pheochromocytoma

Suggested Readings
- Edmonds S, Fein DM, Gurtman A. Pheochromocytoma. Pediatr Rev. 2011;32(7):308-310. DOI:http://dx.doi.org/10.1542/pir.32-7-308

Question 187

A 4-year-old boy is brought to the emergency department by his father after he found an empty bottle of acetaminophen in his bedroom. His father is uncertain how many pills were in the bottle. En route to the hospital, the child began having nonbloody and nonbilious emesis. His father reports no chronic medical issues and no ill contacts. He is admitted for monitoring and management. Physical examination 24 hours after admission showed a mildly jaundiced child. The liver edge is palpable 2 cm below the right costal margin. Laboratory results obtained at admission and at 12, 24 and 48 hours after admission are shown in Item Q187.

Laboratory test	Admission	12 hours after admission	24 hours after admission	48 hours after admission
Aspartate amino-transferase (U/L)	40	72	430	5000
Alanine amino-transferase (U/L)	26	55	400	6000
Bilirubin (mg/dL)	0	1	1	2
International normalized ratio	1.0	1.0	1.5	2.2
Fibrinogen (mg/dL)	400	420	440	480
Ammonia (µg/dL)	20	24	30	32
Glucose (mg/dL)	80	95	75	65

Which of the following 24-hour follow-up laboratory finding is MOST consistent with impending liver failure in this child?

A.   serum ammonia

B.   blood glucose

C.   aspartate aminotransferase

D.   fibrinogen levels

E.   international normalized ratio

Question 187   **Preferred Response: E**

Acute liver failure (ALF) occurs because of diffuse damage to the hepatic parenchyma (hepatocytes), resulting in failure of both synthetic and metabolic function. Early in the course of ALF, there is often a flu-like prodrome followed by hepatomegaly, elevated liver transaminases, and then progressive coagulopathy. The coagulopathy does not respond to vitamin K. As hepatic injury progresses, the patient will experience hypoglycemia, cerebral edema, encephalopathy, and renal failure.

The Pediatric Liver Failure Study Group defines ALF in children as:
   1.   Biochemical evidence of liver injury
   2.   No history of chronic liver disease
   3.   Coagulopathy not corrected by vitamin K
   4.   International normalized ratio greater than 1.5 in a patient with hepatic encephalopathy or greater than 2.0 if no hepatic encephalopathy

Hepatic encephalopathy (HE) is difficult to assess in children, but is critical to the identification of pending acute liver failure. There is a scale to assist with identifying the stages of HE (Item C187).

Item C187. Hepatic Encephalopathy in Children Younger Than 4 Years of Age.

Stage	Clinical Signs	Reflexes	Neurologic Signs
Early (I and II)	Inconsolable crying, sleep reversal, inattention to task	Hyperreflexic	Untestable
Middle (III)	Somnolence, stupor, combativeness	Hyperreflexic	Untestable in most
Late (IV)	Comatose, arouse with painful stimuli (Iva) or no response (IVb)	Absent	Decerebrate or decorticate

Modified and reprinted with permission from Bucuvalas J, Yazigi N, Squires RH. Acute liver failure in children. *Clin Liver Dis*. 2006; 10:149-168

The etiology of ALF is varied and includes:
•   Toxin and medications
   •   Acetaminophen, anticonvulsants, isoniazid, amiodarone, mushrooms, and ecstasy
•   Metabolic

- Galactosemia, tyrosinemia, mitochondrial disease, neonatal hemochromatosis, fatty acid oxidation defects, urea cycle defects, hereditary fructose intolerance, and Wilson disease
- Immune
  - Autoimmune hepatitis, hematophagocytic lymphohistiocytosis
- Infection
  - Hepatitis A and E, herpes simplex virus, Epstein-Barr virus, Enterovirus, sepsis
- Ischemia
  - Shock, Budd-Chiari, Congenital heart disease
- Malignancy
- Idiopathic (50%)

The 4-year-old boy in the vignette has ALF caused by acetaminophen toxicity. Impending hepatic failure is identified by increasing coagulopathy. Patients with liver failure will have hypoglycemia caused by failure of synthesis and release. Ammonia levels are elevated associated with increased glutamine and poor clearance. Fibrinogen levels are more often low, associated with disseminated intravascular coagulation. Transaminases are elevated in this child because of the acetaminophen toxicity and associated hepatitis. Elevated transaminases are associated with inflammation and are not a good predictor of liver failure.

PREP Pearls
- Evidence of coagulopathy is a danger sign of impending acute liver failure.
- Acute liver failure is associated with hepatic encephalopathy, which is defined as having 4 stages.
- Acute liver failure is associated with hypoglycemia.
- The etiology of 50% of acute liver failure in children is idiopathic.

ABP Content Specifications(s)
- Recognize the signs and symptoms of impending hepatic failure

Suggested Readings
- Clemente MG, Schwarz K. Hepatitis: general principles. Pediatr Rev. 2011;32(8):333-340. DOI:http://dx.doi.org/10.1542/pir.32-8-333.

Question 188

A 9-month-old infant diagnosed with achondroplasia presents to your office for a health supervision visit. The parents also want to discuss recurrence risk of the achondroplasia for future pregnancies. Both parents have achondroplasia.

Of the following, recurrence risks for achondroplasia for future live offspring of these parents is BEST estimated at

A.   25%

B.   50%

C.   66%

D.   75%

E.   100%

Question 188   **Preferred Response: C**

Achondroplasia is the most common inherited skeletal dysplasia that results in disproportionate small stature with affected individuals having short arms and legs, macrocephaly, frontal bossing, and midfacial retrusion (Item C188A). Normal intelligence and life span are expected, though hypotonia in infancy can delay motor milestones. Craniocervical junction compression can increase the mortality risk in infancy. Hydrocephalus can be present at times, requiring a ventriculoperitoneal shunt to alleviate increased intracranial pressure in some affected patients. Achondroplasia is an autosomal dominant disorder due to FGFR3 gene mutations.

Item C188A. Child with achondroplasia. Courtesy of L. Parsley.

Approximately 80% of individuals with achondroplasia have parents with average stature with the mutation due to a de novo gene mutation unique to that individual. With a de novo gene mutation, the parents would have a low risk of having another affected child. Twenty percent of individuals with achondroplasia have at least 1 affected parent. An individual with achondroplasia whose reproductive partner has normal stature would have a 50% risk of having a child with achondroplasia (Item C188B).

Item C188B. Risk of an Individual Having Achondroplasia When One Parent is Affected and the Second Parent is of Normal Stature.

Key:

A = Achondroplasia dominant allele
a = Wild-type allele (normal)
RED shading = Affected offspring
GREEN shading = Unaffected offspring

Cross:
aa x Aa

	a	a
A	aA	aA
a	aa	aa

When both parents have achondroplasia, the risk to the offspring is 50% for having a child with achondroplasia, 25% of having a child with average stature and unaffected, and 25% for having a homozygous achondroplasia conception, which is a lethal condition and would not result in a live birth. Therefore, in the case vignette, given the 25% chance of an intrauterine lethal condition, it would be a 66% (or two-thirds) chance of having a liveborn neonate with achondroplasia (Item C188C).

Item C188C. Risk of Having an Affected Child When Both Parents Have Achondroplasia.

Key:

A = Achondroplasia dominant allele
a = Wild-type allele (normal)
GREEN shading = Affected offspring
BLUE shading = Unaffected offspring
RED shading = Lethal condition

Cross:
Aa x Aa

	A	a
A	AA	aA
a	Aa	aa

PREP Pearls
- Achondroplasia, the most common skeletal dysplasia resulting in disproportionate small stature, is an autosomal dominant condition caused by FGFR3 gene mutation.
- Individuals affected with achondroplasia have short arms and legs, macrocephaly, frontal bossing, and midfacial retrusion. Normal intelligence and life span are expected, though hypotonia in infancy can delay motor milestones.
- When both parents have achondroplasia, the risk to the offspring is 50% for having a child with achondroplasia, 25% of having a child with average stature and unaffected, and 25% for having a homozygous achondroplasia conception, which is a lethal condition and would not result in a live birth.

ABP Content Specifications(s)
- Recognize the inheritance pattern of achondroplasia

Suggested Readings
- Pauli RM. Achondroplasia. GeneReviews.http://www.ncbi.nlm.nih.gov/books/NBK1152/#achondroplasia.Genetic_Counseling.
- Trotter TL, Hall JG. Health supervision for children with achondroplasia. Pediatrics. 2005;116(3):771-783. DOI:http://dx.doi.org/10.1542/peds.2005-1440.

Question 189

This is the first health supervision visit for a healthy 12-month-old girl to your practice. When you ask for a copy of the infant's immunization record, the mother becomes anxious and states that she doesn't believe in immunizations. She adds that she has a niece with autism, so she has no intention of allowing her own children to receive vaccines.

Of the following, the BEST approach to address this issue is to

A.   administer the vaccines that are overdue today

B.   discharge the patient from your practice

C.   label the patient's record to avoid future discussions of this topic

D.   notify child protective services due to concerns of medical neglect

E.   provide information about the risks and benefits of each vaccine

Question 189   **Preferred Response: E**

Parental concerns about vaccine safety are common and every practitioner must be comfortable addressing these questions. All patients and their parents should be informed about the risks and benefits of each vaccine, including those who voice hesitancy like the mother in the vignette. In addition, federal law mandates the provision of the vaccine information statements before vaccine administration.

Vaccine hesitancy is associated with perceived risk. As vaccine-preventable diseases become less common and media attention to claims about vaccine safety spreads, the risk-benefit perception for an individual patient has changed. It is important to use some simple strategies when communicating with vaccine-hesitant families. The practitioner should partner with family members in decision-making on behalf of the child, offering the opportunity to ask questions, listening to and acknowledging concerns in a nonconfrontational manner, clarifying and reaffirming accurate beliefs about immunizations, and correcting any misconceptions. Information should be personalized. It is helpful to take a positive approach, focusing on the number of lives saved by immunizations and explaining that vaccines benefit both individual children and communities through herd immunity. The practitioner should provide the vaccine information statement for each vaccine at every immunization visit and document discussions about the benefits and potential for adverse events, revisiting the immunization discussion at subsequent appointments. Parents generally view their pediatric health care provider as an important source of information, so ongoing open discussions may successfully assuage their vaccine concerns.

Despite one's best efforts to educate patients and their families about the effectiveness of vaccines, the incidence of adverse events related to vaccines, and the potential for true morbidity and mortality associated with natural disease, some will decline vaccination. When this occurs, it is recommended that the practitioner document the discussion, and request that the parents sign a waiver affirming their decision not to vaccinate. The American Academy of Pediatrics (AAP) has made available a "Refusal to Vaccinate" form which can be placed in the patient's medical record.http://www2.aap.org/immunization/pediatricians/refusaltovaccinate.html

Vaccines may not be administered without consent. Parents have the right to make informed decisions for their children, so refusal to vaccinate is not considered medical neglect in healthy children and should not be reported to child protective services. In general, it is recommended that physicians continue to care for the patient and family. However, if the practitioner is too uncomfortable to continue care, there is still a legal and ethical obligation to not abandon the patient. The family must be given reasonable notice of the intent to terminate the relationship and ample opportunity to arrange for alternative medical care.

PREP Pearls
- Practitioners should listen to concerns about immunizations, clarify and reaffirm accurate beliefs, and correct any misconceptions.
- Open and ongoing discussions regarding vaccine concerns should be held in an effort to partner with families and encourage vaccination.

- Parents have the right to refuse to vaccinate their children, in which case it is recommended that the provider request the parents to sign a waiver.

ABP Content Specifications(s)
- Plan an appropriate approach to addressing the needs of the vaccine-hesitant family

Suggested Readings
- American Academy of Pediatrics. Clinical report: reaffirmation: responding to parents who refuse immunization for their children. Pediatrics. 2013;131(5):e1696. DOI: http://dx.doi.org/10.1542/peds.2013-043

Question 190

A 14-month-old boy whose parents recently emigrated from India presents to your office for a health supervision visit. The boy is at the third percentile for weight and for height. He is pale and his liver is palpable 3 cm below the costal margin. His parents bring with them laboratory results from India that are shown in Q190.

Test	Result
White blood cell count	11,000/µL (11 x 10⁹/L)
Hemoglobin	5.2 g/dL (52 g/L)
Mean corpuscular volume	59 fL
Hemoglobin A	0%
Hemoglobin A2	21%
Hemoglobin F	79%

If managed appropriately, this patient's disease course is MOST likely to be complicated by

A.   acute chest syndrome

B.   iron deficiency anemia

C.   iron overload

D.   stroke

E.   ulcers of the lower legs

Question 190   **Preferred Response: C**

The size of red blood cells (RBC), as measured by the mean corpuscular volume (MCV), is in large part determined by the content of hemoglobin within the RBC. Any deficiency in the components of hemoglobin will, therefore, result in a low MCV. The 2 primary components of hemoglobin that can be deficient are iron or the globin protein. Hemoglobin A, the normal adult variant, consists of 2 β-globin chains and 2 α-globin chains, with the β-globin gene located on chromosome 11 and the α-globin gene on chromosome 16. Mutations resulting in reduced production of either α-globin or β-globin result in various thalassemia phenotypes and present with a microcytic anemia. The child in the vignette has a severe microcytic anemia and his hemoglobin electrophoresis pattern shows the absence of hemoglobin A (α2 β2), with only hemoglobin A2 (α2 δ2) and F (α2 γ2) present. This means that he has 2 dysfunctional β-globin genes, and therefore has β-thalassemia major. The most appropriate management of thalassemia major is chronic blood transfusions, typically every 3 to 4 weeks, to maintain a hemoglobin greater than 10 g/dL (100 g/L). With each transfusion of packed red blood cells (PRBC) comes a large iron load (each mL of PRBC delivers 0.75 mg of iron). The human body has no mechanism for eliminating excess iron, so iron accumulates with each transfusion. Patients with thalassemia who are treated with frequent transfusions are therefore at high risk for complications associated with iron overload.

The complications associated with chronic iron overload include endocrinopathies such as hypothyroidism, diabetes, hypogonadism, cardiomyopathy, and liver failure. Iron overload can be managed or even prevented through the use of aggressive chelation therapy. The most commonly used chelator is deferasirox, an oral, once a day medication. Some patients are unable to be adequately chelated with deferasirox and require the subcutaneous or intravenous administration of deferoxamine to maintain iron balance. The gold standard for assessing iron overload is liver biopsy, although newer techniques using specially calibrated magnetic resonance imaging are increasingly used. Although iron overload can be clinically monitored through measurement of serum ferritin, the serum ferritin level may be affected by inflammation and can vary widely. Magnetic resonance imaging of the liver and heart, calibrated with the appropriate programing (T2*), can be used for noninvasive monitoring and quantification of iron overload in centers where this technology is available.

Acute chest syndrome and stroke are complications of sickle cell disease and do not occur in β-thalassemia major. Iron overload is the complication associated with frequent transfusions of PRBC, not iron deficiency. Leg ulcers are a complication of diabetes mellitus or sickle cell anemia, not thalassemia major.

PREP Pearls
- The absence of hemoglobin A (α2 ß2) on hemoglobin electrophoresis with only hemoglobin A2 (α2 δ2) and F (α2 γ2) present is diagnostic of ß-thalassemia major.
- Thalassemia major is managed with chronic red blood cell transfusions, and the resultant iron overload is the most common complication of its therapy.
- Iron overload causes endocrinopathies, hepatic dysfunction, and cardiomyopathies.

- Complications of iron overload and death and can be prevented through aggressive chelation.

ABP Content Specifications(s)
- Recognize the clinical and laboratory findings associated with thalassemia major
- Plan the appropriate diagnostic evaluation of suspected thalassemia

Suggested Readings
- Bell MD. Consultation with the specialist: red blood cell transfusions. Pediatr Rev. 2007;28(8):299-304. DOI:http://dx.doi.org/10.1542/pir.28-8-299.

Question 191

You are seeing an 11-year-old boy with a complaint of intermittent vision loss. He says that his vision loss occurs suddenly in both eyes, lasts for a few minutes, and then resolves spontaneously. He saw an optometrist 2 weeks ago and the family was told that there appeared to be nothing wrong. He has no other physical complaints, including no headaches. His parents note that there has been a significant amount of stress in the home recently. He often helps to care for an ailing grandparent who is living with the family. Although his mother seems worried about his complaints, the boy himself seems unconcerned. He has missed about 3 weeks of school over the last 2 months because of this complaint. His parents both have a history of chronic medical symptoms that limit their ability to work.

Of the following, the MOST appropriate next step would be to

A.   order a brain magnetic resonance imaging without contrast

B.   perform a detailed neurologic examination

C.   refer for family counseling

D.   refer the boy to psychiatry

E.   send the patient for a lumbar puncture

Question 191   **Preferred Response: B**

This intermittent and brief vision loss experienced by the boy in this vignette is most likely a psychosomatic or other disorder for which no organic origin will be found (conversion disorder or malingering). Sudden vision loss in both eyes lasting for a few minutes followed by a sudden return of full vision without any other associated findings does not suggest an organic disorder. Another clue to the non-organic origin of the complaint is the patient's relative lack of concern about the symptom, which most people would find fairly distressing. In patients with conversion disorders, this relative lack of concern is sometimes referred to as "la belle indifference." We may theorize secondary gain, as his visual symptoms keep him home, where he can help care for his ailing grandmother.

Despite the very low likelihood of finding any biological abnormalities on physical examination, the best next step in care would be to perform a physical examination complete with extra attention to the neurological system. Performing this examination serves to show that the boy has been noticed and taken seriously, and shows the family that you are not brushing them off. Once a provider has performed a thorough physical examination and can reassure the family that no physical abnormalities have been found, psychosocial issues can be addressed further.

Delivering an "it's all in your head" message is usually counterproductive, as it may lead patients or families to insist on more testing and more specialists to prove they were right.
Instead, the principles of treatment are to:
1.   provide reassurance that the child appears healthy, as you cannot find evidence of anything serious or permanent going wrong with his body
2.   create a positive expectation that the symptom will improve in time on its own, that things like "stress" can cause unusual symptoms in the body and that these symptoms are time-limited
3.   address any stress in his life either through household changes, psychological therapy, or both
4.   discourage any situations that lead to secondary gain from his symptomatic complaints

You would like this family to get the message that despite what he may still intermittently say he experiences that his life should go on as normal.

Ordering a brain magnetic resonance image, while not physically harmful, would be very expensive and is medically unnecessary. Performing a lumbar puncture is an invasive and stressful procedure, for which there is no clinical indication in this case. Referring the family for family counseling might be an appropriate option if family dysfunction is discovered to be a core problem. As family dysfunction has not been revealed to be the cause of his problems, a family therapy recommendation is unlikely to be well received or pursued by the family at this time. Referring him to psychiatry for having a conversion disorder without first performing a physical examination is likely to be counterproductive. This is because the family may get stuck in a "the doctor didn't believe me/take me seriously" situation. After the medical evaluation has been performed, followed by the delivery of an assessment and recommendations, the family may

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decide that they would like help in managing the patient's anxious or stressful adjustment reactions and then can be referred to psychiatry.

PREP Pearls
•   It is important not to label obvious conversion disorder symptoms as psychiatric without first performing a thorough physical examination and making an interpersonal connection about taking their complaints seriously.
•   Medical and laboratory tests performed in the setting of a conversion disorder are monetarily wasteful and do not provide any lasting reassurance to the patient or family.
•   Extensive testing may promote, rather than alleviate, anxiety in the patient and family.

ABP Content Specifications(s)
•   Recognize the various features associated with conversion disorders
•   Identify the various features associated with psychosomatic disorders
•   Formulate an appropriate differential diagnosis of conversion symptoms
•   Plan an appropriate evaluation of psychosomatic disorders

Suggested Readings
•   Mink JW. Conversion disorder and mass psychogenic illness in child neurology. Ann NY Acad Sci. 2013;1304:40-44. DOI:http://dx.doi.org/10.1111/nyas.12298.

Question 192

A 5-year-old boy is admitted to the hospital with a 2-week history of intermittent fever. He has been having trouble walking today and is complaining of pain in his left knee. He is alert and well perfused. His heart rate is 120 beats/min, respiratory rate is 18 breaths/min, and blood pressure is 100/65 mm Hg. On physical examination, you note that he has conjunctival petechiae, but no conjunctivitis or rhinorrhea. He has small, nontender lymph nodes in the anterior cervical chain; he has no jugular venous distension. His neck is supple. His oropharynx is clear, his chest is clear, his cardiac examination shows a regular rate and rhythm, and a normal S1 and S2 with a 2/6 systolic murmur at the left mid-axillary line and the fourth intercostal space. The abdominal examination shows a palpable spleen, but no hepatomegaly. The femoral pulses are 2+. His left knee is swollen, erythematous, and warm to the touch. He is not able to straighten his leg. He has not had any medications.

His laboratory workup shows:

White blood cell count is 10,500/µL (10.5 x 10^9/L), 80% neutrophils, 18% lymphocytes, 2% monocytes
Hemoglobin, 13 g/dL (130 g/L)
Hematocrit, 39%
Platelet count, 390 x 10^3/µL (390 x 10^9/L)
Erythrocyte sedimentation rate, 120 mm/h

Of the following, the MOST important diagnostic test that should be performed to evaluate the etiology of this patient's symptoms is

- A.   abdominal computed tomography
- B.   blood cultures
- C.   C-reactive protein
- D.   chest radiograph
- E.   knee magnetic resonance image

Question 192   **Preferred Response: B**

The child in this vignette most likely has infective endocarditis (IE). All of the suggested diagnostic tests may be useful for this child, but the blood culture is the most important to do first to establish the etiology of the child's illness. Blood cultures are more sensitive in making a diagnosis of IE than an echocardiogram. A C-reactive protein is an inflammatory marker, but not specific to IE. An echocardiogram, especially a transthoracic study, may be negative even when there is IE. The chest radiograph may be useful to look for evidence of infected pulmonary emboli and the resultant multiple infarcts, but would not establish the etiology of the infection. Magnetic resonance imaging of the knee may confirm the knee as a source of initial infection, but does not confirm the additional diagnosis of IE. Abdominal computed tomography may show the enlarged spleen but will not confirm IE.

The Duke criteria for IE are shown in Item C192A and Item C192B. Clinical signs of IE include fever, splenomegaly without hepatomegaly, conjunctival hemorrhages, a new murmur in the mitral position, retinal and splinter hemorrhages, Osler nodes (tender lesions on the pads of the fingers and toes), and Janeway lesions. The child in this vignette does not have signs of congestive heart failure or a murmur to suggest severe valvular regurgitation. His laboratory work is significant for leukocytosis and elevated erythrocyte sedimentation rate.

Item C192A. Definition of Infective Endocarditis According to the Proposed Modified Duke Criteria, With Modifications Shown in Boldface.

Definite infective endocarditis

Pathologic criteria

(1) Microorganisms demonstrated by culture or histologic examination of a vegetation, a vegetation that has embolized, or an intracardiac abscess specimen; or

(2) Pathologic lesions; vegetation or intracardiac abscess confirmed by histologic examination showing active endocarditis

Clinical criteria*

(1) 2 major criteria; or

(2) 1 major criterion and 3 minor criteria; or

(3) 5 minor criteria

Possible infective endocarditis

(1) **1 major criterion and 1 minor criterion**; or

(2) **3 minor criteria**

Rejected

(1) Firm alternate diagnosis explaining evidence of infective endocarditis; or

(2) Resolution of infective endocarditis syndrome with antibiotic therapy for ≤4 days; or

(3) No pathologic evidence of infective endocarditis at surgery or autopsy, with antibiotic therapy for ≤4 days; or

(4) Does not meet criteria for possible infective endocarditis, as above

*See Table **C192B** for definitions of major and minor criteria.

Item C192B. Definition of Major and Minor Criteria for the Diagnosis of Infective Endocarditis (Duke Criteria With Modifications Shown in Boldface.

Major criteria

Blood culture positive for infective endocarditis (IE)

Typical microorganisms consistent with IE from 2 separate blood cultures:

Viridans streptococci, *Streptococcus bovis*, HACEK group, *Staphylococcus aureus*; or

Community-acquired enterococci, in the absence of a primary focus; or

Microorganisms consistent with IE from persistently positive blood culutres, defined as follows:

At least 2 positive cultures of blood samples drawn >12 hours apart; or

All of 3 or a majority of ≥4 separate cultures of blood (with first and last sample drawn at least 1 hour apart)

Single positive blood culture for *Coxiella burnetii* or antiphase I IgG antibody titer >1 : 800

Evidence of endocardial involvement

Echocardiogram positive for IE **(TEE recommended in patients with prosthetic valves, rated at least "possible IE" by clinical criteria, or complicated IE [paravalvular abscess]; TTE as first test in other patients)**, defined as follows:

Oscillating intracardiac mass on valve or supporting structures, in the path of regurgitant jets, or on implanted material in the absence of an alternative anatomic explanation; or

Abscess; or

New partial dehiscence of prosthetic valve

New valvular regurgitation (worsening or changing of pre-existing murmur not sufficient)

Minor criteria

Predisposition, predisposing heart condition or injection drug use

Fever, temperature >38°C

Vascular phenomena, major arterial emboli, septic pulmonary infarcts, mycotic aneurysm, intracranial hemorrhage, conjunctival hemorrhages, and Janeway's lesions

Immunologic phenomena: glomerulonephritis, Osler's nodes, Roth's spots, and rheumatoid factor

Microbiological evidence: positive blood culture but does not meet a major criterion as noted above* or serological evidence of active infection with organism consistent with IE

Echocardiographic minor criteria eliminated

TEE, transesophageal echocardiography; TTE, transthoracic echocardiography

*Excludes single positive cultures for coagulase-negative staphylococci and organisms that do not cause endocarditis.

Reprinted with permission from Li JS, Sexton DJ, Mick N, et al. Proposed modification to Duke criteria of diagnosis of infective endocarditis. *Clin Infect Dis*. 2000;30(4):633-638

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The patient described in this vignette may have 1 major criterion, if the mitral regurgitation murmur is new, and 1 minor criterion, conjunctival hemorrhages, and would therefore be in the category of possible IE. Multiple blood cultures are very important in being able to make a firm diagnosis. The number of blood cultures needed and the volume of blood needed in each age group varies. In a large meta-analysis in the adult population (23,313 patients), 2 sets of 3 blood culture bottles (each 10 mL samples) done within 30 min was as effective as 3 sets of blood cultures. In this setting, 2 aerobic and 1 anaerobic cultures were found to increase the yield, and allowed for declaration of a positive blood culture when 2 were required to rule out a contaminant. The most frequent pathogens were Staphylococcus aureus, Escherichia coli, Klebsiella pneumoniae, Enterococcus, and coagulase-negative Staphylococcus. This volume of blood is not realistic in younger pediatric patients.

Recommendations for infants and children have been made based on age and weight. Priority is placed on aerobic cultures in the smaller infants:

For premature infants less than 1 kg, 1 blood culture with 2 mL of blood
For infants between 1.1 kg and 2 kg, 2 sets of blood cultures each 2 mL
For infants 2.1 to 12.7 kg, 1 set of 4 mL and 1 set of 2 mL
For children 12.8 to 36.3 kg, 2 sets of 10 mL each
For children over 36.3 kg, 2 sets of 20 to 30 mL, each which corresponds to the adult recommendations

Echocardiography is an important tool in characterizing IE and in determining whether or not a patient may need surgical intervention. In the adult literature, up to 50% of patients require surgical intervention in the acute phase of IE, especially if the vegetation is large, growing, or associated with brain emboli or valve dehiscence (in the case of prosthetic valves) and accompanied by refractory congestive heart failure. Infectious disease consultation is very helpful to plan the acute and long term antibiotic management of patients who are managed medically.

In summary, clinical findings are a crucial part of diagnosis in IE. Patients at high risk deserve close scrutiny for physical signs of endocarditis, especially changes in their cardiac examination such as a new regurgitant murmur in the setting of febrile illness. The mortality for infective endocarditis is significant. Multiple blood cultures with adequate volume of blood are more sensitive in making the diagnosis than an echocardiogram. The echocardiogram will be crucial in the decision to obtain surgical consultation.

PREP Pearls
- Blood cultures are more sensitive in making the diagnosis of endocarditis than the echocardiogram.
- A new regurgitant murmur, when present, is the most significant physical finding in endocarditis.

ABP Content Specifications(s)

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- Recognize the clinical findings associated with infective endocarditis and provide appropriate initial management

Suggested Readings
- Baron EJ, Miller JM, Weinstein MP, et al. A guide to utilization of the microbiology laboratory for diagnosis of infectious diseases:2013 recommendations by the Infectious Diseases Society of America (IDSA) and the American Society for Microbiology (ASM)(a). Clin Infect Dis. 2013;57(4):e22-e121. DOI: http://dx.doi.org/10.1093/cid/cit278.
- Gerwitz M, Tawburt K. Infective endocarditis and prevention. In: Allen HD, Driscoll DJ, Shaddy RE, Feltes TF, eds. Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. 8th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2012:1363-1376.
- Li JS, Sexton DJ, Mick N, et al. Proposed modification to Duke criteria of diagnosis of infective endocarditis. Clin Infect Dis. 2000;30(4):633-638. DOI: http://dx.doi.org/10.1086/313753.
- Patel R, Vetter EA, Harmisen WS, Schleck CD, Fadel HJ, Cockerill FR III. Optimized pathogen detection with 30 compared to 20-milliliter blood culture draws. J Clin Microbiol. 2011;49(12):4047-4051. DOI: http://dx.doi.org/10.1128/JCM.01314-11.
- Prendergast B, Tornes P. Valvular heart disease: changing concepts in disease management: surgery for infective endocarditis: who and when? Circulation. 2010;121(9):1141-1152. DOI: http://dx.doi.org/10.1161/CIRCULATIONAHA.108.773598.

Question 193

You are seeing an 8-year-old boy in your office for a health supervision visit. The child has been developing normally. The mother mentions that he bruises easily, but has no other concerns. On physical examination, the boy has smooth, velvety skin with several small bruises of various ages. There are no petechiae. You are able to passively dorsiflex his fifth fingers past 90 degrees, flex his thumbs to the forearms, and hyperextend his knees and elbows past 10 degrees. The boy is able to place his palms flat on the floor with his knees fully extended.

Of the following, the clinical characteristic that would BEST support your suspected diagnosis is

A.   cigarette paper scarring

B.   long bone overgrowth

C.   occipital horns

D.   proximal muscle contractures

E.   skin laxity

Question 193   **Preferred Response: A**

The boy in the vignette has Ehlers-Danlos syndrome (EDS) characterized by a Beighton joint hypermobility score of 9 signifying generalized joint hypermobility and smooth velvety skin. The Beighton score is a simple system to quantify joint laxity and hypermobility. A score of 5 or higher out of a possible 9 points indicates joint hypermobility. One point is assigned for each side with the following findings: passive dorsiflexion of 5th finger >90 degrees, passive flexion of thumbs to the forearms, hyperextension of the elbows beyond 10 degrees, and hyperextension of the knees beyond 10 degrees. One additional point is given for the ability to rest the palms on the floor with forward flexion of the trunk with knees fully extended. In EDS, the skin is fragile, especially over pressure points. Patients with EDS have delayed wound healing which leads to widened and atrophic scars known as cigarette paper scars. There are 6 subtypes of EDS (Item C193).

Item C193. Ehlers-Danlos Subtypes.

Classic	• autosomal dominant • skin hyperextensibilty • atrophic scarring • smooth velvety skin • joint hypermobility
Classic-like	• autosomal recessive • skin hyperextensibilty • atrophic scarring • smooth velvety skin • joint hypermobility
Hypermobility type	• generalized joint hypermobility • mild skin involvement • joint dislocations • chronic joint pain
Vascular type	• excessive bruising • thin skin with prominent vascular pattern • skin is not hyperextensible • arterial and intestinal fragility and rupture • characteristic facies • can have acrogeria (reduction or loss of subcutaneous fat and collagen of the hands and feet), hypermobility of small joints, tendon and muscle rupture, and pneumothorax
Vascular-like	• skin hyperextensibility • scarring • easy bruising • arterial rupture • osteopenia • thin skin • joint hypermobility
Cardiac valvular	• autosomal recessive • severe cardiac valvular disease • skin hyperextensibility • scarring • easy bruising • blue sclerae • increased bone fragility

Modified and reprinted with permission from Callewaert B, Malfait F, Loeys B, De Paepe A. Ehlers-Danlos syndromes and Marfan syndrome. *Best Pract Res Clin Rheumatol.* 2008: 22 (1):165-189

Patients with EDS can have repetitive subluxations, sprains, and chronic joint pain. Muscle hypotonia may cause a delay in motor development. Easy bruisability and bleeding are associated with EDS. There are problems associated with fragile connective tissue such as poor wound healing, hernias, cervical insufficiency, and rectal prolapse. Pregnant EDS patients are considered high risk.

Preventive guidelines are available for patients with EDS including padding and protective bandages for patients with skin disease, and a recommendation for closure of wounds in 2 layers without tension. These patients may require longer periods for wound healing. If there is significant bruising, contact sports should be avoided. Ascorbic acid supplementation may decrease the bruising. If hypotonia, joint pain, or motor delay is present, physical therapy can be beneficial. Joint pain associated with EDS and hypermobility may be managed with nonsteroidal anti-inflammatory drugs (NSAIDs) and physical therapy, however, the use of NSAIDs may need to be limited in patients with significant bruising. For patients with joint hypermobility, contact sports and other sports that produce joint strain should be avoided, but other exercises such as swimming may be beneficial for muscle strengthening. Echocardiography is recommended before age 10 years and should be repeated periodically based on abnormalities found. Patients with vascular subtypes should avoid contact sports, weight lifting, drugs that interfere with platelet function, and invasive vascular surgery.

Long bone overgrowth is associated with Marfan syndrome. Occipital horns syndrome (OHS) is characterized by wedge-shaped calcifications of the trapezius and sternocleidomastoid muscle at the attachment to the occipital bone, lax skin, joint hypermobility, bladder diverticula, inguinal hernias, and vascular tortuosity. Distal joint hypermobility associated with proximal muscle contractures and muscle weakness is characteristic of Ullrich disease or scleroatonic muscular dystrophy. In Ehlers-Danlos syndrome, the skin is hyperextensible and will quickly return to its normal position when stretched. This is in contrast to cutis laxa and De Barsy syndromes, where redundant skin hangs and slowly returns to its normal state. In these syndromes wound healing is normal.

PREP Pearls
- Ehlers-Danlos syndrome has several subtypes, and typically presents with skin hyperextensibilty, atrophic scarring, smooth velvety skin, and joint hypermobility.
- Ehlers-Danlos syndrome patients should be counseled regarding wound healing and injury prevention.
- Echocardiography is recommended before age 10 years in patients with Ehlers-Danlos syndrome and should be repeated periodically based on abnormalities found

ABP Content Specifications(s)
- Recognize the clinical findings associated with Ehlers-Danlos syndrome

Suggested Readings

- Callewaert B, Malfait F, Loeys B, Paepe AD. Ehlers-Danlos syndromes and Marfan syndrome. Best Pract Res Clin Rheumatol. 2008;22(1):165-189. DOI: http://dx.doi.org/10.1016/j.berh.2007.12.005.

Question 194

A 15-year-old female gymnast presents to your office for a health supervision visit. The adolescent's mother is concerned that she has not been sleeping well. The girl's weight is at the fifth percentile and height is at the 50th percentile. Her physical examination is unremarkable. Her heart rate is 96 beats/min and her blood pressure is 128/76 mm Hg. You suspect that the adolescent is using a performance enhancing drug.

Of the following, the substance the adolescent is MOST likely using is

A.   androstenedione

B.   coenzyme Q

C.   erythropoietin

D.   methylphenidate

E.   phentermine

Question 194   Preferred Response: D

The gymnast in the vignette is using methylphenidate as a weight-loss agent. Although her heart rate is in the normal range, it is higher than expected for a well-trained athlete. For activities like gymnastics and dance, a lean physique is often seen as desirable and participants have an increased rate of stimulant use to promote weight loss and increase energy levels.

There is a common perception that athletes use performance-enhancing substances (PES) mainly to enhance muscle bulk. Participants in sports such as football, body building, and wrestling, where a muscular physique is perceived as advantageous, are likely to use anabolic steroids and other compounds that promote weight gain and increased muscle mass. However, there are many types of PES, and athletes may use medications and supplements for various reasons. Performance-enhancing substances are medications or supplements that improve performance in athletic activities or appearance. PES include medications taken for nontherapeutic purposes, or in excess of therapeutic dose, and medications that cause weight loss or gain or alter body composition. Supplements and vitamins, when used in excess of the recommended dose, and substances taken to enhance oxygen-carrying capacity are other examples of PES. In addition, some athletes use medications to prevent detection of another substance used to enhance performance or appearance; these medications are also considered PES. Studies of children and adolescents have demonstrated increased PES use over the past 2 decades. Increased youth participation in sports and increased competitiveness of youth sports leagues are likely factors influencing rates of PES use.

Participation in a sport that emphasizes a particular physique (eg, football, wrestling, or gymnastics) is a risk factor for PES use. Other risk factors include dissatisfaction with body type and exposure to media portrayals of an "ideal" shape. Male athletes are more likely to use substances such as creatine and anabolic steroids that are thought to promote a muscular physique.

Androstenedione is a testosterone precursor with mild androgenic effects, though this substance may have estrogenic effects in the presence of other, stronger androgens. Coenzyme Q is thought to improve energy level and exercise tolerance, but is not commonly used by athletes and does not have stimulant side effects. Abdominal discomfort is the most common side effect. Some athletes use erythropoietin (EPO) to enhance oxygen-carrying capacity. Use of EPO can cause hyperviscosity of the blood, which can lead to thrombosis, but would not cause the low weight or sleep disturbance seen in the patient in this vignette. Phentermine is a stimulant and would have effects that are similar to methylphenidate, but is much less commonly used by athletes.

PREP Pearls
- Participation in a sport that emphasizes a particular physique (eg, football, wrestling, or gymnastics) is a risk factor for performance-enhancing substance use.
- Athletes participating in sports in which a lean physique is seen as desirable have an increased rate of stimulant use to promote weight loss and to increase energy levels.

- Participants in sports in which a muscular physique is perceived as advantageous are more likely to use anabolic steroids and other compounds that promote weight gain and increased muscle mass.

ABP Content Specifications(s)
- Plan the appropriate evaluation when use of performance-enhancing drugs or nutritional supplements is suspected
- Recognize the clinical findings associated with the use of performance-enhancing drugs or nutritional supplements

Suggested Readings
- AAP Committee on Sports Medicine and Fitness. Use of performance-enhancing substances. Pediatrics. 2005;115(4):1103-1106. DOI: http://dx.doi.org/10.15

Question 195

A previously healthy 2-year-old boy presents to your office with several days of tactile fever and nasal congestion interfering with sleep, clear rhinorrhea, and cough. On physical examination, his temperature is 39°C, heart rate is 120 beats/min, and respiratory rate is 24 breaths/min. He is fussy, rubbing his eyes and ears, but is consolable. His voice is hoarse and his posterior oropharynx is erythematous without exudates. You note clear rhinorrhea and erythema of the nasal mucosa. The tympanic membranes are mildly hyperemic and retracted. There is shotty, bilateral cervical lymphadenopathy. Auscultation of the lungs reveals diffuse coarse upper airway sounds. The remainder of the physical examination is unremarkable.

Of the following, the MOST appropriate therapy to recommend for this child is

A.   acetaminophen

B.   dextromethorphan

C.   diphenhydramine

D.   guaifenesin

E.   pseudoephedrine

Question 195   **Preferred Response: A**

The boy in the vignette has a common cold as evidenced by several days of fever, cough, congestion, and rhinorrhea with an otherwise reassuring physical examination. Because of fever and fussiness, the most appropriate therapy to recommend for this child is acetaminophen. Both the Food and Drug Administration and the American Academy of Pediatrics advise that over-the-counter (OTC) cough and cold medications including antihistamines (diphenhydramine), decongestants (pseudoephedrine), antitussives (dextromethorphan), and expectorants (guaifenesin) should not be used in children younger than 2 years; consensus opinion extends this recommendation to children younger than 6 years.

Decongestants, OTC antihistamines, antitussives, and expectorants are most commonly sold in preparations containing multiple ingredients and often including antipyretics and analgesics. Although symptomatic relief is the goal, evidence of efficacy in children is lacking. In young children, the risk of adverse effects (eg, altered mental status, tachycardia, ataxia) is highest. There have been a significant number of events of accidental overdose due to the combination of ingredients in these OTC cough and cold products.

Viruses are the most common cause of upper respiratory tract infections in children (Item C195). Clinical findings may include cough, congestion, sneezing, rhinorrhea, and fever during the initial days of illness. The rhinorrhea may be clear or become yellowish-green within a few days. In adults, illness typically lasts 5 to 7 days. Symptoms in children with upper respiratory tract infection usually persist for at least 10 days, but lessen over time. The development of secondary bacterial infection (eg, acute bacterial sinusitis, otitis media, pneumonia) is suggested by the evolution or worsening of symptoms, especially fever, over several days.

Item C195. Common Causes of Upper Respiratory Tract Infections in Children.

Viruses	Bacteria (secondary infection)
Adenovirus	Group A *Streptococcus*
Coronaviruses	*Haemophilus influenzae*
Enteroviruses	*Moraxella catarrhalis*
Influenza virus	*Streptococcus pneumoniae*
Parainfluenza viruses	
Respiratory syncytial virus	
Rhinoviruses (>50%)	

Courtesy of D. Palazzi

PREP Pearls
- Over-the-counter cough and cold medications are of no benefit in young children with upper respiratory tract infection.
- Over-the-counter cough and cold preparations can cause serious adverse effects in young children.
- Viruses are the most common cause of upper respiratory tract infections in children.
- Rhinovirus causes most upper respiratory tract infections in children.
- Fever, cough, congestion, and rhinorrhea are the most common symptoms of upper respiratory tract infection in children.

ABP Content Specifications(s)
- Identify the etiology of an upper respiratory tract infection
- Understand the natural history of an upper respiratory tract infection

Suggested Readings
- American Academy of Pediatrics. American Academy of Pediatrics urges caution in the use of over-the-counter cough and cold medicines. http://www.generaterecords.net/PicGallery/AAP_CC.pdf.

Question 196

A 3-year-old, previously healthy girl presents to your office after she suddenly began crying and complaining to her mother of burning in her mouth and throat. Just before these symptoms began, the girl and her 4-year-old cousin were at home having a tea party with their dolls. The girls had decorated their tea party table with some leaves taken from a houseplant in the room (Item Q196).

Item Q196

Item 196. Houseplant. Reprinted with permission from the University of Illinois Extension.

On physical examination, the girl is nontoxic appearing and crying. She is afebrile and her vital signs are within normal limits for her age. Her lips are slightly swollen and she is intermittently drooling, although she was able to drink some water on the way to your office without difficulty. Her lungs are clear and she is breathing comfortably without stridor or signs of respiratory distress. Her skin is warm and dry without rash, and her extremities are well-perfused. The remainder of your physical examination shows no other abnormalities. The girl takes no regular medications and has no known allergies.

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

A.   administration of activated charcoal, orally

B.   administration of diphenhydramine, orally

C.   administration of epinephrine, intramuscularly

D.   administration of ibuprofen, orally

E.   consultation with a pediatric gastroenterologist for endoscopy

Question 196   **Preferred Response: D**

The girl in the vignette presents for management of acute mouth and throat pain that began after she was exposed to leaves from a philodendron plant. Although she has lip swelling and intermittent drooling, she displays no signs of significant airway compromise or respiratory difficulty. Oral administration of ibuprofen is the next best step in her management.

It is important for all pediatric providers to recognize the signs and symptoms that can arise from ingestion of a toxic plant, and to understand the principles underlying appropriate management of these ingestions. Children may be exposed to various potentially toxic plants, both within their home environments and outdoors. Although these exposures rarely result in serious toxicity, providers should be aware of the small number of plants that can have serious clinical effects (Item C196).

Item C196. Common Toxic Plants and Clinical Features of Exposure.

Plant	Toxic Part	Toxin	Clinical Features
Ricinus communis (castor bean)	All parts, especially seeds	Ricin	GI: Vomiting, diarrhea, severe dehydration
Cicuta (water hemlock)	All parts, especially root	Cicutoxin	GI: Nausea, vomiting CNS: Seizures
Digitalis purpurea (foxglove), *Nerium oleander* (oleander), *Convallaria majalis* (lily-of-the-valley)	All parts, especially seeds	Cardiac glycosides	GI: Nausea, vomiting CNS: Sedation Cardiac: PR prolongation, QT shortening, bradycardia, ventricular arrhythmias
Rhododendron, Kalemia (mountain laurel), *Pieris japonica*	All parts	Grayanotoxin	GI: Nausea, vomiting CNS: Sedation Cardiac: Bradycardia, atrioventricular block, sinus arrest, hypotension
Aconitum napellus (monkshood), *Delphinium* (larkspur)	All parts	Aconitine	CNS: Sedation, headache Cardiac: Ventricular tachycardia, torsade de pointes
Conium maculatum (poison hemlock), *Nicotiana* (tobacco)	All parts, especially seeds and roots	Nicotine, coniine (cholinergics)	GI: Salivation, nausea, vomiting CNS: Seizures, fasciculations, paralysis Cardiac: Bradycardia Other: Bronchospasm, diaphoresis
Datura (jimson weed), *Atropa belladonna* (nightshade)	All parts, especially flowers, seeds, fruit	Hyoscyamine, scopolamine (anticholinergics)	CNS: Agitation, hallucinations Cardiac: Tachycardia, hypertension
Dieffenbachia, Philodendron, Calacdium, rhubarb	All parts, especially leaves	Oxalates	GI: Oral mucosal irritation, esophagitis, gastritis, esophageal stricture

CNS=central nervous system, GI=gastrointestinal system

Courtesy of M. Wright

Plant exposures account for about 5% of calls to US poison control centers annually, with the vast majority affecting children younger than 6 years. Among small children, plant ingestions generally occur inadvertently as children innocently explore their environments. Older children and adolescents may ingest specific plants intentionally because of their psychoactive effects.

The specific plant causing symptoms for the girl in the vignette, philodendron, is a common houseplant that causes mucous membrane irritation because of the presence of tiny needle-shaped calcium oxalate bundles (raphides) throughout its structure. Other common houseplants containing calcium oxalate raphides include dumb cane (Dieffenbachia), caladium, calla lily (Zantedeschia), peace lily (Spathiphyllum), Jack-in the-pulpit (Arisaema atrorubens), and elephant ear (Colocasia esculenta). When chewed, the calcium oxalate crystals in these plants penetrate mucous membranes, causing immediate pain and inflammation, which is relatively mild in most cases. Recommended treatment for symptomatic children is generally supportive, and consists mainly of cleansing exposed skin and mucous membranes with water and/or mild soap, administration of analgesics, and oral administration of cold fluids, popsicles, or ice.

For children presenting with compromise of the airway, breathing, or circulation after exposure to any poisonous plant, advanced life support measures—including endotracheal intubation to secure the airway when indicated—are paramount. In the United States, regional poison control centers are invaluable resources that are available to clinicians at all times for guidance related to toxic exposures, including exposure to potentially toxic plants. Pediatric providers may contact their regional poison control center at 1-800-222-1222 for information related to signs and symptoms that may arise from exposure to specific plants, as well as for guidance in managing these exposures.

Administration of activated charcoal is generally not recommended for treatment of philodendron ingestion, because the symptoms of this ingestion are typically minor, and activated charcoal does little to alleviate them.

Oral diphenhydramine administration would not be the best initial step in management after philodendron ingestion, because the pain and mucosal irritation caused by this plant arises from microtrauma from calcium oxalate crystals and is not mediated by histamine. Diphenhydramine may help to relieve pruritus in patients who have had toxic plant exposures, but pain is the main symptom displayed by the girl in the vignette; oral analgesics would be the best next step in her management.

Intramuscular epinephrine would not be recommended for treatment of the girl in the vignette, because her symptoms are arising from mucosal irritation by calcium oxalate crystals in the philodendron and not from a systemic allergic reaction.
For children with mild symptoms of ingestion of philodendron and related plant species who can tolerate oral intake without difficulty, consultation with a gastroenterologist would not be indicated.

PREP Pearls
- Philodendron is a common houseplant that causes mucous membrane irritation because of the presence of tiny needle-shaped calcium oxalate bundles (raphides) throughout its structure.
- Pediatric providers should be aware of those plants that can have serious toxic effects.
- Recommended treatment for children who are symptomatic after philodendron ingestion is supportive, consisting of cleansing exposed skin and mucous membranes, administration of analgesics, and oral administration of cold fluids, popsicles, or ice.
- Regional poison control centers are available at all times for guidance related to toxic exposures, including exposures to potentially toxic plants.

ABP Content Specifications(s)
- Recognize the signs and symptoms following the ingestion of a potentially toxic plant, and manage appropriately

Suggested Readings
- Carlson AG, Krenzelo EP. Poisonous plants. In: Erickson TB, Ahrens WR, Aks SE, Baum CR, Ling LJ, eds. Pediatric Toxicology: Diagnosis and Management of the Poisoned Child. New York, NY: McGraw-Hill; 2005:541-547.
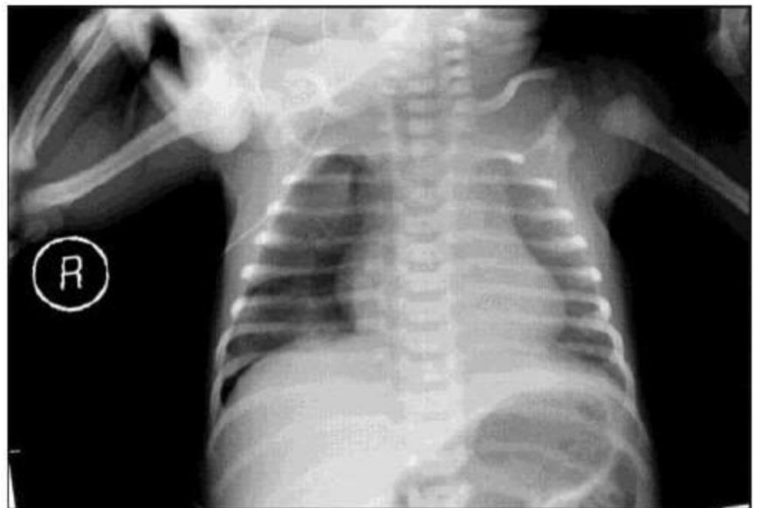
Question 197

You are asked to assess a newborn, 30 minutes after delivery, for evaluation of tachypnea. The newborn was delivered by vaginal delivery complicated by late decelerations and thick meconium-stained amniotic fluid. He was vigorous at birth, therefore direct suctioning of the trachea was not performed. He developed tachypnea and mild intermittent grunting 10 minutes after delivery, which has persisted. Vital signs include a temperature of 37.2°C, heart rate of 160 beats/min, respiratory rate of 80 breaths/min, blood pressure of 60/38 mm Hg, and oxygen saturation of 89% in room air. Physical examination is remarkable for subtle nasal flaring, slightly diminished breath sounds on the right, and meconium staining of the nails and umbilical cord. Arterial blood gas results show a pH of 7.21, PCO2 of 45 mm Hg, PO2 of 54 mm Hg, bicarbonate of 15 mEq/L (15 mmol/L), and base deficit of –8. A chest radiograph is obtained (Item Q197).

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

A.   administer sodium bicarbonate

B.   infuse a normal saline bolus

C.   initiate oxygen therapy via hood

D.   monitor cardiorespiratory status

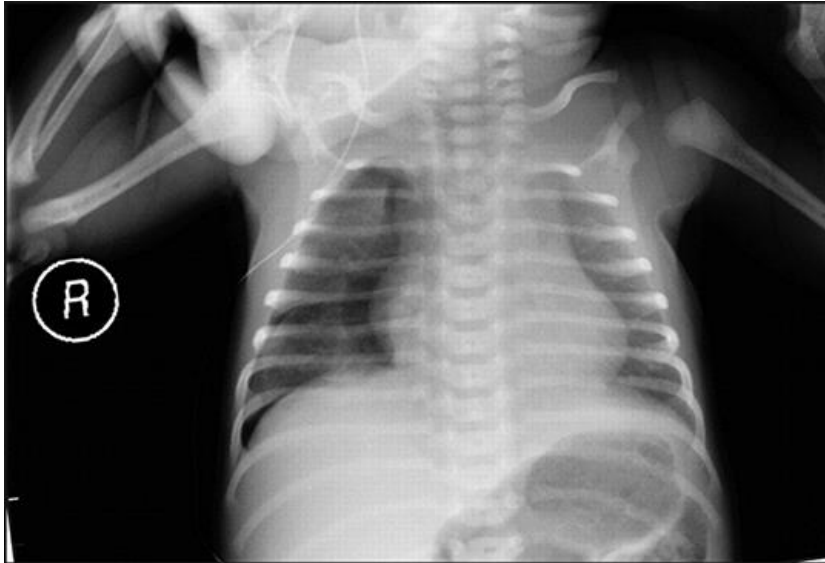E.   perform needle thoracentesis

Item Q197. Chest radiograph for the newborn described in the vignette. Reprinted with permission from Caldwell AL, Goodpasture M, Alden M. Index of suspicion in the nursery. NeoReviews. 2010;11(4):e206 -e210

Question 197   **Preferred Response: C**

The neonate in the vignette has the physical examination and radiographic findings of a right-sided pneumothorax (Item C197), with mild respiratory distress and oxygen saturations of 89% in room air. Oxygen therapy should be initiated via hood to maintain adequate oxygen saturation. A pneumothorax may occur when alveoli overdistend and rupture, releasing air into the pleural space between the lung and chest wall. A spontaneous pneumothorax can occur in up to 2% of full-term neonates with no lung disease, attributed to the high opening pressures generated by crying after delivery. The incidence of pneumothorax increases to 10% in meconium aspiration syndrome (MAS), in which meconium trapped in the airway acts as a ball valve. Air gets into, but not out of the alveoli, leading to overdistention and rupture. The risk of developing a pneumothorax is further increased by mechanical ventilation or continuous positive airway pressure superimposed on an underlying lung disease such as respiratory distress syndrome, pneumonia, or MAS.

Item C197

Item C197. Moderate-sized right pneumothorax on plain radiograph. Reprinted with permission from Caldwell AL, Goodpasture M, Alden M. Index of suspicion in the nursery. NeoReviews. 2010;11(4):e206 -e210.

Spontaneous pneumothorax in a healthy full-term neonate often goes undetected and resolves without intervention. Physical examination findings suggestive of a pneumothorax include tachypnea, grunting, and cyanosis with decreased breath sounds, chest wall asymmetry, and shifting of the point of maximal cardiac impulse. The diagnosis is confirmed on chest radiography, though transillumination may be performed in an emergent situation. Symptomatic neonates with pneumothorax should be monitored closely for cardiorespiratory compromise. If the neonate is hemodynamically stable, supportive therapy is appropriate. Nitrogen washout therapy with 100% oxygen had been recommended until recently, because of the belief that it would accelerate resolution. Newer studies have demonstrated that providing just enough supplemental oxygen to maintain adequate oxygen saturation is equally effective and does not carry the risks of hyperoxia.

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Evidence of a tension pneumothorax including poor perfusion, hypotension, bradycardia, increasing respiratory distress, or worsening metabolic acidosis requires immediate drainage of the pleural air by needle thoracentesis or chest tube placement. Needle thoracentesis may be the only intervention needed for a neonate who does not require mechanical ventilation. Studies have demonstrated that neonates receiving ventilation at low ventilator settings do not always require chest tube placement after needle aspiration and may be monitored closely for reaccumulation of the pneumothorax.

The infant in the vignette has no evidence of hypovolemia or hypotension that would require a normal saline bolus. Sodium bicarbonate is not indicated because the mild metabolic acidosis is likely the result of perinatal events and will resolve over time without treatment.

PREP Pearls
- A spontaneous pneumothorax can be found in up to 2% of full-term neonates with no lung disease.
- Physical examination findings suggestive of a pneumothorax include tachypnea, grunting, and cyanosis with decreased breath sounds, chest wall asymmetry, and shifting of the point of maximal cardiac impulse.
- Evidence of a tension pneumothorax including poor perfusion, hypotension, bradycardia, increasing respiratory distress, or worsening metabolic acidosis requires immediate drainage of the pleural air by needle thoracentesis or chest tube placement.

ABP Content Specifications(s)
- Recognize the characteristic clinical and radiographic appearance of pneumothorax in a newborn infant, and manage appropriately

Suggested Readings
- Clark SD, Saker F, Schneeberger MT, Park E, Sutton DW, Littner Y. Administration of 100% oxygen does not hasten resolution of symptomatic spontaneous pneumothorax in neonates. J Perinat. 2014;34:528-531. DOI:http://dx.doi.org/10.1038/jp.2014.55.

Question 198

A 3-year-old, previously healthy, fully immunized boy presents to the emergency department after he had a seizure at home. He had a fever that morning and nonbloody diarrhea. The boy's mother reports that she gave him acetaminophen and some juice. When she laid him down for a nap, his whole body stiffened and then started jerking rhythmically. This event lasted approximately 2 minutes. Afterwards, he was limp and unresponsive. She called 911 and he was brought by ambulance to the emergency department. On physical examination, his temperature is 38.9°C, blood pressure is 90/48 mm Hg, heart rate is 122 beats/min, and respiratory rate is 38 breaths/minute. He cries when you attempt to examine him, but is consoled by his mother. His general physical and neurological examination results are unremarkable.

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

A.   obtain an electroencephalogram

B.   obtain magnetic resonance imaging of the brain

C.   obtain serum electrolytes

D.   perform a lumbar puncture

E.   provide anticipatory guidance

Question 198   Preferred Response: E

The boy in the vignette had a simple febrile seizure and is recovering as expected. No further diagnostic testing is needed. There is approximately a 30% chance of recurrent febrile seizure in this clinical scenario. In most cases of a first seizure, anticonvulsant medications are not indicated.

Further diagnostic testing would be indicated if an underlying illness were clinically suspected. For example, if there were clinical signs of meningitis such as persistent altered mental status or meningismus, or signs of dehydration, then lumbar puncture or serum electrolytes should be considered, respectively. If the seizure had a focal onset, such as starting on 1 side of the body, then neuroimaging and electroencephalography would be appropriate. If there were postictal hemiparesis or signs of increased intracranial pressure, such as gaze palsy or papillitis, then computed tomography of the head would be the best test to perform. For a new focal seizure without signs of increased intracranial pressure, magnetic resonance imaging of the brain is the most sensitive test to detect a focal lesion.

PREP Pearls
- For a simple febrile seizure in a typically developing, previously healthy child, no further diagnostic evaluation is needed.
- Magnetic resonance imaging of the brain is the most sensitive neuroimaging test for causes of seizure, but computed tomography of the brain should be ordered if there is urgency because of signs of increased intracranial pressure.

ABP Content Specifications(s)
- Plan the appropriate management of a first seizure

Suggested Readings
- AAP Subcommittee on Febrile seizures. Febrile seizures: guideline for the neurodiagnostic evaluation of the child with a simple febrile seizure subcommittee on febrile seizures. Pediatrics. 2011;127;389. DOI:http://dx.doi.org/10.1542/peds.2010-3318.

Question 199

A full-term male 3.9-kg newborn develops jitteriness, tremors, and tachypnea 12 hours after birth. The pregnancy was uncomplicated with no maternal history of infection or diabetes mellitus. He is afebrile and his blood pressure is normal. His physical examination is unremarkable. Serum glucose is 38 mg/dL (2.1 mmol/L). The remainder of his laboratory evaluation, including electrolytes, is unremarkable.

Despite frequent feedings, hypoglycemia is recurrent and the newborn is started on dextrose containing intravenous fluids. Over the next 48 hours, his glucose infusion rate approaches 15 mg/kg per minute to maintain glucose levels above 70 mg/dL (3.9 mmol/L). After 14 days, intravenous fluids are still unable to be weaned without recurrence of hypoglycemia. At this point, during a hypoglycemic event, the newborn is given glucagon 1 mg intravenously and, 20 minutes later, his glucose improves to 87 mg/dL (4.8 mmol/L). Serum ketones and free fatty acids are not present.

Of the following, the BEST next treatment for this patient is

A.   carnitine, 50 mg/kg per day

B.   diazoxide, 5 mg/kg per day

C.   growth hormone, 0.05 mg/kg per day

D.   hydrocortisone, 50 mg/m2 per day

E.   octreotide, 15 µg/kg per day

Question 199   **Preferred Response: B**

The newborn in the vignette has hypoglycemia that persists over time. There are many causes of transient hypoglycemia, such as hypoglycemia that occurs in infants of diabetic mothers, in small-for-gestational age infants with little stored glucose reserve, and with perinatal stress. However, the persistence of this boy's symptoms over a 2-week period suggests a more permanent cause of disease. This infant was diagnosed with hyperinsulinism (persistent hyperinsulinemic hypoglycemia of infancy, or PHHI).

The clues that PHHI is the cause of hypoglycemia in this case include the supraphysiologic glucose requirements, as shown by the high glucose infusion rates and a positive response to glucagon administration (a rise in glucose of over 30 mg/dL [1.7 mmol/L] after glucagon is given). Additional diagnostic criteria include suppressed beta-hydroxybuterate (ketones) and free fatty acids, and a measureable insulin level at the time of hypoglycemia. Infants with PHHI are often large for gestational age. The diagnosis of PHHI is not usually considered until approximately 2 weeks, because it can take that long for the aforementioned transient causes to resolve.

The therapeutic goal for all hypoglycemic disorders is to maintain euglycemia, because a delay in treatment confers a high risk of brain damage. Thus, in infants in whom PHHI is suspected, a continuous glucose infusion should be rapidly established. Often a high glucose infusion rate is needed, requiring a central line for venous access. The long-term treatment for PHHI is diazoxide, an antihypertensive agent that opens the K-ATP channel of beta cells, thereby decreasing insulin secretion. The recommended dose is 5 to 15 mg/kg per day, divided 2 or 3 times daily. Side effects include hypertrichosis and fluid retention.

A less commonly used treatment for PHHI is octreotide, which decreases insulin secretion by hyperpolarization of the beta cells and inhibition of calcium channels. Octreotide is associated with frequent treatment failure because of the development of tachyphylaxis. Recently, octreotide was associated with the occurrence of necrotizing enterocolitis, therefore some centers no longer recommend its use in neonates. For patients who fail medical treatment for PHHI, pancreatectomy may be needed.

Other causes of hypoglycemia include panhypopituitarism, which can cause growth hormone deficiency and/or adrenal insufficiency. Neither of these disorders should present with a supraphysiologic glucose requirement; in such patients, the hypoglycemia does not resolve with glucagon infusion as seen in the newborn in the vignette. Thus, administration of growth hormone or hydrocortisone would not be the correct treatment for this patient.
Metabolic disorders including disorders of fatty acid synthesis, congenital disorders of glycosylation, various congenital syndromes, glycogen storage diseases, and others may cause hypoglycemia. Some of these disorders may respond to carnitine treatment. However, these are unlikely diagnoses in this case, because they generally do not require supraphysiologic dosing of glucose.

PREP Pearls
- Persistent hyperinsulinemic hypoglycemia of infancy (PHHI) is characterized by hypoglycemia that persists beyond age 2 weeks, measurable insulin levels at the time of hypoglycemia, suppression of free fatty acids and ketones, and a positive response to glucagon administration (glucose rises > 30 mg/dL [1.7 mmol/L]).
- PHHI may require supraphysiologic glucose dosing.
- The recommended long-term treatment of PHHI is diazoxide, which opens the K-ATP channel of beta cells, thereby decreasing insulin secretion.
- A delay in treatment of PHHI confers a high risk of brain damage. Ideally blood glucose concentrations should be maintained in the normal range while undergoing diagnostic evaluation.

ABP Content Specifications(s)
- Plan the appropriate immediate and long-term management of hyperinsulinism, while considering the long-term prognosis
- Recognize the clinical features associated with hyperinsulinism

Suggested Readings
- Lord K, De León DD. Monogenic hyperinsulinemic hypoglycemia: current insights into the pathogenesis and management.Int J Pediatr Endocrinol. 2013;2013(1):3. DOI: http://dx.doi.org/10.1186/1687-9856-2013-3.
- Mazor-Aronovitch K, Gillis D, Lobel D, et al. Long-term neurodevelopmental outcome in conservatively treated congenital hyperinsulinism. Eur J Endocrinol. 2007;157(4):491-497. DOI: http://dx.doi.org/10.1530/EJECopyright © 2016 American Academy of Pediatrics. All rights reserved.

Question 200

A 9-year-old boy is brought for evaluation because of snoring nightly that wakes his parents in the next room. He has been diagnosed with attention-deficit/hyperactivity disorder. He has witnessed apneic periods, followed by gasping and partial awakening. He is tired and difficult to awaken in the morning. He has nocturnal enuresis, which has not responded to bed alarms or limitation of liquids prior to bed. On physical examination, he is morbidly obese with a body mass index greater than the 98th percentile for age. His tonsils are 3+ and symmetric. He is sent for a polysomnographic study, which reveals an apnea-hypopnea index of 30. The SpO2 nadir is 70%. Based on these results, the child is scheduled for tonsillectomy.

The parents ask about the clinical relevance of their son's sleep apnea and the risks of tonsillectomy.

Of the following, you are MOST likely to inform the parents that

    A.   attention-deficit/hyperactivity disorder is unrelated to sleep disordered breathing

    B.   nocturnal enuresis is unrelated to sleep disordered breathing

    C.   the patient is at high risk for anesthesia-related complications and should be admitted to the intensive care unit after tonsillectomy

    D.   their son will not require continuous positive airway pressure, as long as tonsils are removed in a timely manner

    E.   tonsillectomy is expected to result in complete and long term resolution of this patient's sleep-disordered breathing

Question 200   **Preferred Response: C**

Obstructive sleep apnea (OSA) in children has been defined as a "disorder of breathing during sleep characterized by prolonged partial upper airway obstruction (obstructive apnea) that disrupts normal ventilation during sleep and normal sleep patterns."

The prevalence of OSA in the general pediatric population may approximate 6%. Boys are affected at a higher rate than girls. Obesity is also a recognized risk factor. Snoring is frequent in affected patients, but individuals with OSA should be differentiated from their counterparts with habitual snoring but without obstructive events. Of note, patients with habitual snoring may be at risk for development of OSA with age or with excessive weight gain. It is recommended that clinicians inquire as to symptoms and sequelae of obstructive sleep apnea in all children who habitually snore (snoring 3 or more nights per week) and that polysomnography (PSG) be performed in children with suggestive symptoms or risk factors for OSA. In addition to snoring, children with OSA may have witnessed apneic events; alternatively, they may gasp or snort with frequent awakenings and disrupted sleep that leaves them poorly rested during the day.

Nocturnal enuresis is also associated with sleep disordered breathing and represents emptying of the bladder during partial awakenings. Even with a thorough history, it is important to recognize that clinical history and physical examination are often poor predictors of respiratory PSG findings. The true prevalence of obstructive sleep disordered breathing is likely significantly underestimated.

Associated physical findings in patients with OSA are numerous. The most common risk factor is adenotonsillar hypertrophy. Other associated physical findings include obesity, laryngomalacia, micrognathia, macroglossia, craniofacial abnormalities, dental malocclusion, and hypotonia.

The functional symptoms and signs of sleep disordered breathing or OSA are variable, but include deficits in cognitive and neuropsychological performance. Multiple studies have demonstrated difficulties with attention, learning, executive function, and school performance in affected children. Behavioral abnormalities have also been extensively described; the most common manifestation is hyperactivity, but attention-deficit/hyperactivity disorder, hypersomnolence, somatization, and depression have all been reported.

In children who are suspected as having OSA, an attended PSG study is recommended. An apnea index of greater than 1 or an apnea-hypopnea index (AHI) of 1.5 is considered abnormal. For a child who is 12 years of age or younger, surgical or medical treatment is generally favored in patients with an AHI of 5 to 10 (mild-to-moderate OSA) or more than 10 (moderate-to-severe OSA). It is recommended that children having either an SpO2 nadir of less than 80% on preoperative PSG (or during recovery after surgical intervention) or an AHI greater than or equal to 24 be admitted postoperatively because of an elevated risk for respiratory compromise.

Treatment options for OSA include adenotonsillectomy if there is confirmed OSA with evidence of nasopharyngeal obstruction from adenoidal or tonsillar tissue. In children who are not surgical

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candidates, positive pressure ventilatory support may be indicated in the form of continuous positive airway pressure (CPAP) or bi-level positive airway pressure (bi-level PAP) during sleep. As adherence and mid face growth defects may be problematic in children on chronic noninvasive positive pressure ventilation (NIPPV), surgical intervention is warranted when an anatomic abnormality is thought to be contributing to OSA.

While obesity is a significant risk factor for OSA, it is not a contraindication to adenotonsillectomy (AT) if there is evidence of adenotonsillar hypertrophy and associated narrowing of the nasopharyngeal airway. Obese children with OSA demonstrate significant improvement in sleep disordered breathing and in quality of life after AT. However, OSA does not resolve in the majority of affected children and close follow-up is required to ensure adequate ventilation (including NIPPV, if necessary). In these children, a program of dietary and exercise modification is advocated for gradual weight loss.

PREP Pearls
- Health care providers should complete a thorough evaluation for sleep-related symptoms and for sequelae of sleep disordered breathing (SDB) in children with habitual snoring.
- Obesity is a significant risk factor for SDB and obstructive sleep apnea (OSA).
- Adenotonsillar hypertrophy is a common anatomic risk factor for OSA and may exacerbate OSA in obese individuals.
- Obesity is not a contraindication for adenotonsillectomy, but surgical management is unlikely to result in full resolution of SDB and close follow-up is strongly recommended.
- Children with apnea-hypopnea index greater than or equal to 24 on polysomnography are at elevated risk for anesthesia complications and should be monitored in an inpatient setting after surgery.

ABP Content Specifications(s)
- Plan an appropriate evaluation for obstructive sleep apnea
- Recognize complications associated with obstructive sleep apnea
- Plan appropriate management of obstructive sleep apnea

Suggested Readings
- Costa DJ, Mitchell R. Adenotonsillectomy for obstructive sleep apnea in obese children: a meta-analysis. Otolaryngol Head Neck Surg. 2009;140(4):455-460. DOI: http://dx.doi.org/10.1016/j.otohns.2008.12.038.

Question 201

A 14-year-old adolescent fell while riding in a bicycle road race 3 days ago. He struck his head, shoulder, and forearm, but he did not lose consciousness. He had a normal neurologic examination in the emergency department on the day of the fall. You are seeing him for follow-up. While discussing bicycle safety with you, he reports that his helmet hit the ground hard, but there are only a few scratches on the plastic cover and the rigid foam component appears intact.

Of the following, the BEST advice to give this adolescent about bicycle helmets is

   A.   bicycle helmet safety standards are voluntary

   B.   bicycle helmets should fit at the top of the forehead, and be tilted so that the occiput is covered

   C.   helmets for all road sports have the same design

   D.   his current helmet should be replaced regardless of its appearance

   E.   undamaged bicycle helmets can be safely used for at least 10 years

Question 201   **Preferred Response: D**

Bicycle helmet use has been shown to substantially decrease the risk of head and brain injuries (not concussion) that result from bicycle crashes. Bright Futures recommends that counseling about bike helmets begin at the 2-year health supervision visit and continues for the rest of childhood and adolescence. To be effective, safety helmets must be appropriate for the activity, fit properly, be worn appropriately, and be replaced regularly. The American Academy of Pediatrics Committee on Injury and Poison Prevention states that a bicycle helmet that has sustained a substantial blow should be discarded and replaced; in particular, they note that if the head hit a hard surface, especially if there are marks on the shell, the helmet must be discarded. Therefore, the adolescent in the vignette should replace his helmet regardless of the appearance of the foam lining. Even if there is no impact to the bicycle helmet, it should be replaced at least every 5 years because the helmet materials may deteriorate over time. When wearing a bicycle helmet, it should be positioned so that it fits low on the forehead and is parallel to the ground. Fit may be improved by the use of pads so that the helmet fits snugly in this position.

Since 1999, the US Consumer Product Safety Commission (CPSC) has mandated safety standards for bicycle helmets. Previously, the American National Standards Institute, Snell Memorial Foundation, or the American Society for Testing and Materials endorsed voluntary standards, and old helmets may carry this certification. However, all currently approved helmets should have a sticker documenting CPSC approval, usually located on the inner liner. Bicycle helmets can also be used for scooter-riding, roller and in-line skating, ice skating, and sledding, but for other sports, different helmets are required in order to protect the participant from the types of blows anticipated from those activities. The CPSC website provides a table listing the appropriate helmet for each recreational activity (http://www.cpsc.gov/en/Safety-Education/Safety-Guides/).

PREP Pearls
- Anticipatory guidance about bicycle helmet use should begin by the 2-year-old well child visit.
- The US Consumer Product Safety Commission (CPSC) mandates safety standards for bicycle helmets and a sticker documenting that approval should be found on any helmet purchased.

Question 202

You are on a community health coalition along with staff from the local health department. A recent health department report showed an increase in the chlamydia rate among teenagers. Several coalition members insist that there should be a virginity pledge campaign in the local schools to address this issue.

Of the following, the MOST accurate statement is

A.   comprehensive sexuality education programs are recommended for sexual risk reduction

B.   condom distribution programs have been linked to increased sexual risk-taking behaviors

C.   more than 75% of high school students are sexually active

D.   national sexuality education standards are enforced by publicly funded schools

E.   virginity pledges have been proven effective for the prevention of sexually transmitted infections

Question 202   **Preferred Response: A**

Despite declining rates in recent years, the United States still has higher rates of teen pregnancy than similarly industrialized countries. According to the 2013 Youth Risk Behavior Survey, 46.8% of high school students report having ever had sex. Adolescents and young adults aged 15 to 24 years have the highest age-specific rates of gonorrhea and chlamydia.

The Community Preventive Services Task Force recommends comprehensive risk reduction programs as an effective strategy to reduce adolescent pregnancy, HIV, and sexually transmitted infections. Comprehensive educational programs include information about abstinence as the best way to prevent pregnancy and sexually transmitted infections, but also include medically-accurate information about contraception and condom use.

In the early 1990s, there were several virginity pledge programs that were designed to prevent sexual risk taking behaviors among teenagers. Some studies have found that such pledges may delay sexual initiation among some adolescents; however, other studies have found that adolescents who broke their pledge used contraception less than their nonpledge peers. Researchers have found that adolescents who received comprehensive sexuality education were significantly less likely to report a pregnancy than those who received abstinence-only education. The National Sexuality Education Standards provide guidance on the core content for developmentally appropriate sexuality education for students in grades K through 12. These standards, however, are not uniformly implemented.

Condom distribution programs have been shown not only to increase condom use, but also to promote delayed sexual initiation among youth.

PREP Pearls
- The Community Preventive Services Task Force recommends comprehensive risk reduction programs as an effective strategy to reduce adolescent pregnancy, HIV, and sexually transmitted infections.
- Comprehensive educational programs include information about abstinence as the best way to prevent pregnancy and sexually transmitted infections, but also include medically-accurate information about contraception and condom use.
- The National Sexuality Education Standards provide guidance on the core content for developmentally appropriate sexuality education for students in grades K through 12.

ABP Content Specifications(s)
- Understand the influence of abstinence-only programs on sexual activity in adolescents

Suggested Readings
- Alford S, Advocates for Youth. Sex education programs: definitions & point-by-point comparison. Advocates for Youth website. http://www.advocatesforyouth.org/publications/publications-a-z/655-sex-education-programs-definitions-and-point-by-point-comparison.

Question 203

A full-term newborn is jittery 3 days after delivery. The pregnancy was notable for limited prenatal care, with the mother intermittently participating in a rehabilitation program because of a history of substance abuse. The nurse describes the newborn as not feeding well with periods of increased fussiness. On physical examination, the temperature is 37.3°C, heart rate is 140 beats/min, respiratory rate is 64 breaths/min, and blood pressure is 60/40 mm Hg. The newborn has excoriations on the nose and chin, tachypnea without signs of distress, slightly increased tone, and moderate tremors when disturbed. The blood glucose level is 60 mg/dL (3.3 mmol/L). Maternal urine toxicology screening was positive for cocaine and tetrahydrocannabinol.

Of the following, the agent MOST likely to be related to the newborn's clinical symptoms is

A. alcohol
B. cannabis
C. cocaine
D. methadone
E. methamphetamine

Question 203   **Preferred Response: D**

The symptoms seen in the newborn in the vignette are most likely related to opiate withdrawal. The maternal use of opiates (eg, heroin, methadone, and buprenorphine) during pregnancy may be associated with withdrawal in the newborn. Neonatal abstinence syndrome (NAS) describes the spectrum of central nervous system, gastrointestinal, and autonomic nervous system signs and symptoms seen in affected newborns (Item C203A). Affected newborns may present with symptoms at birth or up to 10 to 14 days later. Maternal history and drug testing may be used to identify at-risk newborns, but it is important to note that synthetic opioids such as methadone are not detected on many routine urine toxicology screens.

Item C203A. Clinical Findings Seen in Neonatal Abstinence Syndrome.

Central Nervous System	Gastrointestinal	Autonomic Nervous System
Crying	Excessive sucking	Hyperthermia
Decreased sleep	Poor feeding	Sneezing
Tremors	Emesis	Tachypnea
Increased tone	Loose stools	Sweating
Excoriations		Seizures

Courtesy of S. Izatt

Many drugs used by mothers during pregnancy cross the placenta and affect the developing fetus, including agents associated with substance abuse. The potential effects of these agents include congenital malformations, prematurity, low birthweight, abnormal state regulation, and neonatal withdrawal (Item C203B). Alcohol use is associated with congenital birth defects and abnormal brain development, with the exposed fetus at risk for fetal alcohol spectrum disorder (FASD). Congenital birth defects associated with FASD include cardiac defects (ventricular septal defect, atrial septal defect), cleft palate, and renal anomalies (renal agenesis, renal hypoplasia).

Item C203B. Effects on the Newborn of In Utero Substance Exposure.

	Prematurity	Small for Gestation	Neurobehavioral Symptoms	Neonatal Withdrawal	Congenital Malformations
Alcohol	Yes	Yes	Yes	Yes	Yes
Benzodiazepines	Yes	Yes	Yes	Yes	Yes/No
Cocaine	Yes	Yes	Yes	No	Yes/No
Marijuana	No	No	Yes	No	No
Methamphetamines	Yes/No	Yes	Yes	Yes	No
Opioids	Yes/No	Yes/No	Yes	Yes	No

Yes/No = both have been reported.

Adapted and reprinted with permission from: Jansson LL, Velez ML. Infants of drug-dependent mothers. *Pediatr Rev.* 2011;32:5-13.

Newborns exposed to alcohol, barbiturates, and methamphetamines in utero may also display symptoms of withdrawal. The severity of barbiturate withdrawal is similar to that seen with opioids, and typically presents 4 to 7 days after birth. Withdrawal from alcohol or methamphetamines begins within hours after birth. The symptoms of alcohol and methamphetamine withdrawal are much less severe than those of opiate withdrawal. Although no specific treatments exist for nonopioid withdrawal in infants, clinicians must consider polysubstance exposure in newborns born to substance abusing mothers.

The physical examination findings of excoriations, tachypnea, increased tone, and moderate tremors seen in the newborn in the vignette 3 days after delivery suggest opioid withdrawal. Methamphetamine and alcohol withdrawal are less likely, as they typically occur shortly after delivery. Although the urine toxicology screen of the mother in the vignette was positive for cocaine and tetrahydrocannabinol, these agents are not typically associated with the classic findings of neonatal abstinence syndrome.

PREP Pearls
- Newborns exposed to opioids, alcohol, barbiturates, or methamphetamines in utero are at risk for symptoms of withdrawal.
- Neonatal abstinence syndrome (NAS) describes central nervous system, gastrointestinal, and autonomic nervous system signs and symptoms seen in newborns with drug withdrawal.
- Synthetic opioids, such as methadone, are not detected on many routine urine toxicology screens.

ABP Content Specifications(s)
- Recognize the clinical and laboratory features associated with neonatal withdrawal syndrome as a result of maternal drug use
- Recognize the impact of maternal drugs of abuse on a fetus
- Recognize the effects on the fetus and infant of maternal medication use during pregnancy and labor

Suggested Readings

American academy of pediatrics   658

- Burgos AE, Burke BL. Neonatal abstinence syndrome. NeoReviews. 2009;10:e222-e229. DOI: http://dx.doi.org/10.1542/neo.10-5-e222.
- Jansson LM, Velez ML. Infants of drug-dependent mothers. Pediatr Rev. 2011;12:5-13. DOI: http://dx.doi.org/10.1542/pir.32-1-5.
- Jones HE, Kaltenbach K, Heil SH, et al. Neonatal abstinence syndrome after methadone or buprenorphine exposure. N Engl J Med. 2010;363:2320-2331. DOI: http://dx.doi.org/10.1056/NEJMoa1005359.
- Kocherlakota P. Neonatal abstinence syndrome. Pediatrics. 2014;134:e547-e561. DOI: http://dx.doi.org/10.1542/peds.2013-3524.

Question 204

A 5-year-old, previously healthy boy presents to the emergency department with a 2-cm laceration above his left eyebrow, sustained after hitting his head on a granite countertop. His vital signs include a temperature of 37°C, heart rate of 140 beats/min, blood pressure of 90/50 mm Hg, respiratory rate of 30 breaths/min, and oxygen saturation of 100% on room air. He is awake, alert, and anxious appearing. The repair requires sedation because of the boy's age, anxiety level, and proximity of the laceration to the eye. He was given a dose of intravenous (IV) midazolam 0.1 mg/kg, but he remains awake, screaming, and thrashes his head when approached. He is given a second dose of IV midazolam 0.1 mg/kg and falls asleep. His vital signs after the second dose are a temperature of 37°C, heart rate of 90 beats/min, blood pressure of 68/40 mm Hg, respiratory rate of 20 breaths/min, and oxygen saturation of 95% on room air. Upon verbal stimulation, the boy opens his eyes, mumbles a few words, and falls back asleep. His extremities are warm with a capillary refill time of 1 second.

Of the following, the MOST appropriate next step is the IV administration of

A.   0.9% normal saline bolus, 20 mL/kg

B.   atropine, 0.01 mg/kg

C.   epinephrine infusion, 0.05 µg/kg per min

D.   flumazenil, 0.01 mg/kg

E.   naloxone, 0.01 mg/kg

Question 204   **Preferred Response: A**

After receiving 2 doses of midazolam, the boy in the vignette became hypotensive, but is otherwise maintaining his airway and is not on the verge of hemodynamic collapse. The best choice among the options is intravenous administration of a 0.9% normal saline bolus of 20 mL/kg.

Moderate sedation is often required for laceration repairs in children from toddler to early school age, because of anxiety as well as pain. According to the American Academy of Pediatrics, moderate sedation is a state of decreased level of consciousness with appropriate response to physical stimulation or verbal commands. Monitoring should include continuous pulse oximetry, visual assessment of ventilation, and noninvasive blood pressure measurement every 5 minutes. For the boy in the vignette, the intended effect was not achieved after 1 dose of midazolam, as evidenced by continued screaming and thrashing, therefore a second dose was given. Adverse effects of benzodiazepines can include hypotension, bradycardia, and respiratory depression. The following equation can be used to estimate minimum systolic blood pressure for a young child: Systolic blood pressure = 70 mm Hg + (2 × child's age in years)

The child in the vignette has a systolic blood pressure of 68 mm Hg, which qualifies him as hypotensive. However, because he is not profoundly hypotensive, there is no indication of significant overdose, and he has adequate clinical perfusion, the boy is not likely on the verge of cardiovascular collapse.

An important mechanism of hypotension caused by benzodiazepines is dilation of systemic arterioles and venules. The resulting increase in vascular capacitance can decrease preload, thereby reducing cardiac output. Administration of a fluid bolus will increase preload and therefore cardiac output, thereby increasing the blood pressure. Atropine is not recommended in this case because the child does not have bradycardia. Although epinephrine would increase blood pressure by causing vasoconstriction and increase cardiac output by its inotropic properties, it should be reserved for more profound shock states. Flumazenil is a reversal agent for benzodiazepines, but is not routinely recommended because of rebound effects and a risk of seizures. Naloxone is an opioid reversal agent, and would not be helpful in benzodiazepine overdose.

PREP Pearls
- Benzodiazepines can cause hypotension by vasodilation, which decreases cardiac preload and therefore cardiac output.
- Flumazenil is a reversal agent for benzodiazepines, but is not routinely recommended because of rebound effects and a risk of seizures.
- Monitoring during moderate sedation should include continuous pulse oximetry, visual inspection of breathing, and noninvasive blood pressure readings.

ABP Content Specifications(s)
- Plan the appropriate pre-sedation protocol for a patient who is about to undergo moderate sedation

- Understand the indications and contraindications for moderate sedation
- Recognize the side effects associated with an overdose of commonly prescribed sedatives, and manage appropriately

Suggested Readings

- American Academy of Pediatrics Committee on Drugs. Guidelines for monitoring and management of pediatric patients during and after sedation for diagnostic and therapeutic procedures: addendum. Pediatrics. 2002;110:836-838. DOI:http://dx.doi.org/10.1542/peds.110.4.836.

Question 205

A 7-year-old, previously healthy Hispanic girl presents to your office for evaluation of daily fever that has lasted 10 days. She has had associated night sweats, abdominal pain, and a 4 lb weight loss. Her exposures include 2 dogs and 2 birds at home and consumption of unpasteurized cheese from Mexico. Vital signs show a temperature of 39.5°C, respiratory rate of 22 breaths/min, blood pressure of 118/69 mm Hg, and a weight of 27 kg. Her physical examination is unremarkable. Laboratory data shows:

- White blood cells, 4,400/µL (4.4 x 10^9/L)
- Hemoglobin, 11.5 g/dL (115 g/L)
- Hematocrit, 32.2%
- Platelets, 180 ×10^3/µL (180 x 10^9/L)
- Differential, 23% segmented neutrophils, 26% bands, 45% lymphocytes, 6% monocytes
- Alanine aminotransferase, 55 U/L
- Aspartate aminotransferase, 59 U/L
- Urine analysis, negative

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

A.   abdominal ultrasonography

B.   blood culture

C.   chest radiograph

D.   Epstein-Barr virus serologies

E.   urine culture

Question 205   **Preferred Response: B**

The patient in this vignette has an important risk factor for brucellosis, consumption of unpasteurized dairy products, which should prompt the reader to consider Brucella as the etiology of her fever of unknown origin and obtain a blood culture. Imported unpasteurized dairy products, especially from Mexico, are a large source of Brucella infection in the United States. Transmission can also occur by inoculation of infected animal fluids into cuts, mucous membranes, or through inhalation. The diagnosis of Brucella is made by isolating the organism in culture or obtaining Brucella-specific serology. In the acute phase of the infection, blood cultures can often yield the organism.

Although there are various definitions of fever of unknown origin (FUO), some experts consider FUO as fever of at least 8 days' duration in which the diagnosis in not readily apparent after a thorough history and physical examination. Of all etiologies, infections account for the largest proportion of cases of FUO. In the United States, 100 to 200 cases of brucellosis are reported annually. Children are estimated to account for up to one-third of all cases of brucellosis in endemic countries, although underreporting is possible.

Common clinical manifestations of brucellosis include fever, sweating, osteoarticular complaints, hepatosplenomegaly, and transaminitis. Arthritis is typically monoarticular and affects the knee and hip most commonly. Adults are more likely to have involvement of other sites, including the sacroiliac joint or axial skeleton. Bone marrow suppression can occur, as evidenced by the mild leukopenia observed in this patient. Brucella is an intracellular pathogen that can evade immune responses, therefore prolonged (minimum of 6 weeks) combination antimicrobial therapy is needed for effective treatment and to prevent relapses.

While abdominal ultrasonography may show enlargement of the liver and spleen, this would not be a specific finding. Abdominal ultrasonography could be helpful in the diagnosis of hepatosplenic cat scratch disease caused by Bartonella henselae, where microabscesses may be visualized. However, this patient does not have feline exposure, making this a much less likely possibility.

Pneumonia can be occult and should be considered in cases of FUO. Chest radiograph can additionally help identify hilar lymphadenopathy and chest masses. However, this patient lacks respiratory complaints or pulmonary findings on physical examination, making pneumonia less likely. Additionally, given her exposure history, Epstein-Barr virus infection is less likely. However, Epstein-Barr virus should be considered in cases of FUO, especially those with transaminitis. Lastly, the negative urinalysis makes the likelihood of urinary tract infection, diagnosed with urine culture, low.

PREP Pearls
- Indolent infections, such as Brucella, should be considered in the differential diagnosis of fever of unknown origin, especially in the context of consumption of unpasteurized dairy products. A blood culture can reveal the diagnosis in the acute phase of infection.

- Common clinical manifestations of brucellosis include fever, sweating, osteoarticular complaints, hepatosplenomegaly, and transaminitis.
- Brucella is an intracellular pathogen that can evade immune responses, therefore prolonged (minimum of 6 weeks) combination antimicrobial therapy is needed for effective treatment and to prevent relapses.

ABP Content Specifications(s)
- Understand the importance of considering brucellosis in the differential diagnosis of fever of unknown origin

Suggested Readings
- American Academy of Pediatrics. Brucellosis. In: Kimberlin DW, Brady MT, Jackson MA, Long SS, eds. Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:268-270.

Question 206

A 7-year-old girl presents to the emergency department with complaints of "pain in her private area" and "blood in her underwear." According to the mother, the child slipped today while playing on monkey bars. On physical examination, her vital signs show a temperature of 37.8°C, heart rate of 70 beats/min, respiratory rate of 16 breaths/min, and blood pressure of 90/60 mm Hg. Her physical examination is significant for a medium- to large-sized vulvar hematoma with superficial abrasions (Item Q206).

Item Q206
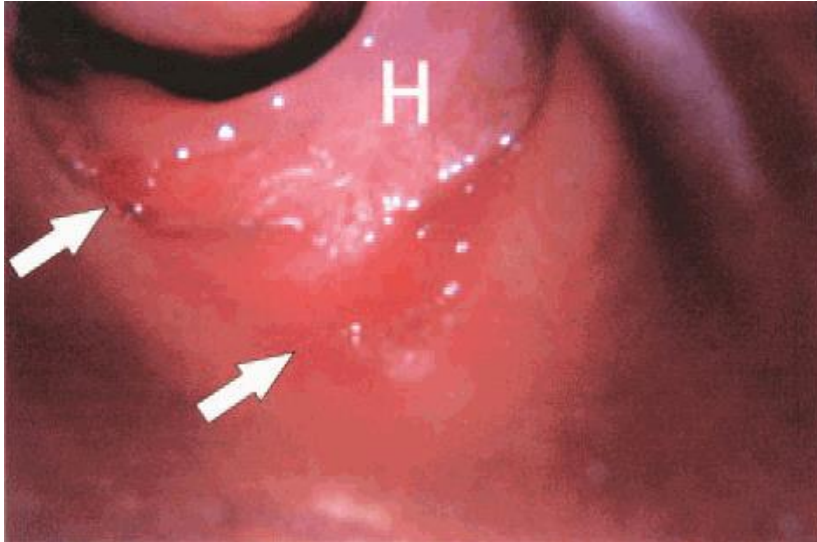
Item Q206. Findings for the girl described in the vignette. Reprinted with permission from Johnson CF. Pediatr Rev. 2006; 27:17

You discuss the differential diagnosis of straddle injury versus sexual abuse with the medical students.

Of the following, the MOST accurate statement regarding this patient's presentation and clinical findings is

A.   blood stains on the underwear cannot be explained by the patient's history

B.   large vulvar hematomas are unlikely due to straddle injuries

C.   sexual abuse is the likely explanation for the patient's presentation and findings

D.   vaginal lacerations are not seen in straddle injuries

E.   vulvar hematomas are common with straddle injuries in girls

Question 206   **Preferred Response: E**

Straddle injuries are injuries of the urogenital area occurring when a child falls and straddles an object, striking with the force of his or her weight. In children presenting with history of injury to the perineum, it is important to differentiate between accidental and nonaccidental (sexual abuse) trauma.

A detailed history and physical examination is warranted in all cases of children with genital trauma. The extent of perineal injury on examination should correlate with the history to confirm nonsexual trauma as the cause of the patient's injury. Caregivers of patients with accidental genital trauma generally seek immediate medical attention. The verbal child can give a supportive history of the accident and there may be witnesses (other family members) to the event. Physical examination in the presence of a chaperone should be performed in a patient with a history of perineal injury. In premenarcheal girls, the genital examination is best performed in the supine (frog leg) and the prone (knee chest) position. The prone (knee chest) examination is indicated in cases with suspected vaginal trauma. The examination may be difficult in a patient with a recent history of trauma and pain from the trauma. In these cases, local anesthesia (application of 2% lidocaine) or procedural sedation (such as in patients requiring sutures) is indicated.

Vulvar hematomas and superficial lacerations (vulvar and vaginal) are the usual straddle injuries in girls. Vulvar hematomas may vary in size following a straddle injury. The vulvar area in young girls is highly vascular, with loose subcutaneous tissues increasing the risk for large hematoma formation. Therefore, complaints of bleeding or blood stains on the underwear are common in patients with straddle injuries and vulvar hematomas. Urinary retention may be associated with vulvar hematomas and the physician needs to ensure proper voiding before discharging the patient home. Patients with large hematomas and urinary retention may need temporary bladder drainage. Most hematomas are usually managed conservatively with adequate pain control, rest, ice packs, and tub baths. Patients are advised to rest on their side or use a foam or air-filled rubber doughnut (while sitting) to avoid pressure injury of the swollen external genitalia. Surgical intervention may be needed in patients with injury to the pelvic floor, urethra, or increasing hematoma size despite adequate conservative management.

Straddle injuries may lead to unilateral and superficial lacerations of the vagina and vulva. The patient in the vignette has lacerations in the right hymenal wall and fourchette. Identification of vaginal lacerations from accidental trauma is important, as penetrating injuries (from sexual abuse) are usually associated with vaginal lacerations in children. Bleeding from hymenal injuries is often minimal and usually requires no treatment. Lacerations of the vaginal mucosa may need suturing under anesthesia.

Complaints to adults or caregivers of uncomfortable experiences from being touched on the genitalia, inappropriate sexualized behaviors (excessive masturbation, adult words associated with sexuality, simulation of sexual behavior with siblings or toys), symptoms of vaginal discharge, genital lesions suggestive of sexually transmitted disease, and genital or anal injuries on physical examination are suspicious for underlying sexual abuse. In sexual abuse-associated

genital trauma, most children do not present until weeks or months after the abuse. In patients, U- or V-shaped clefts (notches) of the posterior rim (from 3 o'clock to 9 o'clock), indicative of healing after a laceration and attenuation or decreased width (less than 1 mm) of the posterior hymen, are suggestive for underlying sexual abuse. It is important to note that only a small percentage of sexually abused children have an abnormal genital or anal finding. Clinicians should also be aware of the age-related hymenal changes and normal anatomic variations of the hymen, which may be confused with features of sexual abuse. Midline sparing (linea vestibularis), developmental variants (fenestrated hymen, failure of midline fusion), labial adhesions, and dermatologic conditions such as lichen sclerosus and pemphigoid may be confused with features of sexual abuse.

For the patient in the vignette, the presenting history, symptoms (of blood in the underwear), and physical examination (vulvar hematoma and acute superficial lacerations) are consistent with straddle injury.

PREP Pearls
- Vulvar hematomas and superficial lacerations are consistent with straddle injuries in girls.
- Identification of vaginal lacerations from accidental trauma is important because sexual abuse is commonly associated with vaginal lacerations.
- Only a small percentage of sexually abused children have an abnormal genital or anal finding.
- A knowledge of the age-related hymenal changes and normal anatomic variations of the hymen is important, as these may be confused with features of sexual abuse.

ABP Content Specifications(s)
- Plan the initial evaluation and management of a patient with genital trauma

Suggested Readings
- Dowd MD, Fitzmaurice L, Knapp JF, Mooney D. The interpretation of urogenital findings in children with straddle injuries. J Pediatr Surg. 1994;29(1):7-10.

Question 207

You are seeing a 12-year-old girl in follow-up. She has a 3-month history of worsening right upper quadrant abdominal pain and bloody diarrhea. She reports 2 to 4 liquid, bloody bowel movements daily. She also reports nausea and a 10 lb weight loss. She denies recent travel, reptile exposure, or ill contacts.

Recent laboratory studies have included:
•   Hemoglobin, 8.6 g/dL (86 g/L)
•   Electrolytes, normal
•   Heme, positive stool
•   Stool culture, negative

Of the following, the BEST next step in diagnosis is

A.   abdominal ultrasonography

B.   colonoscopy

C.   computerized tomography of the abdomen and pelvis

D.   enteroclysis

E.   magnetic resonance imaging enterography

Question 207   **Preferred Response: B**

The girl in the vignette has a history of diarrhea, bloody stools, weight loss, and anemia. Her most likely diagnosis is inflammatory bowel disease. The next step in evaluation of this child is to refer for colonoscopy to obtain tissue for diagnosis. The differential diagnosis of gastrointestinal bleeding varies by age and origin of the bleeding (Item C207).

Item C207. Differential Diagnosis of Gastrointestinal Bleeding by Age.

Age Group	Upper Gastrointestinal Bleeding	Lower Gastrointestinal Bleeding
Neonates	Hemorrhagic disease of the newborn Swallowed maternal blood Stress gastritis Coagulopathy Peptic ulcer disease Vascular anomaly Cow's milk protein sensitivity	Anal fissure Necrotizing enterocolitis Malrotation with volvulus Intussusception Allergic colitis Infectious colitis Hirschsprung disease
Infants aged 1 month to 1 year	Esophagitis Gastritis Peptic ulcer disease Vascular anomaly Cow's milk protein sensitivity Mallory-Weiss tear Intestinal duplications Esophageal or gastric varices Foreign body Hereditary telangiectasia	Anal fissure Allergic colitis Infectious colitis Intussusception Gangrenous bowel Malrotation with volvulus Meckel diverticulum Ischemic colitis Vascular lesions
1 year to 12 years	Peptic ulcer disease Gastritis Mallory-Weiss tear Intestinal duplications Esophageal or gastric varices Foreign body Caustic ingestion Vasculitis (HSP) Hereditary telangiectasia	Polyps Hemorrhoids Meckel diverticulum Anal fissure Infectious colitis Intussusception Malrotation with volvulus Ischemic colitis Vascular lesions Henoch–Schönlein purpura (HSP) Hemolytic uremic syndrome Inflammatory bowel disease Lymphonodular hyperplasia
Adolescents	Esophagitis Peptic ulcer disease Gastritis Mallory-Weiss tear Intestinal duplications Esophageal or gastric varices Foreign body Caustic ingestion Vasculitis (HSP) Hereditary telangiectasia	Polyps Hemorrhoids Inflammatory bowel disease Infectious diarrhea Vascular lesions Inflammatory bowel disease Meckel diverticulum Infectious colitis Henoch–Schönlein purpura Hemolytic uremic syndrome

Courtesy of C. Waasdorp Hurtado

All gastrointestinal (GI) bleed evaluations should begin with assessment of vital signs and hemodynamic stability with resuscitation as clinically indicated. A complete history and physical examination follows stabilization and should include visualization of a stool sample. Testing for fecal occult blood to assess for the presence of blood is essential whenever there is a concern for

GI bleeding. Stool cultures and Clostridium difficile toxin should be completed to evaluate for infectious etiologies. A complete blood cell count (CBC) and coagulation studies will assess for anemia and coagulopathy. If blood loss is active or ongoing, serial CBC studies may be needed to determine need for more urgent interventions. Imaging studies may include an abdominal radiograph to evaluate for foreign bodies and free air, and an upper gastrointestinal (UGI) series to evaluate for anatomic abnormalities, including duplication cysts and abdominal ultrasonography with Doppler to assess for portal hypertension. Tagged red blood cell scans and angiography can be used, but are not effective at localization if the rate of blood loss is low, as is typical in children. A Technetium-99 scan can identify a Meckel diverticulum. Endoscopy of the GI tract can aid in both diagnosis and therapy, particularly when bleeding occurs from the upper GI tract.

The vignette describes a child with a history and laboratory studies that are most consistent with inflammatory bowel disease (IBD) without evidence of an acute abdomen. A colonoscopy will be the best test to aid in the diagnosis. Abdominal ultrasonography, computed tomography of the abdomen and pelvis, magnetic resonance imaging enterography, and enteroclysis would all evaluate the GI anatomy and may identify inflammation, but will not confirm the diagnosis of IBD.

PREP Pearls
- Gastrointestinal bleeding occurs at all ages.
- History and physical examination can give some clues to etiology.
- Testing for fecal occult blood should be used to confirm the presence of blood.
- Colonoscopy is used to confirm the diagnosis of inflammatory bowel disease.

ABP Content Specifications(s)
- Distinguish among the etiologies of occult blood and bright red blood per rectum
- Plan the appropriate evaluation of rectal bleeding
- Formulate an age-appropriate differential diagnosis for rectal bleeding

Suggested Readings
- Dubinsky M. Special issues in pediatric inflammatory bowel disease. World J Gastroenterol. 2008;14(3):413-420.http://www.wjgnet.com/1007-9327/14/413.pdf.

Question 208

A 4-year-old boy presents to your practice with developmental delays, unusual movements, and cerebral palsy. He was a full term spontaneous vaginal delivery without complications during pregnancy or delivery. Initial development was normal for the first few months, which then slowed compared to his peers. The mother states he had low tone and delays noted by 6 months of age. He currently is at the developmental level of a 1-year-old child. He can sit, but does not walk. He has twisting movements and abnormal posturing of his extremities. Recently, he began biting his fingers and lips, as well as head banging. The neurologic examination reveals dystonia, spasticity, extensor plantar reflexes, and increased deep tendon reflexes. You suspect a genetic disorder. You order a chromosomal microarray and a karyotype that are both unremarkable. Due to the recent changes in behaviors, his neurologic examination, and history, the BEST next test to aid in diagnosis in this child is

A.   total homocysteine

B.   urinary urate-to-creatinine ratio

C.   urine mucopolysaccharides

D.   urine porphyrins

E.   very long chain fatty acids

Question 208   **Preferred Response: B**

The child in the vignette has Lesch-Nyhan syndrome, which is an uric acid metabolism disorder that presents with motor dysfunction resembling cerebral palsy, behavioral disturbances, cognitive impairment, and hyperuricemia. Therefore, the best next test to aid in the patient's clinical diagnosis would be a urinary urate-to-creatinine ratio. At birth, patients appear normal, but by 3 to 6 months of age, hypotonia and developmental delay become evident. Most patients can eventually sit, but rarely do they learn to walk. In the first several years, unusual motor movements manifest, including dystonia, choreoathetosis, and opisthotonos. Spasticity, hyperreflexia, and extensor plantar reflexes that mimic cerebral palsy also become apparent. The behavioral disturbances and cognitive decline emerge between 2 and 3 years of age. A pathognomonic clinical finding of Lesch-Nyhan disorder is self-injurious behaviors, as seen in the child in this vignette. Uric acid overproduction leads to deposition of uric acid crystals in the bladder, kidneys, and ureters over time. Gouty arthritis can also occur. Some mothers even report orange crystals in their affected sons' diapers. The index of suspicion is raised when developmental delay is concurrently seen with hyperuricemia or nephrolithiasis. The urinary urate-to-creatinine ratio, the best screening test, should be greater than 2.0 in a child who is younger than 10 years of age who has Lesch-Nyhan syndrome. Hyperuricosuria and hyperuricemia (serum uric acid concentration > 8 mg/dL [476 µmol/L]), while often present, are not sensitive or specific enough to confirm the diagnosis. Diagnostic confirmation is made via analysis of the hypoxanthine-guanine phosphoribosyltransferase enzyme activity, which should be less than 1.5% in individuals with a diagnosis of Lesch-Nyhan syndrome. The only known gene associated with Lesch-Nyhan syndrome is HPRT1. It is inherited in an X-linked recessive manner. Therefore, males who carry the gene change are affected, but females are carriers and are typically unaffected.

Treatment is directed at controlling the uric acid production with allopurinol for the urate nephropathy, gouty arthritis, tophi, and nephrolithiasis; however, this treatment has no impact on the behavioral and neurologic symptoms of the disorder. Baclofen and benzodiazepines can be used for the spasticity. Patients typically require therapies, including habilitative, behavioral, and psychiatric therapy. Protective equipment may be necessary to reduce trauma secondary to self-injurious behaviors.

Total homocysteine would be a good screening test for homocystinurias that are characterized by a Marfanoid phenotype, developmental delay, ectopia lentis, severe myopia, tall stature, and thromboembolism that are not consistent with this patient's phenotype.
Urine mucopolysaccharides would be indicated in a child with developmental regression and progressive coarsening of the facial features. Urine porphyrins should be obtained in a patient with life-threatening acute neurovisceral attacks of severe abdominal pain, tachycardia, hypertension, mental status changes, convulsions, peripheral neuropathy, and hyponatremia. Very long chain fatty acids are an excellent screening test for peroxisomal disorders that present with sensorineural hearing impairment, ocular abnormalities (retinopathy, cataracts, optic nerve atrophy), developmental delay, and a classic dysmorphic appearance.

PREP Pearls
- Lesch-Nyhan syndrome is an uric acid metabolism disorder that presents with motor dysfunction resembling cerebral palsy, behavioral disturbances, cognitive impairment, self-injurious behaviors, and hyperuricemia.
- A pathognomonic clinical manifestation of Lesch-Nyhan disorder is self-injurious behaviors.
- The best screening test is an urinary urate-to-creatinine ratio that should be greater than 2.0 in a child who is younger than 10 years of age who has Lesch-Nyhan syndrome. Hyperuricosuria and hyperuricemia (serum uric acid concentration > 8 mg/dL [476 µmol/L]), while often present, are not sensitive or specific enough to confirm the diagnosis.

ABP Content Specifications(s)
- Recognize the clinical features associated with a disorder of uric acid metabolism
- Recognize the clinical features associated with a disorder of uric acid metabolism

Suggested Readings
- Fu R, Ceballos-Picot I, Torres RJ, et al. Genotype-phenotype correlations in neurogenetics: Lesch-Nyhan disease as a model disorder. Brain. 2014;137(Pt 5):1282-1303. DOI: http://dx.doi.org/10.1093/brain/awt202.
- Jinnah HA, Ceballos-Picot I, Torres RJ, et al. Attenuated variants of Lesch-Nyhan disease. Brain. 2010;133(Pt 3):671-689. DOI: http://dx.doi.org/10.1093/brain/awq013.

Question 209

A 1-month-old male infant is brought to your office by his mother for his first health supervision visit. Pregnancy, labor, and delivery were uncomplicated. He was born appropriate for gestational age at 39 weeks of gestation and was discharged at 3 days of age. He stayed 1 extra day in the hospital for mild jaundice and delayed passage of his first stool. He is primarily breastfed and has been gaining weight along the 25th percentile for his age. He passes stools every 3 days and his mother thinks he may have constipation, but states that another pediatrician assured her that this was normal for breastfed babies. The mother denies any significant family history, though you note that she has iris heterochromia. On physical examination, the infant has mild abdominal fullness, but no organomegaly or palpable masses. On digital rectal examination, the anal canal is tight and a small amount of stool squirts out as you withdraw your finger from the anus. The remainder of his physical examination is unremarkable.

Of the following, the BEST test to confirm the suspected diagnosis for this infant is

A.   celiac panel

B.   immunoreactive trypsinogen level

C.   rectal suction biopsy

D.   sweat chloride test

E.   thyroid panel

Question 209   **Preferred Response: C**

The newborn in the vignette has the characteristic history and physical examination findings of congenital aganglionic megacolon or Hirschsprung disease. Although findings on abdominal radiography, contrast enema, or anorectal manometry support the diagnosis, rectal suction biopsy is the diagnostic gold standard for Hirschsprung disease. The history and physical examination findings that suggest the diagnosis in this case include delayed passage of meconium stool, decreased frequency of stools during the first month after birth, mild abdominal fullness, and the "squirt sign" on digital rectal examination. In addition, the patient's mother has heterochromia of her irises, which should lead the practitioner to consider the association of Hirschsprung disease and Waardenburg syndrome.

Delayed passage of meconium, defined as beyond 48 hours after birth, can be indicative of a serious problem such as bowel atresia or obstruction, imperforate anus, meconium plug, or Hirschsprung disease. It is important for pediatric health care providers to recognize the potential significance of this delay. Neonates who pass their first meconium stool after 48 hours but before 72 hours, and are otherwise well appearing, should undergo a thorough physical examination. The conditions that may be commonly associated with delayed passage of meconium should be carefully considered and excluded. These infants should be followed closely and evaluated promptly if they develop symptoms of abdominal distention, bilious vomiting, or constipation. Any neonate who fails to pass meconium within the first 72 hours after birth should be evaluated for Hirschsprung disease.

Hirschsprung disease is a motility disorder caused by the absence of parasympathetic ganglion cells, because of the failure of neural crest cells to migrate completely during intestinal development. Hirschsprung disease is more prevalent in several genetic syndromes, including Down syndrome, Bardet-Biedl syndrome, multiple endocrine neoplasia type 2, Smith-Lemli-Opitz, and Waardenburg syndrome. Type IV Waardenburg syndrome (also known as Waardenburg-Shah syndrome) has signs and symptoms of both Waardenburg syndrome and Hirschsprung disease. Mutations in the SOX10,EDN3, or EDNRB genes cause type IV Waardenburg syndrome. These genes are important for the development of nerve cells in the large intestine in addition to melanocyte development. Mutation in any of these genes results in hearing loss; changes in the pigmentation of skin, hair, and eyes; and intestinal problems related to Hirschsprung disease. Often there is an autosomal dominant pattern of inheritance, but an autosomal recessive pattern may occur as well.

Celiac disease or gluten-sensitive enteropathy typically presents in infants between 6 and 24 months of age, after the introduction of gluten into the diet. The signs and symptoms suggestive of gluten sensitivity are chronic diarrhea (or rarely, constipation), anorexia, abdominal distention, chronic abdominal pain, and failure to thrive.

Meconium ileus in the newborn is almost always caused by cystic fibrosis. These infants fail to pass meconium stools, and often have marked abdominal distention with bilious emesis soon after birth. Occasionally, infants with cystic fibrosis may have lesser degrees of meconium impaction and a presentation that is more benign. The diagnosis of cystic fibrosis is made

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through immunoreactive trypsinogen levels on the newborn screening test or sweat chloride levels in the older infant or child.

Congenital hypothyroidism should be considered in infants who present with prolonged jaundice and constipation. However, these infants will have additional signs and symptoms, which the infant in this vignette did not have, such as lethargy, hypothermia, feeding problems, poor weight gain, macroglossia, umbilical hernia, large fontanels, hypotonia, and dry skin.

PREP Pearls
- Passage of meconium occurring beyond the first 48 hours after birth is considered delayed.
- Delayed or absent passage of meconium can indicate a serious problem, such as bowel atresia or obstruction, imperforate anus, meconium plug, or Hirschsprung disease.
- Neonates who pass their first meconium stool between 48 and 72 hours after birth, but are otherwise well-appearing, should undergo a thorough physical examination, with close follow-up and further evaluation performed as indicated.
- Any neonate who fails to pass meconium within the first 72 hours after birth should be evaluated for Hirschsprung disease.

ABP Content Specifications(s)
- Recognize disorders associated with delayed or absent passage of meconium

Suggested Readings
- Branski D, Dennis NR, Neale JM, Brooks LJ. Hirschsprung's disease and Waardenburg's syndrome. Pediatrics. 1979;63(5):803-805. http://pediatrics.aappublications.org/content/63/5/803 .

Question 210

An 18-month-old boy presents to the emergency department with a 3-day history of high, spiking fevers and malaise. On physical examination, his temperature is 39.2°C, heart rate is 142 beats/min, and blood pressure is 90/48 mm Hg. He is alert and cranky, but consolable by his mother. He is flushed, but has no rash. He appears well-hydrated. His liver and spleen are not enlarged. He has no retractions or nasal flaring. His lungs are clear to auscultation. His complete blood cell count results are shown:

White blood cell count   5,600/µL (5.6 x 10^9/L)
Hemoglobin   11.2 g/dL (112 g/L)
Platelet count   467 x 10^3/µL (467 x 10^9/L)
Absolute neutrophil count   320/µL

Of the following, over the next 48 hours, this child is MOST likely to develop

A.   a diffuse petechial rash and epistaxis

B.   an erythematous, nonpruritic rash over the trunk and neck

C.   lymphoblasts on his peripheral smear

D.   a purpuric rash rising up the legs to the buttocks

E.   renal failure

Question 210   **Preferred Response: B**

Neutropenia can be classified as mild (absolute neutrophil count [ANC] 1,000-1,500/µL), moderate (ANC 500-1,000/µL), severe (ANC 200-500/µL), or very severe (ANC < 200/µL). The child in the vignette presents with high fevers, malaise, and flushing, as well as isolated, severe neutropenia. His complete blood cell count is remarkable for having a normal hemoglobin level and platelet count. Although these signs and symptoms are nonspecific, the most likely etiology is a viral illness. While many common viral infections can cause a transient neutropenia due to maturation arrest, the presentation in the vignette is consistent with an infection with human herpesvirus 6, the causative agent for roseola (exanthema subitum). This common viral infection typically presents in infants with high, spiking fevers for several days accompanied by irritability. Once the fever breaks, an erythematous, nonpruritic rash develops. Human herpesvirus 6 can cause a transient neutropenia.

A diffuse petechial rash and epistaxis would be most associated with severe thrombocytopenia. Isolated, severe neutropenia should not impact hemostasis or increase the risk of bleeding.

While leukemia can present with low blood cell counts and fever, it would be unusual for it to present as isolated neutropenia. Leukemia is a space-occupying lesion that reduces the bone marrow medullary space available for normal hematopoiesis. This can result in pancytopenia. The normal hemoglobin, platelet count, and absence of a palpable spleen all would suggest that leukemia is not the etiology of this child's acute illness.

A purpuric rash rising up the legs to the buttocks would suggest Henoch-Schönlein purpura (HSP). This is an inflammatory disease of the small blood vessels. While a rash is the most typical sign, it can also cause renal failure. Henoch-Schönlein purpura is not associated with high, spiking fevers or neutropenia. The clinical picture for the boy in this vignette is not consistent with HSP. Although the neutropenia in this vignette is most likely caused by viral suppression of neutrophil production and is most likely transient, other possible causes of the neutropenia include autoimmune neutropenia and congenital neutropenia. Autoimmune neutropenia is most common in the first 2 years of life. Children with autoimmune neutropenia tend not to develop invasive bacterial infections and it typically resolves spontaneously over months to years. To assess the risk of neutropenia being a presentation of congenital neutropenia (for example, severe congenital neutropenia), it would be important to ascertain whether the child has ever had a normal neutrophil count. No mention of prior bacterial infections was made in the vignette, however, it is certainly possible that an 18-month-old boy could present with an undiagnosed congenital neutropenia. Severe congenital neutropenia (SCN) is most often caused by a mutation in the ELANE gene (19p13.3). If there is suspicion for SCN, the gene can be sequenced to establish the diagnosis.

Although the child in the vignette is not reported to be taking any medications, there are many drugs that have neutropenia as a potential adverse effect. These include many antimicrobials, anti-epileptics, chemotherapeutics, immunosuppressives, and many others. It is important to review every medication and supplement the patient is taking to ascertain the risk of drug-induced neutropenia. Often, the treatment is to stop or dose reduce the offending agent.

PREP Pearls
- Neutropenia with a normal hemoglobin and normal platelet count during a febrile illness is most likely transient viral suppression.
- Severe congenital neutropenia and autoimmune neutropenia should be included in the differential diagnosis of an 18-month-old patient presenting with severe neutropenia.
- Severe congenital neutropenia is often caused by a mutation in the ELANE gene (19p13.3).

ABP Content Specifications(s)
- Understand the association of drug therapy with neutropenia
- Recognize the association of common viral infections with transient neutropenia
- Understand the infection risks associated with neutropenia

Suggested Readings
- Boxer LA. Neutrophil abnormalities. Pediatr Rev. 2003;24(2):52-62. DOI: http://dx.doi.org/10.1542/pir.24-2-52.
- Segel GB, Halterman JS. Neutropenia in pediatric practice. Pediatr Rev. 2008;29(1):12-24. DOI:http://dx.doi.org/10.1542/pir.29-1-12.
- Walkovich K, Boxer LA. How to approach neutropenia in childhood. Pediatr Rev. 2013;34(4):173-184. DOI:http://dx.doi.org/10.1542/pir.34-4-173.

Question 211

You have been taking care of a 10-year-old boy for most of his life, along with his 3 siblings. Unfortunately, this young boy developed acute myelogenous leukemia around 8 years of age. Throughout his treatment, you have continued to be in close contact with his family. Today, you received a notice from the local oncology team that he died from complications of his disease. In addition to dealing with your own internal reactions to this loss, you begin to wonder how best to interact with his family going forward.

Of the following, the MOST appropriate action for you to take with this family is

A.   ask if the family would come in to see you in about a month

B.   avoid bringing up any reminders of this boy at future appointments with his siblings

C.   explain how the Kübler-Ross grief stages will be occurring for each of his siblings

D.   offer the advice that children younger than 5 years of age should not attend a funeral

E.   refrain from expressing any tearful emotions when interacting with his parents

Question 211   **Preferred Response: A**

The death of the 10-year-old boy in this vignette will likely provoke very different reactions from his parents, 2 siblings, and care providers. How care providers approach the death of a patient is likely to be influenced by personal feelings rather than just what the family may need the most from the provider. If the provider is not aware of this, personal discomfort with a child's death can lead to actions that they might not otherwise make. For instance, because of personal discomfort, a provider may avoid discussing a child's death. Personal feelings of anger or guilt about the death of a patient are appropriate to share with colleagues who may have experienced the same thing.

What helps a grieving family is for a pediatrician to listen and support them during their process of loss and bereavement. Scheduling an appointment with the family about 1 month after the death of the child provides a good time to address coping concerns, and to reinforce with parents that their decisions were thoughtful and based on what was best at the time. Providers should also address the siblings directly in order to be someone for them who is not embarrassed to talk about the deceased child and the changes in their lives.

The Kübler-Ross stages of grief are very commonly referenced in popular culture, but should not be viewed as a dogmatic step by step process for everyone going through grief. While the "stages" of denial, anger, bargaining, depression, and acceptance are clearly recognizable grief reactions, not everyone experiences them. In fact, a more current view is that most children and adults are quite resilient to loss, particularly if they live in a supportive personal environment, and therefore might not exhibit any external symptoms of grief. There is neither any specific need for people to progress through the Kübler-Ross stages of grief, nor a reason to believe there is something wrong with a child that does not exhibit significant grief reactions.

The decision of whether or not a child should attend a funeral is often a challenging one for families. There is no right or wrong answer, nor should it be based on the child's age alone. Conversant children can be given the option of attending, but should first be told what they are likely to see and hear. If a child does attend a funeral, a supportive person should be available to them to be responsive to their needs and who would be willing to leave with them should the child wish to do so. Funerals are occasions for the living to process their loss and not every child will benefit from that experience.

Parents usually appreciate a treatment provider expressing emotion about a child's death. A pediatrician's affect and attitude can leave a lasting impression, particularly at the time of death. Whether or not their care provider expresses emotion, there is significant value in just being present with the family to provide support without having to worry about finding just the right thing to say.

Children understand death through their developmental level, personal experiences, and the family's religious and cultural beliefs. Children younger than 2 years of age may not understand death, but they do understand separation and the emotions of others. Preschool age children will develop an understanding about death as different from being alive, but might still see death as

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temporary or wonder what dead people "do." Elementary school age children understand that because others can die that they too can die, generally understand the permanence of death, and some may have a stage of being preoccupied with the concept of death.

PREP Pearls
- Scheduling an appointment with the family about 1 month after the death of a child provides a good time to address coping concerns.
- Children can be given the option of attending of attending a funeral, but should first be told what they are likely to see and hear while they are there.

ABP Content Specifications(s)
- Recognize the stages of grief for a patient and family in response to the death of a loved one
- Understand the response to death in patients of various ages, and manage appropriately
- Understand the pediatrician role after the death of a patient
- Understand the various responses of family members to a life-threatening or terminal illness

Suggested Readings
- Linebarger JS, Sahler OJZ, Egan KA. Coping with death. Pediatr Rev. 2009;30(9):350-356. DOI:http://dx.doi.org/10.1542/pir.30-9-350.

Question 212

A 10-month-old male infant is brought to the office with 6 days of fever and a new rash. His mother states that he has been difficult to feed for 2 days because he is so irritable. He usually breastfeeds for 20 min every 3 hours, but has only been feeding for 5 min every 2 hours. He has had 2 wet diapers in the last 24 hours. He has not been given any medication.
No one else is ill at home. The infant's family is Asian American and he is in day care 3 days a week.

Vital signs show a temperature of 39.5°C rectally, respiratory rate of 30 breaths/min, heart rate of 160 beats/min, and a blood pressure of 90/65 mm Hg. Physical examination shows an alert, but very fussy infant. His conjunctiva are injected, but there is no purulent drainage. The lips are red and cracked. There are no mouth ulcers. His tongue is red with white papilla. The neck, chest, and cardiac examinations are unremarkable. The liver edge is at the right coastal margin. No spleen is palpated. His hands and feet have mild edema. There is an erythematous maculopapular rash on his trunk and arms.
Laboratory results are as follows:
- White blood cells, 10,500/µL (10.5 x 10^9/L) with 65% neutrophils, 25% lymphocytes, 10% atypical lymphocytes
- Hemoglobin, 9.5 g/dL (95 g/L)
- Hematocrit, 30.1%
- Erythrocyte sedimentation rate, 60 mm/h
- C-reactive protein, 4.5 mg/L
- Urinalysis shows white blood cells
- Alanine aminotransferase, 50 U/L
- aspartate aminotransferase, 45 U/L

The patient is admitted to the hospitalist service.

Of the following, the BEST next combination of diagnostic and therapeutic maneuvers is

A.   blood culture and intravenous azithromycin

B.   echocardiogram and intravenous immunoglobulin

C.   lumbar puncture and intravenous ceftriaxone

D.   throat culture and intravenous penicillin

E.   urine culture and intravenous ampicillin

Question 212   **Preferred Response: B**

The patient described in this vignette has the symptoms of Kawasaki disease (KD) and is in a high risk demographic group for development of complications. The laboratory data is consistent with KD including pyuria, elevated inflammatory makers, and evidence of hepatic involvement. For the boy in this vignette, the best next step in diagnosis and therapy is to obtain an echocardiogram and treat with intravenous immunoglobulin (IVIG).

As the etiology is still not known, there is no definitive diagnostic test available for KD and the diagnosis must be made on clinical grounds with supporting laboratory tests. Kawasaki disease is more common in boys. The major symptom of KD is fever for at least 5 days. Four additional clinical signs and symptoms are needed to make the diagnosis of typical KD, including cervical lymphadenopathy, red and cracked lips or strawberry tongue, nonpurulent conjunctivitis, a polymorphous rash, and redness and edema of hands and feet, with peeling of the fingers and toes in the later part of the illness. If more than 4 of the principal symptoms are present along with fever, the diagnosis may be made before the fifth day of the illness.

The differential diagnosis of KD includes scarlet fever secondary to group A streptococcal infection or a viral infection such as adenovirus or enterovirus. In KD, one would not expect to see evidence of purulent pharyngitis. If less than 5 clinical criteria are met, the diagnosis may be especially challenging, and the patient may have atypical or incomplete KD (Item C212A). In this situation, an echocardiogram is helpful in making the diagnosis. Echocardiogram is not essential to make the diagnosis of typical KD when there are adequate criteria. The urgency in making the diagnosis of KD is related to the possible development of an inflammatory process that includes the coronary arteries. In KD, there is a 25% risk of development of coronary artery aneurysms (CAA) (Item C212B), which can be decreased to 4% by the administration of IVIG and high dose aspirin (ASA).

The highest risk for development of CAA is in the first 2 weeks of the illness, and administration of IVIG is needed within the first 10 days of febrile illness to help prevent that sequelae. Coronary artery (CA) findings are not required to make the diagnosis of KD, although this may be helpful in making a diagnosis in patients with atypical KD. Extreme irritability, sterile pyuria, and thrombocytosis are frequently seen in KD, but are not 1 of the 5 principle findings used for the diagnosis of KD.

Prior to the use of IVIG, thrombocytosis with platelet counts up to 1 million/µL were seen and thought to contribute to the damaging effects of coronary artery vasculitis. Patients with persistent fever despite treatment with IVIG are at a higher risk of CAA development. A second dose of IVIG, infliximab, or steroids, as well as continuation of high dose ASA, are among the recommendations in this situation. In less complicated cases where the patient becomes afebrile after initial IVIG administration, the high dose ASA is decreased to low dose ASA.

Recommendations for further treatment and follow-up depend on the presence or absence of CAA or the less severe finding of CA ectasia. Follow-up at 2 weeks and 6 weeks after discharge is recommended, as a minimum, even if no coronary involvement was seen initially.

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If CAA have been identified, more intense follow-up will be needed. Cardiac stress testing and angiography may be required in these cases. If the CAA are large, anticoagulation will be needed. Over time, CAA can cause CA stenosis and myocardial ischemia.

A rapid diagnosis of KD must be made because of the risk of CAA. Diagnosis and treatment of KD with IVIG should not be delayed while waiting for an echocardiogram. Treatment with IVIG will help prevent development of CAA. A very high risk group for development of CAA are younger male children of Asian heritage, such as described in the vignette. With or without CAA, patients require cardiology follow-up.

For uncomplicated KD, an echocardiogram is recommended at 2 weeks and again at 6 to 8 weeks. Echocardiograms after this period of time, if initially normal, are not likely to show any new coronary artery abnormalities. Long term planning includes delaying live virus vaccines, such as measles and varicella, for 11 months after treatment with IVIG.

The infant in this vignette does not have respiratory symptoms, therefore azithromycin for pneumonia would not be the ideal treatment for him. Irritability would create concern for meningitis, but the other clinical findings would not make that the most likely diagnosis. A urinary tract infection would not cause the combination of findings described. Kawasaki disease does share several similar clinical findings with scarlet fever. The rash in that case is more likely to be "sand paper" in appearance. This and the additional findings of pyuria without bacteria in this infant would make a throat culture and penicillin not the next step in this patient.

PREP Pearls
- Kawasaki disease (KD) is a clinical diagnosis that needs to be made as soon as possible to allow initiation of intravenous immunoglobulin treatment and prevent development of coronary artery aneurysms.
- An echocardiogram is needed to evaluate for coronary artery involvement, but not to make the diagnosis, except in cases that do not meet criteria for typical KD.

ABP Content Specifications(s)
- Identify cardiac complications associated with Kawasaki disease and how to prevent their occurrence

Suggested Readings
- American Academy of Pediatrics. Kawasaki disease. In: Kimberlin DW, Brady MT, Jackson MA, Long SS, eds. Red Book:2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:494-500.
- Burns JC, Kushner HI, Bastian JF, et al. Kawasaki disease: a brief history. Pediatrics. 2000;106(2):e27.http://pediatrics.aappublications.org/content/106/2/e27.full.pdf+html

Question 213

You are seeing a 12-year-old girl in your office with complaint of generalized pain and back pain that is worsened by exercise. The girl has muscle aches and joint pain that she is not able to localize well. She has noticed recurrent ankle swelling that lasts a few hours with each episode. She rates the pain as a 3 out of 10 at its mildest, and 7 out of 10 at its worst. The girl has pain daily, which is not associated with time of day, and is worsened by exercise. Her review of symptoms is positive for fatigue, headache, difficulty sleeping, abdominal pain with cramping, alternating loose stools and constipation, and occasional mouth sores. She has missed several days of school because of pain, but has kept up with her schoolwork. When asked about changes at home, the girl states that her parents are going through a divorce.

Of the following, the BEST next step in the evaluation and management of this girl would be a(n)

A.   erythrocyte sedimentation rate

B.   human leukocyte antigen-B27 test

C.   referral to a cognitive behavioral therapist

D.   referral to a gastroenterologist

E.   referral to a physical therapist

Question 213   **Preferred Response: A**

For the girl in the vignette, the best choice is to order an erythrocyte sedimentation rate. Any patient with unexplained pain deserves an evaluation for disorders that may be causing those symptoms. The patient in the vignette has an amplified pain syndrome (APS), for which there is no diagnostic test. Pain syndromes can mimic other diseases, and symptoms may overlap those of other conditions such as juvenile idiopathic arthritis, systemic lupus erythematosus, multiple sclerosis, infections, and cancers. Therefore, any patient with a suspected diagnosis of APS must have a workup that rules out these other conditions. Testing for human leukocyte antigen B-27 (HLA-B27) would be appropriate if ankylosing spondylitis (AS) were suspected, however, this patient's back pain is worsened by exercise. In contrast, the inflammatory back pain seen in AS is improved with exercise and is not generalized. Cognitive behavioral therapy has been shown to improve chronic pain, but should not be initiated until organic causes of pain have been ruled out. Inflammatory bowel disease can cause joint pain, but usually does not present with constipation and a laboratory evaluation would likely be performed before referral to a gastroenterologist. Although physical therapy can benefit patients with chronic musculoskeletal pain, other conditions must be ruled out before referral for therapy.

The pain associated with APS can be localized or widespread. There may be skin color changes or swelling that comes and goes. The onset of APS may be acute or triggered by a preceding injury, and symptoms may increase overtime. Patients with APS often are affected by widespread musculoskeletal pain; chronic fatigue; sleep disturbance; and cognitive and mood disorders. Patients with chronic pain often display other somatic symptoms such as headache, dysautonomia, subjective soft tissue swelling, and irritable bowel syndrome. There are several subtypes of amplified pain syndromes.

Complex regional pain syndrome presents as continuing pain that is disproportionate to the inciting event. Some cases are preceded by nerve injury, but this may be absent. Categories include sensory dysfunction, such as hyperalgesia and/or allodynia, and vasomotor dysfunction, presenting with temperature asymmetry or sweating changes, and patient-reported edema. There must be no other diagnosis that better explains these signs or symptoms.

Juvenile fibromyalgia has 4 major diagnostic criteria including generalized musculoskeletal aching in 3 or more sites for 3 or more months, absence of an underlying cause for pain, normal laboratory tests, and 5 or more tender points. There are 10 minor criteria, including chronic anxiety; fatigue; poor sleep; chronic headaches; irritable bowel syndrome; subjective soft tissue swelling; numbness; pain modulated by physical activity, weather factors, and anxiety or stress. To meet the criteria for the diagnosis of juvenile fibromyalgia, the patient must have 4 major and 3 minor criteria, or the first 3 major criteria along with 4 painful sites and 5 minor criteria. Diffuse idiopathic pain is characterized by generalized musculoskeletal aching at 3 or more sites for 3 or more months and exclusion of any disease that explains the symptoms.

Localized idiopathic pain is characterized by pain localized to 1 limb that persists for 1 week with treatment or 1 month without treatment, no history of trauma or disease that can explain the symptoms.

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Several conditions may occur concurrently with APS, including irritable bowel syndrome, chronic fatigue syndrome, interstitial cystitis, chronic headache, functional abdominal pain, or conversion disorders. Psychosocial factors such as stress, injury, resilience, academic factors, peer relationships, family relationships, parenting style, and the medical system's response to the pain can have a significant effect on the level of pain in APS. Several psychological factors can also have an effect on pain in APS patients, such as pain avoidance or acceptance, catastrophizing, coping, cognition, intelligence, motivation and perception.

The approach to treatment for APS should be multidisciplinary. Helpful approaches include exercise, desensitization, stress management, counseling, and self-regulation with purposeful relaxation.

There is little evidence available to guide the treatment of pediatric pain with medication. Several of the medications commonly used effectively in adults are "off label" in the pediatric population. Medications commonly used to treat pediatric pain include acetaminophen, nonsteroidal anti-inflammatory drugs, and Cox-2 inhibitors. Some anticonvulsants, such as gabapentin and pregabalin, have been used to effectively treat chronic pain. Antidepressants have been prescribed, but there is no significant evidence of effectiveness. The most commonly used antidepressants are amitriptyline and nortriptyline. Duloxetine has been used, but is not labeled for pediatric use. Opioids are used to treat pain in patients with cancer, but have limited use for other types of chronic pain, and safety data regarding their long-term use are limited. Other medications, with limited evidence regarding effectiveness, include clonidine, dexmedetomidine, lidocaine, and mexiletine.

PREP Pearls
- Amplified pain syndromes are diagnoses of exclusion. Other conditions must be ruled out before diagnosis.
- Multifactorial treatment including exercise, desensitization, stress management, and counseling should be used in treating patients with amplified pain syndromes.
- Amplified pain syndromes often occur concurrently with irritable bowel syndrome, chronic fatigue syndrome, interstitial cystitis, chronic headache, functional abdominal pain, or conversion disorders.

ABP Content Specifications(s)
- Understand the effects of a patient's developmental stage on tolerating and dealing with pain
- Recognize the behavioral and psychosocial effects of chronic pain syndromes
- Plan the appropriate management of pain in patients of various ages
- Recognize the clinical features commonly associated with chronic pain syndromes

Suggested Readings

- Goodard JM Chronic pain in children and young people. Curr Opin Support Palliat Care. 2011;5(2):158-163. DOI:http://dx.doi.org/10.1097/SPC.0b013e328345832d.
- Hoffart CM, Wallace DP Amplified pain syndromes in children: treatment and new insights into disease pathogenesis. Curr Opin Rheumatol. 2014;26(5):592-603. DOI: http://dx.doi.org/10.1097/BOR.0000000000000097.

Question 214

You are speaking with a group of pediatric residents about exercise in children. You emphasize the importance of routinely discussing the benefits of exercise with young patients and their families.

Of the following, the MOST accurate statement to include in your discussion is that

    A.   children with siblings are more likely to engage in physical activity

    B.   increased physical activity in children correlates with lower rates of coronary artery disease in adults, even if they are not physically active as adults

    C.   physical activity during childhood is correlated with improved bone mineral density in adulthood

    D.   school-based physical activity results in a sustained increase in physical activity outside of school hours

    E.   the majority of high school students in the United States participate in 60 or more minutes of moderate physical activity each day

Question 214   **Preferred Response: C**

Peak bone mineralization occurs during childhood and adolescence. Physical activity during early childhood is correlated with improved bone mineral density in adolescence and adulthood. The Centers for Disease Control and Prevention recommend that school-aged children perform 60 minutes per day of moderate to vigorous physical activity. Only one-third of high school students meet these guidelines, with female, black, and Latino adolescents the least likely to achieve this. In addition, there is a drop-off in physical activity between adolescence and adulthood. Physically active children are more likely to be physically active adults.

Recess, walking, or biking to school, after-school activity programs, and other community recreation programs can all boost physical activity levels. Children are more likely to participate in organized sports if their parents support their athletic endeavors and when their peers participate in sports. Lower socioeconomic status has been correlated with lower rates of sports participation, whereas the availability of inexpensive, easily accessible sports opportunities in the community has been positively correlated with sports participation. Pediatricians can promote physical activity by checking that gross motor skills are being learned in early school-age children and by providing developmentally appropriate activity guidelines and resources to families.

Children with siblings are no more likely than only children to engage in regular physical activity. While physically active adults have lower rates of heart disease, there is no direct link between physical activity during childhood and coronary disease in adulthood. Studies have shown that some school-based physical activity programs can improve fitness, increase the amount of time children and teens participate in physical activity, and lead to decreased screen time, but these effects are not sustained when the school programs end.

PREP Pearls
- Physical activity during early childhood is correlated with improved bone mineral density in adolescence and adulthood.
- Most children and adolescents do not meet recommended levels of physical activity.

ABP Content Specifications(s)
- Understand the importance of regular exercise to promote good general health
- Identify factors that influence participation in contact sports by healthy children and adolescents

Suggested Readings
- Dobbins M, Husson H, DeCorby K, LaRocca RL. School-based physical activity programs for promoting physical activity and fitness in children and adolescents aged 6 to 18. Cochrane Database Syst Rev. 2013;2:CD007651. DOI:http://dx.doi.org/10.1002/14651858.CD007651.pub2.

Question 215

A 6-year-old boy presents to your office with a several month history of worsening nasal congestion, thick nasal discharge, and cough disrupting sleep, despite multiple courses of antibiotics. The boy has asthma and uses an inhaled steroid appropriately for maintenance therapy, but recently has been having frequent exacerbations. Although recent testing did not reveal allergies, he has been using nasal steroid sprays and oral antihistamines. He has a history of multiple upper respiratory tract infections and foul smelling stools.

On physical examination, the boy's temperature is 37°C, heart rate is 110 beats/min, respiratory rate is 20 breaths/min, blood pressure is 95/65 mm Hg, and weight is 22 kg (tenth percentile). Examination of the head and neck reveals deviation of the nasal septum to the right, hypertrophy of the nasal turbinates with mucopurulent discharge, and nasal polyps in the left naris. The tympanic membranes are normal. The posterior oropharyngeal mucosa has a cobblestone appearance and the tonsils are enlarged without exudates. Auscultation of the lungs is significant for a prolonged expiratory phase with mild, intermittent wheezing throughout.

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

A.   adenoidal and tonsillar hypertrophy

B.   defective phagocytosis

C.   dysfunction of a transporter protein

D.   HIV infection

E.   poorly controlled asthma

Question 215   **Preferred Response: C**

The boy in the vignette has a history (chronic respiratory tract problems and infections, foul-smelling stools) and physical examination findings (nasal polyps, sinusitis, prolonged expiratory phase and wheezing, poor growth) suggestive of cystic fibrosis. Cystic fibrosis is caused by a mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) protein, which is found in all exocrine cells and regulates the transport of chloride and other ions. In addition to lower respiratory and gastrointestinal complications, most children with cystic fibrosis develop chronic sinusitis, and up to 35% have nasal polyposis. Sinusitis in children with cystic fibrosis or immune compromise can be resistant to cure.

Chronic sinusitis is an inflammatory disorder of the sinuses and nasal passages that lasts 12 weeks or longer. In children, the signs and symptoms of chronic sinusitis include mucopurulent nasal drainage, cough, nasal congestion or obstruction, and facial fullness or pain. Many factors may contribute to or exacerbate chronic sinusitis including allergic rhinitis, immunodeficiency or other systemic diseases (eg, granulomatosis with polyangiitis), defects in mucociliary clearance (eg, cystic fibrosis), environmental irritants, recurrent viral infections, and anatomic abnormalities.

Although allergic disorders are commonly implicated in chronic sinusitis, the boy in the vignette does not have a proven allergy and has not responded to nasal corticosteroid sprays and oral antihistamines. Allergic fungal sinusitis is an uncommon form of chronic sinusitis, often mistaken for a paranasal sinus tumor, caused by an allergic reaction to aerosolized environmental fungi in an immunocompetent individual. Surgery and immunomodulation are required for treatment.

Adenoidal hypertrophy is associated with Eustachian tube dysfunction and rhinosinusitis, however tonsillar hypertrophy is not. The efficacy of adenoidectomy to treat chronic sinusitis is uncertain, and should only be considered after evaluating the child for underlying conditions (eg, immune deficiency, ciliary defect).

Immune disorders that cause defects in phagocytosis can present with sinusitis (eg, leukocyte adhesion defect, Shwachman-Diamond syndrome), but they are rare and typically also associated with cutaneous and gastrointestinal infections in the first years after birth. Humoral immune disorders, such as immunoglobulin A deficiency and common variable immunodeficiency, occur more commonly and can present with disease isolated to the respiratory tract. Infection with human immunodeficiency virus (HIV) can cause recurrent bacterial infections, but it would be unlikely that chronic, refractory bacterial infection would occur in the respiratory tract without other manifestations of HIV infection (eg, diarrhea, wasting, fungal infections, viral infections). There is a strong association between asthma and chronic sinusitis; approximately 20% of patients with chronic sinusitis have asthma and nearly two-thirds of patients with asthma have sinus disease. Evidence demonstrates that treatment of sinusitis may improve asthma symptoms, but the converse is unproven.

Acute rhinosinusitis usually is caused by a viral infection. In select patients, a secondary bacterial infection develops. Streptococcus pneumoniae, Haemophilus influenzae, and Moraxella catarrhalis are the most common pathogens causing acute bacterial sinusitis. In patients with chronic sinusitis, Staphylococcus aureus and oropharyngeal anaerobes can contribute to disease. Pseudomonas aeruginosa most commonly occurs in immunocompromised hosts and patients with cystic fibrosis. Patients with diabetes and immune compromise can develop invasive fungal sinusitis. Rarely, sinusitis caused by Mycobacteria or parasites occurs in patients with HIV infection. The treatment of chronic sinusitis includes promoting sinus drainage, reducing inflammation, and antimicrobial therapy directed toward the causative pathogens. Medical therapy typically is prolonged (≥3 weeks), and sinus surgery may be indicated for severe or refractory disease.

PREP Pearls
- Cystic fibrosis is caused by a mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) protein, which is found in all exocrine cells and regulates the transport of chloride and other ions.
- Chronic sinusitis is an inflammatory disorder of the sinuses and nasal passages that lasts 12 weeks or longer.
- The signs and symptoms of chronic sinusitis include mucopurulent nasal drainage, cough, nasal congestion or obstruction, and facial fullness or pain.
- Factors that may contribute to or exacerbate chronic sinusitis include allergic rhinitis, immunodeficiency, mucociliary clearance defects, environmental irritants, recurrent viral infections, and anatomic abnormalities.

ABP Content Specifications(s)
- Recognize factors predisposing to chronic sinusitis in patients of various ages

Suggested Readings
- Cherry JD, Mundi J, Shapiro NL. Rhinosinusitis. In: Cherry J, Demmler-Harrison GJ, Kaplan SL, Steinbach WJ, Hotez P, eds. Feigin & Cherry's Textbook of Pediatric Infectious Diseases. 7th ed. Philadelphia, PA: Elsevier Saunders; 2014:193-203.
- Desrosiers M, Evans GA, Keith PK, et al. Canadian clinical practice guidelines for acute and chronic rhinosinusitis. Allergy Asthma Clin Immunol. 2011;7:2. DOI: http://dx.doi.org/10.1186/1710-1492-7-2.

Question 216

A community hospital calls the pediatric emergency department where you are working seeking to transfer an 11-year-old boy with a grade IV laceration to his left kidney. This injury was sustained when the boy fell 10 feet from a tree, landing on his left side. The transferring physician asks for your recommendation regarding insertion of an indwelling bladder catheter to monitor the boy's urine output prior to transporting him to your hospital.

Of the following, the MOST accurate statement regarding this patient is that

    A.   a bladder catheter should be inserted before performing a rectal examination

    B.   bladder catheterization is contraindicated if prostate enlargement is detected

    C.   bladder catheterization should not be performed if bruising to the perineal area is noted

    D.   the presence of red blood cells on urinalysis is a contraindication to bladder catheterization

    E.   urologic consultation is recommended before bladder catheterization

Question 216   **Preferred Response: C**

The boy in the vignette sustained a left kidney laceration after falling from a tree. The most accurate statement regarding his management is that bladder catheterization should not be performed if bruising to the perineal area is present.

All pediatric providers should recognize the contraindications to bladder catheterization after acute trauma. Insertion of urinary catheters in patients with traumatic injuries can help to relieve urinary retention, decompress the bladder, and allow for monitoring of urinary output, which serves as an index of tissue perfusion. Urinary catheters should not be inserted, however, if any of the following contraindications are present: gross hematuria, inability to void, unstable pelvic fracture, blood at the urethral meatus, perineal ecchymoses or scrotal hematoma, or a high-riding or nonpalpable prostate on rectal examination. For patients presenting with any of these contraindications, retrograde urethrography must be performed to confirm an intact urethra before urinary catheter insertion.

A rectal examination should always be performed in patients presenting after abdominal and pelvic trauma beforeinsertion of a bladder catheter. Goals of the rectal examination include assessment of sphincter tone and integrity of the rectal mucosa, determination of prostate position in male patients (high-riding prostate suggests urethral disruption), assessment for gross blood (which may indicate bowel perforation), and identification of pelvic fractures. In female patients, a vaginal examination should also be performed to exclude lacerations from bony fragments from pelvic fractures or from penetrating wounds, prior to urinary catheterization. Prostate enlargement is not a contraindication to bladder catheterization. High-riding position of the prostate or the inability to palpate the prostate indicates disruption of the urethra and are contraindications.

The presence of red blood cells on urinalysis in patients presenting for evaluation of traumatic injuries who do not have gross hematuria or blood at the urethral meatus is not a contraindication for bladder catheterization.

Urologic consultation is not needed for patients without any of the previously discussed contraindications to urinary catheter placement.

PREP Pearls
- Insertion of urinary catheters in patients presenting with traumatic injuries can help to relieve urinary retention, decompress the bladder, and allow for monitoring of urinary output.
- Urinary catheters should not be inserted in patients who have experienced an acute trauma and have any of the following contraindications: gross hematuria, inability to void, unstable pelvic fracture, blood at the urethral meatus, perineal ecchymoses, scrotal hematoma, or a high-riding or nonpalpable prostate on rectal examination.
- A rectal examination should always be performed in patients presenting after abdominal and pelvic trauma before insertion of a bladder catheter.

ABP Content Specifications(s)
- Recognize the contraindications to bladder catheterization following acute renal trauma

Suggested Readings
- American College of Surgeons Committee on Trauma. Abdominal and pelvic trauma. Advanced Trauma Life Support Student Course Manual. 9th ed. Chicago, IL: American College of Surgeons; 2012:122-140.
- Garcia CT, Thompson VT. Genitourinary trauma. In: Fleisher GR, Ludwig S, eds. Textbook of Pediatric Emergency Medicine. 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2010:1316-1327.

Question 217

You are called to see a full-term newborn who is found to have abdominal distension with delayed passage of stool at 28 hours after birth. The pregnancy was remarkable for diet-controlled gestational diabetes and echogenic bowel on the second trimester ultrasonography. The newborn has been breastfeeding well with good urine output and no passage of stool. Physical examination reveals an alert, active newborn with a markedly distended, firm abdomen, and an externally patent rectum. In the course of the subsequent evaluation, a barium enema is performed (Item Q217).

Item Q217

Item Q217. Radiograph for the newborn described in the vignette. Courtesy of Donald Frush, MD.

Of the following, the test that is MOST likely to reveal the underlying cause of this newborn's clinical and radiographic findings is a

A.   chromosome analysis

B.   maternal hemoglobin A1c level

C.   rectal biopsy

D.   sweat chloride test

E.   urine cytomegalovirus culture

Question 217   **Preferred Response: D**

Sweat chloride testing is likely to reveal cystic fibrosis as the underlying cause of the clinical and radiographic findings seen in the newborn in the vignette. The prenatal finding of echogenic bowel is nonspecific and may be seen in up to 1% of fetuses on second trimester ultrasound. Although in most cases, the infant is normal at birth, echogenic bowel can be associated with pathologic conditions including meconium ileus and chromosome anomalies. This infant's barium enema (Item C217A) demonstrates meconium mixed with air in the right lower abdomen, a finding highly suggestive of meconium ileus, which most often occurs in infants with cystic fibrosis. Fifteen percent of newborns with cystic fibrosis have meconium ileus.

Item C217A

Item C217A. Barium enema demonstrating stool mixed with air in distal ileum and distal microcolon suggesting meconium ileus. Courtesy of Donald Frush, MD.
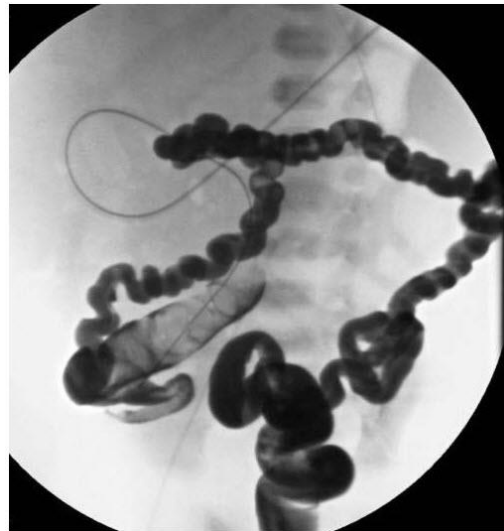
More than 99% of healthy full-term newborns will pass a stool within 48 hours after delivery. Intestinal obstruction should be suspected in a newborn with delayed passage of stools, abdominal distension, and vomiting. The initial assessment should include a physical examination and plain abdominal radiography. This is often unable to distinguish an emergent condition such as midgut volvulus from a condition responsive to conservative care, therefore, a newborn suspected of having an intestinal obstruction should be evaluated by a pediatric surgeon. The decision, made in consultation with both a pediatric surgeon and a radiologist, about whether to perform an upper gastrointestinal series or a contrast enema will be guided by the suspicion of either an upper or lower bowel obstruction.

Lower bowel obstruction is associated with delayed passage of stool (Item C217B). With the exception of anorectal malformations, a contrast enema is generally the best diagnostic study to perform. A recent review supports an association of cystic fibrosis with meconium ileus, but not meconium plug syndrome. If present, the location of meconium plugs will distinguish meconium plug syndrome/small left colon syndrome from meconium ileus. Because 13% of patients with meconium plug syndrome are subsequently found to have Hirschsprung disease, newborns with meconium plugs should have further evaluation if issues with passage of stool persist.

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Item C217B. Diagnoses and Findings Associated with Delayed Passage of Stools in the Newborn.

Diagnosis	Findings
Meconium plug syndrome	Meconium plugs located in the colon; plugs may be passed with rectal stimulation
Meconium ileus	Abdominal distension at birth; plugs located in the ileum with microcolon distally
Small left colon syndrome	Small caliber descending colon that resolves over time; 50% of cases are infants of diabetic mothers
Hirshsprung disease	Transition zone seen on barium enema in 75% of patients; delayed passage of contrast. Rectal punch biopsy demonstrates absence of ganglion cells
Anorectal malformation	Abnormal physical examination, ultrasound, or contrast imaging

Courtesy of S. Izatt

The newborn in the vignette has clinical and radiographic features suggestive of meconium ileus, which is likely the result of cystic fibrosis, and a sweat test should be performed for diagnosis. Chromosome disorders can be associated with bowel obstruction, but typically not with meconium ileus. A maternal hemoglobin A1c level would not be useful, because the barium enema does not demonstrate localized small caliber of the descending colon as would be seen in small left colon syndrome. There is no evidence of a transition zone on the barium enema that would suggest Hirschsprung disease; therefore a rectal biopsy would be unlikely to yield a diagnosis. Congenital cytomegalovirus has been associated with a pseudo-Hirschsprung disease presentation, but this is not supported by the findings on the barium enema.

PREP Pearls
- Intestinal obstruction should be suspected in a newborn with delayed passage of stools, abdominal distension, and vomiting.
- A newborn suspected of having an intestinal obstruction should be evaluated by a pediatric surgeon, because the physical examination and plain abdominal radiography may not distinguish an emergent condition such as midgut volvulus from a condition responsive to conservative care.

ABP Content Specifications(s)
- Recognize the clinical and laboratory features associated with intestinal obstruction in a newborn infant, and manage appropriately

Suggested Readings
- Keckler SJ, St Peter SD, Spilde TL, et al. Current significance of meconium plug syndrome. J Pediatr Surg. 2008;43:896-898. DOI: http://dx.doi.org/10.1016/j.jpedsurg.2007.12.035.
- Levine D. Overview of echogenic masses and calcifications in the fetal abdomen. UpToDate. Available online only for subscription.

Question 218

A 2-day-old male neonate is being cared for in the well-baby nursery. His mother tells the nurse that she is concerned about his vision. She notices that he stares to the side for 20 to 30 seconds every couple of hours. There are no facial twitches, cyanosis, or other symptoms. He was born by normal spontaneous vaginal delivery at 39 weeks' gestational age, with no complications during pregnancy, labor, or delivery. His Apgar scores were 8 and 9 at 1 and 5 minutes, respectively. His birth weight was 3.2 kg and head circumference was 34.5 cm. His physical and neurologic examinations are unremarkable. The neonate is transferred to the neonatal intensive care unit where treatment with antibiotics and acyclovir are started. His initial laboratory evaluation is normal, including complete blood cell count with differential, comprehensive metabolic panel, serum herpes simplex virus polymerase chain reaction (PCR), and urinalysis. Lumbar puncture results are normal, including cerebrospinal fluid protein, glucose, cell count with differential, and herpes simplex virus PCR. Blood, urine, and cerebrospinal fluid cultures are pending.

Of the following, the MOST likely to reveal the cause of the neonate's symptoms is a(n)

A.   magnetic resonance imaging study of the brain

B.   ophthalmology consultation

C.   urine cytomegalovirus culture

D.   urine homovanillic acid and vanillylmandelic acid levels

E.   urine organic acid analysis

Question 218   **Preferred Response:A**
The neonate in the vignette is having episodes of sustained tonic horizontal eye deviation. This is a common manifestation of seizure in full-term neonates. Initial laboratory evaluation shows no evidence of infection, electrolyte abnormality, or acidosis, so the most likely cause of seizure in this infant is brain malformation or brain injury. Of the response choices, the test most likely to reveal a cause for his symptoms is magnetic resonance imaging (MRI) of the brain.
In the neonatal period, seizures commonly manifest with abnormal eye movements, such as sustained lateral gaze, staring, or nystagmus. Other manifestations include focal facial twitching, focal limb jerking, lip smacking, or unilateral limb posturing. Electroencephalogram can be normal in neonates, even during a seizure, because of immature brain development and immature myelination.

The most common cause of neonatal seizure is hypoxic ischemic encephalopathy. Other common causes of neonatal seizure include intracranial hemorrhage, infection, and brain malformation. Less common causes of seizure in the neonatal period include neonatal stroke, hypoglycemia, electrolyte abnormalities (eg, hypocalcemia, hyponatremia, or hypernatremia), inborn errors of metabolism, and epilepsy syndromes.

There are many treatable causes of neonatal seizure, and time to diagnosis and treatment can be critical, therefore the initial diagnostic evaluation is broad. This evaluation includes laboratory testing for infection and electrolyte and metabolic abnormalities. Brain infection can be devastating; therefore empiric antibiotics and antiviral treatment should be started urgently. Anticonvulsant treatment should also be started when clinical suspicion for seizure is high. Brain imaging is almost always performed in these cases; head ultrasonography or computed tomography is often performed first because of their availability, but MRI is preferred.
For the neonate in the vignette, ophthalmology consultation is unlikely to be helpful in determining the cause for seizures. Urine cytomegalovirus (CMV) culture would be appropriate if there were signs suggestive of congenital CMV infection, such as microcephaly, hepatomegaly, characteristic skin rash, thrombocytopenia, or abnormal liver function tests. Urine homovanillic acid and vanillylmandelic acid are elevated in neuroblastoma, which presents with opsoclonus, myoclonus, and ataxia. Opsoclonus is an involuntary, conjugate, random, jerky eye movement, which this neonate does not have. Urine organic acids can be abnormal in some inborn errors of metabolism, which can present with neonatal seizures. However, the lack of acidosis makes an organic acidemia unlikely in this case.

PREP Pearls
- Tonic horizontal eye deviation is a common manifestation of seizure in full-term neonates.
- The diagnosis of seizure is often made clinically in preterm and full-term neonates, because an electroencephalogram can be normal even during a seizure at this age.

ABP Content Specifications(s)
- Understand the various etiologies of neonatal seizures and plan appropriate diagnostic evaluation
- Recognize the clinical findings associated with neonatal seizures and manage appropriately

Suggested Readings
- Gillam-Krakauer M, Carter BS. Neonatal hypoxia and seizures. Pediatr Rev. 2012;33;387. DOI:http://dx.doi.org/10.1542/pir.33-9-387.
- Mosley M. Neonatal seizures. Pediatr Rev. 2010;31;127. DOI: http://dx.doi.org/10.1542/pir.31-3-127.

Question 219

An 8-year-old boy with type 1 diabetes mellitus diagnosed 2 years ago presents for a follow-up visit. Review of systems is significant for increased fatigue and constipation, but he is otherwise doing well.

The boy's physical examination is significant for height that has dropped from the tenth percentile last year to less than the third percentile this year. His examination is otherwise unremarkable. There is no goiter, skin changes, or abnormal findings at his insulin injection sites. His hemoglobin A1c level is 6.9%.

Of the following, the MOST likely explanation for his poor growth is

A.   autoimmune hypothyroidism

B.   celiac disease

C.   gastroparesis

D.   pernicious anemia

E.   thyroid cancer

Question 219   **Preferred Response: A**

In addition to presenting with the typical features of type 1 diabetes mellitus, including polyuria, polydipsia, and polyphagia, children with type 1 diabetes are also at risk of developing additional autoimmune diseases. Thyroid disease is the most common autoimmune disorder associated with type 1 diabetes. About one-quarter of children with type 1 diabetes have thyroid autoantibodies at the time of diagnosis or will develop them within a few years of diagnosis. The presence of thyroid autoantibodies is predictive of thyroid dysfunction. Some studies have shown subclinical hypothyroidism to be associated with an increased risk of symptomatic hypoglycemia and reduced linear growth, so careful follow-up of thyroid function in children with type 1 diabetes is warranted.

Current American Diabetes Association (ADA) guidelines recommend that practitioners consider screening children with type 1 diabetes for antithyroid peroxidase and antithyroglobulin antibodies soon after diagnosis. In addition, thyroid-stimulating hormone (TSH) concentrations should be measured soon after diagnosis, once metabolic control has been established. TSH and thyroxine levels while a child is actively experiencing diabetic ketoacidosis or just diagnosed with type 1 diabetes may be most consistent with sick-euthyroid syndrome, and therefore will not be helpful. If these tests are sent shortly after diagnosis and are normal, the ADA guidelines recommend that practitioners consider rechecking them every 1 to 2 years, especially if the patient develops symptoms of thyroid dysfunction, thyromegaly, an abnormal growth rate, or unusual glycemic variation.

The International Society for Pediatric and Adolescent Diabetes (ISPAD) 2009 Consensus Guidelines for Children and Adolescents with Diabetes also recommend screening of thyroid function by measuring TSH and thyroid antibodies at diagnosis and every second year thereafter in asymptomatic individuals without goiter or in the absence of thyroid autoantibodies. More frequent assessment is indicated if antibodies are present or symptoms develop (eg, poor growth or goiter). It is important for pediatricians to recognize that goiter may not be present in children with thyroid disease.

Many other autoimmune conditions are associated with type 1 diabetes. Celiac disease occurs with increased frequency in patients with type 1 diabetes (1%-16% of individuals compared with 0.3%-1% in the general population). Symptoms of celiac disease include diarrhea, weight loss or poor weight gain, growth failure, abdominal pain, chronic fatigue, malnutrition due to malabsorption, and unexplained hypoglycemia or erratic blood glucose concentrations. The ADA recommends that practitioners consider screening children with type 1 diabetes for celiac disease soon after the diagnosis, as well as in cases with a positive family history of celiac disease, symptoms consistent with the diagnosis, or frequent unexplained hypoglycemia or deterioration in glycemic control. ISPAD guidelines are more stringent, and recommend screening at the time of diagnosis, annually for the first 5 years and every second year thereafter. Although the child in this vignette could have celiac disease, hypothyroidism occurs far more commonly and thus is the most likely explanation for his poor growth.

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Gastrointestinal neuropathies, such as gastroparesis, should be considered in individuals with erratic glucose control or with upper gastrointestinal symptoms without other identified cause. However, this is rare in childhood, usually taking many years to develop. Pernicious anemia is also uncommon in children. It is seen more commonly in adults with type 1 diabetes, occurring in up to 8.5% of those with another autoimmune disease, such as hypothyroidism. Similarly, thyroid cancer is relatively uncommon in children.

The degree of glucose control and duration of diabetes mellitus are linked to long-term complication rates. In long-term (20-year) follow-up of patients with type 1 diabetes mellitus, extensive clinical trials have shown that 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. Other outcomes improve as well. Monitoring for associated disorders and long-term glucose control is important to minimize these complications.

PREP Pearls
- Autoimmune thyroid disease is the most common autoimmune disorder associated with diabetes.
- Routine assessment of thyroid autoimmunity and thyroid function should be performed every 1 to 2 years in children with type 1 diabetes, and sooner if the child develops symptoms of thyroid disease, such as poor growth.
- Goiter may not always be present in children with thyroid disease.

ABP Content Specifications(s)
- Recognize the clinical features associated with type 1 diabetes
- Understand the natural history of type 1 diabetes
- Plan the appropriate diagnostic evaluation for new-onset type 1 diabetes

Suggested Readings
- American Diabetes Association. Standards of medical care in diabetes--2014. Diabetes Care. 2014;37(suppl 1):S14-80. DOI: http://dx.doi.org/10.2337/dc14-S014.
- 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: http://dx.doi.org/10.1056/NEJMoa1111732.
- Kordonouri O, Maguire AM, Knip M, et al. ISPAD Clinical Practice Consensus Guidelines 2009 Compendium: Other Complications and Associated Conditions. Pediatr Diabetes. 2009;10(suppl 12):204-210. Available at:http://onlinelibrary.wiley.com/doi/10.1111/j.1399-5448.2009.00573.x/full.
- Perros P, Singh RK, Ludlam CA, Frier BM. Prevalence of pernicious anaemia in patients with Type 1 diabetes mellitus and autoimmune thyroid disease. Diabet Med. 2000 Oct;17(10):749-751. DOI: http://dx.doi.org/10.1046/j.1464-5491.2000.00373.x.
- Vergouwe Y, Soedamah-Muthu SS, Zgibor J, et al. Progression to microalbuminuria in type 1 diabetes: development and validation of a prediction rule. Diabetologia. 2010;53(2):254-262. DOI: http://dx.doi.org/10.1007/s00125-009-1585-3.

Question 220

A 7-month-old infant is brought to your office for a follow-up evaluation. She was recently hospitalized for the third time with bronchiolitis. On each occasion, she has been treated with a short acting ß-agonist and systemic steroids. There is no history of pneumonia or recurrent otitis media. The infant was full term at birth. The parents report that they have heard "whistling and rattling" in their infant's chest since she was 1 week of age. The infant suffers from gastroesophageal reflux and has been treated with ranitidine.

On physical examination, you note a thriving infant in no respiratory distress. The respiratory pattern is comfortable, but the respiratory rate is mildly elevated at 48 breaths/min. You note a monophonic coarse wheeze, which is more notable at the left chest. The remainder of the examination is unremarkable. You review the most recent chest radiograph and recognize a double aortic arch.

Of the following, the study MOST likely to confirm your suspected diagnosis is

A.   barium esophagram with pressure study

B.   bronchoscopy with bronchoalveolar lavage

C.   computed tomography of chest with contrast

D.   echocardiography with Doppler

E.   upper gastrointestinal series with small bowel follow through

Question 220   Preferred Response: C

The 7-month-old infant in this vignette has a clinical history that most strongly suggests a vascular malformation as the etiology of their airway symptomatology. External compression of the airway is a relatively common, but often underrecognized etiology of childhood respiratory symptoms. Prior to obtaining the definitive diagnosis of a vascular ring or pulmonary sling, many infants and children are diagnosed with and treated for more common etiologies of wheezing (eg, asthma, tracheomalacia). A high index of suspicion for vascular malformations causing airway and esophageal compression must be maintained in infants and children who present with recurrent wheezing, stridor, dysphagia, or apnea. When vascular malformations are present and causing airway compression, symptoms are classically poorly responsive to standard medical therapies. The age of an affected infant may be used as a guide. A diagnosis of bronchiolitis or asthma is highly atypical in a newborn and persistent symptomatology in the first few weeks of life should suggest the presence of a congenital disorder.

The normal aortic arch is left-sided. Anomalies of the anatomy of the aortic arch are best understood through a review of embryogenesis. During early fetal development, 6 pairs of primitive aortic arches are formed in sequence. The final anatomic structure is a result of both regressive and derivational components. The arches that persist normally in humans are the fourth and sixth. Fourth arch derivatives contribute to a portion of the normal left aortic arch. The proximal portions of the sixth arches become the mediastinal portion of the pulmonary arteries and the distal portions form the ductus arteriosus, which generally involutes after birth but which may persist.

Vascular anomalies that may be associated with airway and esophageal compression include:
- Double aortic arch
- Right aortic arch
- Left aortic arch with aberrant right subclavian artery or right descending aorta and right ligamentum arteriosum
- Anomalous innominate artery
- Cervical aortic arch
- Pulmonary artery sling

A vascular ring occurs when the trachea and esophagus are entirely encircled by vascular structures. Vascular rings comprise approximately 1% of congenital cardiovascular anomalies. A double aortic arch, where the right- and left-sided aortic arches surround the trachea and esophagus, is the most common etiology of vascular ring and accounts for 50% to 60% of occurrences. A vascular ring may also be created by a right aortic arch with aberrant left subclavian artery and left ligamentum arteriosum. Infants with vascular rings generally present early in life with wheezing, stridor, and dysphagia caused by this anatomic compression.

An anomalous innominate artery arises either more distally or more leftward as compared to normal and compresses the trachea anteriorly. In a pulmonary artery sling, the left pulmonary artery originates from the right pulmonary artery and then courses to the left between the trachea

and the esophagus; compression most commonly occurs at the distal trachea or right mainstem bronchus. The esophagus may be compressed, but esophageal function is generally maintained.

Prenatal diagnosis of congenital vascular rings and slings has advanced with the utilization of 3 vessel tracheal views on fetal ultrasonography and fetal echocardiography. The postnatal diagnostic evaluation of suspected airway compression by vascular malformations includes plain chest radiography, barium esophagoscopy, echocardiography, bronchoscopy, and computed tomography (CT). Plain chest radiographs showing abnormal laterality of the aortic arch should increase the clinical suspicion in an infant or child with suggestive symptomatology, but further imaging will be required to confirm the diagnosis. For the infant in this vignette, CT will provide excellent visualization of airway compression and is the most likely of the choices to confirm the diagnosis. More recently, magnetic resonance (MR) imaging has become the preferred study in defining vascular anatomy, but is not one of the choices.

Barium esophagram will demonstrate posterior or bilateral esophageal indentations related to double and right-sided aortic arches, but the sensitivity of testing is quite low. Echocardiography may show the anatomy of a vascular ring, but is not able to provide imaging of the airways. Bronchoscopy will often reveal tracheal or mainstem bronchus narrowing, but a specific etiology will not be provided. In addition, bronchoalveolar lavage is not expected to be instructive in this patient. Lastly, while an upper gastrointestinal series may reveal esophageal compression with a vascular ring, it is relatively insensitive compared to CT. Moreover, there is no indication for a small bowel follow through study in this infant.

Cardiac MR imaging is increasingly utilized in the evaluation of vascular malformations. Imaging of systemic and pulmonary vessels, the heart, and the tracheobronchial tree are provided without exposure to ionizing radiation and three dimensional reconstructions allow optimal surgical planning. Magnetic resonance imaging is more time consuming and requires longer sedation times compared to CT.

PREP Pearls
- Vascular malformations and related compressive airway lesions should be considered in all infants with recurrent or chronic airway symptomatology, and particularly in those where symptom onset has been atypically early in life or where response to standard medical therapies has been poor.
- The double aortic arch is the most common etiology of a vascular ring.
- Although a plain chest radiograph, barium esophagram, or echocardiogram may provide support for a diagnosis of a compressive vascular malformation, further imaging with computed tomography or magnetic resonance is generally required for definitive diagnosis and surgical planning.

ABP Content Specifications(s)
- Plan the appropriate clinical and laboratory evaluation of vascular anomalies that affect the airway

Suggested Readings
- Haramati LB, Glickstein JS, Issenberg HJ, Haramati N, Crooke GA. MR imaging and CT of vascular anomalies and connections in patients with congenital heart disease: significance in surgical planning. Radiographics. 2002;22(2):337-349..
- Kellenberger CJ. Aortic arch malformations. Pediatr Radiol. 2010;40(6):876-884. DOI:http://dx.doi.org/10.1007/s00247-010-1607-9.
- Kussman BD, Geva T, McGowan FX. Cardiovascular causes of airway compression. Paediatr Anaesth. 2004;14(1):60-74. DOI: http://dx.doi.org/10.1046/j.1460-9592.2003.01192.x.
- Li S, Luo G, Norwitz ER, et al. Prenatal diagnosis of congenital vascular rings and slings: sonographic features and perinatal outcome in 81 consecutive cases. Prenat Diagn. 2011;31(4):334-346. DOI:

Question 221

A family seeks your advice about a planned wilderness camping trip in Belize with their 4-year-old and 10-year-old children. In addition to endemic dengue, there has been an outbreak of malaria in the area they will visit. You provide prescriptions for malaria prophylaxis and discuss options for insect bite prevention with them.

Of the following, the MOST accurate advice to give this family is

   A.   botanical insect repellents are the only products recommended for skin application for children younger than 5 years of age

   B.   DEET use is associated with a high incidence of seizures in young children

   C.   low concentration (7%) DEET has the same efficacy and duration of action as higher concentration (20%-50%) formulations

   D.   skin application of permethrin is effective in dengue prevention

   E.   20% icaridin and 20% DEET have similar efficacy

Question 221   **Preferred Response: E**

Malaria and dengue fever are among the most common vector borne diseases for travelers. Mosquitoes transmit both of these diseases. A critical issue to address for the family in the vignette is how to prevent mosquito and other insect bites. DEET (N,N-dimethyl-meta-toluamide) has been used as a topical insect repellent since the 1950s and is considered the gold standard for disease prevention. While the safety record for DEET is quite good, there have been some case reports of adverse events, including cutaneous and allergic reactions, cardiovascular effects, and, most notably, central nervous system symptoms including headaches, lethargy, confusion, encephalopathy, and seizures. Most of these case reports involved adults and overuse or ingestion of DEET. Despite the rarity of these adverse reports (less than 50 case reports in > 50 years of use), many people are concerned about DEET toxicity and have sought alternatives. Icaridin, formerly known as picaridin or KBR 3023, was introduced more recently and has the advantage of a more acceptable odor and texture profile. At a concentration of 20%, both icaridin and DEET provide similar levels of protection against several species of mosquito. The World Health Organization recommends icaridin as the first line product for repelling malaria-carrying mosquitoes. However, the manufacturer does not recommend icaridin for use by children younger than 2 years of age.

DEET is active against several species of mosquito, and it has been tested for protection against other insects, including ticks and bed bugs, but its effectiveness against these other arthropods is less clear. Efficacy and duration of action of DEET increases with concentration, and it is most effective when used in concentrations between 10% and 35%. Activity plateaus and even decreases at concentrations above 30% to 50%. It is considered safe for use by infants and by pregnant and lactating women. The American Academy of Pediatrics recommends use of DEET in concentrations less than 30% on the exposed skin of infants and children 2 months of age or older.

Several other products have been developed for use against both mosquitoes and other arthropods. Botanicals are particularly sought after because many consider them safer and they are biodegradable, which synthetic compounds such as DEET are not. Most botanicals are made from plant essential oils derived from leaf or fruits. Oil of lemon eucalyptus has efficacy equal to lower concentration DEET products (7%-15%) and a duration of action of 4 to 7 hours for preventing mosquito bites. It is also effective against flies and gnats, but not against ticks. It is not recommended for infants younger than 6 months of age. Permethrin, a synthetic pyrethroid, is effective against ticks, mosquitoes, and other arthropods, as well as for treatment of head lice and scabies. While not recommended for skin application for arthropod bite protection, permethrin is very useful for treating clothing, camping gear, and bed nets. It must be reapplied after every 5 washings to remain effective. Permethrin is more effective than DEET in protecting against ticks.

PREP Pearls
- •   DEET is a highly effective mosquito repellant that is used in concentrations between 10% and 35%; it is considered safe for topical use by infants 2 months of age and older, as well as pregnant and lactating women.

- Icaridin is equally effective against mosquitos compared to DEET; the World Health Organization recommends it as the first line mosquito repellant product, but is not approved for children younger than 2 years of age.
- Despite concerns and case reports, DEET has a good safety profile.
- Some botanicals provide activity against mosquitoes equivalent to low concentration DEET.
- Permethrin applied to clothing, bed nets, and camping gear is effective against ticks, mosquitoes, and other arthropods. It is not recommended for skin use for arthropod protection.

ABP Content Specifications(s)
- Advise parents regarding the appropriate use of topical insect repellants in children

Suggested Readings
- American Academy of Pediatrics. Insect repellents. Healthy Children website.http://www.healthychildren.org/English/safety-prevention/at-play/Pages/Insect-Repellents.aspx.
- Katz TM, Miller JH, Hebert AA. Insect repellents: historical perspectives and new developments. J Am Acad Dermatol. 2008;58(5):865-871. DOI: http://dx.doi.org/10.1016/j.jaad.2007.10.005.

Question 222

The mother of a 19-year-old young man with moderate intellectual disability calls you because she wants resources for her son. She recently heard about Intermediate Care Facilities for Individuals with Intellectual Disabilities (ICF/IID) offered by the Centers for Medicare and Medicaid Services and asks for more information.

Of the following, the MOST accurate statement to tell the mother is

A. ICF/IID is a mandated benefit that requires comprehensive health care and rehabilitation services

B. ICF/IID facilities provide services for pediatric patients only

C. individuals can apply for and qualify for ICF/IID services even if they are not eligible for Medicaid

D. individuals may receive day treatment services at a center that is considered an ICF/IID

E. to qualify for ICF/IID, individuals must receive or need intensive training, treatment, and health services

Question 222   **Preferred Response: E**

Intellectual disability (ID) is used to describe a group of symptoms that includes severe deficits in an individual's developmental skills in the areas of cognitive, language, motor, auditory, psychosocial, and adaptive function. Individuals with ID require varying levels of support.

To receive Medicaid reimbursement, Intermediate Care Facilities for Individuals with Intellectual Disabilities (ICF/IID) must comply with federal standards regarding management, client protections, staffing, active treatment services, client behavior, facility practices, health care services, physical environment, and dietetic services. Additionally, institutions must primarily be a site for the diagnosis, treatment, or rehabilitation for individuals with ID. To qualify for ICF/IID, individuals must receive or need intensive training, treatment, and health services. These facilities must provide a protected residential setting with 24-hour supervision and ongoing evaluation, planning, coordination, and integration of services.
Individuals receiving ICF/IID services must financially qualify for Medicaid and reside at the facility. Services are not restricted to pediatric patients.

PREP Pearls
- Intermediate Care Facilities for Individuals with Intellectual Disabilities must provide a protected residential setting with 24-hour supervision and ongoing evaluation, planning, coordination, and integration of services.
- Implementation and availability of these services varies by state and region.

ABP Content Specifications(s)
- Understand the state and federal statutes that govern the care of intellectually challenged adolescents

Suggested Readings
- Centers for Medicare & Medicaid Services, US Department of Health and Human Services. Intermediate Care Facilities for Individuals with Intellectual Disabilities (ICF/IID). Centers for Medicare & Medicaid Services website.http://www.cms.gov/Regulations-and-Guidance/Legislation/CFCsAndCoPs/Intermediate-Care-Facilities-for-Individuals-with-Intellectual-Disabilities-ICF-IID.html.

Question 223

A 6-year-old girl is struggling in first grade. Her teacher suggested that she be evaluated for attention-deficit/hyperactivity disorder. In your office, the girl is very shy and sits quietly in a chair, playing a game on a tablet computer for the entire visit. Her parents tell you she sometimes does not hear them or forgets what she is supposed to be doing. They have also noticed staring spells, lasting 5 to 10 seconds. These sometimes occur when she is in the middle of a sentence, and when the spell is over, she picks right up where she left off and completes the sentence. Her physical and neurological examinations are unremarkable.

Of the following, the MOST likely to reveal the cause of the girl's symptoms is a(n)

A.   electroencephalogram

B.   fasting blood glucose

C.   magnetic resonance imaging of the brain

D.   Spence children's anxiety scale

E.   Vanderbilt attention-deficit/hyperactivity disorder rating scale

Question 223   **Preferred Response: A**

The girl in the vignette is having absence seizures. Of the response choices listed, the best way to diagnose these is electroencephalography. Absence seizures are characterized by behavioral arrest for 5 to 10 seconds, with a quick return to normal mental status when the seizure is over. Automatisms such as eyelid fluttering, mouth movements, or eye rolling may be seen, but there is no convulsion or postictal state. Absence seizures typically start around 5 years of age, and can occur 50 or more times per day. They can be elicited in clinic, for diagnostic purposes, by sustained hyperventilation, such as blowing on a pinwheel for at least 2 minutes. Electroencephalography will confirm the diagnosis. Treatment with an anticonvulsant is recommended, with ethosuximide typically being the first choice.

Other causes of inattention do not typically cause the clinical presentation seen in the girl in the vignette. The altered mental status seen in hypoglycemia lasts longer and does not include automatisms, and seizures are typically generalized tonic-clonic convulsions, not absence seizures. So it is unlikely that fasting blood glucose levels will lead to the correct diagnosis in this girl. Similarly, anxiety does not cause brief periods of behavioral arrest with automatisms, so evaluation with an anxiety scale would not be helpful in this case. Testing this girl for attentional disorder would also not be helpful in revealing this girl's diagnosis.

In absence seizures the brain is almost always structurally normal, therefore brain imaging does not need to be performed for the girl in the vignette.

PREP Pearls
- Absence seizures typically present at about 5 years of age.
- Absence seizures are characterized by behavioral arrest for 5 to 10 seconds, sometimes with an automatism such as eye fluttering, then a quick return to baseline.

ABP Content Specifications(s)
- Recognize the clinical findings associated with absence seizures, and manage appropriately

Suggested Readings
- Floet AMW, Scheiner C, Grossman L. Attention-deficit/hyperactivity disorder. Pediatr Rev. 2010;31(2):56-69. DOI:http://dx.doi.org/10.1542/pir.31-2-56.
- Sidhu R, Velayudam K, Barnes G. Pediatric seizures. Pediatr Rev. 2013;34(8):333-342. DOI:http://dx.doi.org/10.1542/pir.34-8-333

Question 224

An 18-month-old girl presents to the emergency department because of increasing lethargy. Two hours earlier, she was found playing with an open pill bottle containing her grandmother's clonidine tablets. The family is uncertain whether she ingested any pills and the girl did not have any pill fragments in her mouth. Her vital signs show a temperature of 37°C, heart rate of 80 beats/min, blood pressure of 65/25 mm Hg, respiratory rate of 20 breaths/min, and pulse oximetry of 100% on room air. Physical examination reveals a sleepy child who opens her eyes and cries with stimulation. Her pupils are 2 mm, equal, and reactive, there is no nystagmus, and her gag reflex is intact. Her lungs are clear. The girl's muscle tone is normal and she exhibits purposeful movements in response to stimulation. Her extremities are cool with a capillary refill of 4 seconds.

Of the following, the MOST appropriate next step is to administer

A.   activated charcoal, 25 g orally

B.   dopamine infusion, 5 µg/kg per min intravenously

C.   sodium bicarbonate, 1 mEq/kg intravenously

D.   syrup of ipecac, 15 mL orally

E.   whole bowel irrigation

Question 224   **Preferred Response: B**

The girl in the vignette has clonidine toxicity, which has caused hypotension, shock, and altered mental status. The most life-threatening condition in this case is hypotension. Of the response choices, dopamine infusion is most likely to ameliorate her hypotension.

Clonidine is primarily an α-2 adrenergic agonist. In the central nervous system, it inhibits presynaptic norepinephrine release and neurotransmission, causing sedation. This mechanism also decreases sympathetic outflow from the vasomotor center, causing decreased arterial blood pressure. Clonidine's action as a partial α-2 antagonist in the medulla also stimulates the excitatory cardiac vagal reflex and the inhibitory baroreceptor reflex, which also contribute to decreased cardiac output. Through its peripheral α-1 agonist activity, clonidine can transiently produce hypertension.

Clonidine is widely used in adults as an antihypertensive medication and for drug withdrawal prophylaxis, and is used in children for its behavioral effects. It is a common agent identified in both intentional and accidental poisonings in children. Hypotension occurs within 30 to 60 minutes of clonidine ingestion. Peak plasma concentrations occur within 3 to 5 hours, and the plasma half-life is 12 to 16 hours. Symptoms of clonidine toxicity can mimic narcotic ingestion, with miosis, lethargy, bradycardia, hypotension, and respiratory depression; some improvement in mental status in response to naloxone can be seen. However, the degree of hemodynamic compromise with bradycardia and hypotension is usually more marked in clonidine ingestion than in opioid toxicity, whereas respiratory depression is typically not quite as severe. Other common ingestions that can cause shock and hypotension include other antihypertensives, benzodiazepines, barbiturates, iron, and tricyclic antidepressants. Treatment generally includes fluid administration, decontamination, and, in more severe cases, administration of inotropic and vasopressor agents. Specific antidotes exist depending on the agent, such as glucagon for calcium channel blockers, and sodium bicarbonate for tricyclic antidepressants.

Treatment of shock and hypotension from a clonidine overdose should include aggressive administration of crystalloid fluids to restore preload in the setting of vasodilation. However, reversing the cardiovascular effects of vasodilation and decreased inotropy from reduced sympathetic outflow may require an adrenergic agonist infusion such as dopamine, epinephrine, or norepinephrine. Activated charcoal and whole bowel irrigation can be considered, but restoration of circulation takes precedence. Sodium bicarbonate is helpful in tricyclic antidepressant toxicity, but not for clonidine. Syrup of ipecac is no longer recommended for childhood poisonings because of a lack of evidence that it improves outcomes and because it may delay administration or reduce the effectiveness of other therapies, including activated charcoal.

PREP Pearls
- Common pediatric medication ingestions causing hypotension include antihypertensives, benzodiazepines, barbiturates, iron, and tricyclic antidepressants
- Clonidine is primarily a central α-2 agonist, causing sedation, respiratory depression, and reduced sympathetic outflow, with the cardiovascular effects of hypotension and bradycardia.
- Clonidine toxicity can mimic opioid toxicity, but usually presents with more severe cardiovascular effects and less severe respiratory compromise.

ABP Content Specifications(s)
- Recognize the signs and symptoms of ingestion of medications that might cause hypotension, and manage appropriately

Suggested Readings
- Fiser DH, Moss MM, Walker W. Critical care for clonidine poisoning in toddlers. Crit Care Med. 1990;18(10):1124-1128. Abstract available at http://www.ncbi.nlm.nih.gov/pubmed/2209040.
- Spiller HA, Klein-Schwartz, Colvin JM et al. Toxic clonidine ingestion in children. J Pediatr. 2005;146(2):263-266. DOI:http://dx.doi.org/10.1016/j.jpeds.2004.09.027.

Question 225

A 12-year-old boy with Crohn disease is hospitalized with fever, abdominal pain, and bloody diarrhea. He was recently hospitalized with blunt abdominal trauma complicated by intestinal perforation. During the past hospitalization, he underwent resection of the terminal ileum and received a course of piperacillin-tazobactam for peritonitis. Current vital signs show a temperature of 39.6°C, respiratory rate of 24 breaths/min, heart rate of 143 beats/min, blood pressure of 120/69 mm Hg, and a weight of 32.6 kg. On physical examination, he is pale and has abdominal distention and diffuse tenderness. Laboratory data shows:

- White blood cells, 17,000/μL (17.0 x 10^9/L)
- Hemoglobin, 7.8 g/dL (78 g/L)
- Hematocrit, 25.9%
- Platelets, 260 x 10^3/μL (260 x 10^9/L)
- Differential, 52% segmented neutrophils, 20% bands, 8% lymphocytes, 17% monocytes, 3% eosinophils
- Clostridium difficile polymerase chain reaction, positive

For this patient, antimicrobial management is BEST guided by

A.   antibiotic susceptibility

B.   episode number

C.   prior surgery

D.   severity of illness

E.   underlying condition

Question 225   **Preferred Response: D**

Severity of infection should guide the management of Clostridium difficile infection. Management of C difficile infection includes discontinuation of the offending agent, if the diarrhea is felt to be antibiotic-related, and targeted treatment of the infection. Management is influenced both by the severity of the presentation and by whether the current episode represents a disease recurrence. Metronidazole is recommended for mild to moderate disease and the first recurrence (second episode). Oral vancomycin is recommended for severe disease and second recurrence (third episode). In adults, severity is measured by several parameters, including the presence of ileus, megacolon, or shock. Supportive laboratory parameters include a white blood cell count of greater than 15,000 cells/μL (15.0 x 109/L) or a rise in the serum creatinine of 1.5 times the normal level. Recurrence of infection can be expected in as many as 30% of patients. Patients who become symptomatic can experience fever, bloody diarrhea, and severe abdominal pain. Severe manifestations can include toxic megacolon that can result in bowel perforation and sepsis.

It is important to know that testing for C difficile infection in infants is not recommended. Additionally, testing for children between 1 and 3 years of age is recommended only in the setting of diarrhea after evaluating for other (eg, viral) etiologies. Approximately one-third of babies 0 to 1 months of age are carriers of C difficile. The carriage rate in children approximates non hospitalized adult rates of 0% to 3% by the age of 3 years. Clinical illness is rarely reported before 2 years of age.

C difficile is a common healthcare-associated pathogen and environmental control is critical in preventing its spread. It is essential that patients with C difficile colitis be placed in contact isolation. Removal of spores from the hands of health care workers is best accomplished with the use of soap and water compared to alcohol-based sanitizer. Cleaning of the environment with sodium hypochlorite is also recommended.

Metronidazole resistance in C difficile is rare and does not influence management decisions. While episode number does affect the choice of therapy, there is nothing to suggest a recurrent episode of infection for the patient in this vignette. While previous surgery and underlying conditions are considered risk factors for acquiring C difficile infections, they do not influence management. High risk surgeries include any manipulation of the gastrointestinal tract, including gastrostomy and jejunostomy tubes. Medical risk factors include underlying bowel disease and impaired humoral immunity.

PREP Pearls
- Management of Clostridium difficile infection is influenced both by the severity of the presentation and whether the current episode represents a disease recurrence.
- Testing for C difficile infection in infants is not recommended and testing for children between 1 and 3 years of age is recommended only in the setting of diarrhea after testing for other (eg, viral) etiologies.
- C difficile is a common health care-associated pathogen and environmental control is critical in preventing its spread.

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ABP Content Specifications(s)
- Institute appropriate infection control measures for Clostridium difficile infection
- Recognize the clinical features associated with Clostridium difficile infection
- Plan appropriate management for a patient with Clostridium difficile infection

Suggested Readings
- American Academy of Pediatrics. Clostridium difficile infection in infants and children. Pediatrics. 2013;13(1):196-200. DOI: http://dx.doi.org/10.1542/peds.2012-2992 .
- American Academy of Pediatrics. Clostridium difficile. In: Kimberlin DW, Brady MT, Jackson MA, Long SS, eds. Red Book:2015 Report of the Committee on Infectious Diseases. 30th ed. American Academy of Pediatrics; 2015:298-301 .

Question 226

A 12-month-old girl is brought to your office for a regular health supervision visit. The parents inform you that the patient was evaluated in the emergency department (ED) 2 days ago for tactile temperatures at home. You review the records from the ED and make note that the patient had a temperature of 38.9°C and an unremarkable physical examination. The patient had a catheterized urine analysis showing a specific gravity of 1.030, a pH of 6.0, 3+ leukocyte esterase, 1+ proteinuria, 1+ blood, and positive nitrites. Her urine microscopy showed 50 to 100 white blood cells/high power field (HPF), less than 5 red blood cells/HPF, bacteria, and no crystals.

You discuss with the parents the sensitivity of urinalysis in diagnosing urinary tract infection (UTI) in children.

Of the following, the positive results in this child with the HIGHEST sensitivity for presumptively diagnosing UTI is

    A.   bacteria on urine microscopy
    B.   leukocyte esterase on urine test strip analysis
    C.   nitrites on urine test strip analysis
    D.   red blood cells on urine microscopy
    E.   white blood cells on urine microscopy

Question 226   **Preferred Response: B**

The American Academy of Pediatrics has recently published guidelines regarding the diagnosis and management of initial urinary tract infections (UTIs) in febrile infants and young children. The guidelines included a meta-analysis of studies in which antimicrobial prophylaxis was compared with no treatment or placebo treatment for children (2 to 24 months of age) with vesicoureteral reflux (VUR). The age group was selected in view of high prevalence of UTI in febrile (defined as temperature of at least 38.0°C or 100.4°F) infants and young children. Factors associated with higher than average likelihood for UTI include:

Temperature greater than 39°C
Absence of another source of infection
Girls younger than 12 months of age
Uncircumcised boys
Fever of greater than 24 hours in boys and greater than 2 days in girls 6)History of urinary tract abnormality

A catheterized or suprapubic aspirated (SPA) urine specimen for both urinalysis and culture before an antibiotic is started should be sent in a febrile infant with no apparent source of infection. Urine culture, on a sample collected by a bag applied to the perineum, has an unacceptably high false-positive rate (88%) and has clinical implications only when cultures yield negative results.

Both urinalysis results that suggest infection (pyuria and/or bacteriuria) and the presence of at least 50,000 colony-forming units/mL of uropathogen cultured from an appropriately collected urine specimen (obtained through catheterization or SPA) are required for confirming the diagnosis of UTI. Urine test strip analysis cannot substitute for urine culture to document the presence of UTI in a patient. The urine test strip analysis should be used in conjunction with culture for diagnosing UTI. As urine culture results are not immediately available, a urine test strip analysis suggestive of UTI diagnosis helps in presumptively treating a patient at initial presentation. Urinalysis can be performed on a bag urine specimen. However, a properly collected urine specimen via catheterization or SPA can be sent for both culture and urinalysis. It is important that the urine specimen be tested within 1 hour after voiding, with maintenance at room temperature or within 4 hours after voiding, with the specimen being kept refrigerated to ensure sensitivity and specificity of the urinalysis.

In urinalysis, identification of leukocyte esterase (marker of pyuria or white blood cells in urine) and nitrite on urine test strip analysis, and urine microscopic examination for white blood cells (WBCs) and bacteria have been commonly used for diagnosing UTI. As seen in Item C226, positive results for leukocyte esterase, nitrites, and urine microscopy has the highest sensitivity (99.8%), followed by leukocyte esterase and nitrites (93%), and leukocyte esterase only (83%) for diagnosing UTI.

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Item C226. Sensitivity and Specificity of Components of Urinalysis, Alone and in Combination.

Laboratory test	Sensitivity (Range), %	Specificity (Range), %
Leukocyte esterase test	83 (67-94)	78 (64-92)
Nitrite test	53 (15-82)	98 (90-100)
Leukocyte esterase or nitrite test positive	93 (90-100)	72 (58-91)
Microscopy, WBCs	73 (32-100)	81 (45-98)
Microscopy, bacteria	81 (16-99)	83 (11-100)
Leukocyte esterase test, nitrite test, or microscopy positive	99.8 (99-100)	70 (60-92)

Reprinted with permission from the American Academy of Pediatrics. Urinary tract infection: clinical practice guideline for the diagnosis and management of the UTI in febrile infants and children 2 to 24 months. *Pediatrics*. 2011;128(3):595-610

In patients with asymptomatic bacteriuria, urine culture is positive and the leukocyte esterase test is negative. Asymptomatic bacteriuria is often observed in school aged and older girls, and less frequently in young infants. Asymptomatic bacteriuria should not be treated with antibiotics, as antimicrobial treatment may do more harm than good. Asymptomatic bacteriuria can be distinguished from true UTI by the presence of pyuria in UTI. False-positive (low sensitivity) results for leukocyte esterase may be seen in patients with fever and from other causes or after vigorous exercise. In the presence of pyuria, a UTI should be confirmed with a positive urine culture result, as pyuria alone does not confirm the diagnosis of UTI.

A positive nitrite test is indicative of the conversion of dietary nitrates to nitrites by urinary pathogens. This reaction requires approximately 4 hours, and is therefore not a sensitive marker for identifying UTI in young children who void frequently. The test is also negative for urinary pathogens (enterococcus) that do not reduce nitrate to nitrite. A positive urine nitrite test has high specificity (98%) and therefore low false-positives.

Urine microscopy requires additional equipment and technical expertise. Standard urine microscopy includes microscopic examination of unstained urine for white blood cells (pyuria > 5 WBCs/high power field or 25 WBC/μL) and bacteria (bacteriuria, presence of any bacteria per high power field). Enhanced urinalysis includes Gram staining of uncentrifuged urine and using a counting chamber for assessment of pyuria (> 10 WBC/μL) and bacteriuria (1 gram-negative rod in 10 oil immersion fields). Enhanced urine analysis has been reported to have higher sensitivity, specificity, and positive predictive value than the standard urinalysis, and is preferred with the availability of equipment and trained personnel.

PREP Pearls
- A catheterized or suprapubic aspirated urine specimen for both urinalysis and culture before an antibiotic is started should be sent in a febrile infant with no apparent source of infection.
- Both urinalysis results that suggest infection (pyuria and/or bacteriuria) and the presence of at least 50,000 colony-forming units per mL of an uropathogen cultured from an appropriately collected urine specimen are required for confirming the diagnosis of urinary tract infection (UTI).
- Positive leukocyte esterase test has high sensitivity, while positive urine nitrites have high specificity for diagnosing UTI.
- In patients with asymptomatic bacteriuria, the urine culture is positive and leukocyte esterase test is negative.

ABP Content Specifications(s)
- Plan the appropriate diagnostic evaluation of a urinary tract infection in children who are and are not toilet-trained

Suggested Readings
- American Academic of Pediatrics. 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: http://dx.doi.org/10.1542/peds.2011-1330.
- Finnell SM, Carroll AE, Downs SM. Technical report: diagnosis and management of an initial UTI in febrile infants and young children. Pediatrics. 2011;128(3):e749-e770. DOI: http://dx.doi.org/10.1542/peds.2011-1332.

Question 227

A 2-year-old boy is brought for evaluation after passing 3 large, maroon-colored bowel movements in the last 12 hours. His parents deny recent fever, nausea, vomiting, diarrhea, or abdominal pain. On physical examination, the patient's heart rate is 140 beats/min and his blood pressure is 80/50 mm Hg. He is somewhat pale, but well appearing with an unremarkable abdominal examination.

Of the following, the BEST next step in diagnosis is

    A. barium enema
    B. computerized tomography of the abdomen
    C. plain radiograph of the abdomen
    D. technetium-99m pertechnetate scintiscan
    E. upper gastrointestinal series with small bowel follow through

Question 227   **Preferred Response: D**

The boy in this vignette has the rapid onset of maroon-colored diarrheal bowel movements and symptoms of anemia. The most likely diagnosis is a Meckel diverticulum and the next step in the evaluation is to obtain a technetium-99m pertechnetate scintiscan. A Meckel diverticulum is an outpouching of the gastrointestinal tract caused by the incomplete obliteration of the omphalomesenteric duct during the seventh week of gestation. The "rule of twos" has been used to describe the classic presentation (Item C227).

Item C227. "Rule of Twos" – Classic Presentation of Meckel Diverticulum.

Occurs in 2% of the population

2 feet proximal to terminal ileum on the antimesenteric border

2 cm in diameter

2 inches in length

2:1 male:female ratio

More common before 2 years of age (45% present before 2 years)

May contain 2 different types of ectopic tissue – gastric and pancreatic

Courtesy of C. Waasdorp Hurtado

Meckel diverticulum may present in several ways, including gastrointestinal bleeding, bowel obstruction, and diverticulitis with or without perforation. In addition, in rare cases, a Meckel diverticulum may be found in a hernia into the vitelline duct, resulting in umbilical drainage.

Approximately 50% of symptomatic Meckel diverticulum contain heterotopic gastric tissue. Acidic secretion in this tissue results in inflammation and ulceration of the diverticulum and adjacent ileum. These patients can present with bloody stools, abdominal pain, and fatigue. Some patients have abdominal pain without bleeding, while others describe rectal bleeding without pain. The physical examination is often unremarkable.

Obstructive symptoms can occur secondary to intussusception, hernia, or volvulus involving the diverticulum. Obstruction is more common in adults than children. Patients present with abdominal pain, distention, nausea, and vomiting. On physical examination, patients have abdominal distention, tenderness with palpation, hypoactive bowel sounds, and sometimes a mass or signs of peritonitis.

Infrequently, symptoms are similar to those of appendicitis with fever, right lower quadrant abdominal pain, nausea, and vomiting. The physical examination is the same as an acute appendicitis.

Evaluation for Meckel diverticulum should include a complete blood cell count and basic metabolic panel to evaluate for dehydration and anemia. Imaging studies may include an abdominal radiograph, contrast study of the bowel, ultrasound, computed tomography, or magnetic resonance imaging of the abdomen and pelvis. These studies are all nonspecific and are best used in cases when obstruction is suspected. For patients with a bleeding presentation, such as the child in this vignette, a technetium-99 pertechnectate scan (Meckel scan) is the best study for evaluation and diagnosis.

The sensitivity and specificity of a Meckel scan are 80% to 90% and 95%, respectively, but this decreases with age. Once identified, excision is the treatment of choice for Meckel diverticulum.

The child in the scenario has a Meckel diverticulum and is presenting with a gastrointestinal bleed, best identified by technetium-99 pertechnectate scan. Barium enema is best used for evaluation of constipation and adds little to an evaluation for acute gastrointestinal bleeding. Computed tomography of the abdomen may identify obstruction and inflammation, but is unlikely to provide diagnosis in this young child. A radiograph of the abdomen will evaluate for a foreign body, pneumatosis, and evidence of obstruction, but will not make the diagnosis of Meckel diverticulum. Maroon-colored stools in this child are indicative of bleeding from the lower gastrointestinal tract, so an upper gastrointestinal series with small bowel follow through is unlikely to provide a diagnosis.

PREP Pearls

- Meckel diverticulum are most likely to present in the first 2 years of life.
- Meckel diverticulum can present with gastrointestinal bleeding, obstruction, or inflammation with perforation.
- Technetium-99 pertechnectate scan is the best test to diagnose a Meckel diverticulum with ectopic gastric tissue.

ABP Content Specifications(s)
- Recognize the clinical features associated with Meckel diverticulum, and manage appropriately

Suggested Readings
- Murphy SM. Management of bloody diarrhoea in children in primary care. BMJ. 2008;336(7651):1010-1015. DOI: http://dx.doi.org/10.1136/bmj.39542.440417.BE.
- Pepper VK, Stanfill AB, Pearl RH. Diagnosis and management of pediatric appendicitis, intussusception, and Meckel diverticulum. Surg Clin North Am. 2012;92(3):505-526. DOI: http://dx.doi.org/10.1016/j.suc.2012.03.011.

- Uppal K, Shane Tubbs R, Matusz P, Shaffer K, Loukas M. Meckel's diverticulum: a review. Clin Anat. 2011;24(4):416–422. DOI: http://dx.doi.org/10.1002/ca.21094.

Question 228

A 3-year-old boy with a history of learning disabilities presents to your office as a new patient with macrocephaly, axillary and inguinal freckling, short stature, and 8 café-au-lait macules over 5 mm in diameter (Item Q228). His blood pressure is 130/88 mm Hg.

Given his genetic disorder, the MOST likely cause of his hypertension is

A.   congenital heart defect
B.   hyperthyroidism
C.   neuroblastoma
D.   renal artery stenosis
E.   Wilms tumor

Question 228   **Preferred Response: D**

The boy in the vignette has neurofibromatosis 1 (NF1), which manifests as multiple café-au-lait macules (CALM) (Item C228), (6 or more CALM lesions over 5 mm in prepubertal individuals and over 15 mm in postpubertal individuals), axillary and inguinal freckling, cutaneous and plexiform neurofibromas, and Lisch nodules in the iris. Learning disabilities, short stature, tibial pseudarthrosis, or optic gliomas can also be present. Most patients meet diagnostic criteria by 8 years of age.

Hypertension is common in NF1 and can occur at any age. In many cases, it is classified as essential hypertension; however, a subset of individuals will have a characteristic NF1 vasculopathy that leads to renal artery stenosis, aneurysms, aortic stenosis, or coarctation of the aorta. Renal artery stenosis could be intrinsic in nature, arising from arterial dysplasia, or extrinsic, caused by a plexiform neurofibroma or other abdominal mass. Therefore, a renovascular or cardiovascular cause of hypertension should be investigated in any patient with an NF1 clinical phenotype. Pheochromocytoma manifesting as hypertension is more common in adults with NF1. Ambulatory blood pressure monitoring should be performed at least once annually in patients with NF1.

Hyperthyroidism and neuroblastoma can cause hypertension, but are not frequent in patients with NF1. Most individuals with neuroblastoma have sporadic neuroblastoma. Congenital heart defects can occur in NF1, more commonly associated with whole gene deletions. Most valvular problems become evident at later ages and are caused by NF1 vasculopathy. The most common valvular problem is pulmonic stenosis.

Wilms tumor is an embryonal malignancy of the kidney that is the most common renal tumor in childhood. It can be found in WAGR (Wilms tumor-aniridia-genital anomalies-retardation) syndrome, Denys-Drash syndrome, Beckwith-Wiedemann syndrome, and hemihyperplasia, but not commonly with NF1.

PREP Pearls
- Neurofibromatosis 1 (NF1) is characterized by multiple café-au-lait macules, axillary and inguinal freckling, cutaneous and plexiform neurofibromas, and iris Lisch nodules.
- Hypertension is common in NF1 patients occurring at any age and requires careful blood pressure monitoring.
- Neurofibromatosis 1 vasculopathy can lead to renal artery stenosis, aneurysms, valvular stenosis, or coarctation of the aorta.

ABP Content Specifications(s)
- Recognize the genetic syndromes associated with hypertension

Suggested Readings

- Friedman JM. Neurofibromatosis 1. GeneReviews. http://www.ncbi.nlm.nih.gov/books/NBK1109/.
- Han M, Criado E. Renal artery stenosis and aneurysms associated with neurofibromatosis. J Vasc Surg. 2005;41(3):539-543.
- Hersh JH. Health supervision for children with neurofibromatosis. Pediatrics. 2008;121(3):633-642. DOI: http://dx.doi.org/10.1542/peds.2007-3364.

Question 229

 3-day-old, full-term female newborn is brought to the office for a scheduled visit. The mother's pregnancy, labor, and delivery were uncomplicated, and the infant was discharged from the hospital 24 hours after birth. Her parents report that she has been doing well, with normal feeding and stooling patterns. However, today they noticed a red "blotchy" rash on her body. The rash seems to come and go. On physical examination, you note an erythematous macular eruption with a few 1- to 2-mm papules and pustules, mostly on the face and trunk (Item Q229). The examination is otherwise unremarkable.

Of the following, the MOST likely cytologic finding associated with this rash is

     A.   eggs or excrement on mineral oil preparation
     B.   eosinophils on Wright stain preparation
     C.   gram-positive cocci on Gram stain preparation
     D.   multinucleated giant cells on Tzanck smear preparation
     E.   pseudohyphae on potassium hydroxide preparation

Question 229   **Preferred Response: B**

The neonate in the vignette has erythema toxicum neonatorum (ETN). ETN is an idiopathic, benign, asymptomatic, self-limited condition that occurs most often in full-term newborns. ETN is usually diagnosed clinically, but if cytologic examination is performed on a pustule smear, a predominance of eosinophils will be revealed on Wright or Giemsa stain. ETN is characterized by evanescent blotchy erythematous macules (few millimeters to centimeters in diameter) that occur in combination with 1- to 3-mm papules and pustules anywhere on the body, especially the face, trunk, and extremities, sparing the palms and soles. ETN usually appears during the first 4 days after birth, but it may be present at birth or occur as late as the second week after birth. It has a tendency to remit and recur during the first 2 weeks after birth. The reported incidence varies from 5% to 70% of newborns and increases with increasing gestational age. ETN does not require treatment.

Differentiation from other pustular eruptions of the newborn is important to rule out more serious infectious or dermatologic conditions. Usually these rashes can be distinguished by the history and clinical findings; however, cytologic findings can be helpful in making the diagnosis. Scrapings of scabies lesions may reveal mites, eggs, or excrement on mineral oil preparation. A gram-positive cocci found on staining of pustular material is typical of staphylococcal folliculitis. A Tzanck smear of herpetic lesions may demonstrate multinucleated giant cells. If pseudohyphae are seen on potassium hydroxide preparation, congenital candidiasis is the most likely cause of the pustular eruption.

PREP Pearls
- Erythema toxicum neonatorum is a benign, asymptomatic, self-limited condition usually diagnosed clinically, that occurs most often in full-term newborns.
- Erythema toxicum neonatorum usually appears in the first 4 days and tends to remit and recur during the first 2 weeks after birth.
- A predominance of eosinophils will be revealed on a Wright or Giemsa stained smear of pustular material from an infant with erythema toxicum neonatorum.

ABP Content Specifications(s)
- Recognize the clinical and cytologic findings associated with erythema toxicum

Suggested Readings
- Udassi S, Marshall S, Acharya R, Tuli SY. Erythema toxicum neonatorum. Cons Pediatricians. 2014;13(11):494 .
- Johr RH, Schachner LA. Neonatal dermatologic challenges. Pediatr Rev. 1997;18(3):86-94 http://www.newnanpediatrics.com/resources/Student-Articles/Neonatal-Dermatologic-Challenges.pdf .
- Paller AS, Mancini AJ. 2011. Cutaneous disorders of the newborn. In: Hurwitz Clinical Pediatric Dermatology: A Textbook of Skin Disorders of Childhood and Adolescence. 4th ed. Philadelphia, PA: Saunders Elsevier; 2011:16-18.

Question 230

You see two 8-week-old infants consecutively in your general pediatrics practice for health supervision visits. The first infant was born at 30 weeks gestational age, while the second infant was born full term. The medical student working with you asks what will happen to the hematocrit of the preterm infant relative to the full term infant.

Of the following, you inform the medical student that, compared with the full term infant, this preterm infant will

    A. reach the nadir of erythrocyte production at the same time, but will recover faster
    B. reach the nadir of erythrocyte production at the same time, but will require longer to recover
    C. reach the nadir of erythrocyte production sooner and will require longer to recover
    D. reach the nadir of erythrocyte production sooner, but will recover faster
    E. take longer to reach the nadir of erythrocyte production and will require longer to recover

Question 230   **Preferred Response: C**

As the fetus develops, the production of erythropoietin increases with time, with the highest production occurring in the final trimester. The production of red blood cells is directly driven by erythropoietin, and as a consequence, a significant portion of the red blood cell mass is produced in the final trimester of pregnancy. As such, premature infants have a lower hematocrit at birth than full term infants. Upon birth, blood oxygen levels increase with the onset of breathing and the closure of the ductus arteriosus. The elevated oxygen level downregulates the production of hypoxia-inducible factor 1, which in turn downregulates the production of erythropoietin. This results in a temporary drop in hematocrit after birth. As premature infants have both a lower hematocrit at birth and an impaired ability to produce erythropoietin, the hematocrit nadir in premature infants occurs earlier and is both deeper and longer than in full term infants. This condition is known as the anemia of prematurity. In full term infants, the hemoglobin typically reaches a nadir of 11 g/dL (110 g/L) at 8 to 12 weeks after birth.

Other factors that can lead to the anemia of prematurity include repeated phlebotomy in sick or premature neonates, a reduced lifespan for the red blood cells, and iron depletion. Although iron depletion is not the cause of the anemia of prematurity, it may impair the ability to recover. Given the reduced iron stores present in the premature infant, it is important to initiate iron supplementation by 8 weeks of life.

PREP Pearls
- The greatest increase in fetal red blood cell mass occurs in the final trimester of pregnancy, so premature infants are born with a lower hematocrit than full term infants.
- Premature infants have impaired production of erythropoietin.
- When the impaired production of erythropoietin is combined with the lower hematocrit at birth, this results in an earlier, deeper, and longer nadir in the hematocrit. This is known as the anemia of prematurity.

ABP Content Specifications(s)
- Recognize the differences in hematocrit values in pre- and full-term infants, and the normal ranges for both

Suggested Readings
- Bishara N, Ohls RK. Current controversies in the management of the anemia of prematurity. Semin Perinatol. 2009;33(1):29-34. DOI: http://dx.doi.org/10.1053/j.semperi.2008.10.006.
- Kett JC. Anemia in infancy. Pediatr Rev. 2012;33(4):186-187. DOI: http://dx.doi.org/10.1542/pir.33-4-186.
- Roth P. Anemia in preterm infants. Pediatr Rev. 1996;17(30):370. http://pedsinreview.aappublications.org/content/17/10/370 .

Question 231

An 8-year-old girl is brought to see you because of school problems. Her school performance has been poor this year, and her teacher reports that she has had poor attention in class. The mother says that for more than 2 months she has resisted and sometimes refused to go to school, and when at school, she sometimes goes to the office crying that she needs to go home. She denies having any peer problems at school. At home, she has developed some sleep problems as well, which the mother has addressed by staying in her room with her until she falls asleep. She has a significant stressor in that her close friend's family moved away this past summer, and she talks about missing her friend. When you talk to the girl, she tells you that she has a specific worry that someone will hurt her mother when she is not there. Her mother emphatically denies that domestic violence is occurring at home.

Of the following, the MOST likely diagnosis is

- A. adjustment disorder
- B. attention-deficit/hyperactivity disorder
- C. oppositional defiant disorder
- D. separation anxiety disorder
- E. specific phobia

Question 231   **Preferred Response: D**

Most children experience developmentally normal separation fears during their preschool years and again upon first entering school. It is when these fears persist for more than 1 month and cause significant dysfunction that an anxiety disorder may be diagnosed. The girl in this vignette is most likely suffering from a separation anxiety disorder. The dysfunction of this disorder typically stems from a preoccupying fear that something bad may happen to them or to someone they love when they are apart. The child may focus their need for reassurance from only one caregiver, typically their mother, which then excludes other family members from that dyad by clinging to that parent's side. Children with this problem may become school refusers, which is different than other more common causes of truancy that do not include a fear of parental separation.

Separation anxiety disorder may include physical symptoms, such as stomach aches or headaches when anticipating or experiencing a separation, which a parent might misinterpret as evidence of a medical disorder. First line treatment includes cognitive behavioral therapy and parental coaching for how to help their child master their fears. When a child has a separation anxiety disorder, there often are other family members with anxiety difficulties.

An adjustment disorder is a temporary state, such as an anxious adjustment reaction to initiating school. Given that this girl's difficulties have lasted more than 2 months, an anxiety disorder would be a better diagnosis. Attention-deficit/hyperactivity disorder is a common cause of school difficulties, but is not likely to generate this degree of school avoidance and need to have close parental contacts. Oppositional defiant disorder may have school refusal as part of the overall oppositional behavior profile, but that diagnosis would not explain the child's extensive anxiety symptoms. Specific phobia is the most common anxiety diagnosis in children and is a result of fear of something specific like snakes, spiders, etc.

PREP Pearls
- Some degree of separation anxiety is often seen in the preschool years and during the first month of school entry, which parents and teachers resolve through reassurance.
- Separation anxiety disorder should be suspected in cases of school refusal or when separation anxiety lasts more than 4 weeks.

ABP Content Specifications(s)
- Recognize co-morbidities commonly associated with phobias and anxiety disorders
- Recognize the clinical findings associated with anxiety disorders in patients of various ages, and manage appropriately

Suggested Readings
- Bagnell AL. Anxiety and separation disorders. Pediatr Rev. 2011;32(10):440-446. DOI: http://dx.doi.org/10.1542/pir.32-10-440.
- 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: http://dx.doi.org/10.1037/a0025933.

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- Mohatt J, Bennett SM, Walkup JT. Treatment of separation, generalized, and social anxiety disorders in youths. Am J Psychiatry. 2014;171:741-748. DOI: http://dx.doi.org/10.1176/appi.ajp.2014.13101337.

Question 232

You are called to evaluate a term female newborn who is noted to have lymphedema. Her prenatal ultrasonography suggested a horseshoe kidney. She is in respiratory distress, requires intubation, and is stabilized from a respiratory standpoint. During intubation, you notice that her neck has thickened folds. On physical examination, her heart rate is 160 beats/min, blood pressure is 90/60 mm Hg in her right arm, and 70/50 mm Hg in her right leg. Her oxygen saturation is 95% in the right arm. She is noted to have a 1/6 systolic murmur at the right upper sternal border. Her femoral pulses are 1+. You are worried about congenital heart disease. You consult cardiology and genetics to help evaluate the newborn.

Of the following, the MOST likely diagnosis is

    A. DiGeorge syndrome
    B. Down syndrome
    C. Kartagener syndrome
    D. Marfan syndrome
    E. Turner syndrome

Question 232   **Preferred Response: E**

The newborn in this vignette has a murmur in the aortic position and a blood pressure gradient between the upper and lower extremities consistent with a coarctation of the aorta. The child also shows evidence of lymphedema and a horseshoe kidney. The most likely diagnosis is Turner syndrome. Turner syndrome is the most common sex chromosome disorder in females, occurring in 1 out of every 2,500 to 3,000 liveborn females. It is most commonly caused by a single X chromosome and is associated with left-sided obstructive cardiac lesions, including coarctation of the aorta and aortic valve disease, such as bicuspid aortic valves. Neonatal lymphedema is a hallmark of Turner syndrome and causes thickened folds in the neck. Eighty percent of neonates born with Turner syndrome will have lymphedema and 35% will have coarctation. Turner syndrome should be suspected in any female who presents with coarctation of the aorta, as 5% are estimated to have Turner syndrome.

Among the other responses, none would be as likely to be seen in females with the constellation of findings described. Patients with Down syndrome can have lymphedema, but more commonly have an atrioventricular (AV) septal defect (or AV canal) and no murmur at birth. They are much less likely to have a coarctation of the aorta. DiGeorge syndrome would be expected in patients with conoseptal defects such as tetralogy of Fallot or an interrupted aortic arch. Kartagener syndrome (lack of normal ciliary function and chronic infections) is associated with heterotaxy lesions (abnormal orientation of internal organs), single ventricle physiology (often with an AV canal type defect), and abnormal pulmonary vein anatomy. Typically, this would not feature aortic position murmurs or lymphedema as seen in the newborn in this vignette. Marfan syndrome is associated with aortic root dilation that is not likely to present in the newborn period. It does not cause coarctation of the aorta.

PREP Pearls
- Turner syndrome is associated with coarctation of the aorta and bicuspid aortic valve and is seen in females only.
- Lymphedema is common in newborns with Turner syndrome.

ABP Content Specifications(s)
- Recognize cardiac conditions associated with Turner syndrome

Suggested Readings
- American Academy of Pediatrics. Care of girls and women with Turner syndrome: a guideline of the Turner syndrome study group. Pediatrics. 2009;123(5):1423. DOI: http://dx.doi.org/10.1542/peds.2009-0394.
- Bondy C, Bakalov VK, Cheng C, Olivieri L, Rosing DR, Arai AE. Bicuspid aortic valve and aortic coarctation are linked to deletion of the X chromosome short arm in Turner syndrome. J Med Genet. 2013;50(10):662-665. DOI: http://dx.doi.org/10.1136/jmedgenet-2013-101720.
- Goldmuntz E, Crenshaw M, Lin AE. Genetic aspects of congenital heart defects. In: Allen HD, Driscoll DJ, Shaddy RE, Feltes TF, eds. Moss and Adams' Heart Disease in

Infants, Children, and Adolescents: Including the Fetus and Young Adult. 8th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2013:617-643.

- Park MK. Obstructive lesions. Park's Pediatric Cardiology for Practitioners. Philadelphia, PA: Saunders Elsevier; 2014:184-205.
- Wong SC, Burgess T, Cheung M, Zacharin M. The prevalence of Turner syndrome in girls presenting with coarctation of the aorta. J Pediatr. 2014;164(2):259-263. DOI: http://dx.doi.org/10.1016/j.jpeds.2013.09.031.

Question 233

You are seeing a 7-year-old boy in your office, which is a medical home with comprehensive care systems available. The boy, recently diagnosed with Becker muscular dystrophy, was referred to neurology 2 weeks ago for a positive Gower sign and elevated muscle enzymes. He has normal intelligence and is able to walk and perform activities of daily living normally. The family is struggling to deal with this new diagnosis. They have concerns regarding his prognosis, educational needs, and care planning.

Of the following, the MOST accurate statement regarding this child is that

A.   the family should contact the Department of Health for community resources
B.   he is not disabled enough to qualify for educational modifications
C.   he will need monthly health maintenance visits
D.   his neurologist should be his primary medical provider
E.   your office will coordinate specialist referrals for disease management

Question 233   **Preferred Response: E**

For the boy in the vignette, who has been diagnosed with a chronic medical condition, the most accurate response choice is that your office will coordinate specialist referrals for disease management. An important role for the general pediatrician treating children with special health care needs (CSHCN) is to provide a medical home that coordinates medical care. The child's primary care provider should be the main source of information about, and referral to, community resources, rather than the Department of Health. Any child with a chronic medical condition can qualify for a 504 educational plan that is not limited by level of disability, and designed to prevent the condition from having a significant impact on the child's academic career. Although the boy in the vignette may need more frequent health maintenance visits than a typical child without special health care needs, monthly visits are more frequent than is necessary for him in his current state of health. The general pediatrician, not the neurologist, should continue to be the child's primary health care provider.

Significant needs and psychosocial factors affect the families of CSHCN. Most will have a significant financial burden because of medical expenses that are not covered by insurance. Household income may be reduced when 1 or both parents are needed as full or part-time caregivers for the child, and are therefore unable to work or must have reduced work hours. Many families have difficulty navigating the medical system. CSHCN are frequently seen by multiple specialists in various settings such as the hospital, emergency room, and outpatient office. Navigating these entry points can be difficult. Other unmet needs of CSHCN include mental health services to help the patient and family cope with the medical condition and its impact on their lives. Many CSHCN also need specialized equipment, including communication and mobility aids. The medical home model offers needed family support along with care coordination. Information for providers regarding setting up a medical home can be found online at http://www.medicalhomeinfo.org and http://www.medicalhomeportal.org/diagnoses-and-conditions.

Families of CSHCN often need to work with the school district to provide appropriate educational opportunities and improve educational outcomes. Children with disabilities will qualify for special education under the Individuals with Disabilities Education Improvement Act of 2004. Families of children eligible for this program work with their schools to develop individualized education plans (IEPs). The parents of children with disabilities who do not qualify for special education services may request specific accommodations such as assistive technology, extra time for school work because of hospitalization, and physical assistance such as wheelchair ramps or special seating arrangements. These resources are available through the Rehabilitation Act of 1973, Section 504. The primary care provider should be able to discuss the programs available with families of CSHCN.

PREP Pearls
- As a medical home, the pediatric practitioner's office should coordinate specialist referrals for disease management.

- Any child with a chronic medical condition can qualify for a 504 educational plan designed to prevent the condition from having a significant impact on the child's academic career.
- Children with disabilities qualify for special education under the Individuals with Disabilities Education Improvement Act of 2004

ABP Content Specifications(s)
- Understand the effects of a child's chronic illness on the family and social relationships
- Identify psychosocial factors associated with chronic and handicapping conditions

Suggested Readings
- American Academy of Pediatrics Council on Children With Disabilities. Provision of educationally related services for children and adolescents with chronic diseases and disabling conditions. Pediatrics. 2007;119:1218-1223. DOI: http://dx.doi.org/10.1542/peds.2007-0885.
- Sadof MD, Nazarian B. Caring for children who have special health-care needs: a practical guide for the primary care practitioner. Pediatr Rev. 2007;28:e36-e42. DOI: http://dx.doi.org/10.1542/pir.28-7-e36.

Question 234

A 17-year-old adolescent presents to your office for a preparticipation physical examination. You inquire about nutritional supplement and drug use. He tells you that he takes creatine to enhance his performance in lacrosse at the recommendation of his coach. He plans to play lacrosse at the college level next year. You discuss the use of creatine and other supplements with him.

Of the following, the MOST accurate statement to include in your discussion is that

      A.   approximately 1% of creatine users develop impaired kidney function
      B.   creatine is unlikely to improve his endurance during the lacrosse season
      C.   creatine use is associated with rhabdomyolysis
      D.   creatine use is banned by the National College Athletic Association
      E.   creatine use increases the risk of cardiac arrhythmias

Question 234   **Preferred Response: B**

Creatine increases lean body mass and can improve performance with short bursts of exercise at maximal intensity. Therefore, creatine can enhance performance in sports that involve brief, high-intensity activities such as the shot put, but it does not improve endurance.

Phosphocreatine recharges adenosine triphosphate (ATP) during short bursts of high-intensity exercise. Creatine supplementation increases phosphocreatine stores, thus augmenting ATP stores. Many young athletes use creatine; studies have reported rates of use between 5% and 30%. The adverse effects of creatine are generally mild; abdominal cramps are the most common. Most athletes, coaches, and health care providers agree that athletes should not use performance-enhancing substances that carry a risk of health impairment. The use of supplements that typically have mild or no side effects, like creatine and caffeine, is an ethical gray area. Many people feel that maximizing physical training and using good dietary practices to improve performance is a better approach than using supplements.

Physicians should discuss supplement use with young patients because children and adolescents may harbor misconceptions about their risks and benefits.

Creatine has rarely been associated with renal failure in case reports; in most cases, the individuals affected had underlying kidney disease. Creatine does not increase the risk of rhabdomyolysis or arrhythmias. Creatine has not been banned by any major sports organization, such as the National College Athletic Association. However, athletes should be counseled that legal supplements can be contaminated with other substances that may be banned.

PREP Pearls
- Creatine can enhance performance in sports that involve brief, high-intensity activities, such as shot put, but it does not improve endurance.
- The use of supplements that typically have mild or no adverse effects, like creatine and caffeine, is an ethical gray area.

ABP Content Specifications(s)
- Recognize and apply ethical principles involved in use of technology for performance enhancement therapies

Suggested Readings
- Castillo EM, Comstock RD. Prevalence of use of performance-enhancing substances among United States adolescents. Pediatr Clin North Am. 2007;54(4):663-675, ix-x. .
- Dandoy C, Gereige RS. Performance-enhancing drugs. Pediatr Rev. 2012;33(6):265-271; quiz 271-262. DOI: http://dx.doi.org/10.1542/pir.33-6-265.

Question 235

An 8-year-old, previously healthy, fully immunized boy presents to your office with bilateral facial swelling that developed over the last 24 hours. Throughout the past week, the patient had a subjective fever, myalgias, and a mild cough. On physical examination, his temperature is 37.5°C, heart rate is 100 beats/min, and respiratory rate is 18 breaths/min. The boy's voice is hoarse. There is bilateral, tender, firm, nonerythematous swelling of the pre-auricular area that extends to the angle of the mandible. His teeth and gingiva are normal, with mild erythema of the buccal mucosa. His posterior oropharynx is erythematous with enlarged tonsils without exudates. He has clear rhinorrhea, erythema of the nasal mucosa, and mildly hyperemic and retracted tympanic membranes. There is shotty bilateral cervical lymphadenopathy. The remainder of the physical examination is unremarkable.

Of the following, the MOST likely etiology of the patient's illness is

- A.   HIV
- B.   Klebsiella species
- C.   mumps
- D.   parainfluenza virus
- E.   Staphylococcus aureus

Question 235   **Preferred Response: D**

The boy in the vignette has bilateral preauricular swelling consistent with parotitis, most likely caused by parainfluenza virus, given his accompanying viral symptoms, immunization status, and history of good health. The differential diagnosis of preauricular swelling includes the following conditions: parotitis, preauricular cyst, sebaceous cyst, lymphadenitis, sialolithiasis, tuberculosis, actinomycosis, and neoplasm. Nearly all of these conditions result in unilateral swelling, but tuberculosis can present with bilateral involvement in the absence of systemic disease.

There are many causes of parotitis in children (Item C235); bilateral involvement in a non–toxic-appearing child is most likely viral in origin. Parotitis can be the first manifestation of human immunodeficiency virus (HIV) infection acquired at birth in an otherwise healthy child, but HIV is a less common cause of bilateral parotitis than other viruses in healthy children. Mumps parotitis continues to occur in individuals who are unimmunized or have waning immunity, but most often is preceded by a prodrome of fever, headache, anorexia, and malaise.

Item C235. Causes of Parotitis in Children.

Infectious	
Bacteria	*Actinomyces* spp
	Bacteroides spp
	Brucella spp
	Escherichia coli
	Francisella tularensis
	Fusobacterium spp
	Haemophilus spp
	Klebsiella spp
	Moraxella catarrhalis
	Peptostreptococcus spp
	Prevotella spp
	Proteus spp
	Pseudomonas aeruginosa
	Pseudomonas pseudomallei
	Salmonella spp
	Staphylococcus aureus
	Group A *Streptococcus*
	Streptococcus pneumoniae
	Treponema pallidum
	Viridans streptococci
Viruses	Adenovirus
	Coxsackieviruses
	Cytomegalovirus
	Echoviruses
	Epstein-Barr virus
	Herpes simplex virus
	Human herpesvirus-6
	Human immunodeficiency virus
	Influenza virus
	Lymphocytic choriomeningitis virus
	Mumps
	Parainfluenza viruses
Mycobacteria	*Mycobacterium avium-intracellulare*
	Mycobacterium tuberculosis
Fungi	*Candida* spp
Noninfectious	
Collagen vascular disease (eg, Sjörgen syndrome, systemic lupus erythematosus)	
Cystic fibrosis	
Drugs (eg, antineoplastic chemotherapy)	
Endocrine disorders (eg, diabetes, hypothyroidism)	
Mechanical (eg, sialolithiasis)	
Metabolic disorders (eg, hepatic disease, hyperlipoproteinemia, hyperuricemia)	
Neoplasms (eg, mixed, mucoepidermoid carcinoma, adenoid cystic carcinoma)	
Sarcoidosis	

Courtesy of D. Palazzi

Acute bacterial parotitis is most commonly caused by Staphylococcus aureus in all age groups. Patients typically present with fever and the sudden onset of swelling, warmth, and tenderness over the parotid gland; disease typically is unilateral, and purulent discharge may be visible from the Stensen duct on the affected side. Acute bacterial parotitis more commonly occurs in neonates, the elderly, and hospitalized patients, with gram-negative (eg, Klebsiella species) and fungal infections occurring in addition to those caused by Staphylococcus aureus. Parotitis caused by anaerobic bacteria (eg, Peptostreptococcus, Prevotella) usually is associated with dental infection.

Patients with common viral causes for parotitis (including mumps) are treated with analgesics, antipyretics, and hydration. Patients with HIV infection should be evaluated for antiretroviral therapy. Treatment of acute bacterial parotitis should be directed toward the most likely causative organisms.

PREP Pearls
- The differential diagnosis of preauricular swelling includes the following conditions: parotitis, preauricular cyst, sebaceous cyst, lymphadenitis, sialolithiasis, tuberculosis, actinomycosis, and neoplasm.
- Mumps parotitis continues to occur in unimmunized individuals or those with waning immunity.
- The treatment of viral parotitis is supportive.
- Staphylococcus aureus is the most common cause of acute bacterial parotitis in all age groups.
- Acute bacterial parotitis is most commonly seen in neonates, the elderly, and hospitalized patients.

ABP Content Specifications(s)
- Plan the appropriate management of a patient with mumps
- Differentiate among the various causes of parotitis
- Formulate a differential diagnosis of preauricular swelling

Suggested Readings
- American Academy of Pediatrics. Mumps. In: Kimberlin DW, Brady MT, Jackson, MA, Long SS, eds. Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015: 564-568.
- Campbell JR. Parotitis. In: Cherry J, Demmler-Harrison GJ, Kaplan SL, Steinbach WJ, Hotez P, eds. Feigin & Cherry's Textbook of Pediatric Infectious Diseases. 7th ed. Philadelphia, PA: Elsevier Saunders; 2014:189-193.

Question 236

You are seeing a 19-month-old boy in your office for follow-up 2 days after a 5-day hospitalization for treatment of orofacial burns. The boy chewed on a frayed extension cord last week and he sustained a 1 x 1.5 cm full-thickness burn to his right oral commissure and lower lip, as well as a small partial-thickness burn to the right side of his chin.

His parents tell you that the boy has been doing well since discharge from the hospital. He has had no fevers and he has been able to tolerate fluids and soft foods. His pain has been well controlled. In your office, the boy's vital signs are normal for his age and he appears well. Physical examination reveals only the healing burns at the right oral commissure and lower lip with eschar formation, and a small healing, partial thickness burn to the right side of his chin. There are no signs of infection. The boy's mucous membranes are moist, and his extremities are warm and well-perfused. The boy's parents ask you about potential for delayed complications from his recent injury.

Of the following, the boy is MOST at risk for

        A.   acute hemolytic anemia within the next 1 to 2 weeks
        B.   acute renal failure within the next 1 to 3 weeks
        C.   progressive sensorineural hearing loss over the next 6 to 12 weeks
        D.   severe bleeding within the next 1 to 2 weeks
        E.   ventricular arrhythmias within the next 1 to 2 weeks

Question 236   **Preferred Response: D**

The young boy in the vignette presents for follow-up 1 week after sustaining electrical burns to his right oral commissure, lower lip, and right chin. Of the response choices, he is most at risk for severe bleeding within the next 1 to 3 weeks. Low-voltage household current injury to the oral cavity and lips typically occurs when a toddler places a household electrical cord in his or her mouth. The resultant injury is a deep burn to the oral commissure with coagulation necrosis. These burns require very close follow-up to prevent severe scarring and contractures. The most serious potential sequela of these burns is severe bleeding from the labial artery when the eschar separates 1 to 2 weeks after the injury. In the past, children with electrical injuries involving the mouth were hospitalized for 2 weeks, but many specialists now manage such cases on an outpatient basis, with close follow-up and thorough education of the caregivers.

All pediatric providers should understand the sequelae associated with electrical burns. Electrical burns are responsible for more than 4,000 emergency department visits and more than 1,500 deaths annually. Many of these injuries involve children. Up to one-third of electrical burns are household burns, which typically occur when young children come into contact with low-voltage (<200 V) alternating current from household appliances, extension cords, or place objects into electrical outlets. Medium (200-1,000 V) and high-current (>1,000 V) injuries are typically seen in adolescents as a consequence of risk-taking behaviors, utility workers, and people exposed to electrical storms.

Electrical burn injuries arise from thermal energy that is released as an electrical current passes through the body. The degree of thermal energy produced depends on the voltage of electric current contacted. The extent of electrical injury depends on the resistance of body structures through which the current passes, the type of current (alternating or direct), the duration of contact with the electrical source, the intensity of the current, and the pathway travelled by the current. Current preferentially flows through body tissues with low electrical resistance, such as blood vessels, nerves, and muscles. Alternating current is more dangerous than direct current, causing muscle tetany because of the continual contraction and relaxation of the muscle with each cycle.

Acute hemolytic anemia is not a typical complication seen after electrical burn injuries.

Acute renal failure may occur in patients with severe burn injuries, because of direct injury to the kidney, renal hypoperfusion, or excessive deposition of myoglobin from injured muscle tissue. It would be very unlikely to develop in this boy, whose injuries were limited to his mouth and right side of the chin, and who is well-appearing with normal urine output 1 week after the injury.

Although victims of severe electrical injuries, including lighting strikes, can present with hearing loss, the development of progressive sensorineural hearing loss in this boy over the next 6 to 12 weeks is extremely unlikely. Damage to the central nervous system including brain, spinal cord, peripheral nerves, and sympathetic fibers may result from electrical injuries. Immediate clinical manifestations may include pain, loss of consciousness, respiratory center paralysis, confusion, visual changes, deafness, sensory deficits, seizures, motor deficits, memory loss, and intracranial

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hemorrhage. Delayed development of peripheral or sensory neuropathy has been observed in some patients after burn injuries, but this is not common.

Acute myocardial injury and life-threatening ventricular arrhythmias can occur as an immediate consequence of electrical injuries. If an electrical current flows across the heart (which occurs more often when the flow takes an arm-to-arm pathway), myocardial injury and cardiac arrhythmias (including ventricular fibrillation or asystole) may occur. However, in the acute setting, a patient with a normal electrocardiogram does not appear to be at significant risk for later development of arrhythmias.

Musculoskeletal sequelae may arise from electrical injuries, with the potential for tissue to become edematous and necrotic, or the development of compartment syndrome. Muscle tetany can result when there is contact with alternating electrical current. Respiratory failure may result if tetany involves the chest wall muscles.

Renal failure can arise from the direct effect of electrical current on kidney tissue, decreased perfusion to the kidney, or renal tubular injury secondary to deposition of myoglobin from extensive muscle damage.

Pulmonary, abdominal, and ocular complications (including the development of cataracts) may also occur, but these are not common sequelae of electrical burn injuries.

In terms of prevention of electrical injuries, it is essential that caregivers keep all electrical outlets covered with safety covers and keep children away from all electrical cords. Adolescents should be counseled on the dangers of climbing electrical structures and taught to avoid other risk-taking behaviors. To avoid potentially life-threatening injury from lighting strikes, children should be taught that during storms they must stay indoors, exit the water immediately, and avoid contact with metal objects.

PREP Pearls
- Injury to the oral cavity and lips from low-voltage household electrical current typically occurs when an infant places a household electrical cord in his or her mouth.
- Oral cavity and lip burns require very close follow-up to prevent severe scarring and contractures. Patients are at risk for severe bleeding from the labial artery when the eschar separates 1 to 2 weeks after the injury.
- Up to one-third of electrical burns are household burns, which typically affect young children who place objects into electrical outlets or come into contact with low-voltage (<200 V) alternating current from household appliances or extension cords.
- Acute myocardial injury and life-threatening ventricular arrhythmias can occur as an immediate consequence of electrical injuries. Patients with normal electrocardiograms in the acute setting do not appear to be at significant risk for developing arrhythmias in the future.

ABP Content Specifications(s)
- Understand the sequelae associated with electrical burns

Suggested Readings
- Joffe MD. Burns. In: Fleisher GR, Ludwig S, eds. Textbook of Pediatric Emergency Medicine. 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2010:1281-1288.
- Pham TN, Gibran NS. Thermal and electrical injury. Surg Clin North Am. 2007;87(1)185-206. DOI: http://dx.doi.org/10.1016/j.suc.2006.09.013.
- Reed JL, Pomerantz WJ. Emergency management of pediatric burns. Pediatr Emerg Care. 2005;21(2):118-129.

Question 237

You are seeing a 1-week-old newborn in your office during a newborn follow-up visit. The mother is worried because her newborn has not begun to "unfold" after being born. The mother tells you that the obstetrician found decreased amniotic fluid on the fetal ultrasonography 6 weeks before her infant was born. Since birth, her newborn always sleeps with her legs flexed up towards her abdomen, with her head tilted to the side. While being held in her mother's arms, you see an active newborn with her head tilted to the right, her chin tilted to the left, and adduction of both forefeet (Item Q237).

Item Q237. Forefoot of the infant described in the vignette. Reprinted with permission from Smith BG. Pediatr Rev. 2009;30(8):287-294.

Of the following, the finding MOST likely to be appreciated during this newborn's examination is

    A.   camptodactyly
    B.   clavicular crepitus
    C.   genu recurvatum
    D.   hip dislocation
    E.   jaw subluxation

Question 237   **Preferred Response: D**

The newborn in the vignette has the physical examination features of moulded baby syndrome (MBS), which include congenital muscular torticollis (CMT), metatarsus adductus, and developmental dysplasia of the hip (DDH). MBS may also present with head molding, postural torticollis, congenital scoliosis, pelvic obliquity with altered hip movement, and malposition of the knees and feet. MBS has been attributed to fetal positioning against the mother's spine, along with prolonged intrauterine constraint later in pregnancy because of factors such as oligohydramnios or a primagravida uterus. MBS has also been called TAC syndrome, referring to the findings of a turned head, adducted hip, and truncal curvature.

The pelvic obliquity seen in MBS leads to an abduction contracture of the hip on the concave side of the newborn, whereas the contralateral side develops an adduction contracture (Item C237). This positioning worsens between 2 and 5 months after birth as the legs become more extended, with an increased risk of hip dislocation on the adducted side. A recent single-center study found that 12% of infants with CMT had DDH. Although the American Academy of Pediatrics does not include CMT as a risk factor in the screening guidelines for DDH, some authors suggest obtaining an ultrasound or anteroposterior pelvis radiograph in affected infants at the same age as standard DDH screening guidelines. Malposition of the knees and feet may occur with MBS. Congenital talipes equinovarus (clubfoot), metatarsus adductus, and tibial torsion have also been associated with intrauterine compression.

Item C237. A newborn with moulded baby syndrome. Reprinted with permission from Buxton RA. The moulded baby syndrome. Curr Orthop. 2001;15(6):439-443.

Torticollis is the most commonly associated finding in infants with plagiocephaly. Infants with torticollis often have some deformation of the occiput in utero, and some authors postulate that affected infants preferentially sleep on these flat occipital surfaces, exacerbating the development of plagiocephaly. Early referral to a physical therapist is recommended to teach caregivers to perform and monitor stretching exercises. Health care providers should also encourage "tummy time" while the infants are awake and monitored, to decrease time spent supine with the head in the infant's "preferred" position.

Genu recurvatum, or congenital dislocation of the knee, is a rare condition that can be either a malformation associated with a genetic condition, such as Larsen syndrome, or an isolated deformation related to oligohydramnios. Jaw subluxation is extremely rare in infants and a few case reports document an association with trauma. Camptodactyly is a flexion deformity of the proximal interphalangeal joints with the fifth finger always affected, usually with a genetic cause, and is autosomal dominant. Clavicular crepitus suggests an underlying fracture that might cause preferential head positioning. None of these findings are commonly associated with MBS.

PREP Pearls
- Moulded baby syndrome (MBS) may include head molding, postural torticollis, congenital scoliosis, pelvic obliquity with altered hip movement, and malposition of the knees and feet.
- MBS has been attributed to fetal positioning against the mother's spine, along with prolonged intrauterine constraint later in pregnancy
- Torticollis is the most commonly associated finding in infants with plagiocephaly.
- Although the American Academy of Pediatrics does not include congenital muscular torticollis in the screening guidelines for developmental dysplasia of the hip, some authors suggest ultrasonography or anteroposterior pelvis radiography in affected infants.

ABP Content Specifications(s)
- Understand how positional deformations and/or malformations develop in a fetus

Suggested Readings
- Buxton RA. The moulded baby syndrome. Curr Orthop. 2001;15:439-443. DOI: http://dx.doi.org/10.1054/cuor.2002.0232.
- Joiner ER, Andras LM, Skaggs DL. Screening for hip dysplasia in congenital muscular torticollis: is physical examination enough? J Child Orthop. 2014;8:115-119. DOI: http://dx.doi.org/10.1007/s11832-014-0572-5.
- Kuo AA, Tritasavit S, Graham JM. Congenital muscular torticollis and positional plagiocephaly. Pediatr Rev. 2014;35:79-87. DOI: http://dx.doi.org/10.1542/pir.35-2-79.
- Rubio AS, Griffet JR, Caci H, Berard E, El Hayek T, Boutte P. The moulded baby syndrome: incidence and risk factors regarding 1,001 neonates. Eur J Pediatr. 2009;168:605-611. DOI: http://dx.doi.org/10.1007/s00431-008-0806-y.
- Smith BG. Lower extremity disorders in children and adolescents. Pediatr Rev. 2009;30:287-294. DOI: http://dx.doi.org/10.1542/pir.30-8-287.

Question 238
A 14-year-old adolescent girl presents to your office with right hand and arm pain for the past month, which is worse at night. The pain is most severe in the palm of her hand and involves her arm up to her shoulder. She has numbness and tingling in her index and middle fingers. The patient states that running warm water over her hand helps. She had been taking ibuprofen, but states that it is not helping anymore. She denies hand or arm weakness. There is no history of trauma. She plays violin in the school orchestra and is having difficulty now because of the pain. Her physical examination is unremarkable, with no skin rashes or discolorations. Her neurological examination is also unremarkable. She has normal sensation and strength in the entire right upper extremity.

Of the following, the treatment MOST likely to improve her symptoms is

- A.   botulinum toxin injections
- B.   cervical discectomy
- C.   nocturnal wrist splinting
- D.   pain clinic referral
- E.   shoulder immobilization

Question 237   **Preferred Response: D**

The girl in the vignette has carpal tunnel syndrome. For mild symptoms, conservative treatment with nocturnal wrist splints can be beneficial. Other conservative measures include oral steroids, local steroid injections, and yoga.

Carpal tunnel syndrome starts with intermittent numbness and tingling in the wrist and first 3 digits of the hand. Pain occurs in the same area and commonly extends up to the shoulder. The absence of neck pain helps differentiate carpal tunnel syndrome from a cervical radiculopathy. As carpal tunnel syndrome progresses, the pain, numbness, and tingling become constant and are often worse at night. Findings on neurologic examination and electromyography/nerve conduction study are often normal until the later stages. The diagnosis is usually made clinically, and if treated in the early stages, progression can be prevented.

The carpal tunnel syndrome seen in the girl in the vignette is likely the result of repetitive hand movements from playing violin. She may need to change her technique or shorten playing times. If the pain is severe and persistent (=6 months), hand muscle weakness or evidence of denervation on electromyography/nerve conduction study may be seen; then surgical decompression should be considered.

Pain clinic referral may provide symptomatic relief; however, this is not the best choice to treat mild symptoms. The other response choices are not effective treatments for carpal tunnel syndrome.

PREP Pearls
- Violin, viola, guitar, and clarinet players are at risk for carpal tunnel syndrome.
- In carpal tunnel syndrome, pain can involve the entire arm but spares the neck.

ABP Content Specifications(s)
- Identify the etiology of peripheral neuropathy in patients of various ages
- Recognize the clinical findings associated with peripheral neuropathy in patients of various ages

Suggested Readings
- Dawson DM. Entrapment neuropathies of the upper extremities. N Engl J Med. 1993;329(27):2013-2018. DOI: http://dx.doi.org/10.1056/NEJM199312303292707.
- O'Connor D, Marshall S, Massy-Westropp N. Non-surgical treatment (other than steroid injection) for carpal tunnel syndrome. Cochrane Database Syst Rev. 2003;(1):CD003219. DOI: http://dx.doi.org/10.1002/14651858.CD003219.

Question 239

A 16-year-old African American adolescent presents to the emergency department in his hometown in Wisconsin, where it is nearing the end of winter. He complains of cramps and contractions in his hands that have lasted for hours. He experienced tingling in his arms and legs during basketball practice intermittently over the last 2 weeks, but assumed this was because of dehydration. He is concerned because the cramping in his hands today did not resolve with fluid intake.

While taking vital signs, the technician in the emergency department accidently left the blood pressure cuff inflated on the patient's arm. After 2 minutes, the adolescent's hand cramping significantly worsened. His wrist and metacarpophalangeal joints flexed and his fingers adducted. You quickly remove the blood pressure cuff and examine the patient. His physical examination is unremarkable, except for facial twitching at the nose and lip, elicited when tapping the angle of the jaw.

Of the following, the measurement MOST likely to reveal the cause of the patient's symptoms is

> A.   1,25-dihydroxyvitamin D
> B.   25-hydroxyvitamin D
> C.   alkaline phosphatase
> D.   creatine kinase
> E.   parathyroid hormone

Question 239   **Preferred Response: B**

The boy in the vignette has symptoms and signs consistent with hypocalcemia. Trousseau sign occurs when a blood pressure cuff is placed around the arm and inflated to a pressure greater than the systolic blood pressure and held in place. In the absence of blood flow, the patient's hypocalcemia and subsequent neuromuscular irritability will induce spasm of the hand and forearm muscles. Chvostek sign is an abnormal reaction to the stimulation of the facial nerve when hypocalcemia is present. When the facial nerve is tapped at the angle of the jaw, the facial muscles on the same side of the face will contract momentarily (typically a twitch of the nose or lips).

The most common reason for hypocalcemia to occur is vitamin D deficiency. Natural sunlight is the major source of vitamin D for children and adolescents, therefore inadequate exposure during the winter months may lead to low vitamin D levels. People with naturally dark skin tone require at least 3 to 5 times longer sun exposure to produce the same levels of vitamin D as those with light skin tone. Thus, for the adolescent in the vignette, vitamin D deficiency would be the most likely cause of hypocalcemia.

The laboratory findings in vitamin D deficiency are summarized in Item C239. Early in the development of vitamin D deficiency, serum parathyroid hormone (PTH) levels increase, which is followed later by increased alkaline phosphatase levels. Calcium and phosphorus may initially be normal. 25-hydroxyvitamin D, with a circulating half-life of 2 to 3 weeks, is the major circulating form and is the best indicator of vitamin D status.

	Vitamin D Deficiency (Early)	Vitamin D Deficiency (Late)
Serum calcium	Normal or low	Low
Serum phosphorus	Normal or low	Low
Serum alkaline phosphatase	Elevated	Very elevated
Serum parathyroid hormone	Elevated	Very elevated
25-hydroxyvitamin D	Low	Very low
1,25-vitamin D	Variable	Variable

Item C239. Biochemical Changes Seen in Vitamin D Deficiency. Courtesy of J Pinsker.

Although 1,25-dihydroxyvitamin D is traditionally taught to be the "active form" of the hormone, its measurement does not reflect vitamin D status. 1,25-dihydroxyvitamin D has a circulating half-life of approximately 4 hours. It circulates at 1,000 times lower concentration than 25-hydroxyvitamin, and the blood level is tightly regulated by serum levels of PTH, calcium, and phosphate. Serum 1,25-dihydroxyvitamin D is frequently either normal or even elevated in those with vitamin D deficiency because of secondary hyperparathyroidism.

Although the patient in this vignette has muscle aches, measurement of creatine kinase is not relevant when considered along with the patient's history and physical examination. Levels of PTH and alkaline phosphatase will both be elevated in the presence of hypocalcemia, however, they can be elevated due to any cause of increased bone turnover, therefore measurement will not identify the specific diagnosis.

The 2006 American Academy of Pediatrics guidelines on calcium and vitamin D intake include recommended sources of nutritional intake to optimize bone health. Guidelines from the Pediatric Endocrine Society also recognize the need to optimize nutritional intake, given the increased use of sunscreen during the summer and limited sun exposure during winter months.

PREP Pearls
- Measurement of 25-hydroxyvitamin D (the storage form of vitamin D) is the definitive test for diagnosing vitamin D deficiency.
- Vitamin D deficiency is the most common cause of low calcium levels in adolescence, especially during the winter months when sun exposure is limited.

ABP Content Specifications(s)
- Recognize the clinical features associated with hypocalcemia in patients of various ages, including that associated with vitamin D deficiency

Suggested Readings
- Greer FR, Krebs NF; American Academy of Pediatrics Committee on Nutrition. Optimizing bone health and calcium intakes of infants, children, and adolescents. Pediatrics. 2006;117(2):578-585. DOI: http://dx.doi.org/10.1542/peds.2005-2822.
- Holick MF, Binkley NC, Bischoff-Ferrari HA, et al; Endocrine Society. Evaluation, treatment, and prevention of vitamin D deficiency: an Endocrine Society clinical practice guideline. J Clin Endocrinol Metab. 2011;96(7):1911-1930. DOI: http://dx.doi.org/10.1210/jc.2011-0385.

Question 241

A 6-year-old boy comes for evaluation after he was diagnosed with amblyopia at a recent optometry evaluation. He was born at term with no complications, has only had a few minor childhood illnesses, and has had no eye trauma. On past routine physical examination, he had a normal cover-uncover test and pupillary light reflex.

Of the following, the MOST accurate statement about this child's amblyopia is that it

A.   can only be treated with eye patching

B.   is most likely to be related to a lesion that obstructs vision

C.   is often not detectable prior to routine vision screening

D.   is unlikely to respond to treatment at this age

E.   only occurs in patients with strabismus

Question 241   **Preferred Response: C**

Amblyopia is a neurologically based decrease in visual acuity that cannot be attributed to ocular structural abnormalities. It most often affects 1 eye, although it can be bilateral. In order for amblyopia to develop, an abnormality of visual input must occur during the critical period of visual cortex development before 7 or 8 years of age. Risk factors for the development of amblyopia are classified as: strabismic, anisometropic (a difference in refractive error between the 2 eyes), ametropic (marked refractive error in both eyes), or deprivation (the result of media opacity such as cataract or caused by adnexal abnormality such as ptosis or obstructing lesion). Astigmatism can also be a contributing condition. The most common risk factors for amblyopia are strabismus and anisometropia, accounting for 37% to 38% each in some studies. Approximately 2% to 4% of US children are affected with amblyopia, and most are asymptomatic in terms of vision until the amblyopia is detected by visual screening. Even if strabismus or an obstructing lesion is obvious, the presence of visual impairment cannot be diagnosed without vision screening.

To date, there have been limited data about the efficacy of vision screening of young, preliterate children for preventing amblyopia. The United States Preventive Services Task Force recommends vision screening at least once between 3 and 5 years of age, but does not currently recommend earlier screening because of the absence of data. However, with the development of handheld autorefractors and photoscreeners, pediatricians can effectively screen children between the ages of 6 months and 3 years, detecting the presence and magnitude of optical (refractive) and physical abnormalities at younger ages. The greatest barriers to implementing screening for these younger ages are cost for equipment, personnel, time, and the lack of assured reimbursement for this service by third party payers. The American Academy of Pediatrics states that it is unlikely that health care providers will adopt this early screening rapidly.

Treatment for amblyopia starts with addressing underlying factors such as strabismus or media opacity. The next step is corrective lenses, and for some children this will provide sufficient treatment to correct the amblyopia. However, many more children will be treated either with occlusion or penalization of the better eye. For 3- to 7-year-old children, patching the good eye for 2 to 6 hours per day produces excellent improvement for both those with moderate amblyopia (visual acuity 20/40-20/100) and severe amblyopia (visual acuity 20/100-20/400). However, compliance is difficult with this regimen and the incidence of bullying against these children is substantial. Instead, many families prefer atropine penalization (ie, blurring vision in the better eye with atropine drops), and for children with moderate amblyopia, the results of this treatment are similar to that of patching. Treatment continues to evolve with ongoing research into the duration needed for patching and the frequency of atropine drop use (weekend only versus daily treatment). While treatment is more effective, the earlier it is started, the evidence suggests that even older children may benefit, particularly if they had not previously received amblyopia treatment. Nearly half of adolescents aged 13 to 17 years at initiation of patching showed 10 letters of improvement or more in visual acuity at the completion of treatment.

PREP Pearls
- Amblyopia is a neurologically based decrease in visual acuity.
- Amblyopia can be secondary to strabismus, refractive errors, or visual deprivation.
- Vision screening is necessary to detect amblyopia, and routine vision screening should begin between 3 and 5 years of age. Newer technology may allow for earlier vision screening.
- Treatment for amblyopia involves treating any underlying condition and providing corrective lenses. Patching or atropine penalization of the better eye is often necessary and has good results.

ABP Content Specifications(s)
- Identify conditions that may lead to the development of amblyopia

Suggested Readings
- American Academy of Pediatrics. Instrument-based pediatric vision screening policy statement. Pediatrics. 2012;130(5):983-986. DOI: http://dx.doi.org/10.1542/peds.2012-2548.

Question 242

A local high school parent-teacher association invites you to speak at its next meeting about drug use among adolescents. During the presentation, a mother asks about trends in substance abuse among adolescents.

Of the following, the MOST accurate statement is

      A.   adolescent substance use is often associated with other risk behaviors
      B.   age of initiation for marijuana use has steadily decreased since 2002
      C.   males are significantly less likely than females to use alcohol on school property
      D.   seventy-five percent of high school students have tried alcohol
      E.   there has been an increase in the number of students who have used alcohol since

Question 242   Preferred Response: A

Alcohol and drug use among young adolescents represent an important public health risk. Substance use can put youth at greater risk for other health problems such as injuries, violence, and sexually transmitted infections. The Youth Risk Behavior Survey (YRBS) assesses substance use among high school students.

According to the 2013 YRBS, 66% of high school students have had at least 1 drink in their lifetime. This represents a decrease from 81% in 1999. Defined as at least 1 drink in the 30 days preceding the survey, 35% of high school students currently drink alcohol. Approximately 19% of students initiated alcohol use before 13 years of age.

Marijuana is a commonly used substance among adolescents. In 2013, 41% of high school students had used marijuana at least once. This represents a decrease from 47% in 1999. Defined as using marijuana at least once in the 30 days preceding the survey, 23% of high school students currently use marijuana. Approximately 9% of students initiated marijuana use before 13 years of age.

In 2011, boys reported using alcohol on school property slightly more than females. The average age of first marijuana use reported in 2011 was 17.5 years, which was similar or slightly older than the average age in recent years.

PREP Pearls
- Substance use can put youth at greater risk for other health problems such as injuries, violence, and sexually transmitted infections.
- According to the 2013 Youth Risk Behavior Survey, 66% of high school students have had at least 1 drink in their lifetime, and 41% of high school students had used marijuana at least once.

ABP Content Specifications(s)
- Identify the approximate initial age for experimentation with drugs of use/abuse
- Recognize general trends in substance use/abuse among children and adolescents
- Understand patterns of use/abuse of drugs with regard to multiple or single drugs

Suggested Readings
- Kann L, Kinchen S, Shanklin SL, et al. Youth Risk Behavior Surveillance — United States, 2013. MMWR Morbid Mortal Wkly Rep. 2014;63(SS04):1-168. http://www.cdc.gov/mmwr/preview/mmwrhtml/ss6304a1.htm.
- Substance Abuse and Mental Health Services Administration, US Department of Health and Human Services. Results from the 2011 National Survey on Drug Use and Health: summary of national findings. Substance Abuse and Mental Health Services Administration website. http://www.samhsa.gov/data/sites/default/files/Revised2k11NSDUHSummNatFindings/Revised2k11NSDUHSummNatFindings/NSDUHresults2011.htm .

Question 243

A 6-year-old boy develops a fever to 39.4°C, cough, and rhinorrhea. After 2 days of symptoms, his parents notice a mass in the middle of his neck and bring him to your office for evaluation (Item Q243). Upon further reflection, his parents recall that the mass presented once before when the boy had a similar infection, but then disappeared after his infection resolved. On physical examination, the neck mass moves upwards with protrusion of the tongue.

Item Q243. Neck mass of the patient in the vignette. Reprinted with permission from Geddes G, Butterly MM, Patel SM, Marra S. Pediatric neck masses. Pediatr Rev. 2013;34(3):115-125.

Of the following, the MOST likely diagnosis is

       A.   branchial cleft cyst
       B.   cystic hygroma
       C.   ectopic thyroid gland
       D.   thyroglossal duct cyst
       E.   thyroid nodule

Question 243   **Preferred Response: D**

The child in the vignette has a midline neck mass most consistent with a thryoglossal duct cyst. Thyroglossal duct cysts are midline anterior neck lesions that generally present after an upper respiratory tract infection, and can be acutely infected. They have a tract or fistula passing through the hyoid bone up to the base of the tongue. On physical examination, thyroglossal duct cysts may move with swallowing. Treatment is always complete excision, including removal of the middle one third of the hyoid bone. Without removal of the hyoid, the thyroglossal duct cyst may recur.

Branchial cleft cysts are the most common congenital neck lesions, accounting for approximately 20% to 30% of all pediatric neck masses. They most often appear on the lateral side of the neck. Complications of branchial cleft cysts include recurrent infection and fistula formation.

Cystic hygromas are spongy, mobile, nontender lesions located in the posterior triangle of the neck, most frequently on the left side. Large lesions can result in airway compromise. If a large cystic hygroma is detected prenatally, delivery should be performed at a center capable of managing the airway and lesion at the time of birth. Fine needle aspiration should be avoided when diagnosing cystic hygromas because hemorrhage into the lesion may cause rapid expansion.

An ectopic thyroid gland may be located anywhere along the path of descent of the thyroid during its embryologic development, most commonly at the base of the tongue. Although these are often midline, they can occur in many locations and present far less commonly as a midline mass compared with a thyroglossal duct cyst. In most cases ultrasonography is necessary to identify the location of an ectopic thyroid gland in a child with hypothyroidism, because there are usually no external signs of the thyroid's location.

Thyroid nodules are relatively uncommon in children. However, they have a high potential for malignancy, estimated to range anywhere from 9% to 50%. Because many children and adolescents with thyroid cancer have metastatic lesions in the cervical lymph nodes at presentation, they most often present with a lateral neck mass.

PREP Pearls
- Thyroglossal duct cysts are anterior neck midline lesions that generally present after an upper respiratory tract infection.
- Branchial cleft cysts are the most common congenital neck lesions, and most often appear laterally.

ABP Content Specifications(s)
- Recognize the clinical features associated with a thyroglossal duct cyst

Suggested Readings

- Geddes G, Butterly MM, Patel SM, Marra S. Pediatric neck masses. Pediatr Rev. 2013;34;115-125. DOI: http://dx.doi.org/10.1542/pir.34-3-115.
- Hopwood NJ, Kelch RP. Thyroid masses: approach to diagnosis and management in childhood and adolescence. Pediatr Rev. 1993;14(12). http://pedsinreview.aappublications.org/content/14/12/481.abstract.

Question 244

You are caring for an 8-year-old hospitalized boy who is receiving chemotherapy for acute lymphoblastic leukemia. He was admitted for fever, neutropenia, and septic shock 5 days ago and has improved after receiving fluid resuscitation, stress dose steroids, intravenous vancomycin and cefepime, and a dopamine infusion. He was weaned off the dopamine after 24 hours. His indwelling central line culture grew Pseudomonas aeruginosa, and after 48 hours, based on sensitivity results, the vancomycin was discontinued. Today, the boy had 8 loose, watery, foul-smelling stools. Stool studies have been sent.

Of the following, the MOST appropriate next step is

- A. discontinuation of cefepime
- B. fluconazole, 12 mg/kg intravenously every 24 hours
- C. ganciclovir, 5 mg/kg intravenously every 12 hours
- D. linezolid, 10 mg/kg orally every 8 hours
- E. metronidazole, 10 mg/kg orally every 6 hours

Question 244   **Preferred Response: E**

The child in the vignette, who has received antibiotic therapy for a central line infection, has pseudomembranous colitis manifested by loose, watery, foul smelling stools. The best treatment for the infection is oral metronidazole.

Pseudomembranous colitis is caused by Clostridium difficile infection of the colon. C difficile is a gram-positive anaerobic bacillus capable of producing a toxin that affects intracellular signaling pathways of colonic epithelium, resulting in inflammation and cell death. Infants younger than 1 year may not develop pseudomembranous colitis from C difficile infection because they may lack the toxin receptor. Any process that disrupts normal gastrointestinal flora, alters immunity, or impairs motility can lead to an infection with a C difficile toxin-producing strain. This can include inflammatory bowel disease, ileus, broad-spectrum antibiotic usage, immunosuppression, and chronic illness. Both the incidence and clinical severity of pseudomembranous colitis have been rising in recent years, especially in the pediatric population. Classic pseudomembranous colitis was strictly a hospital-acquired infection, but its incidence in the community is increasingly recognized.

Broad-spectrum antibiotic use is a common cause of pseudomembranous colitis, though the absence of this history does not rule it out. Antibiotics can kill the normal gastrointestinal flora, leading to selection of resistant organisms and colonization with C difficile. Diagnosis is made by detection of C difficile toxin in the stool. The clinical presentation of pseudomembranous colitis begins within days of colonization, and can range from mild, self-limited diarrhea and cramping to more severe manifestations such as fever, bacteremia, sepsis, abdominal distention, toxic megacolon, and even death.

For the patient in the vignette, risk factors for pseudomembranous colitis include the antecedent history of broad-spectrum antibiotics and immunocompromise. Either oral metronidazole or oral vancomycin is the best therapy for pseudomembranous colitis. Discontinuation of cefepime is not advisable because treatment of his central line infection is necessary. Fluconazole and ganciclovir will not be helpful, because the cause of the diarrhea is not likely to be fungal or viral. Oral linezolid is effective against gram-positive bacteria, but it is not a first-line therapy for C difficile infection because its spectrum is too broad, and it is a bacteriostatic agent.

PREP Pearls
- Pseudomembranous colitis is caused by C difficile infection in the setting of altered immunity, motility, or balance of colonic flora.
- Treatment of pseudomembranous colitis is oral metronidazole or oral vancomycin.
- Reducing the use of broad-spectrum antibiotics is important in the prevention of pseudomembranous colitis.

ABP Content Specifications(s)
- Recognize the association of pseudomembranous colitis with antibiotic therapy, and manage appropriately

American academy of pediatrics   778

Suggested Readings

- Feigin RD. Antimicrobial agent-induced pseudomembranous colitis. Pediatr Rev. 1981;3:147-152. DOI: http://dx.doi.org/10.1542/pir.3-5-147.
- Mezoff EA, Cohen MB. Clostridium difficile infection. In: Kliegman RM, Stanton BF, St. Geme JW III, Schor NF, Behrman RE, eds. Nelson Textbook of Pediatrics. 20th ed. Philadelphia, PA: Saunders Elsevier; 2015:1434-1436 .

Question 245

A 17-month-old girl presents to the emergency department with a limp for 1 week. Her parents have noted progressive swelling of the left knee. Fever has not been documented, but she has been on scheduled ibuprofen since the onset of illness. She has had concurrent nasal symptoms and mild cough. Vital signs show a temperature of 37.2°C, respiratory rate of 24 breaths/min, heart rate of 130 beats/min, blood pressure of 91/53 mm Hg, and a weight of 10.2 kg. On physical examination, she has warmth, a notable effusion, and limited range of motion of the left knee. Laboratory data shows:

White blood cells, 19,300/μL (19.3 x 10^9/L)
Hemoglobin, 10.9 g/dL (10^9 g/L)
Platelets, 303 x 10^3/μL (303 x 10^9/L)
Differential, 24% segmented neutrophils, 3% bands, 68% lymphocytes, 5% monocytes
Sedimentation rate, 50 mm/h
C-reactive protein, 2.3 mg/L
Joint fluid culture, Kingella kingae

Of the following, the BEST treatment for this child's infection is

- A.   cefotaxime
- B.   clindamycin
- C.   linezolid
- D.   trimethoprim
- E.   vancomycin

Question 245   **Preferred Response: A**

Kingella kingae infections are usually susceptible to β-lactam antibiotics, including penicillins and cephalosporins. Of the answers listed, cefotaxime is the correct antibiotic choice for the patient in this vignette. Additional antibiotics with activity against Kingella include aminoglycosides, macrolides, tetracyclines, chloramphenicol, and fluoroquinolones.

Approximately 40% of Kingella are resistant to clindamycin and all are resistant to glycopeptide antibiotics, including vancomycin. Kingella is a gram-negative pathogen. Clindamycin, linezolid, and vancomycin have good gram-positive activity and would be considered in osteoarticular infections caused by gram-positive pathogens such as Staphylococcus aureus; however, these antibiotics have no activity against Kingella. Kingella are also resistant to trimethoprim. However, the combination of trimethoprim with sulfamethoxazole would be effective against Kingella.

Kingella can be an asymptomatic colonizer of the posterior pharynx in 9% to 12% of children between 12 and 24 months of age. Frequently, patients with invasive Kingella infections have viral infections including upper respiratory tract symptoms, gingivostomatitis, or oral ulcers that may allow for invasion of bacteria into the respiratory epithelium and subsequent translocation into the bloodstream.

Kingella is increasingly recognized as a cause of osteoarticular infections, including septic arthritis, osteomyelitis, spondylodiscitis, and tenosynovitis in young children. The majority of infections occur in children between 6 and 48 months of age. Additional manifestations of Kingella infection can include bacteremia and endocarditis (Kingella is one of the HACEK organisms; Haemophilus, Actinobacillus, Cardiobacterium, Eikenella, and Kingella, that can all be found as normal flora in the oral cavity). In comparison with other pathogens that can cause osteoarticular infections and bacteremia, constitutional symptoms, including fever, can be mild or absent in patients with Kingella infection. Similarly, inflammatory laboratory indices are mildly to moderately elevated. Overall, a subacute course is more common.

In young children, in comparison with older children and adults, disease occurs in healthy individuals without underlying conditions. Daycare attendance may be a risk factor for Kingella colonization.

PREP Pearls
- Kingella can be an asymptomatic colonizer of the posterior pharynx of young children.
- Kingella is increasingly recognized as a cause of osteoarticular infections, including septic arthritis, osteomyelitis, spondylodiscitis, and tenosynovitis in young, healthy children.
- Kingella kingae infections are usually susceptible to β-lactam antibiotics, including penicillins and cephalosporins.

ABP Content Specifications(s)
- Recognize the clinical features associated with Kingella kingae infection

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Suggested Readings

- American Academy of Pediatrics. Kingella kingae infections. In: Kimberlin DW, Brady MT, Jackson MA, Long SS, eds. Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:500-501.
- Yagupsky P, Porsch E, St Geme J III. Kingella kingae: an emerging pathogen in young children. Pediatrics. 2011;127(3):557-565. DOI: http://dx.doi.org/10.1542/peds.2010-1867 .

Question 246

A 9-year-old boy presents to your office with the chief complaint of having cola-colored urine for 2 days. His review of systems is significant for an upper respiratory tract infection-like illness 3 weeks ago. On physical examination, the boy has normal growth parameters. He has a respiratory rate of 18 breaths/min, heart rate of 94 beats/min, and blood pressure of 130/90 mm Hg. The remainder of the physical examination is unremarkable. 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.

You suspect acute poststreptococcal glomerulonephritis (PSGN) as the cause of the patient's symptoms and clinical findings. A resident working with you asks how to distinguish PSGN from other forms of glomerulonephritis.

Of the following, you are MOST likely to tell the resident that

    A. duration of antecedent illness and hematuria is similar in poststreptococcal glomerulonephritis and immunoglobulin A nephritis

    B. membranoproliferative glomerulonephritis may be indistinguishable initially from poststreptococcal glomerulonephritis

    C. poststreptococcal glomerulonephritis is associated with activation of the classic complement pathway, whereas alternate complement pathway activation is present in lupus nephritis

    D. recurrent episodes of hematuria are common after recovery from poststreptococcal glomerulonephritis, but not with immunoglobulin A nephritis

    E. serology for streptococcal infections is rarely positive when nephritis is diagnosed in both poststreptococcal glomerulonephritis and membranoproliferative glomerulonephritis

Question 246   **Preferred Response: B**

Glomerulonephritis (GN) refers to the immune-mediated (noninfectious) inflammation of the renal parenchyma. The patient in the vignette has clinical features of acute GN (cola-colored urine and hypertension). Serum chemistries will likely reveal azotemia and electrolyte abnormalities, depending on the severity of renal failure.

The timing of infectious illness and acute nephritis can provide clues to the presenting nephritis. In patients with postinfectious GN, history of illness and acute nephritis is usually separated by 7 to 21 days. However, the onset of acute GN within days of viral infection is seen in immunoglobulin A (IgA) glomerulonephritis, Alport syndrome, and membranoproliferative glomerulonephritis (MPGN).

Further classification of acute GN requires measurement of complement components C3 and C4. Determining whether the patient has hypocomplementemic (associated with a low C3) or normocomplementemic (associated with a normal C3) GN helps in differentiating various nephritides (Item C246). Postinfectious GN, MPGN, and systemic lupus erythematous nephritis (SLE nephritis) are the most frequently identified hypocomplementemic GN in children. The combination of a low C3 level and a normal or slightly decreased C4 level indicates activation of the alternative pathway of complement, as in postinfectious GN or MPGN. In patients with SLE nephritis, there is immune-mediated activation of the classical pathway associated with reductions in both C3 and C4.

Item C246. Classification of Glomerulonephritis (GN) by Complement Level.

Low C3 GN	Normal C3 GN
Postinfectious/Poststreptococcal	Immunoglobulin A GN
Membranoproliferative GN (more common)	Henoch–Schönlein purpura nephritis
Lupus nephritis	Antineutrophil cytoplasmic autoantibody-associated GN
Shunt nephritis	Alport syndrome
Subacute bacterial endocarditis	Membranoproliferative GN (1/3 of cases)

Courtesy of G. Kapur

Postinfectious GN is characterized by immune complex-mediated nephritis after an infectious process. Poststreptococcal GN (PSGN) exclusively relates to a group A, β-hemolytic streptococcal infection. The terms postinfectious GN and PSGN are often used interchangeably because PSGN is the most common acute nephritis in children worldwide. Elevated titers of antibodies to streptococcal antigens are present only in patients in whom postinfectious GN is associated with streptococcal infection. However, in postinfectious GN secondary to nonstreptococcal infections, streptococcal antibody titers will be normal. The streptozyme test measures different streptococcal antibodies and is positive in nearly 95% of patients with pharyngitis, and around 80% of patients with skin infections because of group A, β-hemolytic streptococcus preceding acute nephritis. It includes the anti-streptolysin (ASO), anti-hyaluronidase (AHase), anti-streptokinase (ASKase), anti-nicotinamide-adenine dinucleotidase (anti-NAD), and anti-DNAse B antibodies. After a pharyngeal infection with group A, β-hemolytic streptococcus, the ASO, anti-DNAse B, anti-NAD, and AHase titers are elevated, while only the anti-DNAse B and AHase titers are elevated after a skin infection. The ASO titers are elevated in nearly 80% of patients with PSGN.

Initial urine microscopy shows hematuria, pyuria (glomerular inflammation), and red blood cell casts. Subsequent urine microscopy in patients with postinfectious GN may show persistent microscopic hematuria (which may persist for up to 1 to 3 years in some cases). This has not been associated with a worse prognosis in the patients.

Membranoproliferative GN similar to postinfectious GN can follow a viral illness. It is difficult to differentiate MPGN from postinfectious GN, especially in patients with no history of preceding infection or in patients with short duration between infection and onset of nephritis. As postinfectious GN is more common, these patients are treated as such and followed for resolution of hypocomplementemia and improvement in renal function. Hypocomplementemia usually resolves by 6 to 8 weeks in most cases of postinfectious GN, and persistently decreased C3 levels are suggestive of MPGN. Nephrotic syndrome is more frequently seen in patients with MPGN, as compared to postinfectious GN.

The treatment for acute GN is usually supportive and includes fluid restriction, low sodium diet, and diuretics or vasodilators if patients have edema and hypertension. Renal biopsy is rarely indicated in patients with suspected postinfectious GN. Presence of severe azotemia, rapidly rising serum creatinine (suggestive of rapidly progressive glomerulonephritis), or persistently low complement levels (beyond 6-8 weeks) are indications for renal biopsy in patients with suspected postinfectious GN. Membranoproliferative GN can be differentiated from postinfectious GN by the characteristic renal biopsy findings in each condition.

Patients with IgA nephropathy often present after an upper respiratory infection, similar to the presentation of patients with postinfectious GN. Potential distinguishing features from postinfectious GN include a shorter time between the antecedent illness and nephritis and normal complement levels. History of episodes of gross hematuria prior to the first episode or after the initial episode of nephritis is also suggestive of IgA nephritis, as recurrences are rare in postinfectious GN.

PREP Pearls
- Poststreptococcal glomerulonephritis (PSGN) related to group A, β-hemolytic streptococcal infection is the most common acute nephritis in children worldwide.
- It is difficult to differentiate membranoproliferative glomerulonephritis (MPGN) from postinfectious glomerulonephritis, especially in patients with no history of preceding infection or in patients with a short duration between infection and onset of nephritis.
- Hypocomplementemia resolves by 6 to 8 weeks in most cases of postinfectious glomerulonephritis (GN).
- Persistently decreased C3 levels are suggestive of MPGN.
- A low C3 level in acute GN indicates activation of the alternative pathway of complement (as in postinfectious GN or MPGN).

ABP Content Specifications(s)
- Recognize complications associated with post-streptococcal glomerulonephritis
- Differentiate acute post-streptococcal glomerulonephritis from other forms of glomerulonephritis

Suggested Readings
- 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: http://dx.doi.org/10.1007/s00467-010-1554-6.
- Massengill SF. Hematuria. Pediatr Rev. 2008;29(10):342-348. DOI: http://dx.doi.org/10.1542/pir.29-10-342.
- Varade WS. Nephritis. 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; 2009:2291-2292.

Question 247

An 18-month-old boy is brought to the office for a health supervision visit. He was delivered by spontaneous vaginal delivery at 27 weeks of gestation due to maternal preeclampsia. His birth weight was 980 g. He has been growing and developing normally with no significant medical issues to date.

On physical examination, he is alert and energetic without scleral icterus or jaundice. His height and weight are at the 25th percentage for age. Abdominal examination is notable for hepatomegaly, with his liver edge palpable 4 cm below the right costal margin. The liver is firm and nontender. No splenomegaly is palpated.

Laboratory studies show:

- Aspartate aminotransferase, 35 U/L
- Alanine aminotransferase, 41 U/L
- Bilirubin, 1.5 mg/dL (25.7 µmol/L)
- Direct bilirubin, 0.1 mg/dL (1.7 µmol/L)
- Platelets, 500 x 10^3/µL (500 x 10^9/L)
- α-fetoprotein, 100,000 ng/mL (100,000 µg/L)

Of the following, the BEST next step in evaluation is

A.   abdominal radiograph

B.   magnetic resonance cholangiopancreatography

C.   nuclear medicine liver and spleen scan

D.   positron emission tomography

E.   ultrasonography with Doppler

Question 247   **Preferred Response: E**

The former premature child in this vignette with an enlarged liver and significant elevation in α-fetoprotein most likely has hepatoblastoma. Premature infants are at increased risk for hepatoblastoma, as are patients with Beckwith-Wiedemann syndrome and families with familial adenomatous polyposis coli (FAP). Abdominal ultrasonography with Doppler will confirm a hepatic mass and, of the studies listed, would be the best next step in evaluation.

Hepatomegaly is a nonspecific finding and not a reliable predictor of liver disease. There is significant variability in the size and shape of the liver. If an enlarged liver is suspected, physical examination should assess the liver span at the mid-clavicular line using palpation and percussion. The liver edge should be less than 2 cm below the costal margin at the mid-clavicular line, with the exception of children younger than 2 years of age, when it may be palpable to 3.5 cm below the costal margin. The normal values for liver span are based on gender, age, and body weight, with a variability of +/- 2 to 3 cm. An abdominal ultrasonography can be used to confirm the size of the liver and provides some assessment of density and evaluation for a liver mass, if present. Laboratory studies may include a complete blood cell count, comprehensive metabolic panel, total and direct bilirubin, γ-glutamyl transferase, and an assessment of synthetic function with a prothrombin time and partial thromboplastin time. Evaluations for infectious etiologies and storage disease should be considered based on the history. Liver biopsy may be required to confirm a diagnosis.

The differential diagnosis for hepatomegaly is quite broad and is summarized in Item C247. Splenomegaly occurs with portal hypertension, storage disease, inflammation, infection, and malignancy.

Item C247. Differential Diagnosis for Hepatomegaly.

Inflammation
  Viral: Hepatitis A through E, Cytomegalovirus, Epstein-Barr virus,
    Coxsackievirus
  Bacterial: sepsis, abscess, cholangitis
  Toxin: acetaminophen
  Drugs
  Autoimmune

Inappropriate Storage
  Fat: nonalcoholic fatty liver disease, mitochondrial disease, malnutrition,
    galactosemia, cystic fibrosis, diabetes mellitus, parenteral nutrition
  Glycogen: Glycogen storage disease, diabetes mellitus, parenteral nutrition
  Lipids: Gaucher disease, Niemann-Pick disease, Wolman disease
  Abnormal Proteins: α-1 antitrypsin deficiency, carbohydrate-glycoprotein
    deficiency
  Metals: Wilson disease, hemochromatosis

Infiltrating Disease
  Primary tumors: Hepatoblastoma, hepatocellular carcinoma, hemangioma,
    focal nodular hyperplasia
Secondary or metastatic tumors
Cysts: Parasitic, choledochal, polycystic liver disease
Hemophagocytic syndromes
Extramedullary hematopoiesis

Vascular congestion
  Intrahepatic Obstruction to Hepatic Vein Outflow
   • Venoocclusive disease
  Suprahepatic obstruction
   • Congestive heart failure
   • Pericardial disease
   • Hepatic vein thrombosis (Budd-Chiari)

Biliary Obstruction
  Cholelithiasis
  Biliary atresia
  Congenital hepatic fibrosis
  Caroli disease
  Choledochal cyst
  Tumors
  Idiopathic

Modified and reprinted with permission from Wolf, AD; Levine JV. Hepatomegaly in neonates and
children. *Pediatr Rev.* 2000;21(9):303-310

Abdominal radiographs are not effective at assessing liver size or density. Magnetic resonance cholangiopancreatography is used to assess bile ducts and anatomy, and may be useful in the evaluation of hepatomegaly, but it is not the first imaging modality in the evaluation. Similarly, nuclear medicine liver and spleen scans and positron emission tomography may be used in the evaluation of hepatomegaly to assess blood flow or for tumor evaluations, but are not used to confirm hepatomegaly or as the initial screen for a mass.

PREP Pearls
- Hepatomegaly should be confirmed by physical examination and abdominal ultrasonography.
- Liver size varies with age, sex, and body weight.
- Hepatomegaly may be associated with an acute viral illness.
- Children with hepatomegaly without evidence of recent acute viral illness should be referred to pediatric gastroenterology for further evaluation.

ABP Content Specifications(s)
- Understand the significance of hepatomegaly with or without splenomegaly in children of various ages, and evaluate appropriately

Suggested Readings
- Abu-Wasel B, Walsh C, Keough V, Molinari M. Pathophysiology, epidemiology, classification and treatment options for polycystic liver diseases. World J Gastroenterol. 2013;19(35):5775-5786. DOI:http://dx.doi.org/10.3748/wjg.v19.i35.5775.

Question 248

A 2-month-old infant presents to your office for a health care supervision visit. On examination, you note disproportionate small stature with proximal shortening of the arms and legs, trident hands, hypotonia, a large head, frontal bossing, and midfacial hypoplasia. You suspect a skeletal dysplasia.

Of the following, the MOST likely complication to increase the risk of death in infancy in this patient would be

A.   craniocervical junction compression

B.   hydrocephalus

C.   lumbar spinal stenosis

D.   narrow bell-shaped thorax

E.   obstructive sleep apnea

Question 248   **Preferred Response: A**

Achondroplasia is the most common inherited skeletal dysplasia that results in disproportionate small stature with affected individuals having short arms and legs, macrocephaly, frontal bossing, and midfacial retrusion (Item C248). Normal intelligence and life span is expected, although hypotonia in infancy can delay motor milestones. Craniocervical junction compression can increase the mortality risk in infancy and is the leading cause of mortality in the first year of life. Population-based studies suggest that the risk of death with craniocervical junction compression could be up to 7.5% and is directly related to central apnea, associated with damaged respiratory control centers. Therefore, every infant with achondroplasia should have computed tomography or magnetic resonance imaging of the craniocervical junction in infancy. Additionally, overnight polysomnography should be completed in infancy and interpreted by a pediatric pulmonologist who is an expert in features commonly seen in craniocervical junction abnormalities, such as central hypopnea. A thorough neurologic examination for signs of cervical myelopathy should be performed at each physical examination in infancy and childhood. The best predictors for craniocervical junction constriction include lower limb hyperreflexia or clonus, central hypopnea on sleep study, or reduced foramen magnum size on computed tomography of the craniocervical junction. If symptomatic compression is present, an urgent referral to a pediatric neurosurgeon is required. Careful attention to head circumference, utilizing growth curves standardized for achondroplasia, is mandatory at each well child visit. Hydrocephalus can be present in up to 5% of cases, requiring a ventriculoperitoneal shunt to alleviate increased intracranial pressure.

Item C248. Child with achondroplasia. Courtesy of L. Parsley.

The management of achondroplasia depends on the complications that may arise. Middle ear dysfunction is common, requiring tympanostomy tube placement and close monitoring of hearing. If significant dysfunction is present, a delay in language development can occur. A newborn screening hearing test should be checked initially and repeated by 1 year of age. An orthopedist should investigate any progressive bowing of the legs. A sleep study should be performed during infancy because of the risk for obstructive sleep apnea (OSA). However, many patients develop OSA as an older child or adult, so careful attention to this clinical manifestation is required during each well child visit. Obstructive sleep apnea may be caused by a combination of midfacial retrusion resulting in a smaller airway size, hypertrophy of the lymphatic ring, abnormal innervation of the musculature in the airway, or, in the worst case scenario, craniocervical constriction. Treatment for OSA in patients with achondroplasia may include adenotonsillectomy, weight reduction, continuous positive airway pressure, or tracheostomy in extreme cases.

Significant short stature is universally present with achondroplasia and growth charts specific for patients with achondroplasia should be utilized for routine growth checks. Obesity is common and can worsen the morbidity associated with lumbar stenosis and contribute to joint problems. The development of kyphosis typically occurs in the first 3 years of life, and therefore should be clinically assessed by physical examination every 6 months during that time period. Lumbar spinal stenosis is more common in adulthood. A clinical history and neurologic examination is warranted every 3- to 5-years once an individual with achondroplasia reaches adulthood to look for spinal stenosis.
Screening of developmental milestones throughout infancy and childhood is important. Development attainment should be compared to specific milestone assessments standardized to patients with achondroplasia. A speech evaluation is recommended by 2 years of age if any delays are noted.

Achondroplasia is an autosomal dominant disorder caused by FGFR3 gene mutations. Approximately 80% of individuals with achondroplasia have parents with average stature, with the mutation caused by a de novo gene mutation unique to that individual. An individual with achondroplasia will have a 50% risk of passing it on to their offspring if the reproductive partner is of average stature. When both parents have achondroplasia, the risk of having a child with average stature is 25%, of having achondroplasia is 50%, and of having homozygous achondroplasia, a lethal condition, is 25%.

Of the answer choices in the vignette, craniocervical junction compression would lead to the highest risk of death in infancy due to central hypopnea. Hydrocephalus, lumbar spinal stenosis, and OSA are common complications during childhood, but are not typically associated with a high risk of mortality. Lumbar spinal stenosis is more commonly seen as an adult complication. A hypoplastic thorax is not commonly seen with achondroplasia, but can be seen in other skeletal dysplasias, such as thanatophoric dysplasia, campomelic dysplasia, or achondrogenesis.

PREP Pearls
- Craniocervical junction compression can increase the mortality risk in infancy in patients with achondroplasia.
- Every infant with achondroplasia should have a computed tomography or magnetic resonance imaging of the craniocervical junction, overnight polysomnography, and a thorough neurologic examination for signs of cervical myelopathy. The best predictors for craniocervical junction constriction include lower limb hyperreflexia or clonus, central hypopnea on sleep study, or reduced foramen magnum size on computed tomography of the craniocervical junction.
- Achondroplasia is the most common inherited skeletal dysplasia that results in disproportionate small stature, with affected individuals having short arms and legs, macrocephaly, frontal bossing, and midfacial retrusion. Normal intelligence and life span is expected, although hypotonia in infancy can delay motor milestones.
- Other common complications of achondroplasia include hydrocephalus, delayed motor milestones, sleep apnea, middle ear dysfunction, hearing loss, kyphosis, bowing of the legs, obesity, and lumbar spinal stenosis.

ABP Content Specifications(s)
- Recognize the clinical findings associated with achondroplasia, including complications

Suggested Readings
- Ireland PJ, Johnson S, Donaghey S, et al. Developmental milestones in infants and young Australasian children with achondroplasia. J Dev Behav Pediatr. 2010;31(1):41–47. DOI: http://dx.doi.org/10.1097/DBP.0b013e3181c72052 .
- Pauli RM. Achondroplasia. GeneReviews. http://www.ncbi.nlm.nih.gov/books/NBK1152/ .
- Pauli RM. Achondroplasia. In: Cassidy SB, Allanson JE, eds. Management of Genetic Syndromes. 3rd ed. New York, NY: John Wiley & Sons; 2010:17-37 .
- Trotter TI, Hall JG. Health supervision for children with achondroplasia. Pediatrics. 2005;116(3):771-783. DOI:http://dx.doi.org/10.1542/peds.2005-1440 .
- Wynn J, King TM, Gambello MJ, Waller DK, Hecht JK. Mortality in achondroplasia study: a 42-year follow-up. Am J Med Genet A. 2007;143A(21):2502-2511. DOI: http://dx.doi.org/10.1002/ajmg.a.31919 .

Question 249

You are examining an African-American male newborn at 36 hours of age. He was a full-term newborn without complications. There were no maternal problems during pregnancy. The newborn has been rooming in with his mother and doing well. He is alert, active, and well-appearing, with normal vital signs. The patient's mother states that she noticed tiny pustules on his face, chin, and neckline at birth, but these have begun to disappear. On physical examination, you find a few 2-mm vesicles and white pustules on the trunk and neckline without surrounding erythema or induration. There are several faint brownish macules with a collarette of scale in the perioral area (Item Q249). The remainder of the newborn's physical examination is unremarkable.

Item Q249

Item Q249. Macules, as described for the newborn in the vignette. Reprinted with permission from Krowchuk DP, Mancini AJ, eds. Pediatric Dermatology. A Quick Reference Guide. Elk Grove Village, IL: American Academy of Pediatrics; 2012.

Of the following, the MOST likely diagnosis is

A.   congenital candidiasis

B.   erythema toxicum neonatorum

C.   impetigo neonatorum

D.   miliaria

E.   transient neonatal pustular melanosis

Question 249   **Preferred Response: E**

The neonate described in the vignette has transient neonatal pustular melanosis (TNPM). Transient neonatal pustular melanosis is a benign, self-limited disorder characterized by superficial vesicles or sterile pustules that rupture easily leaving a hyperpigmented macule surrounded by a fine white "collarette of scale." The pustular lesions usually resolve within 24 to 48 hours, whereas the hyperpigmented macules fade gradually over several weeks to months. The distribution of lesions most commonly involves the chin, forehead, neck, lower back, and shins, but may be diffuse. The cause is unknown. Transient neonatal pustular melanosis occurs more commonly in black infants. Wright stain smear of TNPM pustular material demonstrates neutrophils and cellular debris; culture is negative. There are no systemic manifestations and no treatment is needed.

Differentiating this benign condition from other pustular eruptions seen in newborns is important because of the potential for serious illness if the infant has a congenital infection. Laboratory findings can be helpful in corroborating or excluding suspected clinical diagnoses. Congenital candidiasis presents at birth or in the first few days after birth as a diffuse erythematous papular, pustular, exfoliating rash that may involve the palms and soles. Potassium hydroxide preparation of the pustular material reveals pseudohyphae or spores and Candida spp will be found on culture. Erythema toxicum neonatorum is characterized by evanescent blotchy erythematous macules in combination with 1- to 3-mm papules and pustules that occur predominantly on the face, trunk, and extremities, sparing the palms and soles. Cytologic examination of a smear from a pustule will reveal a predominance of eosinophils on Wright or Giemsa stain and culture will be negative. Impetigo neonatorum usually presents as superficial vesicular, pustular, or bullous lesions on an erythematous base. These lesions are easily ruptured, leaving a denuded red moist surface with later crust formation. Gram-positive cocci and neutrophils will be seen on Gram stain and a culture will yield streptococci or staphylococci. Pinpoint 1- to 2-mm clear vesicles without surrounding erythema are characteristic of miliaria crystallina. Myriads of tiny pink papules are characteristic of miliaria rubra. Miliaria is most often distributed in the intertriginous regions, on the face, or areas that have been occluded, such as the upper back, and usually is associated with warming of the infant. Lymphocytes may be seen on Wright stain or Gram stain of a smear of the contents of a miliaria vesicle and culture is negative.

PREP Pearls
- Transient neonatal pustular melanosis (TNPM) is characterized by sterile superficial vesicles or pustules that rupture easily, leaving hyperpigmented macules surrounded by a fine white "collarette" of scale.
- The pustular lesions of TNPM resolve spontaneously within 24 to 48 hours, leaving hyperpigmented macules that fade gradually over several weeks to months.
- Neutrophils and cellular debris are seen on Wright stain smear of TNPM lesions.

ABP Content Specifications(s)
- Recognize the clinical findings associated with transient neonatal pustular melanosis

- Differentiate the laboratory findings associated with transient neonatal pustular melanosis from those of staphylococcal pustules

Suggested Readings

- Johr RH, Schachner LA. Neonatal dermatologic challenges. Pediatr Rev. 1997;18(3):86-94. DOI:http://dx.doi.org/10.1542/pir.18-3-86.
- Paller AS, Mancini AJ. Cutaneous disorders of the newborn. In: Hurwitz Clinical Pediatric Dermatology: A Textbook of Skin Disorders of Childhood and Adolescence. 4th ed. Philadelphia, PA: Elsevier Saunders; 2011:18-19.

Question 250

A 16-year-old female adolescent presents to your office with a swollen and tender left lower leg. She had been previously well, with no fevers, weight loss, or rash. She denied any trauma or known insect bites. She has been on the track team and has denied ever smoking. She has not been sexually active, has not been taking birth control pills, has not been on any long trips, and has not been immobilized for any period of time. On physical examination, she is well appearing. Her temperature is 37°C, her heart rate is 82 beats/min, blood pressure is 108/78 mm Hg, and her respiratory rate is 20 breaths/min. Her height and weight are both at the 50th percentile for age. Her left lower leg is visibly larger than her right and is tender to the touch posteriorly. She has normal pedal pulses bilaterally and a normal gait.

You refer her for ultrasonography of the left lower extremity, which reveals a nonocclusive venous thrombus in the popliteal vein.

Of the following, further evaluation and management of this patient should include

   A.   anti-ß2 glycoprotein antibodies, pulse oximetry, and anticoagulation with enoxaparin

   B.   factor V Leiden, a chest radiograph, and anticoagulation with aspirin

   C.   factor V Leiden, a spiral chest computed tomography, and, if both are normal, no anticoagulation

   D.   protein C and S levels, a magnetic resonance angiogram of the head, and anticoagulation with enoxaparin

   E.   protein C and S levels, ultrasonography of the right lower extremity, and anticoagulation with aspirin

Question 250   **Preferred Response: A**

Awareness of the short and long term consequences of thrombosis in children and adolescents has been increasing. The teenager in the vignette presents with findings typical for a lower leg deep vein thrombosis (DVT). Factors that increase the risk for thrombus formation in an otherwise well adolescent are shown in Item C250. While the patient does not have any identifiable risk factors in her history, it is imperative to evaluate her for a heritable or acquired thrombophilia. Such an evaluation should include levels of protein C and S, an assessment for the presence of the factor V Leiden mutation, gene sequencing to assess for the presence of a prothrombin mutation, and testing for the presence of antiphospholipid antibodies. The risk of having a pulmonary embolus as a consequence of a lower extremity DVT can be readily assessed clinically. The presence of chest pain and any family history of thrombosis should be queried in the history. The physical examination should include a heart rate, respiratory rate, blood pressure, and pulse oximetry. If there is no history of chest pain and the vital signs including pulse oximetry are all normal, the risk of a pulmonary embolus is low, and no further evaluation of the lungs is needed. The most appropriate management of a lower extremity DVT is the initiation of therapeutic anticoagulation. Enoxaparin is easier to manage, requires far less monitoring than anticoagulation with warfarin, and has generally become the first line therapy for children with thromboses. There are newer oral anticoagulation agents (including factor X inhibitors and thrombin inhibitors) that are currently in clinical trials, but are not yet ready to become part of standard medical practice.

Item C250. Risk Factors for Deep Vein Thrombosis.

- Estrogen-containing birth control pills
- Pregnancy
- Smoking
- Overweight or obesity
- Prolonged immobility:
    - i)   Long car rides / flights
    - ii) Studying with a leg folded under the buttocks
    - iii) Post-surgery, trauma, or hospitalization
- Inflammatory bowel disease
- Cancer
- A family history of thrombosis
- A heritable thrombophilia
    - i)   Factor V Leiden
    - ii) Prothrombin gene mutation
    - iii) Protein C deficiency
    - iv) Protein S deficiency
    - v)   Antithrombin III deficiency
- An acquired thrombophilia – antiphospholipid antibodies

Courtesy of J. Fish

The anti-β2-glycoprotein and anticardiolipin antibodies are known risk factors for thrombosis. Circulating phospholipids are required to form a normal clot. The presence of auto-antibodies to phospholipids increases their thrombogenicity. In extreme forms, the presence of antiphospholipid antibodies can lead to sudden, life-threatening end organ damage through synchronous thrombus formation in vital organs, frequently with a consumptive coagulopathy and bleeding. When this occurs, it is known as the catastrophic anti-phospholipid antibody syndrome (APLAS). The treatment of catastrophic APLAS includes anticoagulants, plasma exchange, and immune suppression with corticosteroids or other immune suppressants.

For the patient in the vignette, the best combination of next steps would include testing for antiphospholipid antibodies, pulse oximetry, and anticoagulation with enoxaparin. A chest radiograph would not be warranted, as a chest computed tomography would be far more sensitive than a chest radiograph to assess for a pulmonary embolus. Anticoagulation with aspirin alone would be inadequate therapy for a DVT. There would be no reason to image the patient's brain in this scenario. The only risk for a central nervous system embolus would be if there was a large patent foramen ovale or other right to left intracardiac shunt. Given the rarity of that occurrence and the absence of any central nervous system symptoms, a brain magnetic resonance image would not be warranted.

PREP Pearls
- Patients who present with a new thrombosis should have a workup for heritable thrombophilia including factor V Leiden, prothrombin gene mutation, protein C and S levels, and antithrombin III levels.
- Patients who present with a new thrombosis should have a workup for acquired thrombophilia including an evaluation for antiphospholipid antibodies (lupus anticoagulant, anticardiolipin antibodies, and anti-β2-glycoprotein antibodies).
- Anticoagulation with enoxaparin is easier and requires less monitoring than warfarin.

ABP Content Specifications(s)
- Plan the appropriate evaluation and management of an acquired bleeding or thrombotic disorder

Suggested Readings
- Hunt BJ. Pediatric antiphospholipid antibodies and antiphospholipid syndrome. Semin Thromb Hemost. 2008;34(3):274-281. DOI: http://dx.doi.org/10.1055/s-0028-1082271.
- Roth M, Manwani D. Thrombotic disorders. Pediatr Rev. 2011;32(1):41-43. DOI: http://dx.doi.org/10.1542/pir.32-1-41.
- Yang JY, Chan AK. Pediatric thrombophilia. Pediatr Clin North Am. 2013;60(6):1443-1462. DOI:http://dx.doi.org/10.1016/j.pcl.2013.09.004.

Question 251

You are seeing a 10-year-old girl who has been performing repetitive cleaning behaviors over the past 3 to 4 months. She repeatedly cleans and organizes her bedroom and frequently washes her hands. She is experiencing dysfunction because of these behaviors, which interfere with her ability to fall asleep at night and get to school. The frequent washing is causing red cracked skin on her hands. The mother expresses significant frustration about dealing with these behaviors, and says that their home life is now strained because of it.

Of the following, the BEST next step in care is

A.   obtain anti-streptolysin O titer

B.   prescribe clomipramine

C.   prescribe clonazepam

D.   refer for cognitive behavioral therapy

E.   refer for parent/child interaction therapy

Question 251   **Preferred Response: D**

The girl in the vignette is exhibiting classic signs of obsessive compulsive disorder (OCD). Cleaning and organizing rituals are the most common symptoms in children. This may include hand washing to the point of causing cracked skin, as in the patient in this vignette. Treatment for OCD in children is highly effective and includes a combination of cognitive behavior therapy (CBT) and selective serotonin reuptake inhibitor (SSRI) treatment. For those with less severe symptoms, or for those children and families who are hesitant to initiate use of a SSRI, starting treatment with CBT alone is quite appropriate. Steps used in CBT include teaching the child and family about the disorder, helping the child learn to recognize obsessive compulsive thoughts as urges they can choose to follow or not, and then practicing making the choice not to give in to the OCD urges and selectively ignoring these thoughts. Through practice, the child will learn to tolerate their biologically triggered OCD thoughts, to "talk back" to their OCD thoughts, and remove the OCD's control over their behaviors. While OCD often occurs in children who have a family history of anxiety disorders, OCD is not seen as a "learned" anxiety that comes from living in an anxious household, but rather has a highly biologically driven etiology.

An anti-streptolysin O (ASO) titer might be included within an OCD workup that is suspecting a pediatric autoimmune neuropsychiatric disorder associated with Streptococcus (PANDAS) or streptococcal immune reaction condition having caused the symptoms, but it would not be the best next step in care for several reasons. The first is that the ASO titer only indicates that the child may have experienced a streptococcal infection in the preceding 6 months, which is an extremely common circumstance among healthy children and not indicative of any current disorder. Secondly, associating an autoimmune etiology for a child's OCD does not currently open up any other valid treatment options; clinical care is essentially the same regardless of whether or not the OCD may have been triggered in an autoimmune fashion. A third reason is that this child has no clinical history of acute onset of OCD symptoms in the month immediately following a streptococcal infection, which would be the expected time course of an autoimmune-triggered OCD syndrome.

Prescribing the tricyclic antidepressant clomipramine would be an appropriate treatment for OCD that has proven to be resistant to other treatments. For instance, after there have been 2 SSRI treatment failures and a failure of CBT, a clomipramine trial would be appropriate. While clomipramine is sometimes more effective on OCD symptoms than the SSRIs, because it has significantly more medical adverse effect risks, it is not considered a first line treatment option. Clonazepam is sometimes used for anxiety treatment that has not responded to an SSRI or other nonaddictive, non-tolerance—generating therapies because it can be administered with once daily dosing. However, benzodiazepines are neither a first or second line medication treatment option for OCD, as there are many other evidence-based treatment options. Clonazepam is most appropriately used by specialists in behavioral pediatrics who have first tried other treatment approaches.

Parent-child interaction therapy is an evidence-based treatment for young child behavior management problems. It is not a management strategy for OCD. An essential part of CBT for

OCD in children is to help parents understand what is happening with their child, and coach them on how best to help their child as they struggle to manage their obsessive impulses.

PREP Pearls
- Obsessive compulsive disorder (OCD) is a very biological rather than social environment generated disorder.
- Obsessive compulsive disorder treatment with cognitive behavior therapy (CBT) and selective serotonin reuptake inhibitor (SSRI) treatment is highly effective.
- While there are rare cases in which OCD may have been triggered by an autoimmune reaction to infection (such asStreptococcus), the best clinical treatment for the OCD is still the use of SSRIs and CBT.

ABP Content Specifications(s)
- Recognize the various environmental and biological contributors to the development of obsessive-compulsive disorder
- Recognize the clinical findings associated with obsessive-compulsive disorder in patients of various ages, and manage appropriately

Suggested Readings
- Franklin ME, Sapyta J, Freeman JB, et al. Cognitive behavior therapy augmentation of pharmacotherapy in pediatric obsessive-compulsive disorder: the Pediatric OCD Treatment Study II (POTS II) randomized controlled trial. JAMA. 2011;306(11):1224-1232. DOI: http://dx.doi.org/10.1001/jama.2011.1344.
- Stewart SE, Hezel D, Stachon AC. Assessment and medication management of paediatric obsessive-compulsive disorder.Drugs. 2012;72(7):881-893. DOI: http://dx.doi.org/10.2165/11632860-000000000-00000.

Question 252

A 4-year-old boy was noted to have a heart murmur when seen for a febrile illness by your partner 2 weeks ago. The murmur was described as a 3/6 systolic murmur. He is here today for follow-up and is well. He has no history of syncope, palpitations, or exercise intolerance. The family history is negative for any individuals with sudden death, cardiomyopathy, or early coronary artery disease.

Physical examination shows a heart rate of 76 beats/min, respiratory rate of 18 breaths/min, and blood pressure of 90/65 mm Hg. The lungs were clear to auscultation, the cardiac examination is significant for a regular rate and rhythm, and S1 and S2are normal. There is a 2/6 systolic murmur at the left lower sternal border. There is no radiation to the neck or axilla. There is no rub or gallop. On abdominal examination, there is no hepatosplenomegaly. His femoral pulses are 2+ and normal.

Of the following, the additional finding that would be MOST consistent with an innocent murmur in this child is

A.   low pitched diastolic murmur at the right mid sternal border

B.   mid systolic murmur at the left third intercostal space

C.   soft continuous murmur heard at the left mid scapula

D.   systolic ejection murmur at the right upper sternal border

E.   vibratory systolic murmur at the apex

Question 252   **Preferred Response: E**

The Still's murmur (C252) is a very common innocent murmur in childhood. It is heard best at the left lower sternal border, toward the apex, becomes softer when the child is asked to sit up from the supine position, has a vibratory, somewhat musical quality, and does not radiate into either the pulmonary (left upper sternal border) or the aortic (right upper sternal border) area. The child in this vignette has a Still's murmur.

Innocent murmurs are common in childhood. There are 4 distinct types of innocent murmurs in children:
- Still's murmur (vibratory murmur heard at the left mid sternal border and apex)
- Pulmonary flow murmur in infancy (heard in the left upper sternal border and into the axilla)
- Pulmonary flow murmur (heard in the left upper sternal border in later childhood)
- Venous hum (a holosystolic murmur heard in the supraclavicular area on either the right or left side, which may change or become inaudible when the child's head is turned to the left or right)

Other than the venous hum, innocent murmurs are systolic, less than 3/6 in intensity, and do not obscure S1. If a murmur does obscure S1, it is occurring when no blood flow should be audible, as this is the phase of isovolumic contraction when the atrioventricular valves have closed and the aortic and pulmonic valves have not yet opened.

Differentiation of an innocent murmur from a pathologic murmur is usually done by history and physical examination. If there are symptoms of decreased cardiac output, cyanosis, palpitations, or chest pain, there is a higher likelihood of cardiac pathology.

The other murmurs offered as response choices have qualities that are not innocent. Diastolic murmurs suggest a regurgitant lesion, such as aortic and pulmonary insufficiency. A systolic murmur in the mitral position would suggest mitral regurgitation. A continuous murmur heard over the scapula suggests a patent ductus arteriosus. A systolic murmur in the right upper sternal border (aortic position) would suggest aortic stenosis.

The low pitched diastolic murmur at the right mid sternal border is consistent with tricuspid inflow and, although not usually audible, might be heard with a large atrial septal defect. A mid systolic murmur in the third left intercostal space suggests mitral regurgitation. A murmur in the right upper sternal border is never considered normal, as this is the aortic position. A patient with hyperdynamic circulation, as seen with anemia or infection, can have a murmur in the left upper sternal borders that is louder than the usual innocent pulmonary flow murmur. A vibratory murmur, which is an innocent finding that is also musical and is heard at the left lower sternal border and toward the apex, is consistent with a Still's murmur.

PREP Pearls
- Diastolic murmurs are not innocent.
- Murmurs that obscure S1 or are greater than 3/6 in intensity are not innocent.
- Innocent murmurs are common in children. There are 4 distinct types of innocent murmurs in children:
  o Still's murmur (vibratory murmur heard at the left mid sternal border and apex)
  o Pulmonary flow murmur in infancy (heard in the left upper sternal border and into the axilla) oPulmonary flow murmur (heard in the left upper sternal border in later childhood)
  o Venous hum (a holosystolic murmur heard in the supraclavicular area on either the right or left side, which may change or become inaudible when the child's head is turned to the left or right)

ABP Content Specifications(s)
- Plan the appropriate evaluation of an innocent murmur, and manage appropriately

Suggested Readings
- Etoom Y, Ratnapalua S. Evaluation of children with heart murmurs. Clin Pediatr (Phila). 2014;53(2):111-117. DOI:http://dx.doi.org/10.1177/0009922813488653.
- Frank JE, Jacobe KM. Evaluation and management of heart murmurs in children. Am Fam Physician. 2011;84(7):793-800.
- Menashe, V. Heart Murmurs. Pediatr Rev 2007; 28 (4):e19-e22. DOI: http://dx.doi.org/10.1542/pir.28-4-e19.
- Park MK. Physical examination: auscultation. Park's Pediatric Cardiology for Practitioners. 8th ed. Philadelphia, PA: Saunders Elsevier; 2014:9-40.
- Punnoose AR, Burke AE, Golub RM. Innocent (harmless) heart murmurs in children. JAMA. 2012;308(3):305. DOI:http://dx.doi.org/10.1001/jama.2012.6223.
- Teitel DF, Cassidy SC, Fineman JR. History and physical examination. In: Allen HD, Driscoll DJ, Shaddy RE, Feltes TF, eds.Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. 8th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2013:69-81.

Question 253

A 13-year-old adolescent boy presents to your office with complaint of joint pain that started 3 months ago. He complains of lower back, left hip, right knee, and left ankle pain that is worse in the morning and improves with exercise. Rest makes his back pain worse. On physical examination, the patient has tenderness over the right tibial tuberosity and the left calcaneus. There is swelling at the insertion of the Achilles tendon. He has pain with passive movement and decreased range of motion of his left hip.

Of the following, the additional feature MOST supportive of your suspected diagnosis is a

A.   decreased lumbar flexion (positive Schober test)

B.   first-degree relative that is human leukocyte antigen-B27 positive

C.   first-degree relative with psoriasis

D.   history of illness 2 weeks before onset of symptoms

E.   paraspinal tenderness

Question 253   **Preferred Response: A**

The 13-year-old boy in this vignette, with a complaint of inflammatory back pain that is worse in the morning and worsens with rest but improves with exercise, most likely has ankylosing spondylitis (AS). Among the response choices, decreased lumbar flexion (a positive Schober test) best supports this diagnosis. Inflammatory back pain in AS usually has an insidious onset. Patients may also have alternating buttock pain and nighttime back pain that occurs in the second half of the night. The clinical criteria used for the diagnosis of AS include low back pain for 3 months that is improved by exercise and not relieved by rest, limitation of lumbar spine motion in the sagittal and frontal planes, and decreased chest expansion relative to normal values for age and sex. Diagnostic criteria for AS require the child to meet at least 1 clinical criterion and have radiologic evidence of sacroiliitis (grade 3-4 if unilateral or grade 2-4 if bilateral).

Spondyloarthritis is a classification of several different arthrides that can affect the spine and sometimes the sacroiliac joints. Spinal involvement is often not present at disease onset, but develops over time. Ten percent to 20% of patients with spondyloarthritis develop symptoms in childhood, which is known as juvenile spondyloarthritis (JS). Clinical features of spondyloarthritis include enthesitis, peripheral arthritis, axial arthritis, uveitis, gastrointestinal inflammation, and cardiac manifestations. In spondyloarthritis, the peripheral arthritis is usually an asymmetric arthritis of the large joints of the lower extremity. Hip involvement is more frequent in the pediatric population. Tarsitis is more common in spondyloarthritis than in other types of arthritis. It is difficult to predict which patients will progress from spondyloarthritis to AS.

Enthesitis is inflammation of the entheses, the bone insertion sites for ligaments, tendons, fascia, and joint capsules. Enthesitis causes pain, swelling, and tenderness and presents more commonly in pediatric disease. Enthesitis can also occur with various types of arthritis, as well as overuse or traction injuries such as Osgood-Schlatter syndrome. Enthesitis-related arthritis (ERA) is defined as arthritis or enthesitis with sacroiliac joint tenderness or inflammatory spinal pain and 2 of the following:
- the presence of HLA-B27
- acute anterior uveitis
- onset in a boy older than 6 years
- family history of first-degree relative with AS, ERA, sacroiliitis with inflammatory bowel disease, reactive arthritis, or acute anterior uveitis

Axial arthritis is associated with inflammatory back pain that has an insidious onset, occurs in the second half of the night, improves with exercise but not with rest, and is associated with morning stiffness. There can be buttock pain that alternates, as in spondyloarthritis. Inflammatory back pain is not as common in the pediatric population as it is in the adult population. Sacroiliitis is often the first sign of axial involvement in AS. Silent sacroiliitis is found in 21% of children with spondyloarthritis. Sacroiliitis can usually be seen on plain radiography. However, in cases in which plain radiography is normal and spondyloarthritis is suspected, a magnetic resonance imaging scan of the pelvis with short T1 inversion recovery (STIR) images is best to make the diagnosis.

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Uveitis is the most common extra-articular manifestation of spondyloarthritis, and is more common if patients are HLA-B27 positive. Approximately 33% of patients with AS develop uveitis.

Gastrointestinal inflammation is associated with spondyloarthritis. Approximately 30% of patients with inflammatory bowel disease have peripheral and axial arthritis. Of patients with AS, 60% have subclinical gastrointestinal inflammation, and 6.5% will develop inflammatory bowel disease.

Cardiovascular manifestations of spondyloarthritis can include atherosclerosis, an increased risk for ischemic heart disease and myocardial infarction, aortitis, aortic regurgitation, and aortic and mitral valve thickening. Cardiac manifestations are usually mild in juvenile AS, but risks increase with age. Therefore, these patients should be evaluated for and counseled regarding cardiac risk factors.

Elevated C-reactive protein level can be associated with AS. Other laboratory studies are of limited value in diagnosing AS, but may be useful to rule out other diseases.

A positive HLA-B27 is associated with AS, but is not diagnostic. Six percent to 10% of the white population is HLA-B27 positive, limiting its diagnostic usefulness. Only 20% of people who are HLA-B27 positive and have a family history of AS will go on to develop AS. Although a family history of AS in an HLA-B27–positive patient is a diagnostic factor, HLA-B27 positivity in the absence of a family history of AS is not. A patient with inflammatory back pain with peripheral enthesitis and a first-degree relative with psoriasis would meet criteria for psoriatic arthritis, not AS. A history of illness 2 weeks before the onset of symptoms would more likely be associated with a postinfectious or reactive arthritis. Paraspinal tenderness is more likely to be caused by musculoskeletal pain than muscle strain.

PREP Pearls
- Anklylosing spondylitis is a clinical diagnosis that includes enthesitis, peripheral arthritis, axial arthritis, uveitis, gastrointestinal inflammation, and cardiac manifestations.
- Sacroiliitis is often the first sign of axial involvement in ankylosing spondylitis.
- HLA-B27 is associated with increased risk of uveitis in ankylosing spondylitis.

ABP Content Specifications(s)
- Recognize the clinical and laboratory findings associated with ankylosing spondylitis

Suggested Readings
- Ramanathan A, Srinivasalu H, Colbert RA. Update on juvenile spondyloarthritis. Rheum Dis Clin North Am. 2013;39:767-788. DOI: http://dx.doi.org/10.1016/j.rdc.2013.06.002.
- Tse SML, Laxer RM. New advances in juvenile spondyloarthritis. Nat Rev Rheumatol. 2012;8:269-279. DOI:http://dx.doi.org/10.1038/nrrheum.2012.37.

Question 254

A 15-year-old adolescent presents to your office for evaluation of a neck injury that occurred earlier that day. During football practice, the left side of his neck was struck by another player's leg during a tackling drill. He immediately developed left-sided neck pain, accompanied by pain and weakness in his left arm. You review the recommended evaluation and treatment for this injury with the boy and his family.

Of the following, radiographic studies are MOST strongly warranted in the presence of

A.   bilateral symptoms

B.   an injury that applied traction to the affected side of the neck

C.   tenderness at the location where the boy was hit

D.   weakness in the biceps muscles

E.   weakness that lasts more than 2 hours

Question 254   **Preferred Response: A**

The boy in the vignette has sustained a mild brachial plexus injury, commonly referred to as aburner or a stinger. Individuals with this injury experience unilateral upper extremity symptoms consistent with the innervation pattern for nerves of the upper brachial plexus. Bilateral symptoms or lower extremity symptoms suggest cervical spine injury and should prompt cervical-spine immobilization and radiographic studies, including cervical spine films, and if these are negative, consideration of magnetic resonance imaging.

Stinger injuries are common in contact sports, especially football. Typically the diagnosis can be made based on clinical symptoms and physical examination findings. Recurrence is common. There are 3 mechanisms for these mild brachial plexus injuries: direct force applied to the brachial plexus, as happened to the boy in the vignette; forced lateral bending of the neck causing compression of the cervical nerve roots, leading to ipsilateral symptoms; or traction applied to the nerve roots, causing contralateral symptoms. Athletes typically report pain radiating down the arm. Weakness and paresthesias in the arm and shoulder are also common. Symptoms generally last less than 24 hours.

On physical examination, affected individuals often have weakness of the deltoid, biceps, and rotator cuff muscles. Sensory changes may also be seen. Radiographic studies may be indicated to rule out other injuries, such as a cervical spine fracture or spinal cord injury, but are not needed to diagnose a stinger.

Traction is a common mechanism for mild brachial plexus injuries, so the injury mechanism for the boy in the vignette should not prompt concern. Tenderness at the site of impact is expected for most soft tissue injuries. Biceps weakness is a common clinical sign of a stinger injury. The duration of the boy's symptoms does not suggest a severe injury.

PREP Pearls
- Mild brachial plexus injuries, commonly referred to as burners or stingers, are common in contact sport athletes.
- Bilateral or lower extremity symptoms suggest cervical spine injury and should prompt cervical spine immobilization and radiographic studies.

ABP Content Specifications(s)
- Plan the appropriate management of an acute sports-related neck injury
- Identify the sports in which cervical injury most commonly occurs

Suggested Readings
- Sarwark JF, LaBella C. Pediatric Orthopaedics and Sports Injuries: A Quick Reference Guide. 2nd ed.Elk Grove Village, IL: American Academy of Pediatrics; 2014: 795 pp.
- Standaert CJ, Herring SA. Expert opinion and controversies in musculoskeletal and sports medicine: stingers. Arch Phys Med Rehabil. 2009;90(3):402-406.

Question 255

An 8-year-old boy presents to the emergency department with a 3-day history of right facial swelling and poor oral intake. On physical examination, his temperature is 39°C and vital signs are normal for age. His right cheek is markedly swollen and firm, with mild erythema and moderate tenderness to palpation. Examination of the oropharynx reveals dental caries, malodorous breath, and right mandibular gingival swelling with erythema. There is purulent fluid observed near the base of the first molar and palpation of the tooth elicits pain. The remainder of the physical examination is unremarkable.

Of the following, the BEST antimicrobial therapy for this patient is

A.   ampicillin-sulbactam

B.   cefotaxime

C.   metronidazole

D.   tetracycline

E.   vancomycin

Question 255   **Preferred Response: A**

The boy in the vignette has a pyogenic odontogenic infection presenting with fever and a lower molar periapical abscess with spread of infection to adjacent tissues. Odontogenic infections typically are polymicrobial, with oral facultative streptococci and anaerobic bacteria commonly involved (Item C255). Anaerobic organisms, particularly anaerobic gram-negative rods, predominate in more severe disease, especially that which spreads beyond the teeth and alveolar processes. In addition to surgical drainage of the abscessed tooth, the most appropriate therapy for this patient is ampicillin-sulbactam, because it has activity against β-lactamase–producing anaerobic and aerobic organisms.

Item C255. Anaerobic Bacteria Commonly Found in Odontogenic Infections.

Strict anaerobes
Bacteroides
Fusobacterium
Peptostreptococcus
Porphyromonas
Prevotella
Facultative anaerobes
Streptococcus anginosus group
Viridans streptococci

Courtesy of D. Palazzi

Most orofacial infections arise from periapical, periodontal, or pericoronal dental infection that spreads by the path of least resistance into surrounding (potential) spaces and tissues. Spread of infection into the fascial spaces can result in significant facial swelling and fever, such as that described in the vignette. Odontogenic infections in children usually are localized; significant spread can lead to respiratory compromise and life-threatening complications.

Serious odontogenic infection should be treated with intravenous antibiotics and, if indicated, surgical drainage. Given the polymicrobial nature of infection, with anaerobic organisms playing an important role in severe disease, broad-spectrum antimicrobial therapy with activity against β-lactamase–producing anaerobic and aerobic organisms is optimal.

Cephalosporins, such as cefotaxime, are second-line agents for the treatment of odontogenic infection. They effectively treat β-lactamase–producing oral streptococci and gram-negative pathogens such as Eikenella, but must be combined with metronidazole for anaerobic coverage. Metronidazole has excellent activity against anaerobic pathogens but no activity against aerobic gram-positive and gram-negative organisms. Tetracycline is not recommended for children

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younger than 9 years because it permanently stains teeth by becoming incorporated into newly formed dentin. In addition, there is increasing resistance to tetracycline among gram-positive and gram-negative organisms. Although vancomycin has broad-spectrum activity against aerobic gram-positive organisms (eg, methicillin-resistant Staphylococcus aureus [MRSA], streptococci), it is not necessary in the treatment of odontogenic infection as more narrow-spectrum agents will suffice. Mild odontogenic infection may be treated with oral penicillin, amoxicillin, or clindamycin, but as resistance to these agents is increasing, patients must be monitored closely for clinical improvement.

PREP Pearls
•   Odontogenic infections typically are polymicrobial, with oral facultative streptococci and anaerobic bacteria commonly involved.
•   Broad-spectrum antimicrobial therapy with activity against beta-lactamase–producing anaerobic and aerobic organisms is optimal for the treatment of odontogenic infections.
•   Spread of odontogenic infection into the fascial spaces can result in significant facial swelling and fever; significant spread can lead to respiratory compromise and life-threatening complications.

ABP Content Specifications(s)
•   Understand the association between an anaerobic infection and dental and periodontal disease

Suggested Readings
•   Garcia C, McCracken GH Jr. Antibacterial therapeutic agents. In: Cherry J, Demmler-Harrison GJ, Kaplan SL, Steinbach WJ, Hotez P, eds. Feigin & Cherry's Textbook of Pediatric Infectious Diseases. 7th ed. Philadelphia, PA: Elsevier Saunders; 2014:3182-3241.
•   Simos C, Gonzalez BE. Infections of the oral cavity. In: Cherry J, Demmler-Harrison GJ, Kaplan SL, Steinbach WJ, Hotez P, eds. Feigin & Cherry's Textbook of Pediatric Infectious Diseases. 7th ed. Philadelphia, PA: Elsevier Saunders; 2014:140-155

Question 256

A 3-week-old full-term newborn, with fever and decreased oral intake over the past 24 hours, is admitted to a teaching hospital where you are the supervising pediatrician. Her plan of care includes obtaining blood, urine, and cerebrospinal fluid cultures, and initiating empiric intravenous antibiotic therapy. As you discuss this plan with the admitting resident, you ask how she will manage the newborn's procedure-related pain. The resident replies that she generally does not use any pain management modality when performing procedures in newborns, as they are much less affected by these procedures than older children.

Of the following, the MOST accurate statement about pain management for patients of this age is that

A.   a single modality should be used to manage procedure-related pain

B.   their level of procedure-related pain is overestimated by parents and health care providers

C.   they do not have the cognitive ability to remember painful experiences

D.   they typically display a less pronounced physiologic response to pain than older children

E.   undergoing painful procedures may result in long-term changes in pain response

Question 256   **Preferred Response: E**

The infant in the vignette will undergo a series of painful procedures (venipuncture, intravenous catheter placement, bladder catheterization, and lumbar puncture) as part of an evaluation for potential serious bacterial infection. The most accurate statement related to pain management in infants is that undergoing painful procedures in infancy may result in long-term changes in pain response.

It is imperative for all pediatric providers to understand the effects of patients' developmental stage on their response to pain and to apply this knowledge in implementing appropriate pain management plans for children of various ages. The field of knowledge related to pain response in children and pediatric pain management techniques has advanced significantly over the past 2 to 3 decades. Regarding newborns specifically, an emerging body of literature highlights the critical importance of appropriate pain management. Historically, analgesics were rarely administered to newborns, based on the theory that they experience less pain because of the immaturity of their nervous systems. Until the late 1980s, in fact, newborns often underwent certain surgical procedures without anesthesia.

Recent studies suggest that newborns may actually have an increased sensitivity to pain, which may be attributable to the fact that although their ascending nerve pathways can transmit painful stimuli to the brain, descending inhibitory pathways are not yet established. Data demonstrate that neonates display a more pronounced physiologic response to pain and require higher serum concentrations of analgesics to modulate pain compared with older children. Studies also indicate that repeated exposure to painful stimuli in the neonatal period can increase sensitivity to subsequent painful stimuli as well as routine handling. In addition, infants who experience painful procedures may develop altered responses to future painful episodes, even those that occur well beyond infancy. Pain may even be a contributing factor in the occurrence of intraventricular hemorrhage in preterm infants.

A number of general principles should be applied to managing pain appropriately in children:
- Pain prevention: Pain should be managed prophylactically, if it can be anticipated.
- Assessment: Pain must be adequately assessed for it to be appropriately managed.
- Multimodal approach: Pain should be addressed using various strategies.
- Parental involvement: Parental input and involvement should be incorporated into pain management plans.
- "Non-noxious" routes: Analgesia should be administered through the least painful routes possible.
- Procedure-related pain control: Pain associated with diagnostic and therapeutic procedures should be managed aggressively to avoid increased anxiety and pain related to subsequent procedures and encounters with health care providers.

A single modality approach to the management of procedure-related pain is not the best strategy for infants or older children. A multimodal approach incorporating environmental, behavioral, and pharmacologic interventions is much more effective.

Pain levels related to procedures are generally underestimated rather than overestimated, largely because of the inability of caretakers to reliably assess pain in infants.

Current literature contradicts the theory that infants lack the cognitive ability to remember painful experiences. In fact, recent studies indicate that experiencing pain during infancy may alter a child's responses to future painful episodes. Studies also indicate that compared with older children, newborns may display a greater physiologic response to pain.

PREP Pearls
- Inadequate pain management in infants can lead to significant short- and long-term sequelae.
- Pain should be managed prophylactically whenever possible.
- A multimodal approach to pain management incorporating environmental, behavioral, and pharmacologic interventions is the recommended approach for children of all ages.
- Parental input and involvement should be incorporated into the pain management plans of all children.

ABP Content Specifications(s)
- Understand the effects of a patient's developmental stage on tolerating and dealing with pain

Suggested Readings
- Anand KJ, International Evidence-Based Group for Neonatal Pain. Consensus statement for the prevention and management of pain in the newborn. Arch Pediatr Adolesc Med. 2001;155:173-180. DOI:http://dx.doi.org/10.1001/archpedi.155.2.173.
- Gray L, Garza E, Zageris D, Heilman KJ, Porges SW. Sucrose and warmth for analgesia in healthy newborns: an RCT .Pediatrics. 2015;135:e607-e614. DOI: http://dx.doi.org/10.1542/peds.2014-1073.
- Zempskey WT, Schecter NL. What's new in the management of pain in young children. Pediatr Rev. 2003;24:337-348. DOI: http://dx.doi.org/10.1542/pir.24-10-337.

Question 257

A 33-week gestation newborn who was delivered at home is brought to the emergency department via ambulance. The pregnancy was complicated by limited prenatal care and maternal substance abuse. The mother was unable to describe the events surrounding the labor and delivery. Your quick assessment reveals a heart rate of 90 beats/min and a respiratory rate of 50 breaths/min. Physical examination demonstrates a jittery newborn with decreased perfusion, moderate grunting, and cool extremities.

Of the following, the MOST likely cause of the clinical findings seen in this newborn is

A.   drug withdrawal

B.   hypothermia

C.   intraventricular hemorrhage

D.   sepsis

E.   surfactant deficiency

Question 257   **Preferred Response: B**

The history and clinical findings seen in the newborn in the vignette suggest cold stress from hypothermia. A newborn with cold stress will develop tachypnea and peripheral vasoconstriction. The peripheral vasoconstriction leads to decreased perfusion, poor color, and cool extremities. Sinus bradycardia may also develop as the hypothermia worsens. Biochemical findings associated with cold stress include metabolic acidosis and hypoglycemia.

All newborns are at risk for convective, evaporative, and conductive heat loss at the time of delivery. This heat loss is exaggerated in newborns born prematurely because of decreased epidermal thickness, decreased brown fat, and increased surface area. Optimizing the temperature in the delivery room, gently drying the neonate, and replacing any wet towels and blankets with dry warm blankets can minimize heat loss in the full-term and late-preterm newborn. Unless clinically unstable, full-term and late-preterm newborns should be placed on the mother's chest shortly after birth for both bonding and enhanced thermoregulation.

A radiant warmer should be available in the delivery room for management of newborns who are unstable or premature. This will provide the resuscitation team ready access to the newborn while minimizing heat loss. The amount of heat delivered by the radiant warmer must be carefully monitored. Many radiant warmers allow a temperature probe to be placed on the newborn's skin that allows auto regulation of the overbed heat output. If auto regulation is unavailable, and the radiant warmer temperature is manually set to deliver maximum heat, the newborn's temperature should be monitored frequently to avoid hyperthermia.

The clinical signs and symptoms of cold stress can mimic those of neonatal sepsis or drug withdrawal, though sinus bradycardia is generally not present in those conditions. Severe intraventricular hemorrhage (IVH) can also have similar findings as cold stress, but most IVH cases occur in premature infants born at less than 32 weeks of gestation. Cold stress can lead to surfactant deficiency in affected premature infants, but the mild tachypnea without other signs of respiratory distress in the newborn in the vignette makes surfactant deficiency unlikely.

PREP Pearls
- Newborns are at risk for convective, evaporative, and conductive heat loss at the time of delivery.
- A newborn with cold stress presents with tachypnea and peripheral vasoconstriction. The peripheral vasoconstriction leads to decreased perfusion, poor color, and cool extremities.
- Biochemical findings associated with cold stress include metabolic acidosis and hypoglycemia.
- Unless clinically unstable, full-term and late-preterm newborns should be placed on the mother's chest and gently dried shortly after birth for both bonding and enhanced thermoregulation.

ABP Content Specifications(s)
- Recognize the hazards and benefits associated with the use of radiant warmers for neonates
- Recognize the signs and symptoms of cold stress in a newborn infant, and manage appropriately

Suggested Readings
- American Academy of Pediatrics. Textbook of Neonatal Resuscitation. 6th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2011:329 pp.
- Laptook A, Jackson GL. Cold stress and hypoglycemia in the late preterm ("near-term") infant: impact on nursery of admission. Semin Perinatol. 2006;30:24-27. DOI: http://dx.doi.org/10.1053/j.semperi.2006.01.014.
- Sedin G. To avoid heat loss in very preterm infants. J Pediatr. 2004; 145: 720-722. DOI:http://dx.doi.org/10.1016/j.jpeds.2004.08.065.

Question 258

You are seeing a 6-year-old boy in follow-up from a visit 1 month ago for newly noted intermittent exotropia. He was referred to an ophthalmologist, who found an unremarkable examination and recommended monitoring. The boy's mother notices his eye wandering several times a day now. In addition, she feels he looks excessively tired in the afternoons and she is putting him down for naps, but he says he is not tired at all. He goes to sleep easily at night, sleeps through the night, and wakes up without difficulty. He does not snore. He has not had fevers, headaches, or difficulty with walking or running. His physical examination, including neurological examination, is unremarkable.

Of the following, the correct diagnosis will MOST likely be identified by

A.   magnetic resonance imaging of the brain

B.   measuring acetylcholine receptor antibodies

C.   measuring Epstein-Barr virus antibodies

D.   polysomnography

E.   trial of amphetamine therapy

Question 258   **Preferred Response: B**

The boy in the vignette has myasthenia gravis. His symptoms of intermittent ocular misalignment and ptosis are highly suggestive of this disease. Other symptoms can include intermittent dysphagia or head drop. Limb weakness is rarely a presenting symptom. In the classic presentation of myasthenia gravis, the patient has intermittent symptoms that improve with rest. Most patients feel better after rest or a nap, so the clinician has to determine if the improvement is subjective or if the patient has actual weakness that improves. It can also be difficult to differentiate ptosis from drooping eyelids because of fatigue; children with ptosis do not subjectively feel tired. Because the symptoms fluctuate, the examination findings can be normal at times, as in the boy in the vignette.

The diagnosis of myasthenia gravis is confirmed with serum myasthenia gravis antibody titers. In hospitalized patients, edrophonium can be given intravenously to help confirm the diagnosis, but is not used as commonly as it was in the past. Edrophonium is a short-acting acetylcholinesterase inhibitor which, when given intravenously, will cause a patient with myasthenia gravis to have a sudden improvement in symptoms. The patient may also experience sudden salivation, and possibly defecation or bradycardia because of its parasympathomimetic effects. Another method used to diagnose myasthenia gravis is a nerve conduction study with repetitive stimulation. Repetitive stimulation is a specialized test that confirms the presence of a defect in signal transmission at the neuromuscular junction. This is almost always caused by myasthenia gravis, but the diagnosis must be confirmed with antibody testing. Initial treatment of myasthenia gravis is usually with oral pyridostigmine, a longer-acting acetylcholinesterase inhibitor than edrophonium. Chronic immunosuppression is sometimes necessary.

The boy in the vignette appeared sleepy due to ptosis that worsened in the afternoon, not fatigue, so evaluation for causes of chronic fatigue, such as Epstein-Barr virus antibody titers or polysomnography, are not needed. A trial of stimulants is also not indicated for ptosis. Magnetic resonance imaging of the brain is not helpful in diagnosing disorders of the neuromuscular junction such as myasthenia gravis.

PREP Pearls
- Drooping eyelids can be the result of fatigue or ptosis in patients with myasthenia gravis.
- New onset of intermittent ocular misalignment and intermittent ptosis is suggestive of myasthenia gravis.

ABP Content Specifications(s)
- Recognize the clinical findings associated with myasthenia gravis, and manage appropriately
- Plan the appropriate diagnostic evaluation of myasthenia gravis

Suggested Readings
- Index of suspicion. Pediatr Rev. 2000;21(9):315-320. doi: http://dx.doi.org/10.1542/pir.21-9-315.

- Patterson MC, Gomez MR. Muscle disease in children: a practical approach. Pediatr Rev. 1990;12(3):73-82. DOI:http://dx.doi.org/10.1542/pir.12-3-73.

Question 259

A 12-year-old boy with spastic quadriplegic cerebral palsy, seizure disorder, osteopenia, and intellectual disability sustains a complex fracture of his left femur after an accidental fall. He is subsequently wheelchair bound for 2 months. He is brought to your office for evaluation of new onset anorexia, nausea, abdominal pain, and polyuria.

Current medications include:
- Valproic acid, 500 mg orally twice daily
- Methylphenidate hydrochloride, 36 mg orally once daily
- Vitamin D3, 1,000 IU orally once daily

Of the following, the laboratory test MOST likely to have abnormal results is

A.   platelet count

B.   serum alanine aminotransferase

C.   serum ionized calcium

D.   serum potassium

E.   urine culture

Question 259   **Preferred Response: C**

The boy in the vignette has signs of hypercalcemia resulting from prolonged immobilization. Hypercalcemia occurs more commonly in growing children and adolescents who are immobilized. It is thought to result from increased release of bone calcium in individuals with high bone turnover. Hypercalcemia causes a urinary concentrating defect by inhibiting vasopressin action. Symptoms include polyuria and polydipsia, headache, nausea, and abdominal pain. Mental status changes can also occur. The diagnosis is confirmed by measurement of serum ionized calcium concentrations. Treatment with saline diuresis and loop diuretics usually is sufficient to return calcium levels to normal. When hypercalcemia is resistant to these therapies, bisphosphonates have been very effective.

Liver disease or hypokalemia might cause similar symptoms to those of the child in the vignette, but in the setting of immobilization, hypercalcemia is more likely. A urinary tract infection also might mimic these findings, but the boy has no fever or dysuria. Low platelet count could present with easy bruising or even internal bleeding, but would not be related to immobilization.
Of note, this patient is also taking vitamin D, which could potentially raise calcium levels further. However, he is only taking 1,000 IU of vitamin D per day. Although the recent Institute of Medicine (IOM) Report on Vitamin D recommends only 600 IU of vitamin D per day in a child of this age, the IOM, the Endocrine Society, and the European Food Safety Authority all note the safe upper limit of vitamin D intake to be up to 4,000 IU per day for children 11 years of age or older. Thus, for the child in the vignette, immobility is likely the major contributor to his hypercalcemia, not his vitamin supplementation.

PREP Pearls
- Hypercalcemia can occur after prolonged periods of immobilization, especially in growing children and adolescents.
- The Institute of Medicine recommends a daily allowance of 600 IU per day of vitamin D for children 1 year of age or older.

ABP Content Specifications(s)
- Recognize the clinical features associated with hypercalcemia, including that occurring as a result of immobilization

Suggested Readings
- Ross AC, Taylor CL, Yaktine AL, et al. Dietary Reference Intakes for Vitamin D and Calcium; Food and Nutrition Board,.Dietary Reference Intakes for Calcium and Vitamin D. Washington, DC: National Academies Press. 2011.http://www.iom.edu/Reports/2010/Dietary-Reference-Intakes-for-calcium-and-vitamin-D.aspx.
- Vogiatzi MG, Jacobson-Dickman E, Deboer MD; Drugs, and Therapeutics Committee of The Pediatric Endocrine Society. Vitamin D supplementation and risk of toxicity in pediatrics: a review of current literature. J Clin Endocrinol Metab. 2014;99(4):1132-1141. DOI: http://dx.doi.org/10.1210/jc.2013-3655.

Question 260
You are evaluating a 15-year-old female adolescent with the chief concern of hoarseness of voice. Two weeks prior to presentation, the patient's father reports a history of acute and near complete absence of voice for 1 to 2 days. Although she can now speak, her voice is still abnormal with lower volume, lower pitch, and a persistent hoarse quality. There has been no stridor, cough, wheezing, or fever. Symptoms of gastroesophageal reflux are denied. Nasal symptoms have been notably absent. The patient is an avid cheerleader.
On physical examination, the patient is well appearing in no acute distress. Her voice is "raspy" and low pitched. There is no stridor or respiratory distress. A head, eyes, ears, nose, and throat examination is unremarkable. Nasal mucosa is normal in appearance. The oropharynx is clear. Her neck is supple without palpable lymphadenopathy. Lungs are clear bilaterally without wheezing, crackles, or differential aeration. Cardiac, abdominal, extremity, and neurologic examinations are unremarkable.

Of the following, the MOST likely etiology of this patient's hoarseness of voice is

A.   allergic rhinitis

B.   gastroesophageal reflux disease

C.   laryngeal papillomatosis

D.   vocal abuse

E.   vocal cord paralysis

Question 260   **Preferred Response: D**

The adolescent depicted in the vignette has dysphonia that is likely the result of vocal abuse. The most common etiology for hoarseness of voice in an otherwise healthy adolescent without other suggestive symptomatology is vocal abuse such as the adolescent in this vignette who is a cheerleader. In a study of 142 children with dysphonia, the most common diagnosis was vocal cord abuse. Related and inflammatory vocal cord nodules may be found in association with vocal cord abuse.

Childhood dysphonia generally can be explained by 1 of several underlying mechanisms including infectious, anatomic, congenital, inflammatory, neoplastic, neurologic, and iatrogenic etiologies.

The larynx descends in anatomic location from birth to adulthood. The normal position of the vocal cords is C4 at birth, with descent to C5-C6 by adolescence and further to C7 in adulthood. Laryngeal growth also occurs in concert with linear growth. Laryngeal growth normally accelerates during infancy and preschool years and then again during adolescence. These changes in the structure and function of the pediatric larynx can lead to dysphonia. Infections (most commonly viral pathogens) are common etiologies for acute laryngitis. In these patients, hoarseness of voice is often experienced in addition to more classic symptoms of an upper respiratory tract infection, such as fever, cough, and rhinitis. Vocal symptoms in this setting typically resolve within 7 to 10 days. If symptoms persist, an evaluation of the airway for alternate etiologies of chronic laryngitis is warranted.

Laryngeal stenosis and laryngeal webs or subglottic cysts may narrow the flow of air sufficient to create a "breathy" voice or one with abnormal frequency or resonance. Gastroesophageal reflux (GER) and laryngopharyngeal reflux have been a recent topic of significant interest in the evaluation of children with nonspecific airway symptoms including chronic hoarseness. In addition, the population of children with defined eosinophilic esophagitis and related gastrointestinal and airway symptoms has grown in recent decades. The extent to which diagnostic evaluations for these disorders are pursued is generally dependent on clinical history. An empiric trial of therapies for GER may be considered if visual findings at bronchoscopy or endoscopy are classic. Although young children may have esophageal evidence of GER disease without relatable symptoms such as heartburn, dyspepsia, vomiting, or dysphagia, older patients, such as the adolescent in this vignette, are more likely to exhibit classic symptomatology.

Spasmodic dysphonia is a neurologic condition in which sudden involuntary muscle spasms cause abnormal vocal cord motion and difficulty in producing normal vocalization. Spasms may occur in adduction, abduction, or in a mixed pattern. Spasms are generally absent with laughter and singing and are often more severe during periods of stress. Vocal cord dysfunction (VCD) is described as an abnormal, but nonspasmodic adduction of the vocal cords. Patients with VCD may experience stridor and a feeling of air limitation or dyspnea that is localized to the neck. A psychogenic component has been proposed for both spasmodic dysphonia and VCD.

The most feared etiology of chronic hoarseness of voice is also one of the rarest. Laryngeal papillomatosis is infrequently encountered, affecting 4.5 out of 100,000 children; it is caused by human papilloma virus 6 and 11. Papillomas grow over time, with a predilection for the vocal cords, larynx, and bronchi; progressive growth and a recurrent pattern may cause respiratory insufficiency and death through airway obstruction. This disease is manageable with surgery and antiviral therapy.

Vocal cord paralysis may cause hoarseness and may occur after cardiac surgery or with injury to the recurrent laryngeal nerve during thyroid or other surgical interventions at the cervical region. Alternate etiologies for acute and chronic hoarseness include intubation trauma, allergic rhinitis, and environmental allergens or irritants. Furthermore, in patients treated for asthma, inhalational therapies such as short acting β-agonists and inhaled corticosteroids may cause hoarseness, presumably from local irritation.

In patients with chronic hoarseness, an evaluation of the upper airway by otolaryngology head and neck surgery (OHNS or ENT) is warranted to evaluate for alternate etiologies. If vocal abuse is confirmed, voice rest and therapy may establish more functional speech patterns in order to limit inflammatory response and restore normalcy of voice.

PREP Pearls
- The most common etiology for acute hoarseness is an uncomplicated viral illness with associated laryngitis.
- The most common etiology for chronic hoarseness of voice is vocal abuse.
- Iatrogenic hoarseness may be induced by inhalational therapies for asthma.
- A laryngoscopic evaluation of the upper airway is warranted in patients with chronic hoarseness of voice.

ABP Content Specifications(s)
- Formulate a differential diagnosis of hoarseness
- Recognizes the various causes of hoarseness
- Plan the appropriate evaluation of hoarseness

Suggested Readings
- Connelly A, Clement WA, Kubba H. Management of dysphonia in children. J Laryngol Otol. 2009;123(6):642-647. DOI:http://dx.doi.org/10.1017/S0022215109004599.
- Kelchner LN, Brehm SB, de Alarcon A, Weinrich B. Update on pediatric voice and airway disorders: assessment and care.Curr Opin Otolaryngol Head Neck Surg. 2012;20(3):160-164. DOI:http://dx.doi.org/10.1097/MOO.0b013e3283530ecb.

Question 261

An 8-year-old boy presents to your office for evaluation of sleep problems. His bedtime is 8:30 PM, but he usually falls asleep while watching television around 11:00 PM. Once he falls asleep, he sleeps soundly during the night. He awakens at 6:30 AM, but is often drowsy and difficult to arouse. He does not snore or mouth breathe, wet the bed, or have a history of behavioral health problems. He is performing adequately in school. On physical examination, he has a body mass index that is at the 79th percentile for his age, 2+ tonsils, and patent nares. The remainder of his examination is unremarkable.

Of the following, the MOST appropriate initial management for this child is to

A.   encourage him to "catch up" on sleep on weekends

B.   order a sleep study

C.   prescribe a nasal topical corticosteroid

D.   reduce his physical activity in the late afternoon

E.   remove electronic media access from his bedroom

Question 261   **Preferred Response: E**

Child and adolescent television viewing has long been recognized as having both negative and positive effects. In the last 1 to 2 decades, a host of additional media sources have become readily available to children, reinforcing the need for families to consider what role media will have in their children's lives. Excessive exposure to media such as television, video, cell phones, tablets, and computers has been associated with multiple health and social effects in children, including obesity and metabolic conditions, stress and psychological disorders, poor school performance, and sleep disturbance. At the same time, media can promote learning (eg, shows like Sesame Street) and encourage positive behaviors (eg, the "It Gets Better" campaign). Therefore, the American Academy of Pediatrics (AAP) has set a policy for pediatricians to address media exposure at each health supervision visit and in particular to ask 2 questions: "How much recreational screen time does your child or adolescent consume daily?" and "Is there a TV set or an internet-connected electronic device in the child's or adolescent's bedroom?" These questions provide an opportunity to discuss the benefits and detriments of media use and to suggest appropriate limits. The AAP policy further states that pediatricians should make recommendations to parents, as listed in Item C261. Pediatricians are also urged to work with their local school district to advocate for media education and to promote innovative use of new technology in schools. On a national level, the AAP encourages a dialogue with the entertainment industry and governmental agencies to promote prosocial programming and to limit marketing of tobacco, alcohol, junk food, and fast food to children.

Item C261. American Academy of Pediatrics Policy Recommendations for Recreational Screen Time.

Recommendations to pediatricians	Recommendations to parents
Ask 2 key questions at each health supervision visit: • How much recreational screen time does your child or adolescent consume daily? • Is there a TV set or an Internet-connected electronic device in the child's or adolescent's bedroom? Take a more detailed media history if the patient is overweight or obese, has aggressive behavior, uses tobacco, alcohol, or drugs, or is having difficulty in school	Limit screen time to less than 1 to 2 hours/day No screen use for children younger than 2 years of age Keep television and internet-connected devices out of the child's bedroom Monitor the media, particularly websites and social media sites, that children are using Watch television, movies, and videos with children and adolescents and use it to prompt discussions of family values Establish a family media use plan with mealtime and bedtime curfews, cell phone, and texting rules, and internet and social media use rules

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The child in the vignette has a history typical of delayed sleep phase, therefore further interventions beyond sleep hygiene counseling are not recommended at this time. He does not demonstrate snoring, apnea, mouth breathing, or nasal congestion that might suggest obstructive sleep apnea and has no other symptoms of parasomnias. Therefore, a sleep study should be considered if he does not respond to sleep hygiene intervention or if further symptoms are noted. Since he has no signs of nasal congestion, topical steroids are unlikely to affect his sleep. Allowing catch-up sleep on weekends can actually worsen delayed sleep phase syndrome and thus is discouraged. Appropriate physical activity should be encouraged for all children and exertion during the afternoon is unlikely to affect his sleep.

PREP Pearls
- The presence of a bedroom television has been associated with later bedtimes, difficulty falling asleep, and decreased total sleep hours.
- Electronic media in the child's bedroom has been associated with increased risk for obesity, substance use, and exposure to sexual content.
- The pediatrician should ask 2 key questions regarding media use at each well child visit: "How much recreational screen time does your child or adolescent consume daily?" and "Is there a TV set or an internet-connected electronic device in the child's or adolescent's bedroom?"
- Anticipatory guidance about media exposure should include discussion of screen time limits, appropriate use of social media, and developing a family media plan that includes restrictions on inappropriate or excessive media use while promoting family time.

ABP Content Specifications(s)
- Counsel parents regarding appropriate limits on screen time (eg, TV, computer) for their children

Suggested Readings
- American Academy of Pediatrics. Children, adolescents, and the media. Pediatrics. 2013;132(5):958-961. DOI:http://dx.doi.org/10.1542/peds.2013-2656.
- American Academy of Pediatrics. Policy statement–children, adolescents, obesity and the media. Pediatrics. 2011;128:201-205. DOI: http://dx.doi.org/10.1542/peds.2011-1066.
- Bhargava S. Diagnosis and management of common sleep problems in children. Pediatr Rev. 2011;32(3):91-98. DOI:http://dx.doi.org/10.1542/pir.32-3-91.
- Garrison MM, Liekweg K, Christakis DA. Media use and child sleep: the impact of content, timing and environment.Pediatrics. 2011;128:29-35. DOI: http://dx.doi.org/10.1542/peds.2010-3304.
- Strasburger VC. Pediatricians, schools, and media. Pediatrics. 2012;129(6):1161-1163. DOI:http://dx.doi.org/10.1542/peds.2011-3213.

Question 262

A 16-year-old adolescent presents to the office for a health supervision visit. Upon review of his social history, you discover that his grades have dropped significantly over the last year. He was also recently suspended for coming to school intoxicated. He reports daily marijuana use. He states that he drinks alcohol and smokes marijuana when alone and to feel better about life. His friends have told him to stop using marijuana because he forgets things when he is using it.

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

A.   distributing brochures on the effects of alcohol and marijuana

B.   identifying referral options and facilitating the patient's follow-through with them

C.   informing local law enforcement of the patient's behaviors

D.   providing a brief in-office intervention and scheduling follow-up in 2 months

E.   talking with his parents about restricting his access to alcohol

**Question 262**   **Preferred Response: B**

Guidelines for adolescent preventive services recommend that providers screen adolescents at least annually for substance use. The CRAFFT, a 6-question screening tool, has been validated for use with adolescents. To administer the CRAFFT, providers should first ask adolescents whether they have used alcohol, marijuana, or anything else to get high in the last 12 months. All adolescents are then asked about whether they have ever ridden in a Car with a driver who has been drinking or using drugs, or whether they operated a Car while under the influence. Only adolescents who endorsed using a substance are asked the remaining questions (RAFFT of CRAFFT):

- Do you use drugs or alcohol to Relax?
- Do you drink or use drugs when you are Alone?
- Do you Forget things when you drink or use drugs?
- Have your Friends or Family told you that you need to cut back on your use?
- Have you gotten into Trouble while you were using drugs or alcohol?

Each "yes" response is given 1 point.

Adolescents with a CRAFFT score of 1 should receive brief counseling regarding the effects of substance use. A score of 2 or more is considered positive.

The adolescent in this scenario reports daily marijuana use. According to the Screening to Brief Intervention (S2BI)- based approach to Clinical Screening Brief Intervention and Referral to Treatment (SBIRT, Item C262), which triages adolescents based on risk categories for substance use disorders, this patient would be considered at risk for severe substance use disorder. In addition to assessing for problems and advising the patient to quit, identifying referral options and facilitating the patient's follow through would be the most appropriate next step. Referring providers should track the adolescent's progress through follow-up appointments or phone calls.

IN THE PAST YEAR, HOW MANY TIMES HAVE YOU USED TOBACCO? ALCOHOL? MARIJUANA?

SUD, Substance use disorder

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ITEM C262: The screening to brief intervention-based approach to clinical screening brief intervention and referral to treatment.
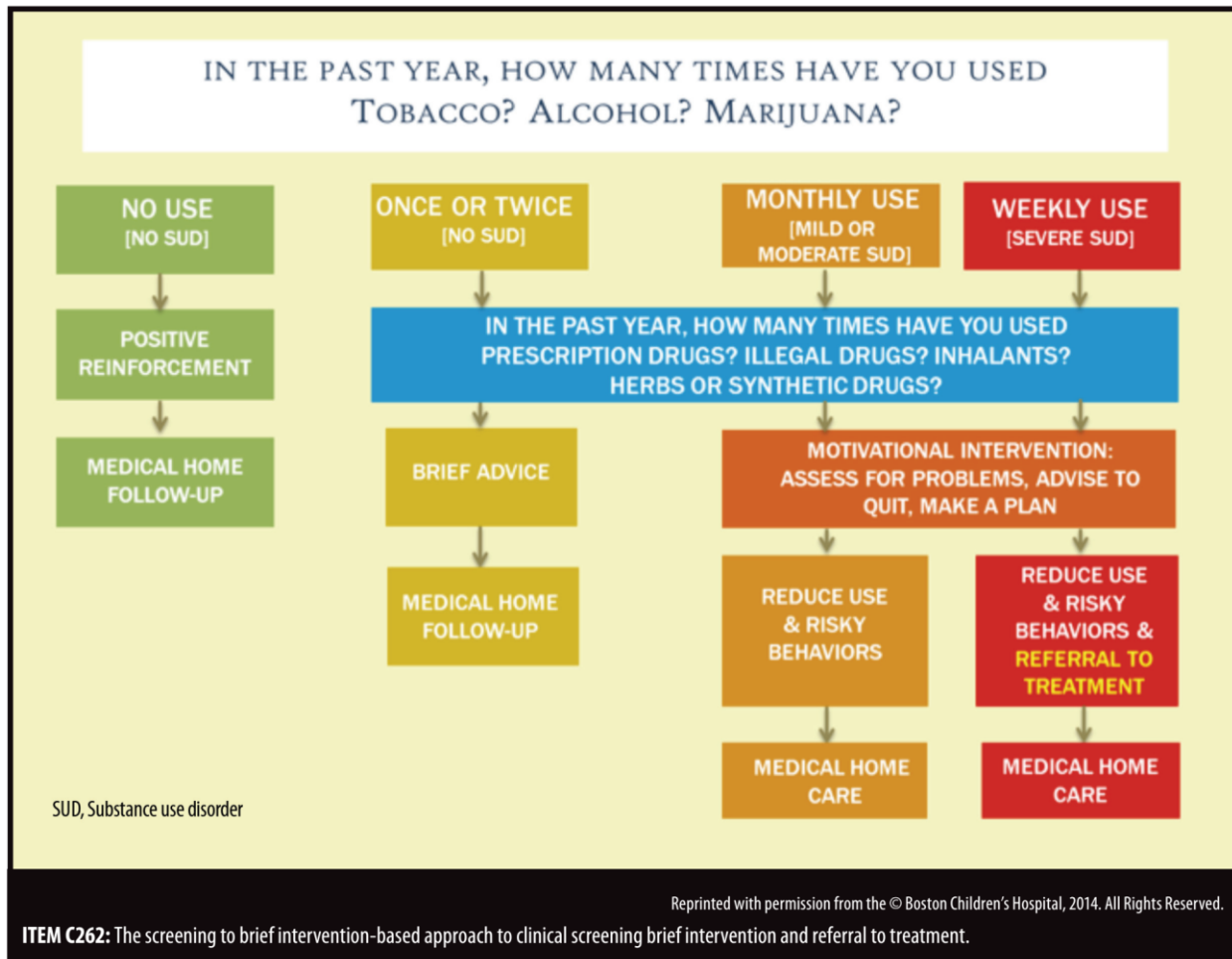
While providing information about the adverse effects of alcohol and drugs may be helpful to this individual, more intensive intervention would be required in this situation. Similarly, working with the parents to restrict the adolescent's access to drugs and alcohol would not be sufficient. If an adolescent with a CRAFFT score of 2 or more has no major problems and has the perceived self-efficacy for behavioral change, a brief in-office intervention may be sufficient, but the typical follow-up would be less than 2 months. Breaking confidentiality for the patient's safety should be considered in this situation; however, notifying law enforcement is not necessarily indicated.

PREP Pearls
- Guidelines for adolescent preventive services recommend that providers screen adolescents at least annually for substance use.
- The CRAFFT screening tool is a 6-item assessment of adolescents' substance use.
- SBIRT is the framework for in-office intervention and referrals when using and interpreting the CRAFFT.

ABP Content Specifications(s)
- Understand the primary care physician's role in preparing an adolescent and his/her family for referral for substance use/abuse treatment
- Understand the importance of periodically reassessing the progress of a patient who has been referred for substance use/abuse treatment, including reassessment for relapse

Suggested Readings
- American Academy of Pediatrics. Substance use screening, brief intervention, and referral to treatment for pediatricians.Pediatrics. 2011;128(5):e1330-e1340. DOI: http://dx.doi.org/10.1542/peds.2011-1754.
- Massachusetts Department of Public Health Bureau of Substance Abuse Services. Provider guide: adolescent screening, brief intervention, and referral to treatment for alcohol and other drug use using the CRAFFT screeing tool. Masseachusetts Department of Substance Abuse Services website.http://www.mass.gov/eohhs/gov/departments/dph/programs/substance-abuse/prevention/screening-brief-intervention-and-referral-to.html.

Question 263

You are caring for a 16-year-old adolescent with cystic fibrosis. His forced expiratory volume in 1 second is moderately decreased at 1.5 L (45% predicted). He is chronically infected with Pseudomonas aeruginosa and methicillin-resistant Staphylococcus aureus. He was most recently hospitalized with acute and non-massive hemoptysis. He does not have pulmonary hypertension and has not required chronic supplemental oxygen. Eight months ago, he was admitted to the intensive care unit when he required noninvasive respiratory support with bi-level positive pressure ventilation during a pulmonary exacerbation. At his parents' request, you begin to discuss lung transplant evaluation and this patient's advance care directives.

Of the following, the MOST appropriate approach to this issue is

A.   any discussions regarding advance directives should be deferred to the patient's pediatric pulmonologist or intensivist

B.   discussions regarding advance directives are premature and should not occur until the patient has experienced additional cystic fibrosis-related complications

C.   earlier rather than later discussion of advance care directives may ease anxiety and improve quality of life in patients with chronic disease

D.   the involvement of palliative care sends a message that the medical team is "giving up" on the patient

E.   the patient should not participate in advance care directive discussions because he has not yet reached the age of legal consent

Question 263   Preferred Response: C

This patient with cystic fibrosis represents a growing population of pediatric patients living with complex and life-limiting illnesses. In the United States, more than 50,000 children die each year, and a significant number of these deaths are attributable to a chronic medical condition. Medical and surgical treatment options have continued to evolve and have resulted in an increased life expectancy for many diseases that previously were associated with death during childhood. However, many illnesses are still associated with considerable morbidity and mortality. As such, all pediatricians should be educated in the development and implementation of care directives; patients and their families should be engaged in the development of these plans and the plans should be updated as needed and as the course of the disease changes. While many pediatric patients with chronic illnesses are under the care of a subspecialist, the development of an advanced directive is not the sole responsibility of the subspecialty physician. Families benefit if the care team (including the general pediatrician) is coordinated in their approach to these discussions.

The Patient Self Determination Act of 1991 mandates that patient preferences as to advance directives (AD) be actively solicited and documented for all adult patients. In contrast, no similar legislation has been directed at children and adolescents. It is generally appreciated that end-of-life discussions allow patients and families to examine their disease and their treatment preferences. When inquiries are made as to end-of-life preferences, literature has shown that the majority of families choose home or hospice as a preferred location of death for their child. In instances where care directives are not obtained, children are more likely to expire in the hospital.

The World Health Organization (WHO), in 2008, defined palliative care for children as "the total active care of the child's body, mind, and spirit," along with "support to the family." Furthermore, they outlined that palliative care "begins when the illness is diagnosed and continues regardless of whether or not a child receives treatment directed at the disease." An effective palliative care approach is multidisciplinary in nature and includes the family, as well as community resources. In keeping with this WHO recommendation, discussions regarding ADs are not dependent on the stage of disease and can proceed during active treatment. The intent of these discussions should not be interpreted as the health care team "giving up."
Even prior to the age of legal consent, it has become apparent that children and adolescents will be more educated and aware of care directives if they are involved and have a voice in these discussions. In clinical research studies, the "assent" of younger children is routinely sought. Most pediatric palliative care has focused on children with HIV and cancer. In a 2014 investigation of US parents' and caregivers' awareness of ADs, 61.6% of respondents reported that they had never heard of ADs and 81.8% had never discussed one previously. Only 3% had created an AD for their child.

The community of pediatric care providers will continue to benefit from a standardized approach to planning for end-of-life care. Best practice guidelines were developed in England in 2007. In the United States, the Children's Hospice and Palliative Care Services Project (ChiPPS) aims to enhance pediatric hospice and palliative care education, access, and quality.

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PREP Pearls
- The approach to end-of-life planning with an advanced directive discussion should take place early in the chronic disease course, with updates to the plan as needed.
- The approach to the end-of-life care planning for a patient with chronic and life-limiting illness is multidisciplinary, and should include various members of the medical and health care team, as well as family and community resources.

ABP Content Specifications(s)
- Recognize and apply ethical principles involved in the patient-parent-pediatrician relationship regarding advance care planning
- Understand the use of advance directives in pediatrics

Suggested Readings
- Brook L, Hain R. Predicting death in children. Arch Dis Child. 2008;93(12):1067–1070. DOI:http://dx.doi.org/10.1136/adc.2007.127332.
- Liberman DB, Pham PK, Nager AL. Pediatric advance directives: parents' knowledge, experience and preferences.Pediatrics. 2014;134(2):e436-e443. DOI: http://dx.doi.org/10.1542/peds.2013-3124.
- Lyon ME, Garvie PA, Briggs L, et al. Is it safe? Talking to teens with HIV/AIDS about death and dying: a 3-month evaluation of Family Centered Advance Care (FACE) planning - anxiety, depression, quality of life. HIV AIDS (Auckl). 2010;2:27–37. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3218704/.
- Lyon ME, Garvie PA, McCarter R, Briggs L, He J, D'Angelo LJ. Who will speak for me? Improving end-of-life decision-making for adolescents with HIV and their families. Pediatrics. 2009;123(2):e199-e206. DOI:http://dx.doi.org/10.1542/peds.2008-2379.
- Lyon ME, Jacobs S, Briggs L, Cheng YI, Wang J. A longitudinal, randomized, controlled trial of advance care planning for teens with cancer: anxiety, depression, quality of life, advance directives, spirituality. J Adolesc Health. 2014;54(6):710-717. DOI: http://dx.doi.org/10.1016/j.jadohealth.2013.10.206.
- World Health Organization. Definition of palliative care for children. World Health Organization website.http://www.who.int/cancer/palliative/definition/en/.

Question 264

A 1-day-old female newborn, with an unremarkable prenatal history, is admitted to the neonatal intensive care unit for increasing lethargy and feeding difficulties. The baby was born at term, by normal spontaneous vaginal delivery, after spontaneous rupture of membranes with clear amniotic fluid. Apgar scores were 8 and 9 at 1 and 5 minutes, respectively. Prenatal screening for infection was negative. The newborn took 20 mL of formula within 1 hour of birth, but has taken less with each subsequent feed. She has to be awakened and stimulated in order to feed. Upon admission, the newborn is minimally arousable with painful stimuli. Her vital signs show a temperature of 37°C, pulse of 150 beats/min, respiratory rate of 20 breaths/min, blood pressure of 60/40 mm Hg, and oxygen saturation of 100% on room air. Her pupils are 4 mm, equal, and sluggish. Her lungs are clear to auscultation bilaterally and her heart has a regular rate and rhythm. The newborn's muscle tone is decreased.

Laboratory results include:
- Sodium, 140 mEq/L (140 mmol/L)
- Potassium, 5 mEq/L (5 mmol/L)
- Chloride, 100 mEq/L (100 mmol/L)
- Bicarbonate, 15 mEq/L (15 mmol/L)
- Glucose, 70 mg/dL (3.9 mmol/L)
- Ammonia, 150 µg/dL (107.1 µmol/L)

Of the following, the test MOST likely to reveal this newborn's diagnosis is

A.   blood culture

B.   brain magnetic resonance imaging

C.   cerebrospinal fluid herpes simplex virus polymerase chain reaction

D.   echocardiography

E.   urine organic acids

Question 264   **Preferred Response: E**

The infant in the vignette has hyperammonemia and progressive lethargy after an unremarkable prenatal history and delivery. This is most likely caused by an inborn error of metabolism, and the test most likely to reveal the diagnosis is urine organic acids.

The many causes of coma in neonates can be divided by organ system, and the differential diagnosis for each case can be narrowed based on the clinical presentation. Infectious causes are common, and can include bacterial, viral, and parasitic etiologies. There is often an antecedent history of prolonged rupture of membranes, maternal fever, or a positive screen for group B Streptococcus. Pathogens of neonatal sepsis that are not routinely screened for include Escherichia coli, Listeria monocytogenes, and herpes simplex virus. Congenital heart disease is another important cause of poor feeding and lethargy in neonates. Although cyanotic congenital heart disease is usually apparent at birth, infants with patent ductus arteriosus–dependent lesions such as hypoplastic left heart syndrome, interrupted aortic arch, and critical aortic stenosis may appear acyanotic and otherwise normal at birth, but deteriorate within the first few days when the ductus arteriosus closes. Fetal tachycardia and cardiomyopathy can also cause progressive poor feeding and lethargy. None of these cardiac problems would cause hyperammonemia. Maternal intoxication, perinatal asphyxia, hypoglycemia, and birth trauma are causes of neonatal coma that can be identified through maternal or perinatal history; however, none of these would present with hyperammonemia.

The other main category of causes of neonatal coma is inborn errors of metabolism. These infants appear normal at birth because in utero the metabolite causing encephalopathy, usually a small molecule that can readily cross the placenta, is cleared by the mother's metabolism. For these infants the time of onset of signs and symptoms of a metabolic crisis will vary, but in severe cases can occur within hours or days. Poor feeding, lethargy, respiratory depression, and hypotonia are common initial signs. Acute encephalopathy presenting in an infant with an unremarkable history and unremarkable results of routine blood tests, cultures, and chest radiography should alert the clinician to consider an inborn error of metabolism. Acidosis, hyperammonemia, and hypoglycemia can cause coma, intracranial hypertension, and hemodynamic compromise. Suspected metabolic disease should prompt urgent evaluation of pH, lactate level, electrolytes, liver function tests, ammonia, and glucose level. More common metabolic disorders (Item C264) include organic acidemias, urea cycle defects, glycogen storage diseases, galactosemia, and fatty acid oxidation defects. The diagnosis can often be made with evaluation of urine organic acids.

Item C264. Inborn Errors of Metabolism Associated With an Acute Crisis.

Diagnostic Clues		
Primary	**Secondary**	**Suggested Disorders**
Acidosis	± Hypoglycemia ± Lactic acidosis ± Ketosis ± Elevated ammonia Increased anion gap	Various organic acid disorders

	Significant lactic acidosis
	Normoglycemia
		Mitochondrial disorders
		Pyruvate dehydrogenase
		  deficiency
		Alpha-ketoglutarate
		  dehydrogenase deficiency
		Pyruvate carboxylase
		  deficiency

	Significant lactic acidosis Hypoglycemia	Glycogen storage type I Fructose-1,6- bisphosphatase deficiency
	Normal anion gap Normal lactate No ketosis	Renal tubular acidosis
Hyperammonemia	Alkalosis or normal pH Normal lactate	Urea cycle disorders
	Reye-like illness (hypo- glycemia, elevated liver transaminases, normal ketones)	Fatty acid oxidation defects
	Acidosis ± Lactic acidosis ± Ketosis ± Hypoglycemia Increased anion gap	Various organic acid disorders
Hypoglycemia	Acidosis ± Ketosis ± Lactic acidosis ± Increased ammonia Increase in anion gap	Various organic acid disorders
	Hepatomegaly ± Lactic acidosis	Glucogen storage disorders
	No ketosis No acidosis Normal lactic acid	Hyperinsulinemia Fatty acid oxidation defects
	Hyponatremia Hypotension	Adrenal insufficiency
	Signs of liver failure	Tyrosinemia Glycogen storage disease type IV Galactosemia Niemann Pick type C

Reprinted with permission from Levy PA. Inborn errors of metabolism: part 1: overview. *Pediatr Rev*. 2009;30:131

While blood culture and evaluation for herpes simplex virus could reveal an infectious diagnosis for the infant in this vignette, a metabolic cause of this presentation is more likely. Brain magnetic resonance imaging can detect brain malformations that can cause encephalopathy, but those infants generally have an abnormal neurologic status immediately after birth. Echocardiography would be important if congenital heart disease were suspected, but such infants generally do not have hyperammonemia.

PREP Pearls
- Common causes of lethargy and coma in a neonate include infection, cardiac abnormalities, and metabolic disorders.
- Newborns with inborn errors of metabolism, cardiomyopathy, or patent ductus arteriosus (ductal) dependent cardiac lesions may appear normal at birth then develop poor feeding and lethargy in the first few days after birth.
- Neonatal hyperammonemia can be caused by urea cycle defects, fatty acid oxidation defects, and organic acidemias.

ABP Content Specifications(s)
- Formulate a differential diagnosis of lethargy and coma in a neonate

Suggested Readings
- Donn SM, Banagale RC. Neonatal hyperammonemia. Pediatr Rev. 1984;5:203-208.http://pedsinreview.aappublications.org/content/5/7/203 .
- Levy PA. Inborn errors of metabolism: part 1: overview. Pediatr Rev. 2009;30:131-138. DOI:http://dx.doi.org/10.1542/pir.30-4-131.
- Levy PA. Inborn errors of metabolism: part 2: overview. Pediatr Rev. 2009;30:e22-e28. DOI:http://dx.doi.org/10.1542/pir.30-4-e22.

Question 265

An 11-month-old female infant presents to the emergency department with fever and lethargy. She was seen by her primary care physician 8 days ago with upper respiratory tract symptoms and a cough. Vital signs show a temperature of 39.8°C, respiratory rate of 60 breaths/min, heart rate of 178 beats/min, blood pressure of 118/57 mm Hg, and a weight of 10.09 kg. On physical examination, she is lethargic and has a full anterior fontanelle. Laboratory data shows:

- White blood cells, 12,400/μL (12.4 x 10^9/L)
- Hemoglobin, 11 g/dL (110 g/L)
- Platelets, 450 x 10^3/μL (450 x 10^9/L)
- Differential, 27% segmented neutrophils, 30% bands, 38% lymphocytes, 5% monocytes
- Cerebrospinal fluid (CSF) white blood cells, 2,200/μL (98% segmented neutrophils)
- CSF red blood cells, 24/μL
- CSF glucose, 20 mg/dL (1.1 mmol/L)
- CSF protein, 375 mg/dL
- CSF Gram stain, gram-negative diplococci

Of the individuals who had contact with this patient, the BEST candidate for chemoprophylaxis is the

A.   12-year-old sibling who was vaccinated 1 year ago

B.   aunt who lives next door and is 24 weeks pregnant

C.   pre-school classmate whom she last saw 10 days ago

D.   unvaccinated resident who took the history in the emergency department

E.   vaccinated nurse who obtained a nasal wash 8 days ago

Question 265   **Preferred Response: A**

The patient in this vignette has meningitis with gram-negative diplococci, most likelyNeisseria meningitidis. The index patient's sibling is a household member and is a high risk close contact and should receive chemoprophylaxis. Immunization status does not play a role in the decision to provide chemoprophylaxis. This is because licensed vaccines are not 100% effective. Therefore, the sibling's vaccination status is irrelevant.

Antimicrobial prophylaxis is recommended for close contacts of cases with invasive meningococcal disease, including household members or childcare center providers with whom contact occurred at any time during the 7 days before symptom onset, and anyone with direct exposure to the index case's oral secretions in the 7 days before symptom onset. Another qualifying exposure includes air travel; a passenger seated directly next to an index case on a flight lasting at least 8 hours should receive chemoprophylaxis.

Chemoprophylaxis is not recommended for contacts of patients with N meningitidis isolated from nonsterile sites (eg, conjunctival or oropharyngeal swab cultures) who do not have invasive disease. Additionally, persons with contact with a high risk contact and not with the index case do not require chemoprophylaxis. Family members that do not reside within the same household, such as the aunt that lives next door, also do not need chemoprophylaxis.

While a childcare center contact is considered a close contact, exposure 10 days prior to symptom onset precludes concern for infection in this patient. The incubation period for N meningitides is 1 to 10 days and symptomatic infection usually occurs in less than 4 days. Healthcare personnel with direct exposure to the patient's secretions are candidates for chemoprophylaxis. The resident who took the history in the emergency department does not require chemoprophylaxis. Obtaining a nasal wash, as in response choice E, would be considered direct exposure to respiratory secretions. However, this sample was obtained more than 7 days before the symptom onset, so the nurse does not require chemoprophylaxis.

Chemoprophylaxis should be administered within 24 hours. Agents used for chemoprophylaxis include rifampin, ceftriaxone, and ciprofloxacin. Index cases that are treated with an agent other than ceftriaxone need to be decolonized with 1 of the recommended chemoprophylactic drugs after their course of treatment for invasive disease to eliminate nasopharyngeal colonization. When an outbreak caused by a serogroup covered by licensed vaccines occurs, immunization is an adjunctive prophylaxis measure.

PREP Pearls
- Antimicrobial prophylaxis is recommended for close contacts of patients with invasive meningococcal disease, including household members, daycare, or childcare center contacts with whom contact occurred at any time during the seven days before symptom onset, and anyone with direct exposure to the index case's oral secretions in the 7 days before symptom onset.
- Chemoprophylaxis for Neisseria meningitidis should be administered within 24 hours with rifampin, ceftriaxone, or ciprofloxacin.

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- Healthcare personnel without direct exposure to the patient's secretions are not candidates for chemoprophylaxis.

ABP Content Specifications(s)
- Plan appropriate prophylaxis for individuals exposed to Neisseria meningitidis

Suggested Readings
- American Academy of Pediatrics. Meningococcal infections. In: Kimberlin DW, Brady MT, Jackson MA, Long SS, eds. Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015:547-558.
- Crohn AC, MacNeil JR, Clark TA, et al. Prevention and control of meningococcal disease: recommendations of the advisory committee on immunization practices. MMWR Morbid Mortal Wkly Rep. 2013;62(RR02):1-28.http://www.cdc.gov/mmwr/preview/mmwrhtml/rr6202a1.htm.

Question 266

A 10-year-old boy presents to your office with the chief complaints of increased thirst, micturition, and nighttime bed wetting for the last 6 months. According to his mother, the patient attained bladder and bowel control by 4 years of age. The patient has also been complaining of headaches on waking up in the morning, which have been increasing in severity for the last week. He has also had vomiting associated with headaches on waking up for the last 2 days. There is no history of encopresis. They moved to a new home and the patient is attending a new school for the last year. He has been an above average student. 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 is in no pain currently.
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.   psychogenic polydipsia

B.   small bladder

C.   stress

D.   urinary tract infection

E.   urine concentration defect

Question 266   **Preferred Response: E**

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 10-year-old boy in the vignette has recent onset of increased micturition and associated secondary nocturnal enuresis, which needs further evaluation.

Polyuria is characterized by an increased total urine volume resulting from an underlying defect in water balance. This presents with the excretion of large volumes of dilute urine, as seen in diabetes mellitus (osmotic diuresis), diabetes insipidus (anti-diuretic hormone disorders), and psychogenic polydipsia. It is important to note that children with polyuria may have nocturia or nocturnal enuresis; however, the more frequently reported symptoms of frequency, nocturia, or enuresis may not be associated with increased urinary volume (or polyuria).

Recent onset of increased thirst, micturition, and nocturnal enuresis (and nocturia) is indicative of increased urine volume in the patient. His urinalysis shows a specific gravity of 1.005, which also points towards an underlying urine concentration defect associated with increased urine volume. Absence of glucose in the urine rules out diabetes mellitus as the cause of his polyuria.

Diabetes insipidus (DI) occurs secondary to either decreased secretion of anti-diuretic hormone (ADH) known as central DI, or renal resistance to ADH effects known as nephrogenic DI. Patients with DI present with polyuria, polydipsia, and increased thirst. Patients with DI have decreased urine osmolality and associated serum osmolality that may be normal (older patients able to increase their water intake) or increased (in patients unable to increase their water intake, as in neonates or those with developmental delay). A detailed neurologic examination, including examination of the spine, is vital in any patient presenting with recent onset abnormal voiding patterns. The history of headaches more prominent on waking up in the morning and association with vomiting is indicative of intracranial pathology, as noted for the patient in this vignette. Central DI is characterized by decreased ADH secretion. It could be idiopathic (most common) or secondary to central nervous system tumors, infiltrative lesions (histiocytosis), and trauma (surgical or nonsurgical).

Nephrogenic DI in children is secondary to a mutation in either the ADH-receptor (AVPR 2) or the aquaporin 2 channels. The AVPR 2 mutations have an X-linked inheritance, account for 90% of cases, and males are more severely affected than females. Water restriction, used to differentiate nephrogenic DI from central DI, is not indicated in neonates and infants. This differentiation is made through evaluation of the response to vasopressin. Vasopression can be administered intravenously (1 μg in infants and 2 μg in older children) and the response is assessed by evaluating serum electrolytes and urine osmolality in 1 to 2 hours. Urine osmolality and serum electrolytes can be rechecked in 3 to 4 hours, as the response can take time in some cases. Intranasal desmopressin acetate (10 μg for infants, 20 μg for older children) can also be used for evaluating response to vasopression. However, it should be noted that intranasal formulations have been associated with risk for developing severe hyponatremia and are no

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longer indicated for the treatment of primary nocturnal enuresis. If there is no response to vasopressin and the urine osmolality remains at less than 100 mOsm/kg (100 mmol/kg) over baseline, the diagnosis of nephrogenic DI is confirmed.

Psychogenic polydipsia presents with hyponatremia associated with a low urine osmolality, consistent with water overload. Maximal urine concentration is usually impaired (500 to 600 mOsm/kg) compared to that in normal patients (800 mOsm/kg or more). Water deprivation in these patients will increase the urine osmolality (> 500 mOsm/kg) and there will be no response to desmopressin because endogenous production and release of ADH is normal.

Recent onset stress such as moving to a new home and school can sometimes lead to secondary nocturnal enuresis in children. However, the patient's symptoms of morning headaches increasing in severity and vomiting are suggestive of an intracranial pathology.

Symptoms of bladder dysfunction (associated with underlying small bladder capacity, overactive bladder, dysfunctional voiding) include urinary frequency, urgency, and urge incontinence. Incontinence in such patients may present with daytime or nocturnal enuresis. Constipation is a commonly associated symptom in patients with bladder dysfunction. 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. 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. Bladder function is evaluated with renal bladder ultrasonography, voiding cystourethrogram, and urodynamic studies (intravesical pressures and volume of fluid during filling, storage, and voiding). In the vignette, urinary tract infection is unlikely in the absence of fever or urinary symptoms such as dysuria, flank pain, or burning micturition. The absence of leukocyte esterase, nitrites, and bacteria on urinalysis also rules out urinary tract infection as the underlying cause of the patient's symptoms.

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 consecutive months.
- Polyuria (excretion of large volumes of dilute urine) may be seen in diabetes mellitus (osmotic diuresis), diabetes insipidus (antidiuretic hormone disorders), and psychogenic polydipsia.
- Recent onset of increased thirst, micturition, and nocturnal enuresis (and nocturia) is indicative of increased urine volume in a patient.
- Diabetes insipidus (DI) occurs secondary to either decreased secretion of antidiuretic hormone (ADH) known as central DI, or renal resistance to ADH effects known as nephrogenic DI.

ABP Content Specifications(s)
- Plan the appropriate management of enuresis of various types
- Plan the appropriate evaluation of enuresis of various types, including that occurring after continence has been achieved

Suggested Readings
- Linshaw MA. Back to basics: congenital nephrogenic diabetes insipidus. Pediatr Rev. 2007;28(10):372-380. DOI:http://dx.doi.org/10.1542/pir.28-10-372.
- Miller RS, Libber Sm, Plotnick LP. Polyuria. 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; 2009:1670-1675.
- Nevéus T, von Gontard A, Hoebeke P, et al. The standardization of terminology of lower urinary tract function in children and adolescents: report from the Standardization Committee of the International Children's Continence Society. J Urol. 2006;176(1):314-324. DOI: http://dx.doi.org/10.1016/S0022-5347(06)00305-3 .
- Saborio P, Tipton GA, Chan JCM. Diabetes insipidus. Pediatr Rev. 2000;21(4):122-129. DOI:http://dx.doi.org/10.1542/pir.21-4-122.

Question 267

A 3-year-old boy presents to your office with a history of recurrent rectal prolapse without visible blood. He began having prolapse with bowel movements 6 weeks ago. It occurs with every bowel movement and resolves without intervention. His bowel movements are described as clusters of grapes. His mother denies fever, recent illness, diarrhea, poor growth, or other chronic medical issues. Physical examination shows height and weight at the 50th percentile for age. His abdomen is mildly distended with positive bowel sounds. His rectal examination identifies a normally placed anus without visible hemorrhoids or fissures.

Of the following, the MOST likely diagnosis for this child is

A.   celiac disease

B.   cystic fibrosis

C.   functional constipation

D.   rectal polyps

E.   ulcerative colitis

Question 267   **Preferred Response: C**

The 3-year-old boy in this vignette has rectal prolapse with each bowel movement, which resolves without intervention. He has stools that are grape-like, indicating constipation. Functional constipation is the most likely diagnosis and reason for his rectal prolapse. Although rectal prolapse can be seen in cystic fibrosis, this child has growth parameters that are at the 50th percentile, making this unlikely. The normal growth parameters and lack of diarrhea make celiac disease an unlikely diagnosis for this child. The stool has not had visible blood and no polyps are seen or palpated on digital rectal examination, making rectal polyps an unlikely cause of his rectal prolapse. Ulcerative colitis is typically associated with diarrhea, not constipation, and is not typically associated with rectal prolapse.

Rectal prolapse is defined as either mucosal or full thickness protrusion of the rectum through the anus. On physical examination, concentric rings of rectal mucosa can be seen (Item C267). Rectal prolapse has a male predominance. The differential diagnosis includes constipation, acute diarrhea, cystic fibrosis, parasitosis, polyps, malnutrition, increased intra-abdominal pressure, and conditions that result in pelvic floor weakness.

Item C267

Item C267. Rectal prolapse. Reprinted with permission from Fleisher GR, MD, Ludwig S, MD, Baskin MN, MD. Atlas of Pediatric Emergency Medicine. Philadelphia, PA: Lippincott Williams & Wilkins; 2004.

Treatment involves management of the underlying cause, when identified. Conservative therapy with manual reduction of the prolapse and aggressive management of the underlying etiology is often successful. Emergent evaluation is indicated if the prolapse cannot be reduced. If prolapse recurrence persists, surgical intervention is recommended.

PREP Pearls
- Constipation is the most common cause of rectal prolapse.
- Conservative medical therapy is often successful.
- Cystic fibrosis and parasites should be ruled out prior to surgical intervention.
- Rectal prolapse has a male predominance.

ABP Content Specifications(s)
- Identify the clinical conditions other than cystic fibrosis that are associated with rectal prolapse

Suggested Readings
- Gourgiotis S, Baratsis S. Rectal prolapse. Int J Colorectal Dis. 2007;22(3):231-243 .
- Melton GB, Kwaan MR. Rectal prolapse. Surg Clin North Am. 2013;93(1):187-198. DOI:http://dx.doi.org/10.1016/j.suc.2012.09.010.

Question 268

You are called to assess a female newborn for unusual limb findings. Pregnancy was remarkable for decreased fetal movement. There was no history of maternal uterine fibroids or oligohydramnios. Delivery was via cesarean delivery secondary to transverse fetal position. Weight and length are appropriate for gestational age. On physical examination, you note that the shoulders are internally rotated, the elbows are extended, the wrists are flexed with ulnar deviation, the fingers are stiff, and the thumbs are positioned in the palms (Q268). On the lower extremities, the hips are bilaterally dislocated with knees extended, and the feet have equinovarus contractures. Breathing is normal and unlabored. Facies are non-dysmorphic.

Of the following, the MOST likely diagnosis is

A.   arthrogryposis multiplex congenita

B.   Crouzon syndrome

C.   perinatally lethal osteogenesis imperfecta

D.   peroxisomal disorders

E.   Prader-Willi syndrome
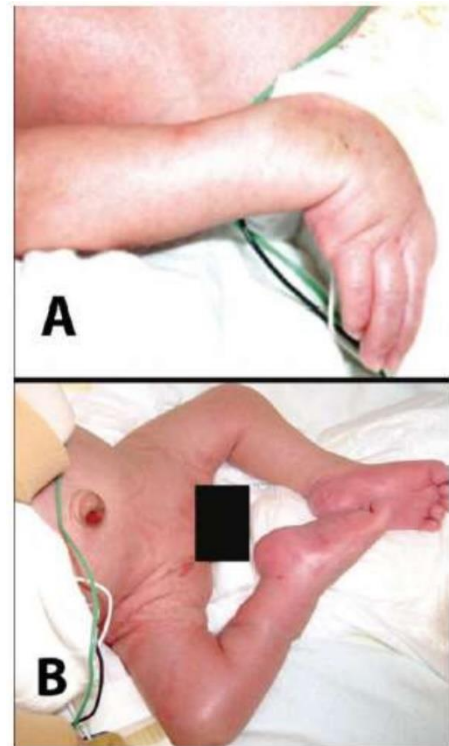
Item Q268A, Item Q268B. Findings for the newborn described in the vignette. Reprinted with permission from Cherian L, Rosenberg JJ. Pediatric Rev. 2010; 31(9):385-387.

Question 268   **Preferred Response: A**

The baby in the vignette has arthrogryposis multiplex congenita (AMC) (Item C268A, Item C268B), but more specifically, a subtype called amyoplasia, which is commonly called the "classic arthrogryposis." Arthrogryposis is a descriptive term for a congenital nonprogressive contracture affecting 1 or more areas of the body. A contracture of a joint could be in a permanently flexed position or a straightened position with restricted movement of the joint. If the arthrogryposis impacts 2 or more different regions of the body, it is referred to as AMC, as in the newborn in this vignette who has multiple affected joints. The shoulders, elbows, knees, wrists, ankles, fingers, toes, and hips are commonly affected. Arthrogryposis multiplex congenita is a heterogeneous condition that could be secondary to disorders of the central or peripheral nervous system, maternal myasthenia gravis, connective tissue disorders leading to decreased fetal movement, vascular causes, uterine crowding caused by uterine fibroids or other uterine anomalies, environmental factors, maternal infections (cytomegalovirus or toxoplasmosis), or teratogenicity. Most cases are sporadic in occurrence; however, more than 400 different genetic disorders can also be associated with AMC. Approximately 50% of cases have a genetic basis, so a thorough family pedigree should be obtained. Motor neuron conditions, including spinal muscular atrophy, mitochondrial cytopathy, and primary myopathy are the most commonly seen genetic disorders associated with AMC. Spinal muscular atrophy (SMA) should be considered given the high population carrier frequency of 1 out of 30 through 1 out of 50. Although most patients with SMA present with hypotonia, up to 20% present with in utero contractures, especially in SMA type 0.

The diagnosis of amyoplasia, as in the newborn in this vignette, should be considered when the following clinical manifestations are present:
- Symmetric congenital rigid contractures
- Internal rotation of the shoulder, fixed extension of the elbows, pronation of the forearm, flexion of the wrist
- Significant equinovarus deformity of the foot
- Marked decrease in muscle mass of limbs
- Lack of flexion creases on fingers and hands
- Dimples over affected joints
- Unaffected, mobile trunk
- Nevus flammeus in the mid-face or glabellar region
- Digital reductions (12%)
- An alert infant with non-dysmorphic facies
- Negative family history for arthrogryposis

Reprinted with permission from Cherian L, Rosenberg JJ. Visual diagnosis: multiple joint contractures in a male newborn. *Pediatr Rev.* 2010;31(9):385-387

ITEM C268: Newborn with arthrogryposis mulitplex congenita, or classic arthrogryposis.

Making an accurate diagnosis of AMC is essential so the provider can monitor for other problems that may be associated with the diagnosis and provide accurate prognostic guidelines. Thirty percent to 40% of patients have early feeding problems, often necessitating gastrostomy tube or nasogastric tube feedings. Feeding problems are typically short lived, lasting until about 4 months of age. Increased rates of bowel atresia and abdominal wall musculature deficiencies have been noted due to vascular pathology. Some individuals have respiratory problems initially. Patients frequently require extensive physical therapy, casting, and orthopedic procedures. Most have gross and fine motor delays, but have normal intelligence and communication skills. Individuals are quite capable of excelling scholastically, but will require extensive parental support because of their physical limitations. Patients are at risk for scoliosis in childhood and arthritis in affected joints starting in the third decade of life. Dilated ureters and transient hydronephrosis are present in some individuals. Mothers often report a lack of fetal movement known as akinesia during pregnancy.

Crouzon syndrome is a genetic disorder characterized by coronal and sagittal craniosynostosis, leading to wide-set bulging eyes, vision problems, a beaked nose, and an underdeveloped upper jaw. They also have dental problems, hearing loss, and occasionally cleft lip and palate, along with normal intelligence. Peroxisomal biogenesis disorders in the newborn period present with hypotonia, poor feeding, distinctive facies, seizures, and liver cysts with hepatic dysfunction. Prader-Willi syndrome is characterized by severe hypotonia and feeding difficulties in early infancy, followed by excessive eating and gradual development of morbid obesity. Patients have cognitive impairment, hypogonadism, short stature, and a distinctive behavioral presentation (temper tantrums, stubbornness, and obsessive-compulsive behaviors). Crouzon syndrome, peroxisomal disorders, and Prader-Willi syndrome do not present with congenital contractures.

Perinatally lethal osteogenesis imperfecta presents with relative macrocephaly, dysmorphic facies, dark blue sclera, intrauterine growth restriction, extreme bowing and shortening of extremities caused by multiple underlying fractures, hypoplastic thorax, and rib fractures, with early death resulting from pulmonary insufficiency.

PREP Pearls
- Arthrogryposis multiplex congenita presents with congenital nonprogressive contractures of 2 or more joints of the body. Patients have symmetric congenital rigid contractures involving internal rotation of the shoulders, fixed extension of the elbows, pronation of the forearm, flexion of the wrist, and significant equinovarus deformity of the foot.
- Patients with arthrogryposis have a marked decrease in muscle mass of limbs along with non-dysmorphic facies.
- Arthrogryposis multiplex congenita is a heterogeneous condition that could be secondary to disorders of the central or peripheral nervous system, maternal myasthenia gravis, connective tissue disorders leading to decreased fetal movements, vascular causes, uterine crowding caused by uterine fibroids or other uterine anomalies, environmental factors, maternal infections (cytomegalovirus or toxoplasmosis), or teratogenicity.

ABP Content Specifications(s)
-      Recognize the clinical features associated with arthrogryposis

Suggested Readings
- Gordon N. Arthrogryposis multiplex congenita. Brain Dev. 1998;20(7):507-511. DOI: http://dx.doi.org/10.1016/S0387-7604(98)00037-0.
- Haliloglu G, Topaloglu H. Arthrogryposis and fetal hypomobility syndrome. Handb Clin Neurol. 2013;113:1311-1319. DOI: http://dx.doi.org/10.1016/B978-0-444-59565-2.00003-4.
- Hall JG, Aldinger KA, Tanaka KI. Amyoplasia revisited. Am J Med Genet A. 2014;164(3):700-730. DOI:http://dx.doi.org/10.1002/ajmg.a.36395.
- Rink BD. Arthrogryposis: a review and approach to prenatal diagnosis. Obstet Gynecol Surv. 2011;66(6):369-377. DOI:http://dx.doi.org/10.1097/OGX.0b013e31822bf5bb.

Question 269

The parents of an 8-month-old male infant bring him to your office for follow-up after a recent episode of otitis media. He completed a 10-day course of amoxicillin and is clinically improved. His fever resolved after 3 days and his activity is back to normal. The infant's mother is concerned because he has loose stools and diaper rash. He is breastfeeding well, but is not eating as much pureed food as he did before the illness. On physical examination, you find erythematous papules and pustules on the penis, scrotum, and inner thighs (Item Q269). There are bilateral middle ear effusions and a few white plaques on the buccal mucosa. The remainder of the physical examination is unremarkable.

Item Q269

Item Q269. Diaper rash as seen in the infant described in the vignette. Reprinted with permission from Singalavanija S, Frieden IJ. Pediatr Rev.1996;16(4):142-147.

Of the following, the BEST treatment for this patient's condition is a(n)

   A.   oral antifungal suspension

   B.   oral broad spectrum antibiotic

   C.   topical antibiotic cream

   D.   topical antifungal ointment

   E.   topical steroid cream

Question 269   **Preferred Response: A**

The infant in the vignette has candida dermatitis in combination with oral candidiasis. An oral antifungal suspension should be used when oral thrush coexists with perineal candidiasis. Candida diaper dermatitis often follows a course of oral antibiotics. The diagnosis is usually based on the characteristic clinical findings: beefy red plaques with satellite papules and pustules typically involving the inguinal creases, lower abdomen, mons, scrotum, and base of penis. Microscopic examination of a potassium hydroxide preparation of a smear from a pustule will demonstrate budding yeast or pseudohyphae, but this need only be performed in the case of recalcitrant diaper dermatitis or a severely ill infant. Topical antifungal treatment for candidiasis localized either to the diaper area or oral mucosa would be appropriate. Oral antifungal treatment is best when these occur concurrently.

The presumptive cause of diaper dermatitis should be identified to select the appropriate treatment. Antibiotics should be used to treat bacterial infections; Streptococcus typically causes a perianal erythematous rash, whereas Staphylococcus may cause cellulitis or bullous impetigo on the buttocks. Topical antibiotic cream or ointment may be used for well-localized superficial bacterial skin infections. Oral antibiotics should be prescribed for widespread lesions, cellulitis, systemic symptoms, or if there is a concurrent bacterial infection, such as otitis media. More severe infection may require parenteral antibiotic treatment.

In contrast to candidiasis, irritant diaper dermatitis typically spares the intertriginous creases and involves the convex surfaces of the buttocks. Both may involve the lower abdomen, proximal thighs, and perineal area. The primary method of treatment for irritant dermatitis is frequent diaper changes to keep the area clean and dry. A protective barrier cream, such as zinc oxide or petrolatum-based preparations, can aid in healing. When applied at every diaper change such creams are effective in forming a barrier to protect the skin from ongoing contact with stool and urine. Topical corticosteroids should rarely be used, and only to treat severely inflamed irritant diaper dermatitis. Only low-potency nonhalogenated topical corticosteroid creams should be used, sparingly, twice daily for no longer than 3 to 5 days.

PREP Pearls
- The treatment of diaper dermatitis should be directed toward the most likely etiology.
- When oral thrush coexists with perineal candidiasis, an oral antifungal suspension should be used to treat both concurrently.
- The mainstay of treatment of irritant diaper dermatitis is to minimize direct skin contact with urine and feces.

ABP Content Specifications(s)
- Recognize the etiology of diaper dermatitis, and manage appropriately

Suggested Readings
- Paller AS, Mancini AJ. Cutaneous disorders of the newborn. Hurwitz Clinical Pediatric Dermatology: A Textbook of Skin Disorders of Childhood and Adolescence. 4th ed. Philadelphia, PA: Saunders Elsevier; 2011:20-23.

- Rasmussen JE. Diaper dermatitis. Pediatr Rev. 1984;6(77):77-82.http://pedsinreview.aappublications.org/content/6/3/77.
- Singalavanija S, Frieden IJ. Diaper dermatitis. Pediatr Rev. 1995;16(4):142-147. DOI:http://dx.doi.org/10.1542/pir.16-4-142.

Question 270

A 6-year-old boy presents to the emergency department with pallor, leg pain, and bruising. The results of a complete blood cell count are shown:

- White blood cell count   11,600/μL (11.6 x 10^9/L)
- Hemoglobin   7.2 g/dL (72 g/L)
- Platelet count   67 x 10^3/μL (67 x 10^9/L)
- % neutrophils   3%
- % leukemic blasts   26%

A blood sample is sent for flow cytometry to determine whether the leukemic blasts are lymphoid or myeloid. The resident working with you asks why this is important to do at this time.

Of the following, this information is necessary because it will impact

A.   prognosis and treatment of the leukemia

B.   prognosis, but not treatment of the leukemia

C.   whether or not emergent leukopheresis is indicated

D.   whether or not the child will need a central venous line

E.   whether or not to leukoreduce (filter) and irradiate transfused blood products

Question 270   **Preferred Response: A**

Leukemia is among the most common types of childhood cancer, representing approximately one-third of the 15,000 childhood cancer diagnoses in the United States each year. Among the childhood leukemia diagnoses, the vast majority are acute leukemia, either lymphoid or myeloid. Acute lymphoblastic leukemia (ALL) is the single most common cancer diagnosis in children and is far more common than acute myeloblastic leukemia (AML). The 2 forms of childhood acute leukemia behave and are treated in very different manners, and have vastly different prognoses. While ALL is among the most curable forms of childhood cancer (standard-risk ALL has an overall survival of well over 90%), AML has, at best, a 60% overall survival. Acute lymphoblastic leukemia is treated with relatively low intensity therapy over a long period of time, while AML is treated with very high intensity therapy over a relatively short period of time. As rapidly initiating therapy for a child with newly diagnosed leukemia is very important, distinguishing whether a child with acute leukemia has AML or ALL as quickly as possible is a high priority. Historically, these were differentiated on the basis of light microscopic appearance and histochemical stains. Current standards of practice utilize flow cytometry to distinguish between ALL and AML.

Flow cytometry maps the surface antigens on cells. The cells are incubated with antibodies to surface markers that are conjugated to fluorochromes. After incubation, the cells are drawn in a single file through the flow cytometer in which various lasers hit the cells. If the wavelength of light emitted by the laser excites the fluorochrome conjugated to the antibody, a different wavelength of light is emitted by the fluorochrome that can be detected by the flow cytometer. If that second wavelength is detected, then the targeted surface marker is present on the cell. Acute lymphoblastic leukemia and AML have markedly different patterns of surface markers, making flow cytometry an effective way to rapidly and accurately distinguish between the two. Flow cytometry can also be used to distinguish between different subtypes of ALL, including B and T cell.

Leukopheresis is indicated in patients who present with hyperleukocytosis, generally a white blood cell count greater than 100,000/μL. The patient in the vignette had a presenting white blood cell count of 11,600/μL, so leukopheresis would not be indicated whether he had ALL or AML. Irrelevant of the diagnosis, the patient will need a central venous catheter to deliver the chemotherapy. Whether the child in the vignette has ALL or AML, he will be immunocompromised, and as such, should receive leukoreduced and irradiated blood products at all times.

PREP Pearls
- Acute lymphoblastic leukemia (ALL) and acute myeloblastic leukemia (AML) have markedly different prognoses and treatments.
- Flow cytometry can rapidly and accurately distinguish AML from ALL.
- Leukopheresis is indicated only in the context of hyperleukocytosis (generally when the white blood cell count is > 100,000/μL).

ABP Content Specifications(s)
- Understand that management of leukemia is dependent on its type

Suggested Readings
- Hutter JJ. Childhood leukemia. Pediatr Rev. 2010;31(6):234-241. DOI: http://dx.doi.org/10.1542/pir.31-6-234.
- Pearce JM, Sills RH. Consultation with the specialist: childhood leukemia. Pediatr Rev. 2005;26(3):96-104. DOI:http://dx.doi.org/10.1542/pir.26-3-96.
- Pui CH, Carroll WL, Meshinchi S, Arceci RJ. Biology, risk stratification, and therapy of pediatric acute leukemias: an update. J Clin Oncol. 2011;29(5):551-565. DOI: http://dx.doi.org/10.1200/JCO.2010.30.7405

Question 271

You have just diagnosed a 9-year-old boy with attention-deficit/hyperactivity disorder, following your usual data-gathering steps from both school and home that helped characterize the problem. After discussing the treatment options, the parents have elected to initiate methylphenidate and plan a follow-up appointment with you in 4 weeks.

Of the following adverse effects, the MOST likely to occur in this patient is

      A.   dysphoria
      B.   hallucinations
      C.   headaches
      D.   hypertension
      E.   tics

Question 271   **Preferred Response: C**

Stimulant medications are very well known for causing appetite suppression and weight loss (for which regular monitoring of the growth curve is very helpful) and difficulty with initiating sleep. In addition to these risks, more than 10% of children using stimulants will also experience headaches, stomach aches, dry mouth, and nausea. Two percent to 10% of children using stimulants will experience irritability, dysphoria, cognitive dulling, obsessiveness, anxiety, tics, dizziness, or blood pressure and pulse changes. Less than 2% of children using stimulants could have a notable, but rare reaction of hallucinations (usually visual or tactile rather than auditory) or manic symptoms; these are typically risks that appear when using stimulants at high doses.

Of the options listed in the vignette, headaches are the most likely to be experienced by this child. Usual care monitoring of stimulant use includes following the child's height and weight, checking blood pressure and pulse after treatment initiation to ensure the child did not have an outlier response, and reviewing treatment response repeatedly at least every 6 months after reaching an effective dosage to assess for loss of benefit as the child grows and develops.

PREP Pearls
- The most common adverse effects of stimulant medication include appetite suppression, insomnia, headaches, stomach aches, dry mouth, and nausea.
- Children receiving stimulant therapy should be monitored at least every 6 months to measure growth, screen for new adverse effects, and identify any loss of treatment effectiveness.

ABP Content Specifications(s)
- Understand the common side effects of medications used to treat attention deficit hyperactivity disorder

Suggested Readings
- Hilt RJ. Monitoring psychiatric medications in children. Pediatr Ann. 2012;41(4):157-163. DOI: http://dx.doi.org/10.3928/00904481-20120307-13.
- Pierce K. Treatment of ADHD. Pediatr Ann. 2011;40(11):556-562. DOI: http://dx.doi.org/10.3928/00904481-20111007-06.