You are evaluating a 5-year-old boy who is hospitalized in the pediatric intensive care unit with findings of poor perfusion, renal failure, and respiratory compromise requiring intubation. You note an abnormality on the cardiac monitor (image).

Of the following, the MOST likely cause of this patient’s electrocardiographic findings is:

A. hyperkalemia  
B. hypernatremia  
C. hypocalcemia  
D. hypokalemia  
E. hyponatremia
Preferred Response: A

Peaked T waves on rhythm strip or electrocardiography (ECG), as shown for the boy in the vignette, are indicators of hyperkalemia. Other electrocardiographic findings might include atrioventricular block, widening of the QRS complex, and degeneration of the ECG complex into a sinusoidal shape. Elevated potassium concentrations appear to have a direct effect on the potassium channels, increasing their activity and speeding membrane repolarization. Hyperkalemia causes an overall membrane depolarization that inactivates many sodium channels. The faster repolarization of the cardiac action potential causes the tenting of the T waves, and the inactivation of sodium channels causes a sluggish conduction of the electrical wave around the heart, which leads to widening of the QRS complex. The serum potassium concentration at which electrocardiographic changes develop is somewhat variable. Hyperkalemia has many causes (Item C5).

Hypernatremia and hyponatremia are not typically associated with rhythm irregularities or electrocardiographic disturbances. Hypocalcemia can result in prolongation of the QT interval. This type of electrical instability puts the patient at high risk for torsades de pointes, a specific type of ventricular fibrillation. Hypokalemia can cause flattened or inverted T waves, ST depression, and a prolongation of the QT interval.

Suggested Reading:

Wrenn KD, Slovis CM, Slovis BS. The ability of physicians to predict hyperkalemia from the ECG. Ann Emerg Med.1991;20:1229–1232

Item C5. Causes of Hyperkalemia

Ineffective Elimination
- Renal insufficiency
- Medications that interfere with urinary excretion
  - Angiotensin-converting enzyme inhibitors and angiotensin receptor blockers
  - Potassium-sparing diuretics
  - Nonsteroidal anti-inflammatory drugs
  - Calcium channel blockers
  - Trimethoprim
  - Pentamidine
- Mineralocorticoid deficiency or resistance
  - Addison disease
  - Aldosterone deficiency
  - Congenital adrenal hyperplasia
  - Type IV renal tubular acidosis

Excessive Release From Cells
- Rhabdomyolysis, burns, or any cause of rapid tissue necrosis, including tumor lysis syndrome
- Blood transfusion or hemolysis
- Shifts/transport out of cells caused by acidosis, low insulin concentrations, beta-blocker therapy, digoxin overdose, or the paralyzing agent succinylcholine

Excessive Potassium Administration
- Medication errors
- Toxic ingestion
A 15-year-old girl presents to your clinic for evaluation of primary amenorrhea. She has a history of mild seasonal allergies and a right radius fracture at age 9. Her mother’s height is 5 ft 9 in and father’s height is 6 ft 2 in. The mother reports that her menarche occurred at 11 years, and the father recalls shaving at 14 years. On physical examination, the girl has Sexual Maturity Rating (SMR) 3 pubic hair and SMR 1 breast development. Bone age radiography shows a skeletal maturity of 13 years. No other abnormalities are noted. Her growth curve is shown in image.

Of the following, the MOST appropriate next step in this girl’s evaluation and treatment is to:

A. initiate oral conjugated estrogen therapy
B. measure serum estradiol
C. measure serum thyroid-stimulating hormone
D. obtain a karyotype
E. perform a bimanual examination
The differential diagnosis of primary amenorrhea includes constitutional delay, eating disorders, chronic disease (typically causing low weight for height), severe androgen resistance, Rokitansky syndrome (absence of uterus), imperforate hymen, gonadotropin deficiency, and primary ovarian failure. Based on the short stature, growth pattern, and absence of breast development in the presence of pubic hair described for the girl in the vignette, gonadal failure due to Turner syndrome should be considered and a karyotype obtained. Indeed, the most common cause of primary ovarian failure in otherwise healthy girls is Turner syndrome. Initiating estrogen therapy in a patient without establishing a clear diagnosis is unwise. Measurement of serum estradiol is unnecessary because the finding of breast Sexual Maturity Rating 1 indicates absence of pubertal estradiol concentrations.

Although thyroid disease can cause menstrual irregularities, other symptoms and signs would be expected, making the measurement of thyroid-stimulating hormone unnecessary. A bimanual examination is not required in the first stages of an amenorrhea evaluation when Turner syndrome is suspected and could be distressing for this girl.

Classic Turner syndrome occurs in girls who have a complete absence of one X chromosome (karyotype 45XO). However, a large percentage of affected girls have mosaic presentations, meaning they have a mix of 45XO and 46XX chromosomes when multiple cells are examined. Such girls often have less of the characteristic stigmata of Turner syndrome (e.g., shield chest, webbed neck, low posterior hair line, short stature). Girls who have mosaic Turner syndrome are more likely to present with primary amenorrhea if they are not diagnosed on the basis of the more classic physical features of the condition.

Of note for the girl in the vignette, her parents are both tall, providing a mid-parental height of 5 ft 9 in, which is approximately the 95th percentile for adult women, but the patient is growing at only the 5th percentile on a standard growth curve. It is important to consider parental height even when children maintain normal growth velocity. The 95th percentile for height on a Turner syndrome growth curve nearly overlaps with the 5th percentile for height in unaffected girls. Accordingly, girls who have Turner syndrome rarely reach the 5th percentile without intervention such as growth hormone or oxandrolone therapy. Growth hormone therapy is approved for short stature in girls who have Turner syndrome, even though they do not have growth hormone deficiency. In this case, the girls' above-average mid-parental height contributed to her ability to reach a height within the reference range for unaffected girls.


Source: 2012 PREP® Self-Assessment, Question 27
A 6-year-old previously healthy boy presents with the recent development of nocturnal dyspnea. On questioning of his parents, you discover that the child has experienced exercise intolerance, two episodes of syncope while running, poor appetite, and a cough without congestion over the past year. His physical examination reveals a heart rate of 120 beats/min, respiratory rate of 26 breaths/min, gallop rhythm, III/VI high-pitched blowing systolic murmur at the apex, hepatomegaly, and diminished pulses. Chest radiography documents an enlarged cardiac silhouette with pulmonary vascular congestion (image), and echocardiography demonstrates a regurgitant mitral valve with a dilated left ventricle and markedly reduced systolic contractility.

Of the following, the MOST likely cause for this child’s dilated cardiomyopathy is:

A. a congenital mitral valve abnormality  
B. Duchenne muscular dystrophy  
C. Friedreich ataxia  
D. rheumatic heart disease  
E. sickle cell disease
The causes of a dilated cardiomyopathy are diverse and include viral myocarditis, arrhythmias, metabolic conditions, muscle disorders, drug toxicities, and congenital cardiac lesions. Among the mitral valve disorders are abnormalities of the valve leaflets or chordal apparatus, which result in either acute neonatal symptoms or chronic, indolent findings that eventually degenerate. Mitral valve disorders can occur in isolation or as a part of a complex of left heart pathology that may include abnormalities of the aortic valve (e.g., bicuspid aortic valve with stenosis or regurgitation), aortic arch (coarctation of the aorta), and left ventricle (hypoplastic left heart syndrome). Mitral valve disease can result in either stenosis or regurgitation. Both can be tolerated by the child for long periods of time, but eventually they worsen and result in left atrial dilation, left ventricular dilation, and elevated pressure back into the lungs (pulmonary hypertension).

As the left ventricle continues to dilate, efficiency of contractility is reduced because fiber cross-linking exceeds the most optimal portion of the Starling curve, and eventually a dilated cardiomyopathy results. Accordingly, the boy in the vignette most likely has a congenital mitral valve abnormality and congestive heart failure (CHF).

Symptoms of CHF in older children include exercise intolerance, fatigue, dizziness or syncope, shortness of breath, palpitations, diaphoresis, abdominal discomfort, and anorexia. Among the signs are tachycardia, rales, poor tissue perfusion, hepatomegaly, and a gallop rhythm. Auscultation may identify a blowing systolic ejection murmur at the apex with radiation to the back from mitral regurgitation. Even in the absence of an intrinsic mitral valve disorder, mitral regurgitation can develop when left ventricular dilation causes a change in geometry that results in poor leaflet coaptation. Echocardiography is the gold standard for diagnosing both mitral valve disorders and a cardiomyopathic process. In children, the mitral valve can be seen in exquisite detail, allowing identification of pathologic features, as described for this boy. Color Doppler interrogation assists with determination of mitral regurgitation or stenosis. The left ventricular cavity can be assessed for chamber dilation, and quantitative determination of contractility is the routine part of echocardiographic assessment.

Duchenne muscular dystrophy often leads to the development of a dilated cardiomyopathy due to the adverse effect of the dystrophin mutation on the cardiomyocyte. However, the child in the vignette is younger than the typical child who develops left ventricular dysfunction (unusual before 10 years of age). In addition, most boys who have Duchenne muscular dystrophy and develop a dilated cardiomyopathy already manifest skeletal muscle involvement. Friedreich ataxia is associated with the development of a cardiomyopathy, but it is a hypertrophic rather than a dilated cardiomyopathy. In addition, a child who has Friedreich ataxia and advanced cardiac findings invariably exhibits ataxia. Mitral valve (as well as aortic valve) thickening and dysfunction are found in rheumatic heart disease. However, this child has none of the other cardinal features of acute rheumatic fever (dermatologic, infectious, neurologic, or joint). Sickle cell disease can result in high-output heart failure due to anemia, but this child does not have the features of a chronic disease state, which very likely would have developed by the age of 6 years.

Suggested Reading:

Madriago E, Silberbach M. Heart failure in infants and children. Pediatr Rev. 2010;31:4-12. DOI: 10.1542/pir.31-1-4

Source: 2012 PREP® Self-Assessment, Question 57
You are evaluating a 2-month-old infant in the emergency department whose parents state that she has had trouble breathing for the past week. The infant was born at term via vaginal delivery and had no prenatal or neonatal complications. The parents explain that for the past couple of days, she appears to be breathing fast and seems to “suck her chest in when she breathes.” Physical examination reveals a thin infant in moderate respiratory distress whose temperature is 37.0°C, heart rate is 150 beats/min, respiratory rate is 50 breaths/min, blood pressure is 74/48 mm Hg, and oxygen saturation is 94% in room air. Her lungs are clear to auscultation, but she has suprasternal and subcostal retractions. She also “bobs” her head with inspiration. The nurse was able to suction the nasopharynx by passing a suction catheter through each nostril, but there was no improvement in the infant’s respiratory status. You order chest radiography (image).

Of the following, the MOST likely cause of this infant’s symptoms is:

A. bronchomalacia  
B. choanal atresia  
C. mediastinal tumor  
D. pneumonia  
E. pneumothorax
Preferred Response: C

Airway obstruction, as described for the infant in the vignette, can be a life-threatening problem that can have a variety of causes, including anatomic abnormalities, trauma, malignancies, infection, and foreign bodies. Croup remains the leading cause of upper airway obstruction in children, followed by foreign bodies. Among the signs of airway obstruction are dyspnea, stridor, cough, gagging, retractions, or respiratory failure. The first priority in evaluating patients who have signs of airway obstruction is to determine the degree of obstruction by observing for audible sounds, effective air movement, and adequate respiratory effort. The infant in the vignette has clear evidence of respiratory distress, as evidenced by the presence of tachypnea and retractions. Chest radiography (Item C79A) demonstrates a widened mediastinum, marked shift of the trachea to the right, and normal lung fields, findings that are consistent with an intrathoracic mass. Follow-up computed tomography scan of the chest demonstrates a mediastinal mass causing severe narrowing of the trachea (Item C79B) and (Item C79C).

Acute respiratory failure due to a new-onset chest mass is uncommon but must be recognized quickly and managed appropriately to prevent fatal consequences. Clinical presentations can vary, depending on the degree of vascular and airway compression by the mass, but 60% of children present with respiratory symptoms. Superior vena cava compression results in head, neck, and upper extremity edema. Initial management is independent of the cause of the mass and consists of prompt attention to the potential for airway compromise, which can occur in up to 20% of patients.

Choanal atresia is the most common congenital anomaly of the nose and can present with a spectrum of breathing patterns ranging from noisy breathing to respiratory distress, but the ability to pass a suction catheter easily through each nostril excludes this diagnosis in this infant.

Bronchomalacia, loss of airway patency due to insufficient cartilage or compression, typically presents in infancy but is characterized by expiratory wheezing. Pneumonia and pneumothorax can both present with acute respiratory failure, but the presence of normal breath sounds on physical examination and results of chest radiography make these diagnoses unlikely.

Suggested Reading:
Gangadharan SP. Evaluation of mediastinal masses. UpToDate Online 18.3. 2010. Available online only for subscription

Loftis LL. Emergent evaluation of acute upper airway obstruction in children. UpToDate Online 18.3. 2009. Available online only for subscription


Source: 2012 PREP® Self-Assessment, Question 79
An 8-year-old girl presents for evaluation of “neck swelling.” Her mother reports that the swelling began approximately 1 year ago and has progressed with time. The swelling is not causing any pain. On physical examination, the girl’s thyroid gland is diffusely enlarged, has a cobblestone texture, but has no discrete nodules (image). Findings for all other systems are within normal parameters. Free thyroxine and thyroid-stimulating hormone measurements are normal.

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

A. assess thyroglobulin
B. assess thyroid peroxidase antibodies
C. assess thyroid-stimulating immunoglobulins
D. order neck ultrasonography
E. perform a thyroid biopsy
The girl described in the vignette has classic signs and symptoms of chronic lymphocytic thyroiditis (Hashimoto thyroiditis). Patients who have chronic lymphocytic thyroiditis are commonly euthyroid (have normal free thyroxine and thyroid-stimulating hormone values) for many years despite having other physical (goiter) or laboratory (autoantibody) signs of autoimmune thyroid disease. Although this girl has thyromegaly, as many as two thirds of patients who have Hashimoto thyroiditis have atrophic disease that results in small or even nonpalpable glands. The most sensitive laboratory test to confirm the presence of autoimmune thyroid disease is measurement of autoantibodies specific to thyroid antigens. In the case of Hashimoto thyroiditis, antibodies to thyroid peroxidase alone have greater than 85% sensitivity. Sensitivity increases to more than 90% if thyroglobulin autoantibodies are also measured.

Thyroglobulin concentrations (not to be confused with thyroglobulin antibody titers) simply reflect the total volume of thyroid tissue and are expected to be elevated in patients who have thyromegaly. Thus, measuring thyroglobulin is not helpful in determining the cause of thyromegaly. Thyroid-stimulating immunoglobulins are an excellent tool for assessing risk for Graves disease (which should result in a large, smooth, and firm gland) but are not specific for Hashimoto disease. On rare occasions, patients who have negative antibodies can still be proven to have Hashimoto thyroiditis by virtue of a biopsy and histologic review of thyroid tissue. However, unless the patient has a discrete nodule that is palpable on physical examination, neither ultrasonography nor thyroid biopsy is indicated during the initial evaluation.

Of note, patients who have any type of autoimmune thyroid disease are at increased lifetime risk for thyroid nodules and thyroid cancers compared with the general population.

Pain on palpation of the thyroid could be indicative of acute or subacute thyroiditis but is rarely noted in those who have Hashimoto thyroiditis.

Suggested Reading:


Source: 2012 PREP® Self-Assessment, Question 80
An 11-month-old boy presents to your office with a 5-day history of fever, nasal congestion, conjunctivitis, and the development of a rash over the past 24 hours. The rash began on his head and neck and spread to his trunk (image) and extremities. The family recently returned from a trip to Ireland. His past medical history is unremarkable, and his immunizations are up to date.

Of the following, the BEST test for diagnosing this child’s condition is:

A. measles immunoglobulin (Ig) M serology
B. nasal aspirate for viral culture
C. rubella IgM serology
D. skin biopsy
E. throat culture for group A *Streptococcus*
Visual Diagnosis Critique 6

Preferred Response: A

The 5-day prodrome of respiratory symptoms, conjunctivitis, and fever followed by development of a rash beginning on the head and neck and spreading to the trunk and extremities reported for the boy in the vignette is suggestive of measles. Malaise and lesions on the buccal mucosa (Koplik spots) (Item C82) just before rash development on the head and neck are other findings suggestive of clinical measles. The Centers for Disease Control and Prevention clinical definition for measles is an illness characterized by generalized rash lasting 3 days with temperature of 38.3°C, cough, coryza, or conjunctivitis and epidemiologic linkage to a confirmed case of measles. The rash of measles, which is due to immune complex deposition, may not occur in immunosuppressed hosts.

Most measles cases in the United States arise from importation from countries where the disease persists.

Many European countries, including Ireland, remain endemic areas for measles. In 2010, Ireland had the second highest incidence rate for measles in Europe. Although immunizations for the boy in the vignette are described as up to date, an 11-month-old child would not have routinely received a measles vaccine unless it was administered as part of the preparation for travel.

Measurement of specific measles immunoglobulin (Ig)M is the most reliable test for diagnosing measles and is available through local health departments with a rapid turnaround time. As a paramyxovirus, measles can be cultivated in tissue culture, but this is not routine in most clinical laboratories.

Rubella IgM serology can be used to diagnosis rubella, but this illness is generally milder than measles and does not have the extent of prodromal symptoms. A skin biopsy can detect IgG complexes of measles antigen but would not be a routine test used for diagnosing typical measles. The rash of scarlet fever may have morbilliform features, but the pattern of downward spread from the head and neck and the prodromal symptoms described are more consistent with measles than group A streptococcal infection.

Management of measles infection is supportive. Complications include otitis media, laryngotraceobronchitis (croup), bacterial tracheitis, bronchopneumonia, and diarrhea. In immunosuppressed children, severe bronchopneumonia may develop. Neurologic involvement may include acute encephalitis (~1 in 1,000 cases) or subacute sclerosing panencephalitis, a rare degenerative disease characterized by behavioral and intellectual deterioration and seizures that occurs 7 to 10 years after acute measles infection. Overall mortality from measles is 1 to 3 in 1,000 cases in the United States, with higher death rates in immunocompromised individuals.

Individuals who received a killed measles vaccine in the 1960s have developed atypical measles, characterized by distal extremity rash, nodular pneumonia, and neurologic symptoms, after exposure to wild-type virus.

Suggested Reading:

- Centers for Disease Control and Prevention. Measles (Rubeola): 2010 Case Definition. 2010. Available online only
- Centers for Disease Control and Prevention. Complications of Measles. 2009. Available online only
You are called to the newborn nursery to evaluate a term newborn for abdominal distention. The infant was born 12 hours ago by vaginal delivery. The pregnancy was complicated by gestational diabetes and polyhydramnios. Spontaneous rupture of the membranes occurred 1 hour before delivery with copious amniotic fluid that appeared to be lightly meconium-stained. The infant is breastfeeding well and has passed one stool since birth. Her mother describes the infant as spitting up about a teaspoon of “green stuff” twice in the past hour. Physical examination reveals an active, alert infant who has a moderately distended abdomen that is not tender to palpation. The rectum appears patent. You obtain a radiograph of the abdomen (image).

Of the following, the MOST appropriate next step is:

A. abdominal computed tomography scan
B. abdominal ultrasonography
C. contrast enema
D. lateral abdominal radiograph with the infant prone
E. upper gastrointestinal radiographic series
Preferred Response: E

The most appropriate imaging to assess an infant who has bilious emesis and an abdominal radiograph suggesting a proximal bowel obstruction, such as the newborn described in the vignette, is an upper gastrointestinal radiographic series (UGI). The infant in the vignette has a radiograph that demonstrates limited passage of air into the proximal small bowel (Item C106A). Bowel obstruction in the newborn is an emergency that requires rapid diagnosis. Antenatal clues may include polyhydramnios and bilious amniotic fluid. Careful attention to clinical findings can help distinguish between proximal and distal bowel obstruction, leading to selection of optimal imaging studies and management.

Proximal bowel obstructions include duodenal atresia, jejunoileal atresia, and malrotation with midgut volvulus. Infants who have duodenal atresia or jejunoileal atresia present with vomiting and abdominal distension. The vomiting may be bilious if the obstruction is distal to the ampulla of Vater. The abdominal radiograph of newborns who have duodenal atresia usually demonstrates the classic double-bubble sign (Item C106B), whereas newborns who have jejunoileal atresia often will have large dilated loops proximal to the obstruction. The classic presentation of a midgut volvulus is bilious vomiting, with an abdomen that may initially be soft until blood flow through the superior mesenteric artery is compromised and ischemia develops. The radiograph of the abdomen is often nonspecific except for dilated bowel loops. UGI should be obtained when the infant’s predominant symptom is bilious vomiting because midgut volvulus may be catastrophic, and early surgical intervention is essential.

Distal bowel obstructions include distal ileal atresia, colonic atresia, meconium ileus, meconium plug syndrome, Hirschsprung disease, and imperforate anus. Affected infants often present with abdominal distension and failure to pass meconium within the first 24 hours after birth; bilious vomiting is uncommon but may be seen. The abdominal radiograph in distal bowel obstruction shows dilated loops of bowel. Insipissated meconium or calcifications may be seen in meconium ileus. Contrast enema may be both diagnostic and therapeutic for meconium ileus and meconium plug syndrome. The finding of a transition zone on contrast enema suggests the diagnosis of Hirschsprung disease and the need for diagnostic rectal biopsy for ganglion cells. The absence of air in the rectum on abdominal radiograph may suggest an imperforate anus, for which a follow-up lateral abdominal radiograph with the infant prone permits assessment of the distal progression of gas and assists in determining the surgical plan.

Abdominal ultrasonography and computed tomography scanning have a very limited role in assessing congenital bowel obstruction in the neonate. Surgical consultation is often needed for definitive diagnosis and treatment.

Suggested Reading:

Source: 2012 PREP® Self-Assessment, Question 106
You are evaluating a 10-year-old boy for a rash and fatigue. He recently returned from visiting family in South Africa, where he experienced an illness characterized by fever and sore throat 2 weeks ago. On physical examination, he is afebrile and his heart rate is 100 beats/min, respiratory rate is 28 breaths/min, and blood pressure is 110/65 mm Hg. He has a macular, erythematous rash on his trunk (image). In addition, you note a III/VI blowing systolic murmur at the apex as well as a II/VI long diastolic murmur at the left lower sternal border. Twelve-lead electrocardiography reveals sinus rhythm with first-degree atrioventricular block. Echocardiography documents valve dysfunction.

Of the following, the MOST likely explanation for this child’s diastolic murmur is:

A. aortic valve insufficiency
B. aortic valve stenosis
C. mitral valve insufficiency
D. mitral valve stenosis
E. pulmonary valve stenosis
Preferred Response: A

The boy described in the vignette has two major (erythema marginatum and carditis) and one minor (prolongation of the PR interval) criteria for the diagnosis of acute rheumatic fever (ARF) (Item C127). In ARF, carditis (an inflammatory process that involves the myocardium, pericardium, and valves) can lead to chronic cardiac conditions of mitral (and occasionally aortic) regurgitation and stenosis. Carditis generally resolves with therapy, but many individuals who have ARF develop permanent damage to the aortic and mitral valves. The valves become thickened, which can lead to both reduced valve excursion (stenosis) and poor leaflet coaptation (regurgitation). The high-pitched diastolic murmur originating at the left sternal border and radiating to the apex described for the boy in the vignette is consistent with aortic insufficiency. An early systolic click often is associated with valve opening. Pulmonary valve stenosis is not associated with rheumatic heart disease.

Aortic valve stenosis leads to a systolic ejection murmur, which is typically best heard at the left sternal border (directly over the valve), with radiation to the neck and right infraclavicular region (along the pathway of the ascending aorta). Often, an audible systolic click is associated with the “snapping open” of the thickened valve leaflets. Mitral valve stenosis results in a diastolic murmur that is generally of low pitch, difficult to discern unless the stenosis is extreme, and best heard along the left axillary line. The murmur of pulmonary valve stenosis is systolic, is best heard along the right sternal border (over the valve), and radiates to the left infraclavicular region (along the path of the main pulmonary artery). An early systolic click often is associated with valve opening. Pulmonary valve stenosis is not associated with rheumatic heart disease.

Suggested Reading:
Cilliers AM. Rheumatic fever and its management. BMJ Clin Evid. 2006;333:1153-1156. DOI: 10.1136/bmj.39031.420637.BE. Available online only for subscription

Source: 2012 PREP® Self-Assessment, Question 127


Item C127. Jones Criteria for Diagnosis of Acute Rheumatic fever

<table>
<thead>
<tr>
<th>Major Criteria</th>
<th>Minor Criteria</th>
<th>Supporting Evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Carditis</td>
<td>Clinical findings</td>
<td>Positive throat culture or rapid test OR Elevation of streptococcal antibody test</td>
</tr>
<tr>
<td>Polyarthritis</td>
<td>Fever, arthralgia</td>
<td>Elevation of acute phase reactants; prolonged PR interval</td>
</tr>
<tr>
<td>Chorea</td>
<td>Laboratory findings:</td>
<td>Elevation of streptococcal antibody test</td>
</tr>
<tr>
<td>Erythema marginatum</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Subcutaneous nodules</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

A 14-year-old girl first developed redness across the bridge of her nose and on her cheeks about 3 months ago. Subsequently, the rash spread to involve more of her face as well as her hands. She now reports increasing fatigue and some weakness; she has difficulty climbing stairs and has needed help brushing her hair. She has had no fever or joint swelling. Physical examination of the appropriately developed teen reveals normal vital signs; a heliotrope rash involving the eyelids, nasal bridge, and cheeks (image A); and erythematous scaling papules on the extensor surfaces of her metacarpal, phalangeal, and interphalangeal joints (image B). She has weakness and tenderness of the proximal musculature and must employ the Gower maneuver to arise from sitting.

Of the following, the test that is MOST likely to contribute to the diagnosis is:

A. cranial computed tomography scan
B. edrophonium (Tensilon®) test
C. electroneurography
D. magnetic resonance imaging of proximal muscles
E. skin biopsy
Preferred Response: D

The girl described in the vignette has the typical facial rash of juvenile dermatomyositis (JDM): a red-to-violet masklike rash involving the cheeks and eyelids and sometimes extending to the ears (ie, heliotrope). She also exhibits Gottron papules, flat-topped erythematous hyperkeratotic papules usually overlying metacarpophalangeal or interphalangeal joints (Item C195). JDM is the most common of the inflammatory myopathies affecting children and adolescents, with an estimated incidence of approximately 2 to 4 cases/million children per year. Diagnostic criteria developed in 1975 require the presence of three or more criteria plus the characteristic rash for a definite diagnosis of JDM or two criteria plus rash for a probable diagnosis. The criteria include: proximal muscle weakness (sparing facial and eye muscles), elevated muscle enzyme values, myopathic electromyography (EMG), and characteristic findings on muscle biopsy.

Recently, a greater understanding of the pathogenesis and the availability of newer technology has changed the diagnostic approach, and efforts are underway to revise the diagnostic criteria. A survey of rheumatologists throughout the world showed that only about 60% of practitioners used EMG or muscle biopsy, opting instead for less invasive testing to confirm the diagnosis. Magnetic resonance imaging of proximal muscles indicating inflammatory muscle changes was the next most useful diagnostic test, according to rheumatologists. Additional tests, including searches for p155 antigen, myositis-specific and -related antibodies, and von Willebrand antigen, may be used, but these factors are not consistently found in each patient. Major histocompatibility screening also finds the presence of HLA-DQA1*0501 in more than 80% of affected patients, but its presence is not required for diagnosis.

The edrophonium test is used in the diagnosis of myasthenia gravis. In that condition, extraocular muscle involvement is the most prominent finding and rash is not a typical component. Nerve conduction studies might be useful in the evaluation of peripheral nerve disease, which manifests with greater distal than proximal weakness. Cranial computed tomography scan might be useful in the diagnosis of centrally caused conditions, but elevated muscle enzyme values described for this girl make myopathy a more likely diagnosis. A skin biopsy might be performed if the rash is atypical, but rheumatologists in a recent survey rated skin biopsy as not usually helpful or necessary for the diagnosis of JDM.

Suggested Reading:
Feldman BM, Rider LG, Reed AM, Pachman LM. Juvenile dermatomyositis and other idiopathic inflammatory myopathies of childhood. Lancet. 2008;371:2201-2212. DOI: 10.1016/S0140-6736(08)60955-1

Source: 2012 PREP® Self-Assessment, Question 195

Item C195
A 10-year-old girl presents with right wrist pain. According to her mother, the girl has been complaining of this pain for 2 weeks, and the pain has persisted despite administration of daily ibuprofen. The girl cannot recall any recent injury and has had no fever or other systemic symptoms. She is right-handed and participates in gymnastics and soccer. Physical examination of her wrist reveals no redness or swelling, but she has moderate tenderness diffusely over the radial aspect and pain with wrist flexion and extension. She has no snuff-box tenderness and her hand is neurovascually intact. You obtain radiographs of the wrist (image).

Of the following, the MOST likely explanation for this child's wrist pain is:

A. a navicular fracture
B. a sprain
C. an overuse injury
D. osteomyelitis
E. septic arthritis
Preferred Response: C

Overuse injury of the distal radial physis is a well-recognized condition among young gymnasts and the most likely diagnosis for the girl described in the vignette. It is believed that the chronic, repetitive physical loading on immature physeal cartilage of the distal radius causes physeal ischemia that, if unaddressed, may lead to osseous necrosis. Although most of these stress injuries resolve with rest and do not result in permanent damage, several case reports documenting premature partial or complete physeal closure with radial growth arrest in these young athletes have raised concern.

The prevalence of radiographically documented stress-related physeal injuries ranges from 10% to 85% of young gymnasts studied. The findings typically include irregular widening of the physes, thickening of the zone of provisional calcification, and subchondral sclerosis, as are seen in the radiograph from the patient in the vignette. Radiographs in patients who have navicular fractures may be normal initially, but after 2 weeks, they demonstrate fracture healing and callus formation. Sprains and septic arthritis would not be expected to produce radiographic changes, and osteomyelitis is clinically unlikely with the absence of systemic symptoms.

Clinically, athletes who have overuse injuries initially present with chronic wrist pain on weight-bearing and progress to decreased wrist range of motion. Physical symptoms precede radiographic changes. Experts suggest that early evaluation of pediatric athletes who develop joint pain is important in identifying stress injuries so that appropriate interventions, including rest until symptoms resolve, modification of training regimens, and strengthening/conditioning routines, can be implemented.

Suggested Reading:


Source: 2012 PREP® Self-Assessment, Question 210
A 14-year-old boy presents with fever, joint aches, and a rash for the past 3 days. He was started on amoxicillin for sore throat a week ago. One of his classmates had a recent diagnosis of strep throat. He has a history of being successfully treated for acute otitis media with amoxicillin a month ago and has been well in the interim. On physical examination, he has swollen lymph nodes, periarticular swelling, and dusky urticaria-like lesions on his skin (image).

Of the following, these findings are MOST likely due to:

A. amoxicillin allergy
B. infectious mononucleosis
C. penicillin allergy
D. scarlet fever
E. serum sickness–like reaction
Visual Diagnosis Critique 11

Preferred Response: E

This teenager has the symptoms of a serum sickness–like reaction to amoxicillin. Serum sickness–like reactions typically involve a constellation of signs and symptoms, which can include arthralgias, lymphadenopathy, and urticarial rash with or without fever. Fever, when present, is typically low-grade. Children with serum sickness–like reactions may present with acute onset of joint pain that often leads to inability to walk. The most characteristic rash is an urticarial or serpiginous macular rash that starts in the anterior lower trunk, groin, periumbilical, or axillary regions, and spreads to involve the back, upper trunk, and extremities. The rash generally lasts a few days to 2 weeks. Ulcers, secondary infection, and scarring do not occur. It has been suggested that the term “serum sickness–like disease” should be replaced by “urticaria with arthritis” to describe this drug-induced syndrome, although this has not become common practice.

In contrast, the cardinal features of classical serum sickness are rash, fever, and polyarthralgia or polyarthritides, which begin to 2 weeks after first exposure to the responsible agent and resolve within a few weeks of discontinuation. Although patients may appear very ill and uncomfortable during the acute febrile stage, the disease is self-limited and prognosis is excellent once the responsible drug is stopped. During classic serum sickness, signs of mild renal dysfunction may be evident; however, renal involvement is unusual in serum sickness–like reactions caused by medications.

Serum sickness may develop more rapidly and severely if a previously immunized patient is reexposed to the culprit antigen. Rather than requiring 7 to 14 days for the development of IgM antibodies, the anamnestic IgG response can begin within 12 to 36 hours. Drugs, particularly antibiotics, are the leading cause of serum sickness–like reactions to antibiotics, cefaclor, and trimethoprim-sulfamethoxazole are most commonly implicated, although many drugs have been associated with these reactions. In children, serum sickness–like reactions are about 15-fold more likely with cefaclor than with other cephalosporins or amoxicillin, even though all are structurally similar β-lactam antibiotics.

Other drug reactions that may mimic serum sickness or serum sickness–like reactions include nonspecific exanthems, urticaria, and generalized hypersensitivity reactions. The development of an IgE-mediated drug allergy can cause the onset of urticaria during a course of therapy. Other symptoms of Type I, IgE-mediated allergic reactions are pruritus, flushing, angioedema, wheezing, laryngeal edema, abdominal distress with emesis or diarrhea, and hypotension. Symptoms usually appear within minutes to hours of drug administration and then escalate rapidly. This child does not have symptoms of penicillin or amoxicillin IgE-mediated allergy.

In general, penicillin is the most commonly reported medication allergy and is self-reported by at least 10% of patients. However, in large-scale studies, approximately 85% to 90% of these individuals are found to not be allergic and are able to tolerate penicillins. Patients may become allergic to the β-lactam ring structure that is common to all penicillins, or to the R-group side chains that distinguish different penicillins. Penicillin skin testing is the preferred method of evaluation of possible type I IgE-mediated penicillin allergy. The positive predictive value of penicillin skin testing is variable with approximately 50% (range, 25%-100%) of skin test-positive patients developing immediate allergic reactions upon being challenged with penicillin. However, the negative predictive value of penicillin skin testing is very high. Serious immediate-type reactions in patients challenged with penicillin after negative penicillin skin testing have not been reported. In vitro tests (radioallergosorbent tests or enzyme-linked immunosorbent assay assays) for penicillin, amoxicillin, and ampicillin are commercially available, but these assays are of limited clinical use because their predictive values have not been defined.

Some patients form IgE antibodies that can recognize the R-group side chains rather than the core ring structure. These individuals develop immediate-type reactions to amoxicillin or ampicillin but are able to tolerate penicillin; these patients are said to be selectively allergic to these aminopenicillins. It is rarely possible to distinguish these 2 types of allergy without skin testing, so patients allergic to penicillin should be advised to avoid all penicillins until they can be definitively evaluated. In the United States, however, selective IgE-mediated allergy to the aminopenicillins appears to be uncommon (<1%).

The child does not have features consistent with scarlet fever. The rash of scarlet fever generally occurs in association with pharyngitis requiring prior exposure to Streptococcus pyogenes. The rash is described as a diffuse erythema that blanches with pressure and numerous small (1-2 mm), papular elevations that give a “sandpaper” quality to the skin. While scarlet fever with pharyngitis can predispose to acute rheumatic fever, joint aches and pains are typically not observed during the course.

Infectious mononucleosis (IM) classically occurs in adolescents and is characterized by fever, severe pharyngitis, and anterior and posterior cervical or diffuse lymphadenopathy. Prominent constitutional symptoms include fatigue, anorexia, and weight loss. Patients who are treated with ampicillin or amoxicillin may develop a characteristic rash described as maculopapular (Item C3). The incidence of rash may be as high as 70% to 90%. The mechanism responsible for the rash is not well understood. Development of a drug-related rash during IM does not appear to indicate a true drug allergy because patients subsequently tolerate ampicillin without an adverse reaction. This child has symptoms for only a few days; his rash is urticarial rather than maculopapular, making IM less likely.

Suggested Reading:
Joint Task Force on Practice Parameters; American Academy of Allergy, Asthma and Immunology; American College of Allergy, Asthma and Immunology; Joint Council of Allergy, Asthma and Immunology. Drug allergy: an updated practice parameter. Ann Allergy Asthma Immunol. 2010;105(4):259-273

Source: 2013 PREP® Self-Assessment, Question 3

Item C3
An 18-month-old boy is brought to your clinic because of redness of his left cheek that his mother noticed yesterday. He has been otherwise well and has had no fever. Physical examination findings reveal a tender, erythematous area of slight induration on his left cheek (image). His mother tells you that the boy recently started sucking ice chips.

Of the following, the MOST likely diagnosis is:

A. cellulitis
B. cold panniculitis
C. dental abscess
D. juvenile xanthogranuloma
E. lipoma
Preferred Response: B

Cold panniculitis, also known as “popsicle panniculitis,” is a lesion of the skin caused by prolonged exposure to cold objects. Sucking on popsicles or ice chips results in the most common presentation on the cheeks, but any area of the body exposed to cold can be affected. The mechanism of pathogenesis is similar to that of fat necrosis and likely occurs because in infants the subcutaneous fat is more apt to solidify than in adults.

Typical lesions are bluish red nodules which may be painful. They usually arise within 1 to 2 days of cold exposure, and they may persist for several weeks. Although rarely required for diagnosis, histologic examination reveals histiocytic and lymphoid cells within fat lobules. Because the lesions resolve within weeks, no treatment is necessary. However, it is advisable to avoid cold exposure if possible.

Cellulitis is suppurative inflammation of the skin caused by bacteria, such as Streptococcus pyogenes and Staphylococcus aureus. A child who has cellulitis is often febrile, and his skin is very tender, erythematous, and warm to the touch. A dental abscess could cause painful swelling of the cheek, but fever and tenderness over the gingiva and affected tooth would be expected. Juvenile xanthogranuloma is characterized by yellowish or brownish nodules on the skin (Item C10), and the lesions are typically present from birth or early infancy. A lipoma, a benign tumor of fat cells, may be found anywhere on the body but typically does not cause tenderness or skin discoloration.

Suggested Reading:


Source: 2013 PREP® Self-Assessment, Question 10
A 2-year-old boy you have followed up since birth presents to your office for a health supervision visit. He was born at term and has had normal growth and development. The mother recently noticed that he looks “cross-eyed” at times. There is no family history of eye abnormalities. On physical examination, his left eye is deviated nasally with a white pupillary reflex (image). There is no proptosis, conjunctival injection, or pain with eye movements. The remainder of results for the physical examination is normal. Results of the complete blood count are within normal limits.

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

A. bilateral retinoblastoma  
B. congenital cataract  
C. leukemia with chloroma  
D. neuroblastoma  
E. unilateral retinoblastoma
Preferred Response: E

One of the most common causes of leukocoria (white pupillary reflex) in children is retinoblastoma (47% of cases); therefore, all children with a new finding of leukocoria should be referred immediately to an ophthalmologist who is experienced in examining children. Other causes include persistent fetal vasculature, retinopathy of prematurity, cataracts, optic disc abnormalities, uveitis, or vitreous hemorrhage.

Retinoblastoma is the most common intraocular tumor of childhood, occurring in approximately 1 in 15,000 live births. The annual incidence is 11 per 1 million children from birth to 4 years old and 0.6 per 1 million children 5 years old and older. The average age at the time of diagnosis is 2 years in unilateral cases and 1 year in bilateral cases. Leukocoria and strabismus are the most common presenting findings. Other findings may include decreased vision, ocular inflammation, vitreous hemorrhage, hyphema, orbital cellulitis, proptosis, glaucoma, eye pain, fever, and a family history of retinoblastoma. The diagnosis of retinoblastoma is usually made on the basis of clinical examination, and the presence of calcification on computed tomography or ocular ultrasonography.

In this vignette, the age of the child, the lack of findings in right eye, and the relatively higher frequency of unilateral disease over bilateral disease make unilateral retinoblastoma the most likely diagnosis in this patient. Leukemia with chloroma, or intraocular leukemic infiltration, is a less likely diagnosis given the normal blood cell count. Neuroblastoma can present in this age group but is not associated with leukocoria. Ocular findings typically seen in neuroblastoma include Horner syndrome (miosis, ptosis, enophthalmos, and anhydrosis), periorbital hemorrhage, proptosis, papilledema, retinal hemorrhage, strabismus, and opsoclonus. Congenital cataracts are also a common cause of leukocoria but are usually apparent at birth or early infancy (Item C12). Cataracts can be associated with congenital infections or various systemic disorders (eg, diabetes mellitus, galactosemia, Turner syndrome, Down syndrome, peroxisomal disorders).

Suggested Reading:

Kaufman PL, Saunders RA. Approach to the child with leukocoria. In: Basow DS, ed. UpToDate Online. Waltham, MA: UpToDate; 2011. Available online only for subscription


Source: 2013 PREP® Self-Assessment, Question 12
A 4-year-old girl enters your practice for the first time. Her mother reports that her birth history was unremarkable and early developmental milestones were within normal limits. However, over the past year her development has seemed to plateau. Her medical history is significant for recurrent ear infections and, more recently, rather loud snoring. On physical examination, you note slightly coarse facial features, a protuberant abdomen with a small reducible umbilical hernia, enlarged liver, and mild contractures of the fingers (image).

Of the following, according to these findings, the test that would be MOST diagnostically helpful is:

A. a lipid profile  
B. plasma amino acids  
C. a sleep study  
D. urine glycosaminoglycans  
E. urine organic acids
The clinical findings described in this vignette are typical for a child with a form of mucopolysaccharidosis (MPS) disorder. Development for many children with an MPS disorder is initially normal, subsequently plateaus, and then regresses because of accumulation of metabolic by-products (glycosaminoglycans). This is typical of some MPS subtypes, including Hurler, Hunter, and Sanfilippo syndromes. Diagnostic screening includes urine testing for glycosaminoglycans and definitive testing for specific enzymes, depending on the findings on the urine test. Recurrent otitis media and snoring secondary to tonsillar and adenoidal hypertrophy are also quite common early features. Later findings include bony changes (dysostosis multiplex), joint contractures (especially in the fingers), hepatosplenomegaly, and secondary umbilical hernias. The child in this case is most likely affected with Hurler syndrome or a slightly milder variant known as Hurler-Scheie syndrome, both of which are associated with deficiency of the enzyme α-L-iduronidase and are inherited in an autosomal recessive manner. Hunter syndrome primarily affects boys because it is inherited as an X-linked recessive condition. Sanfilippo syndrome is a clinically distinct condition associated with a later onset of organomegaly. Classic phenotypic findings, such as the coarsening of facial features and hirsutism seen earlier with Hurler and Hunter syndromes, occur later in Sanfilippo syndrome. Enzyme replacement therapy has been developed for some forms of MPS, including Hurler and Hurler-Scheie, and results in decreased organomegaly, improvement in respiratory function, and resolution of other somatic features. Current enzyme therapy is ineffective in improving cognitive function because these intravenously infused enzymes do not cross the blood-brain barrier.

A lipid profile would not be helpful in determining a diagnosis in this child. A sleep study may demonstrate or uncover obstructive sleep apnea but would not explain the other clinical findings that point to an MPS disorder. Plasma amino acids and urine organic acids are studies used to diagnose other types of inborn errors of metabolism, but these conditions would not present with coarse facies, finger contractures, or an umbilical hernia.
A 14-year-old girl comes in because of a dark rash around her neck that she first noticed 4 or 5 months ago. She has been otherwise healthy. The girl had menarche at 11 years of age, and her periods are fairly regular. Findings on physical examination are normal except for a body mass index of 28.2 and a velvety thickening of the skin on the sides and nape of her neck (image).

Of the following, the MOST appropriate laboratory evaluation for this patient is:

A. antinuclear antibody and rheumatoid factor
B. antithyroglobulin antibody
C. fasting blood glucose and insulin levels
D. potassium hydroxide preparation of a skin scraping
E. testosterone, follicle-stimulating hormone, luteinizing hormone
Preferred Response: C

Acanthosis nigricans (AN) in children and adolescents is most often a manifestation of obesity-related insulin resistance, and its incidence is increasing as the obesity rate among US children increases. AN may be the result of stimulation of receptors, particularly insulin and insulin growth factor-1 receptors, on keratinocytes and dermal fibroblasts by circulating serum insulin. The clinical appearance is of hyperpigmented, hypertrophic, velvety skin first appearing in the neck or axillae, but it also may be seen in the groin and under the breasts. Additional sites may include the flexor surfaces of the extremities, the umbilicus, the face, and perioral and perianal areas. Occasionally, it can involve mucosal surfaces, although this may signal a more unusual underlying condition.

While the majority of affected pediatric patients have obesity-associated AN, there are other categories of the condition. Benign AN is transmitted as an autosomal dominant trait with variable penetrance, and it is unclear as to whether insulin resistance occurs in these children. The onset may be at birth or during childhood or adolescence, and it may be unilateral initially and progress to a more generalized distribution. AN is associated with a number of rare and diverse conditions, including polycystic ovary disease; hypergonadism, insulin resistance, and acanthosis nigricans (HAIR-AN) syndrome; Prader-Willi syndrome; and Bardet-Biedl syndrome (the third suggested reading [Sinha and Schwarz] provides a more complete list of syndromes in which AN is a feature). Fortunately, AN that is associated with malignancy is rare in pediatrics. In this setting, AN has a rapid onset and commonly involves the mucous membranes. AN is most often associated with gastric adenocarcinoma but has been reported in children who have Wilms tumor, osteogenic sarcoma, thyroid papillary carcinoma, and adrenocortical carcinoma. Drug-induced AN has been attributed to nicotinic acid, corticosteroids, methyltestosterone, oral contraceptives, heroin, and some anticonvulsants.

A recent study among primarily minority youth found that 29% of 8- to 14-year-olds who had acanthosis nigricans had abnormal glucose homeostasis, 27% had elevated systolic blood pressure, and 50% had reduced high-density lipoprotein cholesterol concentrations. Even after correction for body mass index, AN was significantly related to abnormal glucose homeostasis. In another study of children and adults, AN predicted the presence of type 2 diabetes mellitus independent of any other risk factor, and AN was associated with elevated fasting insulin levels.

For the patient in the vignette, evaluation of fasting blood sugar and fasting insulin levels would be useful to assess for insulin resistance. Both systemic lupus erythematosus and Hashimoto thyroiditis may occasionally be associated with AN, but, in the absence of other symptoms, this evaluation is unlikely to be helpful. AN is commonly associated with hyperandrogenic states, including polycystic ovary disease, and evaluation for this condition could be considered. However, in the absence of signs of hyperandrogenism (eg, hirsutism) and in the presence of regular menstrual periods, obtaining testosterone, follicle-stimulating hormone, and luteinizing hormone concentrations is not indicated. Since AN does not exhibit an elevated, scaling border, performance of a potassium hydroxide preparation to evaluate for tinea corporis is unnecessary.

Suggested Reading:


Source: 2013 PREP® Self-Assessment, Question 35
You are evaluating a 16-year-old girl who has a 5-day history of progressive cough, shortness of breath, and fever. Her vital signs include a temperature of 39.0°C, a heart rate of 100 beats/min, and a respiratory rate of 35 breaths/min. On physical examination, she has mild respiratory distress, prolonged expiration, and decreased breath sounds over the left chest. You obtain a chest radiograph (image) and decide to admit her to the hospital for further treatment.

Of the following, the MOST appropriate empiric antibiotic regimen to order is:

A. ampicillin and gentamicin
B. azithromycin
C. ceftriaxone
D. cefuroxime and nafcillin
E. doxycycline
Preferred Response: C

Pneumonia (inflammation of the lung) includes both infectious (bacterial, viral, fungal, and parasitic) and noninfectious (aspiration, hydrocarbon, and drug induced) causes. It is one of the most frequently encountered pediatric medical problems, with an estimated annual incidence of 150 million cases worldwide for children less than 5 years of age; the incidence decreases in adolescents. North American providers are estimated to see 5 to 20 cases per year depending on the age range of their patient population. Bacteria cause 20% to 50% of community acquired pneumonias, and the majority of patients recover uneventfully with appropriate treatment. Approximately 10% of patients require hospital admission, and an even smaller percentage of patients develop pleural and parenchymal complications, such as empyema, lung abscess, necrotizing pneumonia, pneumothorax, and bronchopleural fistulas.

The girl described in the vignette has signs and symptoms consistent with a significant respiratory process, including decreased aeration, tachypnea, and prolonged expiration. Her chest radiograph demonstrates a large left-sided pleural effusion and lobar pneumonia that, when combined with her history and physical examination findings, are most consistent with a bacterial infection. Ultrasonography demonstrated a large pleural effusion with multiple septations (Item C47).

In cases of suspected bacterial pneumonia, prompt antibiotic treatment is crucial and the agent(s) selected should cover the most common pathogens, taking into consideration the age of the patient, existence of underlying medical conditions, and local epidemiology. The introduction of pneumococcal and Haemophilus influenzae type B conjugate vaccines has decreased the incidence of invasive disease caused by these bacteria. Prior to the introduction of these vaccines, Streptococcus pneumoniae was estimated to cause up to 44% of bacterial pneumonias, and although less common today, it still represents the major pathogen. Chlamydia pneumoniae and Mycoplasma pneumoniae cause “atypical pneumonia” in 3% to 23% of infants and older children, respectively.

Recent studies have shown a rise in patients with empyema despite the decrease in pneumococcal pneumonia. Staphylococcus aureus, with the majority being methicillin resistant, is partly responsible for the increase, as some studies have shown this organism to be isolated more frequently from patients with empyema than S pneumoniae. Streptococcus pyogenes (group A streptococcus) is a less frequent cause but can be associated with severe necrotizing pneumonia.

Evidence-based guidelines for outpatient and inpatient management of infants and children with community-acquired pneumonia (CAP) have been published. Because of its excellent activity against S pneumoniae, amoxicillin remains the recommended initial outpatient therapy for infants, children, and adolescents who are fully immunized and who have mild to moderate community-acquired pneumonia. Drug allergy or suspicion of atypical pathogens (such as Mycoplasma pneumoniae or Chlamydia pneumoniae) will dictate use of alternative antibiotics such as a second- or third-generation cephalosporin or a macrolide, respectively.

For patients requiring inpatient care, ampicillin or penicillin G is indicated when there is a locally documented lack of high-level penicillin resistance for invasive S pneumoniae. Empiric therapy with a third-generation parenteral cephalosporin (ceftriaxone or cefotaxime) is appropriate for hospitalized infants and children who have complicated pneumonias (such as the girl in the vignette with probable empyema), for infants who are not fully immunized, or in areas where high-level penicillin resistance of pneumococcal strains is present. If S aureus is suspected, vancomycin or clindamycin should be added. Vancomycin has not been shown to be more effective than third-generation cephalosporins in the treatment of pneumococcal pneumonia in North America. If an atypical pathogen is suspected, then an oral or parenteral macrolide should be added to the cephalosporin regimen, but this would be an inadequate monotherapy in this patient. Ampicillin plus gentamicin, cefuroxime plus nafcillin, and doxycycline alone are not appropriate antibiotic regimens for this patient.

Suggested Reading:


A 6-year-old girl is brought to the clinic because of vulvar and anal itching over the last week. Her mother denies vaginal discharge or stool abnormalities. Physical examination findings reveal mild anal erythema and a normal vulva with no vaginal discharge. Microscopic examination of tape placed on the anus reveals the structures shown (image).

Of the following, the MOST appropriate pharmacologic agent to use is:

A. albendazole  
B. anidulafungin  
C. benznidazole  
D. ciprofloxacin  
E. itraconazole
Preferred Response: A

Anal and vulvar itching can be caused by numerous conditions, including infections, local irritation, and various forms of dermatitis. In children, infection with the roundworm Enterobius vermicularis, or pinworm, is a common cause. Adult female pinworms reside in the large intestine and migrate to the perianal and perineal area at night, depositing up to thousands of eggs on the skin (Item C61). Transmission of the eggs from one person to another occurs via the fecal-oral route. Eggs can survive up to 3 weeks and are ingested from fingernails, bedding, toys, or other surfaces; autoinfection occurs as well.

The diagnosis of pinworm infection can be made by either visualizing the adult worms on the perineum at night or placing a piece of transparent tape over the anal area and then examining it microscopically at low power. These tape specimens should ideally be collected over 3 consecutive mornings. Microscopic examination of stool does not usually reveal the eggs.

Three drugs may be used to treat pinworm infection: mebendazole, pyrantel pamoate, and albendazole. For all 3 drugs, a single dose is administered and then repeated in 2 weeks. Parents should also be advised of the risk of reinfection, so morning bathing, frequent washing of bedclothes, and excellent hand washing should be recommended. Anidulafungin is used to treat candidal infections. Benznidazole is used in the treatment of trypanosomiasis. Ciprofloxacin is an antibiotic, and itraconazole is an antifungal medication: neither is effective in treating pinworm infections.

Suggested Reading:


Source: 2013 PREP® Self-Assessment, Question 61

A 16-year-old girl presents with a 4-day history of fever, chills, nonproductive cough, and sore throat and a maculopapular truncal rash. On physical examination, she is well appearing. Her temperature is 39.1°C, her respiratory rate is 24 breaths/min, and her pulse rate is 76/min. Examination of the girl’s head, eyes, ears, nose, and throat reveals an erythematous pharynx without exudates. Her neck is supple and without lymphadenopathy. Respiratory auscultation reveals scattered crackles in her lungs, and cardiac auscultation shows no murmur, rub, or gallop. Her abdomen is without organomegaly, her extremities are without lesions, and results of her neurologic examination are within reference range. There is a pink maculopapular rash primarily on her trunk. You obtain a chest radiograph (image).

Of the following, the BEST test for confirming the cause of this child’s pneumonia is:

A. blood culture
B. cold agglutinins
C. nasopharyngeal aspirate for viral antigens
D. sputum Gram stain and culture
E. throat swab for mycoplasma polymerase chain reaction
Preferred Response: E

The combination of pharyngitis, nonproductive cough, chills, scattered rales, and skin rash in conjunction with the girl’s age and the presence of bilateral infiltrates on chest radiograph is typical of symptomatic Mycoplasma pneumoniae infection. Detection of mycoplasma DNA by polymerase chain reaction on throat swab specimens has recently been demonstrated to be a sensitive and specific method for diagnosing M pneumoniae respiratory infections. The test is becoming increasingly clinically available. If not available, mycoplasma IgG and IgM serology can be obtained. Routine isolation of the organism is not readily available.

It is unlikely that the girl in the vignette has “typical” bacterial pneumonia, such as pneumonia caused by Streptococcus pneumoniae. Additionally, bacteremia is unlikely with bacterial pneumonia at this age. Although cold agglutinins are frequently sent for the diagnosis of Mycoplasma infection, they are neither adequately sensitive nor specific. Therefore, cold agglutinins are of no value and should not be ordered. Nasopharyngeal aspirate for viral antigens would be an appropriate test for detecting respiratory viruses. Although viral infections may cause many of the same symptoms as described in the vignette, the chills, the high fever, and the radiographic findings of bilateral patchy infiltrates are consistent with Mycoplasma infection at this age.

Suggested Reading:


Source: 2013 PREP® Self-Assessment, Question 161
You are examining a term male newborn one day after birth. The pregnancy and birth were uncomplicated. Physical examination is remarkable only for an erythematous patch involving his right forehead, eyelid, and cheek (image). His parents are concerned this could be a “serious birthmark” and ask you whether it will resolve.

Of the following, the statement you are MOST likely to make is:

A. he is at risk for intracranial vascular anomalies
B. he will likely develop other skin lesions such as café au lait macules
C. systemic steroids will be effective in reducing the lesion
D. the lesion will likely worsen initially and then resolve over several years
E. thrombocytopenia is a common finding with this type of skin lesion
Preferred Response: A

The newborn described in the vignette has an erythematous patch that is most consistent with the diagnosis of capillary malformation, also called port-wine stain or nevus flammeus. These lesions are true vascular malformations and therefore are present throughout life (growing with the child proportionately) and are in contrast with hemangiomas, which are usually not present at birth and typically involute after a period of growth.

Capillary malformations may occur anywhere on the body, but those on the face, especially in the distribution described in the boy in the vignette, warrant special attention because up to 10% of patients with V1 distribution are found to have Sturge-Weber syndrome (SWS). SWS is the triad of a facial (V1) capillary malformation, ipsilateral leptomeningeal angiomatosis, and choroidal vascular malformation of the eye. Other neurological problems such as seizures and intellectual disability are common.

Consequently, the infant described in the vignette should undergo evaluation for SWS, which includes magnetic resonance imaging with contrast of the brain and ophthalmologic evaluation; his parents should be informed of his risk of seizures.

Pulsed dye laser therapy is the treatment of choice for capillary hemangiomas, regardless of whether SWS is present. Systemic steroids are not of value, although they may play a role in the management of aggressive hemangiomas. Café au lait macules are commonly seen in patients with other neurocutaneous syndromes, such as neurofibromatosis, but they are not generally associated with capillary malformations. Thrombocytopenia is seen in Kasabach-Merritt syndrome, which is associated with “atypical-appearing” hemangiomas (actually tufted angiomas or hemangioendotheliomas), but it is not associated with SWS or with capillary malformations in general.

Suggested Reading:


Source: 2013 PREP® Self-Assessment, Question 163
You are called to assess a newborn who has pallor and tachycardia immediately after birth. The infant was born at term by vacuum-assisted vaginal delivery because of a prolonged second stage of labor and maternal exhaustion. She emerged limp with a weak cry and required 30 seconds of positive pressure ventilation to improve respiratory effort and heart rate. On physical examination immediately after birth, she has a temperature of 37.0°C, heart rate of 190 beats/min, respiratory rate of 60 breaths/min, and blood pressure of 54/26 mm Hg. She has significant pallor with delayed capillary refill, tachycardia with a gallop, and a fluctuant boggy mass over the posterior occiput (image). Her saturation by pulse oximetry is 95% on room air.

Of the following, the MOST appropriate initial test to order immediately is:

A. a computed tomography scan of the head
B. a hematocrit
C. a plain radiograph of the skull
D. an electrocardiogram
E. an electroencephalogram
Preferred Response: B

The history and clinical presentation of the infant described in the vignette suggest a subgaleal hemorrhage (SGH) with resultant hypotensive shock. A test for hematocrit is needed immediately to determine the requirement for packed red blood cell transfusion. The acute blood loss and hypovolemia are a medical emergency requiring rapid assessment and management. SGH occurs after the rupture of the emissary veins that course between the periosteum of the skull and the galea aponeurotica. This potential space is estimated to be able to contain 50% to 80% of an infant’s blood volume. On physical examination of the head, the typical finding is a dependent, fluctuant mass (often associated with a fluid wave) that is not confined by suture lines. The risk of SGH is increased with vacuum-assisted deliveries that involve multiple applications and pop-offs of the vacuum cup. Some estimate that half these infants may have an associated intracranial hemorrhage (ICH) or skull fracture. In contrast to SGH, isolated ICH is often asymptomatic and can be seen in up to a quarter of term neonates born by vaginal delivery. ICH may be subdural, subarachnoid, intraventricular, or intraparenchymal. Of these, subarachnoid hemorrhage is most commonly found in the symptomatic term infant, whereas subdural hemorrhage is seen most often in the asymptomatic term infant. Clinical findings of apnea, seizure, or unexplained hypotonia in a term infant should prompt the clinician to consider the possibility of ICH. Noncontrast computed tomography (CT) scan remains the preferred first method of investigation, although bedside head ultrasound (HUS) may be performed if the infant is felt to be too unstable to be transported. However, HUS may miss bleeding or mass effect adjacent to the skull. Magnetic resonance imaging of the brain may be performed in a nonurgent manner to further delineate the extent and location of cerebral injury when ICH has been diagnosed. The association between instrumented vaginal delivery and ICH remains controversial. Term infants who have SGH and/or IVH should have serial hematocrit values obtained to monitor for evidence of ongoing blood loss. A complete blood cell count and coagulation studies are needed for both evaluation and ongoing management of affected infants. The most common hematologic condition associated with ICH in term infants is thrombocytopenia, which may be caused by disseminated intravascular coagulation or a genetic, immune-related, or infectious disorder. Coagulation factor deficiencies may be found infrequently with SGH and/or IVH.

For the infant described in the vignette, the finding of tachycardia is likely related to hypovolemia and an electrocardiogram should be considered only if the tachycardia persists after volume replacement. Suspicion of seizure activity or the clinical course should guide the decision to perform an electroencephalogram. Further evaluation of this infant with CT scan of the head or plain radiographs of the skull may be performed once the infant has been stabilized in order to evaluate for ICH or skull fracture.

Suggested Reading:


Modanlou HD. Neonatal subgaleal hemorrhage following vacuum extraction delivery. Internet J Pediatr Neonatol. 2005;5(2)

Rosenberg AA. Traumatic birth injury. NeoReviews. 2003;4(10):e270-e276. doi:10.1542/neo.4-10-e270. Available online only for subscription

Source: 2013 PREP® Self-Assessment, Question 184