Cardiology

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Question 1

A 7-month-old female infant is brought to the Emergency Department. Her parents are concerned about her head appearing swollen. She was discharged from the inpatient unit a week prior to this encounter, after having a successful cavopulmonary anastomosis (Glenn shunt) for single ventricle disease. On examination, the child appears slightly irritable. She is afebrile, her heart rate is 136 beats/minute, respiratory rate is 28 breaths/minute, and transcutaneous oxygen saturation is 88%. Her neck and face appear slightly swollen. There is no pedal edema. On cardiac examination, first and second heart sounds are heard. No murmur is observed. The most likely cause for this patient's clinical findings is:

A. Postoperative atelectasis
B. Superior vena cava syndrome
C. Postoperative renal dysfunction
D. Congestive cardiac failure
E. Drug reaction to antibiotics used in postoperative phase

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Correct Answer: B. Superior vena cava syndrome due to elevated central venous pressure

Superior vena cava syndrome can be iatrogenic and may occur as a complication of a central venous line or cardiac surgery. Patients with single ventricle disease are now successfully palliated in a staged fashion. In many cases, neonatal intervention is followed by a cavopulmonary shunt performed between 4 and 8 months of life. It involves connecting the superior vena cava directly to the right (frequently) or left pulmonary artery (in cases of bilateral superior vena cavae, each is anastomosed to its ipsilateral branch pulmonary artery). Following the repair, the superior vena cava now faces the slightly higher pressure in the branch pulmonary artery. This results in increased cerebral venous pressure, which might cause irritability in the patient. When the pressure in the pulmonary artery is high, there can be swelling of the face and the neck. This might also be the result of any obstruction in the venous system, as seen with thrombosis. Whenever superior vena cava syndrome is suspected, a thrombus in the venous system should always be excluded.

The patient in this vignette is afebrile and does not manifest any respiratory distress. A saturation of 88% is expected following a Glenn shunt. Thus, there is no supportive evidence for postoperative atelectasis. Postoperative renal dysfunction is unlikely to manifest for the first

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time this late after surgical intervention. The edema is localized to the face and neck. This onset would rule out renal dysfunction and congestive cardiac failure. An anaphylactic reaction to the use of drugs is also unlikely to manifest for the first time this late after the intervention.

Take-Home Message
Unilateral swelling of the face, neck, and chest following a Glenn shunt should always raise the suspicion of superior vena cava syndrome that results from obstruction to the venous circulation or increased venous pressure. In addition to obstruction due to thrombosis, extrinsic compression due to tumors and fungal infections should be considered.

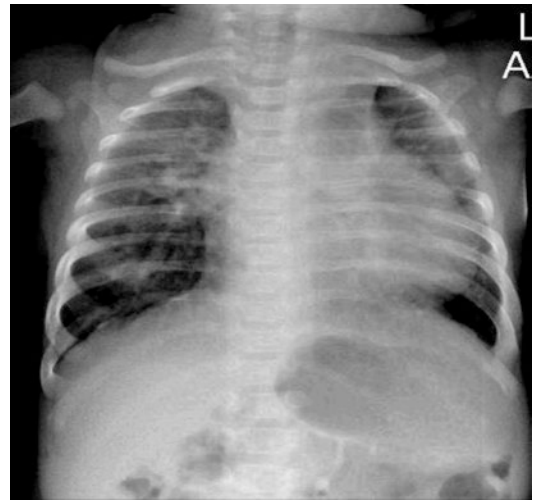
ABP Content Specification
- Evaluation of edema.

Question 2

The parents of a two-week old girl are concerned about her breathing. She was born at full term by a normal vaginal delivery and discharged on the second day of life. At her one-week visit, her pediatrician detected a heart murmur and referred to the ED for further evaluation. She had regained her birth weight but over the past 3 days appears to take longer to complete her feed taking breaks and sweating easily.

On examination, the neonate is observed to be breathing at 50 breaths per minute with nasal flaring. Transcutaneous oxygen saturation is 97%. Peripheral pulses are strong, but the baby has tachycardia at 170 beats per minute. Air entry is equal bilaterally, but crackles are heard at the bases. A pansystolic murmur is heard over the precordium. The liver edge is palpable 3 cm below the right costal margin.

A chest radiograph is obtained and is shown as follows:



At this time, the next best step in the management of this patient would be:

A. Advise the parents to keep the appointment with the pediatric cardiologist.
B. Administer a dose of intravenous furosemide.
C. Obtain a complete sepsis workup, and start intravenous antibiotics.
D. Start the child on nasal oxygen at 2 L per minute.
E. Order a dose of oral digoxin.

Correct Answer: B. Administer a dose of intravenous furosemide.

The neonate in question appears to be in congestive cardiac failure due to a large left-to-right shunt. Her oxygen saturation suggests an acyanotic heart lesion similar to a ventricular septal defect or a patent ductus arteriosus. Neonates with such lesions often become symptomatic

after the pulmonary vascular resistance drops, which usually occurs after 48 hours of life. The large shunt of blood to the pulmonary vasculature results in tachypnea, poor feeding, and failure to thrive. Increase in sympathetic tone results in tachycardia and sweating, especially with feeds. While the accurate diagnosis of the congenital heart lesion and its medical and/or surgical management is appropriately done by the pediatric cardiologist, the child in this vignette is in symptomatic high-output failure and should be treated for the same. Intravenous furosemide would result in diuresis by reducing the preload, decrease pulmonary vascular congestion, and to some degree improve feeding. A single oral dose of digoxin would not be effective in the acute management of this child. The newborn in this vignette has tachycardia, but no other signs to suggest sepsis, and thus a sepsis workup and use of antibiotics is not indicated. The presence of a heart murmur, with supportive radiological evidence of cardiomegaly and pulmonary over-circulation, suggests a left-to-right shunt. The tachypnea in this child is secondary to pulmonary over-circulation. In this scenario, the use of nasal oxygen will not relieve the tachypnea. However, the potent pulmonary vasodilatory properties of oxygen might worsen the shunt and pulmonary over-circulation.

Take-Home Message

Neonates with congestive cardiac failure present with decreased feeding, sweating, irritability, and failure to gain weight. Chest radiograph typically shows cardiomegaly with increased pulmonary markings and vascularity.

ABP Content Specification

- Recognize signs and symptoms of congestive heart failure.
- Plan management of acute congestive heart failure.

Question 3

The triage nurse calls you to assess a six-year-old boy who presents with a two-day history of fever and chest pain. Parents report that he was discharged from the hospital 13 days ago after an uncomplicated surgical repair of an atrial septal defect. You note an anxious child who refuses to lie down on the examination table. His neck veins appear distended. His extremities feel cool to the touch. On palpation, his peripheral pulses are thready. He does not cooperate with an abdominal examination, but his liver feels enlarged. Crackles are audible at the lung bases. You cannot hear his heart sounds very clearly, but there is no murmur or rub. Which of the following abnormal findings on investigation is most likely to be encountered in the patient described in this vignette?

A. A large pericardial effusion on an emergent bedside ultrasound
B. Normal cardiac silhouette with evidence of bilateral pleural effusions on a chest X-ray
C. Marked anemia on a complete blood count
D. Leukocytosis on a complete blood count
E. Supraventricular tachycardia on ECG

Correct Answer: A. A large pericardial effusion on an emergent bedside ultrasound

The boy in this vignette has signs of tamponade from a large pericardial effusion. About 15% of patients who undergo surgical closure of an atrial septal defect may develop a pericardial effusion. The median duration after surgery when the effusion is diagnosed is 13 days. When there is a rapid accumulation of pericardial fluid, there is impaired filling of the ventricles. This results in engorged neck veins and hepatomegaly because of impaired right ventricular filling and pulmonary venous congestion because of impaired left

ventricular filling. Poor cardiac output usually follows, resulting in pallor and thready pulses.

Cardiac tamponade is an emergency. An emergent bedside ultrasound performed by a trained emergency physician can quickly confirm the presence of a pericardial effusion. Resources can then be mobilized to emergently drain the effusion, which can be life-saving.

While bilateral pleural effusions can coexist with a pericardial effusion, the cardiac silhouette will not be normal with a large pericardial effusion. Patients with cardiac tamponade appear pale, but this manifestation is secondary to poor cardiac output. There is no evidence to support an infection in this patient, and the distended neck veins are not seen in supraventricular tachycardia.

Take-Home Message

Cardiac tamponade should be recognized rapidly to prevent death from hemodynamic compromise. Patients with acute tamponade present with dyspnea, tachycardia, tachypnea, signs of hypoperfusion, and shock. Other findings include elevated jugular venous pressure, pulsus paradoxus, and muffled heart sounds. Diagnosis can be made by echocardiography, and treatment is emergent pericardiocentesis.

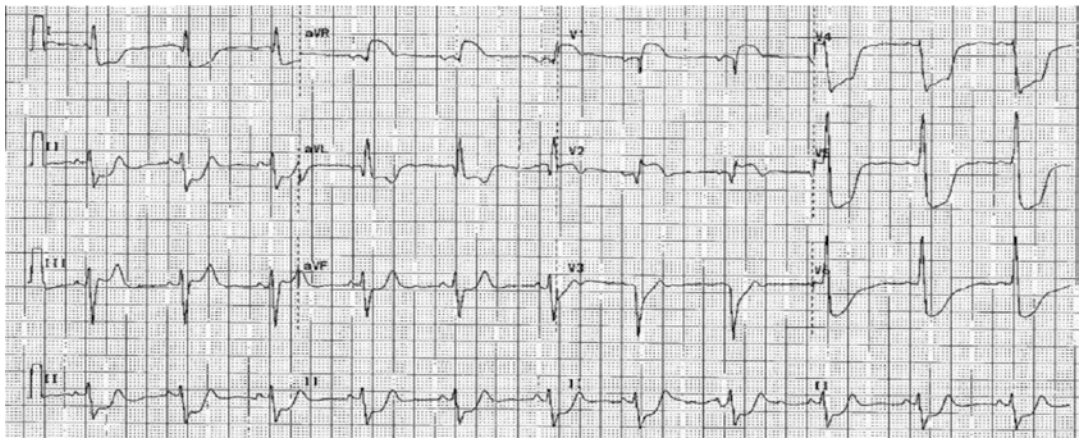
ABP Content Specification

- Recognize the signs and symptoms of pericardial effusion and tamponade.

Question 4

A 16-year-old boy suddenly collapsed while playing basketball in school. His coach rushed to his side, but the student revived spontaneously without requiring any resuscitative measures. Emergency medical services had been summoned, and they brought the student to the emergency department. His transport was uneventful.

You observe a cheerful young adolescent who appears to be in no acute distress. He is afebrile, with a heart rate of 60 beats/minute, breathing at a rate of 14 breaths/minute, and with an automated blood pressure measurement of 108/68 mm Hg in the right arm. He experienced crushing chest pain before his episode of syncope, but is not particularly bothered, because he frequently experiences chest pain while playing basketball. He denied any intercurrent illness, taking prescribed medications or illicit drugs, and any family history of sudden cardiac death. On examination, his peripheral pulses are strong. First, heart sound is normal, while the second is physiologically split. No heart murmur is audible. The nurse obtained a 12-lead electrocardiogram while the patient was in triage, which is now available for review and shown as follows.



The next best step in the management of this patient is:

A. Order a urine toxicology screen.
B. Counsel the patient to maintain adequate hydration before playing basketball.
C. Request an urgent evaluation by a pediatric cardiologist.
D. Order a CT angiogram of the chest.
E. Order a blood test to measure potassium levels.

Correct Answer: C. Request an urgent evaluation by a pediatric cardiologist.

The male athlete in this question has a troubling history of exertional chest pain and now presents with exertional syncope. While chest pain and syncope generally have a benign etiology in the pediatric age group, about 6% of patients do have a cardiac etiology. They frequently present with exertional symptoms. The American Heart Association recommends preparticipation cardiovascular screening for all athletes who give a history of exertional chest pain and syncope. Cardiac causes for exertional chest pain and syncope would include hypertrophic cardiomyopathy, anomalies of the coronary arteries, aortic stenosis, systemic and pulmonary hypertension, myocarditis, and dilated cardiomyopathy. While aortic stenosis is associated with a heart murmur and hypertrophic cardiomyopathy with a dynamic murmur, the other causes may have a normal physical examination at rest. The evaluation of exertional chest pain and syncope can be performed by a pediatric cardiologist on a routine basis, with the caveat that the patient not indulge in competitive sports pending the evaluation. However, the ECG on the patient in this vignette is troubling in that there are marked ST-T changes, indicating an ischemic process. An urgent evaluation by a pediatric cardiologist, including performing an echocardiogram, would be the most beneficial next step. An echocardiogram delineates global or regional ventricular dysfunction and presence of effusions and assess for structural abnormalities like hypertrophic cardiomyopathy or anomalies of coronary origin (namely, anomalous origin of left coronary from the right sinus of Valsalva). Coronary vasospasm induced by cocaine can result in chest pain, though it may occur even without exertion. The purely exertional nature of chest pain in this vignette does not suggest chest pain induced by cocaine abuse. If echocardiography suggests anomalous origin or intramural course of the coronary arteries, then it can be confirmed by CT angiography.

Hyperkalemia results in peaked T wave, widening of the QRS complex followed by pulseless electrical activity. The goal of therapy is to stabilize cell membranes by calcium gluconate or calcium chloride, which counteracts the effects of hyperkalemia on myocardium action potential within minutes. Calcium gluconate is preferred in children as it is less irritating to the veins. The other treatment includes bicarbonate or insulin with glucose, albuterol nebulization, and sodium polystyrene sulfonate. Insulin increases the activity of the Na-K-ATPase pump in the skeletal muscle and drives K into cells. Glucose is administered simultaneously with insulin to prevent hypoglycemia. Sodium polystyrene sulfonate is a resin which exchanges sodium for potassium at a 1:1 ratio and therefore increases potassium excretion.

Take-Home Message

Ischemic heart disease in children is uncommon but should be suspected in children presenting with exertional chest pain. ECG is a good initial screen to identify left ventricular hypertrophy suggestive of hypertrophic cardiomyopathy or manifest ST and or T wave changes indicating ischemia. Patients with electrocardiographic changes should be referred to cardiology for evaluation and prohibited from performing exercise and participating in sports until cleared.

ABP Content Specification

- Recognize ECG abnormalities in children.

Question 5

A 10-year-old girl is brought to the Emergency Department by her parents. She is usually in good health but has been complaining of excruciating pain in different parts of her body over the past few days. Today, she refused to bear weight on her left leg. Her vital signs are as follows: temperature 102.7 °F (39.3 °C), heart rate 132 beats/minute, respiratory rate 20 breaths/minute, and blood pressure 116/68 mm Hg. She is in mild distress. Her left knee appears swollen. It is warm to touch and quite tender. There is no bruising around the knee. Lungs are clear to auscultation. First and second heart sounds are clearly audible. A grade 2/6 pansystolic murmur is heard at the apex of the heart. Abdomen is soft, without any evidence of organomegaly.

The etiology of this patient's symptoms is most likely to be elucidated by which of the following investigations?

A. AP and lateral X-rays of the left knee
B. Elevated antistreptolysin O titer in blood
C. Leukocytosis on synovial aspirate
D. Positive influenza swab
E. Elevated ANA titers in blood

Correct Answer: B. Elevated antistreptolysin O titer in blood

The patient in this vignette fits the profile of rheumatic fever, which occurs most often in children between the ages of 5 and 15 years. It typically occurs several weeks after an episode of streptococcal pharyngitis. The salient features are fever, migratory arthritis affecting major joint groups, an evanescent rash usually over the abdomen, and subcutaneous nodules. Chorea may occur a few months after the pharyngitis. The migratory arthritis refers to the sequential involvement of joints, with inflammation resolving in one joint and then beginning in another joint.

In some patients, pancarditis may occur that affects all three layers of the heart. Valvulitis usually affecting the mitral and aortic valves, causes valvar insufficiency and can manifest with a new murmur.

Since rheumatic fever usually occurs several weeks after the episode of pharyngitis, the rapid strep test or a throat culture may not be positive at the time of evaluation. However, evidence of past streptococcal infection may be deduced from an elevated blood titer of antistreptolysin O. The arthritis in rheumatic fever is flitting and fleeting. The radiograph of the affected joint often reveals nonspecific findings of increase in the joint space. Synovial aspirate will reveal a sterile inflammation, is not diagnostic, and certainly not indicated for making a diagnosis. Rheumatoid arthritis is more likely to affect smaller joints. In a patient who has been in good health, and without a long-standing history of joint pains, one would not consider a diagnosis of rheumatoid arthritis first.

Take-Home Message

Rheumatic fever, although rare, occurs from time to time in the United States, often in clusters. The diagnosis is based on the modified Jones criteria which includes carditis, arthritis, chorea, erythema marginatum, and subcutaneous nodules as major criteria. Arthralgia, fever, a prolonged PR interval, increased acute phase reactants such as ESR, and previous rheumatic fever are minor criteria. Rheumatic fever is diagnosed if there are two major and one minor criterion or one major and two minor criteria. In all situations, there must be evidence of recent streptococcal infections.

ABP Content Specification

- Recognize the signs and symptoms of rheumatic fever.

Question 6

A 17-year-old male is transferred from a juvenile detention facility to the Emergency Department as he was found to be febrile. His past history is significant for multiple arrests for felony to support an intravenous drug addiction. He gives a history of "feeling warm" for many days and has

been given oral aspirin to control his fever. On physical examination, he is in no significant distress. His axillary temperature is 101.4 °F (38.6 °C), heart rate is 112 beats/minute, and blood pressure is 102/68 mm Hg in the right arm. The nasal passages and throat appear normal. There is nonspecific enlargement of lymph nodes in his neck. A 2/6 pansystolic murmur is heard at the left lower sternal border. The abdomen is soft, without any evidence of organomegaly.

At this time, the next best course of action in the management of this patient would be:

A. Arrange for an outpatient appointment with a pediatric cardiologist.
B. Arrange for admission to the inpatient service.
C. Obtain a urine toxicology screen.
D. Obtain a serum salicylate level.
E. Arrange for a psychiatric evaluation.

Correct Answer: B. Arrange for admission to the inpatient service.

This patient likely has infective endocarditis (IE) that needs immediate hospital admission. IE is defined as an infection, usually bacterial, of the endocardial surface of the heart. It occurs with an incidence of 0.3 per 100,000 children per year with a mortality of 11.6%. There has been an increase in the incidence of IE with the greater survival of children following surgical intervention for CHDs. At the same time, improved survival of premature born infants and patients with chronic medical conditions who undergo prolonged hospitalization (frequently with chronic instrumentation) increases the pool of patients with structurally normal hearts who are at risk for IE. Intravenous drug abusers are at risk for developing endocarditis involving right-heart structures.

The classic triad of fever, anemia, and a heart murmur is rarely present. Oftentimes, patients with IE present with nonspecific symptoms. Typically, they have relapsing low-grade fever. There usually is vague myalgia, headache, and arthralgia. If the progression of the infection is rapid, then they may appear toxic. A new or changing heart murmur may signify involvement of one of the valves. In cases of slowly evolving infections, signs relating to peripheral immunologic phenomena, such as the appearance of splinter hemorrhages, Osler nodes, Janeway lesions, and Roth spots, may be noted. Osler nodes are small, painful nodules usually present on the palmar surfaces of the fingers and toes; they may wax and wane. Janeway lesions are hemorrhagic, nonpainful macules which are found primarily on the palms and soles; they are embolic in origin. Roth spots are round, boat-shaped, or flame-shaped hemorrhages in the retina due to septic emboli to the eye.

The patient in this vignette is at risk for developing endocarditis due to his intravenous drug use**.** His heart murmur might be consistent with new onset of tricuspid regurgitation. The best course of action would be to obtain a complete blood count, obtain blood cultures from at least three sites, and start broad-spectrum antibiotic therapy. The classic lesion of endocarditis is the vegetation which may be identified by an echocardiogram, but a negative echocardiogram cannot rule out the diagnosis of endocarditis. An outpatient evaluation by a cardiologist would thus not be appropriate.

Most IE is accompanied by positive blood cultures. A select group of organisms cause most cases of endocarditis. Gram-positive organisms, particularly alpha-hemolytic streptococci (Streptococcus viridans), Staphylococcus aureus, and coagulase negative staphylococci predominate. In native valves, streptococcal involvement is more common, most often affecting the mitral valve. Congenital heart disease accounts for 15% of cases, with bicuspid aortic valve being the most common. S. aureus is generally more common among IVDA patients and involves the tricuspid valve.

While it is possible that the patient might have continued abusing drugs, this would not explain the findings on physical examination. Hyperthermia is a manifestation of severe salicylate toxicity. The patient in this vignette does not have evidence of nausea, vomiting, tinnitus, or

hyperventilation, which would precede hyperthermia. Behavioral intervention by a child psychiatrist would be helpful given the history of drug use but would not manage the acute issues facing this patient.

Take-Home Message

IE presents usually with nonspecific symptoms but should be suspected among IV drug abusers and those that have therapeutic hardware in the heart. While the diagnosis is straightforward in the setting of positive blood cultures and visible vegetations on echocardiography, presumptive treatment may be necessary among those that raise high clinical suspicion, despite absence of these findings.

ABP Content Specification

- Recognize signs and symptoms of infectious endocarditis.

Question 7

A 10-month-old male infant is sent to the Emergency Department from his pediatrician's office. He has been febrile for the past 6 days and has developed a rash over the past day. He lives at home with his parents, who both deny any recent illness. Parents report that child has been irritable since the onset of fever and has had reduced oral intake, but no vomiting or diarrhea. On examination, the child is fussy and febrile at 102.2 °F (39 °C). His eyes appear injected. There is no rhinorrhea. The angles of his mouth appear cracked, while his lips and tongue appear inflamed. A diffuse macular erythematous rash is noted on the trunk. He has palpable axillary lymph nodes. Examination of his ears reveals a mobile tympanic membrane.

At this time, the next best step in the management of this patient is:

A. Complete sepsis workup with blood, urine, and spinal fluid cultures, followed by antibiotic therapy.
B. Reassure the parents that this is a viral illness and prescribe acetaminophen for symptomatic relief.
C. Admit to the inpatient unit for immune globulin therapy.
D. Prescribe amoxicillin for otitis media.
E. Administer intravenous fluids for rehydration.

Correct Answer: C. Admit to the inpatient unit for immune globulin therapy.

The infant in question shows signs strongly suggestive of Kawasaki disease, an acute, self-limited, acquired inflammatory condition characterized by vasculitis of small to medium vessels. The etiology of this acute illness is not well understood. To satisfy the criteria for diagnosis of Kawasaki disease, the patient should have high-grade fever with no obvious cause for at least 5 days and any four of the following 5 criteria: non-purulent bulbar conjunctivitis that spares the limbus, a strawberry tongue, lymphadenopathy, a generalized rash, swelling of the hands and feet, and peeling of the skin around the anus. It primarily afflicts children between 1 and 5 years of age. Laboratory investigations are not diagnostic, but include leukocytosis, and a marked increase in acute-phase reactants, namely, C-reactive protein and erythrocyte sedimentation rate. There is significant thrombocytosis about 3 weeks into the illness. The acute phase is also characterized by hydrops of the gall bladder. Perhaps the most significant effect is on the cardiovascular system. In the acute phase, patients might have myocarditis and a pericardial effusion. About 20% of patients may develop aneurysms of the coronary arteries. Primary treatment is IV immunoglobulin and aspirin. When intravenous immune globulin therapy is instituted prior to 10 days after the onset of fever, the risk of coronary artery involvement drops to about 2%.

The infant in question has had fever without any obvious cause and also manifests supporting criteria for the diagnosis of Kawasaki disease. Elevation in acute phase reactants, and a nonspe-

cific CBC would provide supportive evidence for this diagnosis. The risk of coronary artery involvement would be decreased by prompt institution of immune globulin therapy. An echocardiogram in the acute phase would be helpful in assessing myocardial function and to rule out a pericardial effusion. Absence of coronary artery aneurysms in the acute phase does not rule out Kawasaki disease.

The child in this vignette is ill, without being toxic. Fussiness is common in infants with Kawasaki disease. While a workup for an infectious etiology may not be unreasonable, a spinal tap and administration of antibiotics does not appear to be justified. The infant is febrile, but does not have rhinitis, vomiting, or diarrhea. There are no sick contacts. Under such circumstances, a diagnosis of a viral illness is not appropriate. While fever without an obvious etiology in a 10-month-old should raise suspicion for otitis media, the infant in this vignette has mobile tympanic membranes on examination. Due to stomatitis and glossitis, the infant has poor oral intake and is probably slightly dehydrated. While rehydration would be part of clinical management, investigations for and management of Kawasaki disease would take precedence.

Take-Home Message

The diagnosis of Kawasaki disease should be entertained in children that present with high fever more than 5 days with no evident etiology, with supportive clinical findings. When intravenous immune globulin is administered within 10 days of the onset of fever, the incidence of coronary artery involvement is significantly lower than without treatment.

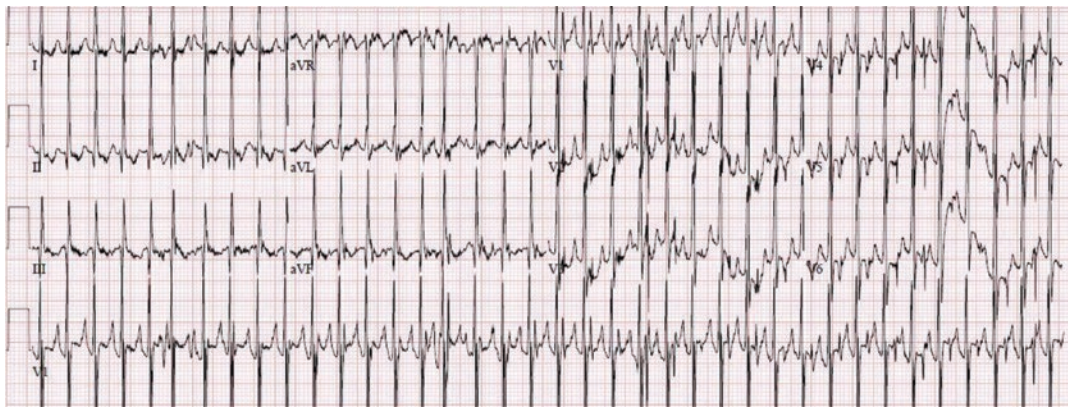
All children who are suspected of having Kawasaki disease require ECG, echocardiogram, and urgent consultation with a pediatric cardiologist.

ABP Content Specification

- Recognize signs and symptoms of Kawasaki disease.

Question 8

A 6-week-old female infant is brought to the Emergency Department by her mother for decreased activity for a few hours and is not feeding as usual. On examination, she is awake, afebrile, and responds appropriately to her mother. Her heart rate is too fast to count. She is breathing at 40 breaths/minute. Her peripheral pulses are weak but palpable, though the automated blood pressure machine is unable to obtain a reading. Other than a fast heart rate, the cardiac examination is unremarkable. A quick examination reveals normal breath sounds, no organomegaly, and normal tone in all extremities. A 12-lead electrocardiogram is obtained and is as shown in the figure.



At this time, the next best step in the management of this infant is:

A. Apply an ice pack to the face, while the nurse attempts to obtain intravenous access.
B. Gently massage the eyeballs of the patient.
C. Obtain intraosseous access to administer fluid resuscitation.
D. Prepare for DC cardioversion with 0.5 J/kg of current.
E. Do a complete sepsis workup and administer broad-spectrum antibiotics.

Correct Answer: A. Apply an ice pack to the face, while the nurse attempts to obtain intravenous access.

The electrocardiogram on the patient in this vignette demonstrates narrow-complex tachycardia (supraventricular tachycardia). When prolonged, it may result in ventricular dysfunction and may manifest with lethargy and poor feeding. The appropriate management of a patient in supraventricular tachycardia depends on the mental status of the patient and the hemodynamic status. If the tachycardia results in hemodynamic collapse, then the appropriate intervention would be synchronized cardioversion. However, the patient in this vignette is awake and responsive. While the automated machine is unable to record a blood pressure, her pulses are palpable. In such a setting, the appropriate intervention would be to pharmacologically cardiovert the rhythm using intravenous adenosine. Adenosine terminates the tachycardia by blocking the atrioventricular node.

While intravenous access is being established, it is acceptable to attempt cardioversion by a vagotonic maneuver such as applying a cold stimulus by an ice pack to the face, bearing down, coughing, gagging, etc. However, noxious modes of increasing vagal tone by massaging the eyeballs should not be applied. While the child may be dehydrated from poor oral intake, abolishing the tachycardia would result in better oral intake and rehydration. Her perfusion and blood pressure would improve. At this time, there is no indication for invasive modes of fluid resuscitation by the intraosseous route. While sepsis does result in tachycardia, the rate in this case is above the limits of sinus tachycardia. P waves are not seen on the ECG, making supraventricular tachycardia the diagnosis as opposed to sepsis.

Wolff-Parkinson-White (WPW) syndrome is suggested by a shortened PR interval, slurred QRS complex (delta wave), and a widened QRS complex. In patients with ECG findings of narrow-complex tachycardia with preexcitation suggestive of WPW syndrome which are refractory to vagal maneuvers and adenosine, procainamide should be used. In unstable patients, synchronized cardioversion should be performed. If WPW is associated with wide-complex tachycardia, atrioventricular nodal blocking agents such as adenosine and digoxin should be avoided.

In patients with wide complex tachycardia who are otherwise stable and do not require immediate cardioversion, adenosine can be used. In such situations, adenosine may convert supraventricular tachycardia to sinus rhythm or differentiate ventricular from supraventricular tachycardia.

Take-Home Message

Narrow-complex tachycardia may manifest with nonspecific symptoms in infants. When confirmed by an electrocardiogram, sinus rhythm can be restored with intravenous adenosine or synchronized cardioversion (in cases of hemodynamic collapse). While setting up to deliver adenosine, non-noxious vagal maneuvers may be used to restore sinus rhythm.

ABP Content Specification

- Recognize and plan the management of ECG abnormalities in children.

Question 9

A 5-month-old male infant is brought by an emergency medical response team to the ED. His parents report that the child has been vomiting once per day and having loose stools for the past 3 days. This afternoon, he woke up “irritable.” He started crying and could not be easily con-

soled. He then progressively turned more and more dusky and finally became unresponsive. By the time emergency responders showed up at the family home, the child had regained consciousness. In the ED, he is noted to be febrile at 101.4 °F (38.6 °C). His heart rate is 156 beats/minute, respiratory rate is 32 breaths/minute, and transcutaneous oxygen saturation is 78% in room air. His fontanelle is slightly sunken, but his lips and eyes are moist. He is comfortable in his mother's arms and is drinking from a bottle. His extremities are warm, but the fingertips appear dusky. Cardiac examination is significant for a harsh 3/6 ejection systolic murmur that radiates to the axillae. His parents report that he was diagnosed with tetralogy of Fallot in fetal life and has regular follow-up with a pediatric cardiologist.

At this time, the most appropriate step in the management of this child is:

A. Arrange for an urgent outpatient follow-up visit with the pediatric cardiologist within 1 week.
B. Initiate workup for IE and start antibiotic therapy.
C. Admit to the inpatient service for observation.
D. IV normal saline bolus of 100 ml/kg over 30 minutes.
E. Discharge the patient home with instructions on oral rehydration therapy.

Correct Answer: C. Admit to the inpatient service for observation.

Tetralogy of Fallot is the most common cyanotic CHD. The degree of cyanosis depends on the severity of right ventricular outflow tract obstruction. In a majority of centers, the surgical approach to this lesion is to perform a complete repair electively at 3–4 months of life. However, some patients with this condition might experience hypercyanotic spells. While the exact etiology of this condition is unclear, it results in an acute increase in right ventricular outflow obstruction, causing increased shunting of desaturated blood into the systemic circulation, which in turn results in a worsening of cyanosis. The increased right ventricular outflow tract obstruction results in acute reduction of pulmonary blood flow. With significant obstruction, the heart murmur disappears, and the patient may lose consciousness. Acidosis and dehydration increase the risk of a hypercyanotic spell.

Management of a hypercyanotic spell involves keeping the patient calm, ensuring euvolemic status, use of supplemental oxygen and the "knee-chest" position, intravenous morphine (0.1 mg/kg) and sodium bicarbonate (1 mEq/kg), and infused esmolol and phenylephrine (neosynephrine) 0.1 mcg/kg/min titrated to effect.

The progression of events in this vignette suggests that the patient had a hypercyanotic spell. While the infant in this vignette appears to have recovered, he is at risk for another spell and should not be sent home. A fluid bolus of 100 ml/kg is out of proportion to the degree of dehydration. While cyanotic patients are at risk for developing IE, the clinical vignette does not support this diagnosis.

Take-Home Message

Tetralogy of Fallot is increasingly diagnosed in fetal life and addressed surgically in an elective fashion at between 4 and 6 months of life in those without severe right ventricular outflow obstruction. However, some of these patients might go on to develop hypercyanotic spells, which would prompt appropriate stabilization and earlier surgical intervention.

ABP Content Specification

- Recognize signs and symptoms and life-threatening complications of congenital cardiac lesions.

Question 10

An obese 17-year-old female presents to the ED with a three-day history of fatigue. She is unable to walk up one flight of stairs to her apartment. She denies any history of fever, rhinitis, vomiting, or diarrhea. Her past history is remarkable for giving birth after a term pregnancy about 3 months ago. Her vital parameters are as follows:

heart rate of 114 beats/minute, respiratory rate of 30 breaths/minute, and blood pressure of 90/62 mm Hg. She appears anxious. Her jugular veins appear slightly distended. Pedal pulses are felt, but there is slight pedal edema. On auscultation, the heart rate is fast, but the heart sounds are slightly difficult to hear. There are bibasilar rales. The patient is unable to cooperate for an abdominal examination. A chest radiograph is obtained, and shows moderate cardiomegaly, with basal congestion. The presentation of the patient in this vignette is consistent with:

A. Peripartum cardiomyopathy
B. Fulminant viral myocarditis
C. Deconditioning secondary to obesity
D. Fatigue associated with caring for a newborn
E. Acute pneumonia

Correct Answer: A. Peripartum cardiomyopathy

Peripartum cardiomyopathy is a rare association with pregnancy and can occur in the last month of pregnancy, up to 5 months after childbirth. While it is more common in women over the age of 30, it can afflict any age group. The causes are poorly understood. The patient in this vignette presents with symptoms of congestive cardiac failure. She is 3 months postpartum and does not have any history of an intercurrent viral infection to suggest fulminant viral myocarditis. While the patient is obese, deconditioning would not explain the distension of jugular veins, pedal edema, bibasilar rales, or the cardiomegaly and pulmonary vascular congestion seen on the chest radiograph. These findings would also not be present in a fatigued mother caring for her newborn. The patient has cardiomegaly and pulmonary vascular congestion on her chest radiograph. She is not febrile. This does not support a diagnosis of acute pneumonia.

Take-Home Message
The clinical findings of congestive heart failure in a postpartum woman of any age should prompt evaluation for peripartum cardiomyopathy.

ABP Content Specification
- Differentiate the etiology of congestive heart failure by age.

Question 11

A 13-year-old boy is brought to the Emergency Department from the school nurse's office. He has been febrile for the past 4 days, with a history of emesis and poor oral intake. This morning, while going from one classroom to another, he fainted. He revived spontaneously and was taken to the nurse's office. On arrival to the ED, he was noted to be febrile at 104 °F (40 °C). His tongue is moist, and skin turgor is maintained. His peripheral pulses are thready. His jugular veins appear distended. On cardiac examination, heart rate is rapid, and sounds are difficult to auscultate. Air entry is equal in both lung fields, though rales are heard at both bases. The liver is palpable 3 cm below the right costal margin. Serum electrolytes are normal. A chest radiograph shows mild cardiomegaly and bilateral lower lobe haziness. At this time, the next best step in the management of this patient is:

A. Intravenous antibiotic therapy
B. One dose of intravenous furosemide
C. Intravenous bolus of 1 L of normal saline over 30 minutes
D. Intravenous infusion of dopamine
E. Inhaled albuterol

Correct Answer: B. One dose of intravenous furosemide

Acute fulminant myocarditis is a rapidly progressive process that follows a viral infection in otherwise healthy children with no personal or family history of cardiac disease. It may present with chest pain, palpitations, syncope, arrhythmia, or sudden cardiac death. While the progression of symptoms is rapid, and the clinical presentation usually dire, when recognized and appropriately treated, the outcomes are generally positive.

The patient in this vignette has become quite symptomatic after a brief illness. He presents with signs of low cardiac output and venous congestion. However, he does not appear to be dehydrated. Anti-congestive therapy with a diuretic such as furosemide would help decrease the pulmonary vascular congestion and improve symptoms. While inotropy with infused dopamine will probably be indicated, empiric treatment with dopamine in a patient who is not in extremis is probably not called for. Though the patient has been vomiting, he does not appear severely dehydrated. Administration of a fluid bolus to a patient clinically suspected of having myocarditis might worsen the pulmonary edema. The patient does not have any evidence to back up the suspicion of pneumonia or a reactive airways attack, so the use of antibiotics and bronchodilators is not supported.

Take-Home Message

Low cardiac output can result from several conditions. It is essential to use clinical findings and investigations to distinguish those that involve intravascular depletion from those that do not. Clinical management is different in each case.

ABP Content Specification

- Know the etiology and recognize the signs and symptoms of myocarditis.

Question 12

A 16-year-old girl presents to the Emergency Department with complaints of severe abdominal pain. She has had two bouts of retching prior to the onset of the pain. She admits to have taken 25 pills of digoxin 4 hours ago, in order to kill herself after fighting with her mother. The mother was upset about her daughter's unplanned pregnancy. The patient's past medical history is significant for supraventricular tachycardia, which is well-controlled with the use of digoxin. On arrival to the ED, the heart rate is 60 beats/minute, respiratory rate is 14 breaths/minute, and blood pressure is 116/68 mm Hg. The patient is awake and alert. She does not appear dehydrated. Serum chemistry, including blood glucose, BUN, and creatinine, is normal, except for a potassium level of 5.2 mEq/L. The serum digoxin level is 0.7 ng/ml. An ECG is obtained and is normal. Urine pregnancy test is positive. At this time, the most appropriate step in the management of this patient is:

A. Administer activated charcoal orally or by a nasogastric tube.
B. Arrange for an outpatient appointment with the adolescent obstetric service.
C. Administer digoxin antibodies intravenously.
D. Admit to the inpatient unit with continuous cardiac monitoring.
E. Admit to the inpatient psychiatry unit.

Correct Answer: D. Admit to the inpatient unit with continuous cardiac monitoring.

Digoxin is a cardiac glycoside with a narrow therapeutic window. Adverse effects can be seen both with acute poisoning and chronic overdose. With acute poisoning, the patient may be asymptomatic for a few hours before developing gastrointestinal symptoms such as nausea, vomiting, and abdominal pain. Chronic overdose usually results in neurological symptoms such as lethargy and mental status changes. In either case, cardiac effects are likely, which include virtually every form of arrhythmia. The approach to the diagnosis of digoxin toxicity would include measurement of serum chemistries, digoxin level, and serial electrocardiograms. An abnormal BUN or serum creatinine would signify renal toxicity. In adults with digoxin toxicity, serum potassium above 5.5 mEq/L is associated with a risk of dying. A digoxin level alone cannot be used to guide management. Patients with acute or chronic digoxin overdose should be monitored in an inpatient setting for cardiac arrhythmias. Beyond the first couple of hours after an acute ingestion, use of activated charcoal may not be helpful. Any clinically significant digoxin overdose can be treated with administration of Fab antibodies to

digoxin. The patient in this vignette does not show any evidence of hypotension or an arrhythmia and thus should be monitored, as opposed to receiving therapy. While the suicide attempt does merit a detailed psychiatry evaluation, the management of the digoxin overdose takes precedence over that of the patient's psychiatric or obstetric conditions.

Hyperkalemia is considered a surrogate indicator for severity in acute digoxin overdose. It increases AV nodal block and can therefore exacerbate conduction delay. Potassium level may predict the outcome in patient with acute digoxin toxicity.

Take-Home Message

In cases of acute digoxin poisoning, a normal digoxin level does not obviate the need for observation. Monitoring the patient for arrhythmias, especially in the setting of hyperkalemia, is important.

ABP Content Specification

- Recognize the signs and symptoms and plan the management of digoxin toxicity.

Question 13

A 6-day-old newborn is brought to the Emergency Department by her parents. She was born after a full-term pregnancy and had an uncomplicated perinatal course. At the time of discharge from the newborn nursery at 3 days of life, a heart murmur was heard. She had a normal ECG, and her transcutaneous oxygen saturation at that time was 98% in room air. An appointment to see a pediatric cardiologist is due in 3 days. However, for the past 2 days, the baby has been tachypneic and feeding poorly. Since this morning, she has also been quite lethargic.

Vital signs in the ED are as follows: temperature 98.6 °F (37 °C), heart rate 175 beats/minute, respiratory rate 38 breaths/minute, and blood pressure 100/64 mm Hg in the right arm. The patient is lethargic, but arousable. Femoral pulses are not palpable. Air entry is heard in both lung fields. Heart sounds are heard, as is a grade 2/6 ejection systolic murmur at the left sternal border. The liver edge is palpable 2 cm below the right costal margin. At this time, the next step in the management of this patient is:

A. Establish intravenous access to start a prostaglandin infusion.
B. Administer broad-spectrum antibiotics.
C. Reassure the parents and ask them to keep the outpatient cardiology appointment.
D. Start an infusion of milrinone at 0.5 mcg/kg/minute
E. Obtain a CT scan of the brain.

Correct Answer: A. Establish intravenous access to start a prostaglandin infusion.

The newborn in this vignette has clinical evidence of coarctation of the aorta. Critical coarctation of the aorta manifests with circulatory failure and shock with closure of the ductus arteriosus. The portion of the aorta distal to insertion of the ductus is hypoperfused after ductal closure. This results in diminished femoral pulse and metabolic acidosis. Since the aorta proximal to the ductal insertion site receives adequate blood flow, blood pressure in the upper extremities might be normal to high, and the oxygen saturation is normal. It may be suspected clinically by a significant upper extremity–lower extremity blood pressure gradient and diminished or absent femoral and pedal pulses.

Coarctation is frequently associated with a bicuspid aortic valve. Initially, the left ventricle is able to pump against the increased afterload it faces, but eventually it fails resulting in symptoms of congestive heart failure. When there is clinical evidence of a ductal-dependent lesion, it is appropriate to institute prostaglandin therapy while awaiting a cardiology evaluation and diagnostic imaging.

While the patient in this vignette is sick enough for a diagnosis of sepsis to be entertained, that is unlikely to result in hepatomegaly. Antibiotic therapy would not be the first medical intervention that is warranted in this case. The poor pedal perfusion is secondary to obstruction at the level of the coarctation. An infusion of mil-

rinone is unlikely to improve lower extremity perfusion. The lethargy demonstrated by the patient is in keeping with the severity of the cardiac condition, and a CT scan of the brain may not be helpful. The management of a newborn with a ductal-dependent cardiac lesion is a medical emergency and cannot wait for a scheduled cardiology outpatient appointment.

Take-Home Message
Prostaglandin therapy is a life-saving intervention in patients with duct-dependent congenital heart lesions. When clinically indicated, a prostaglandin infusion should be started to open the ductus arteriosus and ensure systemic or pulmonary perfusion.

ABP Content Specification
- Understand the pathophysiology of congenital heart disease.

Question 14

A 10-year-old boy recently immigrated from Africa to the United States. His parents bring him to the ED because he has been unable to participate in gym at school. He gets extremely fatigued after running a quarter mile on the track. He becomes very short of breath, and his lips and fingernails turn blue. Vital parameters at triage are a heart rate of 96 beats/minute, respiratory rate of 28 breaths/minute, blood pressure of 100/68 mm Hg, and transcutaneous oxygen saturation of 88% in room air. His extremities are warm and well-perfused, but his fingertips appear unusual. Chest is clear to auscultation, with no added sounds. On cardiac examination, first heart sound is normal, while the second heart sound is single. A grade 2/6 continuous murmur is best heard over the back. The abdomen is soft, and there is no organomegaly.

The next best step in the management of this patient is:

A. Start an intravenous infusion of prostaglandin.
B. Cardiology consultation with echocardiogram
C. Pulmonology consultation with chest CT scan.
D. Orthopedic consultation with radiograph of the fingers.
E. Counseling regarding improving physical conditioning.

Correct Answer: B. Cardiology consultation with diagnostic imaging of the heart.

The boy in this vignette is desaturated and has clubbing of the fingernails. At 10 years of age, this is likely to be untreated cyanotic congenital heart disease or severe pulmonary hypertension secondary to unrepaired acyanotic congenital heart disease. In developed nations, it is rare for hemodynamically significant lesions to remain unaddressed up until 10 years of life. However, the boy recently immigrated from Africa. Since there is only a single second heart sound, and evidence of collateral flow in the lung fields (continuous murmur in lung fields), the lesion here is likely pulmonary atresia and ventricular septal defect with collateral flow to the lung fields. A cardiology consultation with an echocardiogram is the modality most likely to delineate the structural heart lesion.

Prostaglandin infusion is unlikely to be effective at maintaining patency of the ductus at 10 years of age. There is no evidence to support a likely pulmonary cause for the patient's signs. Clubbing is related to desaturation. Radiograph of the fingers is unlikely to delineate the cause for cyanosis.

Take-Home Message
Clubbing is a sign of chronic cyanosis and is frequently secondary to cardiac or respiratory causes. Workup for clubbing should involve investigation for a possible cardiac cause.

ABP Content Specification
- Understand the pathophysiology, signs, and symptoms of congenital heart disease.

Question 15

A 15-year-old girl is brought to the Emergency Department by EMS personnel. She was showering when she felt dizzy. She attempted to steady herself, but was unable to maintain her balance and fell down, hitting her head against the wall. She denied losing consciousness. She had no prior episodes of syncope, palpitations, or other recent illnesses. There is no family history of cardiac disease or sudden death. In triage, her vital parameters are as follows: heart rate 96 beats/minute, respiratory rate 14 breaths/minute, and blood pressure 102/68 mm Hg. She is alert and oriented. A tender bruise is noted on her forehead. Her extremities are warm and well-perfused. Lung fields are clear to auscultation. Heart sounds are heard, with no murmur or rub. There is no organomegaly.

The most appropriate next step in the management of this patient is:

A. CT scan of the brain to evaluate for intracranial hemorrhage.
B. Admit to the inpatient unit for observation.
C. Blood alcohol level.
D. Arranging for outpatient appointment with a pediatric cardiologist.
E. Reassurance about the benign nature of symptoms and discharge.

Correct Answer: E. Reassurance about the benign nature of symptoms and discharge.

Syncope is a very common complaint in the pediatric age group. It is estimated that about 15% of people will have had at least one episode of syncope prior to the end of adolescence. Fortunately, in about 80% of these patients, neurocardiogenic syncope is the cause for symptoms. It is characterized by dizziness and syncope associated with prolonged standing or standing in a hot and humid environment. The outlook for these patients is positive, with no risk of sudden cardiac events. Cardiac causes contribute to a very small percentage of patients with syncope and include structural lesions, arrhythmia, and poor contractility.

The workup of patients presenting with syncope comprises of detailed history-taking and a thorough physical examination. The goal of the evaluation is to identify high-risk patients with underlying heart disease, which may include ECG abnormalities (e.g., long QT syndrome, WPW preexcitation, and Brugada syndrome), cardiomyopathy, or structural heart diseases.

When the history is consistent with neurocardiogenic syncope, and when the family history and physical examination are not contributory, further tests are not required. While an ECG is frequently performed in the scenario, the yield of a positive result is quite low. These patients do not need follow-up with a pediatric cardiologist. The patient should be reassured about the benign nature of symptoms. Lifestyle changes to prevent postural hypotension should be discussed. A brain CT scan is not indicated under the circumstances mentioned in the vignette and should not be obtained. Referral to cardiology or neurology may be indicated for those with recurrent episodes of syncope. There is no indication for inpatient observation.

Take-Home Message

Most cases of syncope in the pediatric age group are benign and are secondary to neurocardiogenic cause (vasovagal). A detailed history of the event is frequently all that is needed to make the diagnosis. A negative family history and physical examination can help support the diagnosis. While the literature likely supports this approach, it would be reasonable to obtain a fingerstick, obtain a pregnancy test in the reproductive group women, and consider CBC (to look for anemia in teenage girls) and an ECG to evaluate for prolonged QT interval or ventricular hypertrophy.

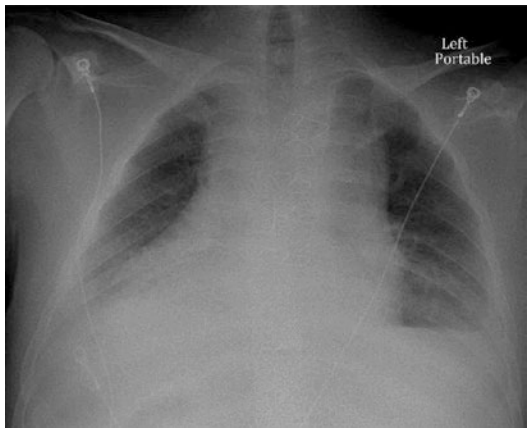
ABP Content Specification

- Understand the causes of syncope.

Question 16

A 5-year-old boy is brought to the Emergency Department by his father. He was discharged 3 days ago following a successful total cavopulmonary connection repair (Fontan operation) for single ventricle disease. His postoperative course was uncomplicated and was discharged home 5 days after surgery. He has a follow-up appointment scheduled with his cardiologist in 3 days. His father is concerned because the boy has been running a low-grade fever for the past 1 day. He is not as active and has difficulty breathing. His vital parameters are temperature of 100.5 °F (38.1 °C, heart rate of 136 beats/minute, respiratory rate of 32 breaths per minute, and blood pressure of 102/64 mm Hg. He has mild nasal flaring and use of suprasternal muscles. Air entry is decreased in the left lung field. Extremities are warm and well-perfused. First and second heart sounds are heard, and no murmur is evident. There is no organomegaly on abdominal palpation. A chest radiograph is obtained and shows haziness in the left hemithorax, with no increase in vascular markings. At this time, the most appropriate intervention is:

A. Discharge the patient home with twice daily oral furosemide.
B. Admit to the inpatient service for further investigation and possible pleurocentesis.
C. Reassure the father and ask him to keep the outpatient appointment.
D. Obtain CBC and blood culture and administer IV antibiotics.
E. Check renal function.



Correct Answer: B. Admit to the inpatient service for further investigation and possible pleurocentesis.

The boy in the vignette clinically has a pleural effusion, which is also borne out by the findings of the chest X-ray. This is a not an unexpected short-term issue following all kinds of cardiac surgical intervention. If the hemodynamic findings preceding a Fontan completion are unfavorable, then recalcitrant pleural effusions are very common. Depending on the rapidity of accumulation of fluid, the findings can include cardiorespiratory collapse to insidious symptoms such as low-grade fever and mild respiratory distress. The boy in this instance appears to be tolerating the effusion well. However, the rapidity with which the fluid has accumulated warrants close follow-up. In cases with hemodynamic embarrassment, the fluid may require emergent drainage followed by oral diuretic therapy. Isolated diuretic therapy without taking into account the speed of accumulation of fluid is not a great option. Likewise, discharging the patient under such circumstances, even with diuretic therapy, does not appear appropriate. The clinical scenario and chest X-ray findings do not suggest an infectious etiology or one related to renal dysfunction.

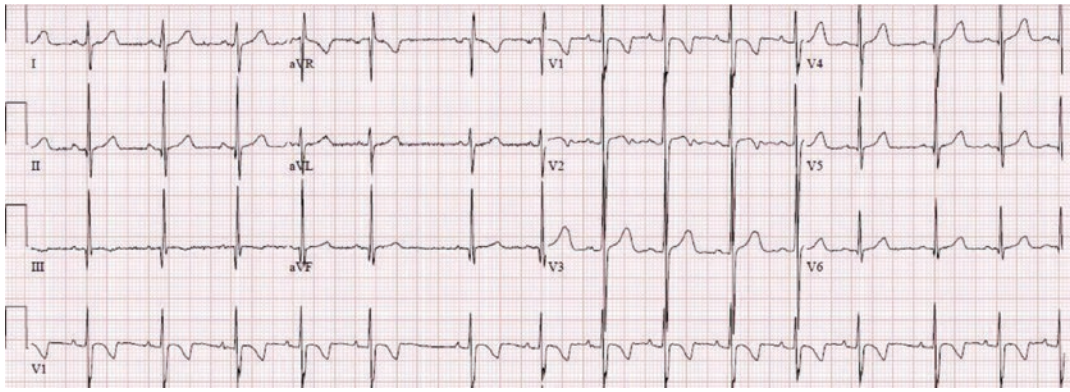
Take-Home Message
Patients recover quickly following open-heart surgical procedures and frequently are discharged home within the first week. Providers need to be cognizant of the potential complications that may manifest after discharge, such as pleural and pericardial effusions.

ABP Content Specification
- Know the etiology of pleural effusion.

Question 17

A 7-year-old girl has been complaining of chest pain for 2 days. She describes the pain as sharp and localized over her sternal region. She experiences the pain mostly while lying in bed. Her past

history is significant for mild-persistent asthma, which has been responding well to inhaled budesonide and albuterol therapy. For the past 2 days, she has had a bad cough and has been wheezing slightly, as per the mother. While getting a routine albuterol nebulizer treatment, she began crying due to acute chest pain. The nurse obtains a12-lead ECG (shown below).



Based on the ECG, the appropriate step in the management of this patient is:

A. Obtain CPK-MB fraction and troponin in view of inverted T waves in the anterior leads.
B. Change therapy to racemic epinephrine.
C. Continue with albuterol therapy, and advance management of acute asthma exacerbation.
D. Call the cardiologist on call for a stat echocardiogram.
E. Obtain a chest radiograph.

Correct Answer: C. Continue with albuterol therapy, and advance management of acute asthma exacerbation.

Chest pain is a common complaint in pediatric practice, but only about 4–6% of pediatric-age patients are likely to have a cardiac cause. Noncardiac causes such as musculoskeletal chest pain, acute asthma exacerbation, and gastroesophageal reflux are far more common. When the personal and family history is reassuring, further laboratory investigations are not necessary. Care must be taken in interpreting pediatric ECGs. While the T waves in the anterior precordial leads are upright for the first 48–72 hours, thereafter, the T waves stay inverted for the next 7–14 years. This should not prompt calls for more testing in these patients. A stat echocardiogram or chest radiograph is not indicated in this scenario. Likewise, racemic epinephrine is not indicated in this patient.

Take-Home Message

Pediatric chest pain is frequently noncardiac in origin. While ECGs are frequently performed as part of the evaluation of chest pain, the normal variations in ECG patterns in children need to be taken into account while interpreting them.

ABP Content Specification

- Know the etiology of chest pain in children and adolescents.

Question 18

A 15-year-old girl presents to the Emergency Department with an episode of syncope. She reports feeling progressively tired for the past month. She denies any viral infection prior to the onset of fatigue. This morning, she had her breakfast and was getting ready for school when she "passed out." Emergency medical personnel were summoned, but she had regained consciousness before they arrived. Her past medical history is

significant for a "heart condition" – her blood vessels are "backward." She reports being evaluated by a cardiologist a few months ago and was told that no intervention was necessary at that time, but that she should follow up at regular six-month intervals. In triage, her vital parameters are heart rate of 48 beats/minute, respiratory rate of 14 breaths/minute, blood pressure of 88/52 mm Hg, and oxygen saturation of 98% in room air. At this time, the most appropriate intervention for this patient is:

A. Make an appointment for an earlier than scheduled follow-up appointment with the cardiologist.
B. Obtain a urine pregnancy test.
C. Check Lyme titers on this patient.
D. Insert a transvenous temporary pacing wire.
E. Obtain an urgent ECG and make preparations for an isoproterenol infusion.

Correct Answer: E. Obtain an urgent ECG and make preparations for an isoproterenol infusion.

The girl in this vignette has significant bradycardia. She also gives the history of her blood vessels being "backward." This would fit with the clinical presentation of corrected transposition of the great vessels. This is a rare anamoly that accounts for 0.5% of all cases of congenital heart disease. It can present either in isolation or in association with other lesions such as VSD or pulmonic stenosis. When occurring in isolation, the defect may not be picked up as patients are not cyanotic, nor do they have a murmur. These patients usually present in adolescence or adulthood with symptoms related to dysfunction of the right ventricle which serves as the systemic ventricle or to bradycardia from high-grade atrioventricular block or complete heart block.

With a heart rate of 48 beats per minute, there is a high likelihood of the patient in this vignette having either high-grade or complete atrioventricular block. However, she is awake and alert and does not need emergent pacing.

If bradycardia is due to complete heart block, then emergency transcutaneous pacing should be performed. If there is refractory bradycardia, then cardiac pacing can be lifesaving, but it should be reserved for unconscious or sedated patients.

The use of a positive chronotropic agent such as isoproterenol could potentially increase the heart rate, blood pressure, and systemic effects while awaiting a permanent pacemaker. A 12-lead ECG will help in identifying the cause of the patient's bradycardia.

Pregnancy can be the cause of dizziness and postural hypotension but is less likely in this patient with a past history of being diagnosed with a heart defect, who also has bradycardia. While Lyme disease can cause atrioventricular block, there is no history of travel, illness, or skin rash to suggest that diagnosis. The patient in this vignette is symptomatic with bradycardia and hypotension and should be managed emergently rather than in an elective outpatient setting in the future with the pediatric cardiologist.

Take-Home Message

Patients with congenitally corrected transposition of the great vessels who have no other cardiac anomaly may present for the first time in adolescence or young adulthood with complete heart block. While the chronic bradycardia is usually well-tolerated, once heart block is established, the patient needs to be evaluated for placement of a permanent pacemaker.

ABP Content Specification

- Recognize signs and symptoms and life-threatening complications of congenital cardiac lesions by age.

Question 19

A 17-year-old girl is brought to the Emergency Department with uterine contractions. She is near-term and had spontaneous rupture of the membranes. She has had regular prenatal care. She has been followed by the maternal-fetal service for hypoplastic left heart syndrome (HLHS) in the fetus. Her labor is fairly rapid, and she is delivered of a 3.2 kg newborn male. After being

dried and warmed, the vital parameters of the newborn are as follows: heart rate 146 beats/minute, respiratory rate 28 breaths/minute, and oxygen saturation 91% in room air. You obtain an arterial blood gas analysis, which shows a PaO_2 of 39 mm Hg. Of the following, the most appropriate intervention at this time would be:

A. Allowing the child to bond with the parents and encourage breastfeeding.
B. Administer oxygen by nasal cannula.
C. Establish an intravenous line and start a prostaglandin infusion.
D. Intubate the child and provide ventilator support.
E. Obtain a CBC and blood culture and start antibiotics.

Correct Answer: C. Establish an intravenous line and start a prostaglandin infusion.

HLHS is a relatively uncommon cyanotic congenital heart lesion that affects around 2000 newborns a year in the United States. The defect is usually detected in fetal life on the basis of an abnormal ultrasound and fetal echocardiogram. About 10% of fetuses with HLHS are noted to have an intact or highly restrictive atrial septum. These fetuses are delivered by a planned C-section, as the neonates are likely to be quite sick and have high mortality rates. In other cases, delivery is planned after 39 completed weeks of gestation. Because this is a mixing lesion with unobstructed pulmonary blood flow, the oxygen saturations are usually in the high 80s to low 90s, with PaO_2 in the arterial blood around 35 to 45 mm Hg. The lesion is ductal-dependent, and hence institution of a prostaglandin infusion to keep the duct open is essential. The oxygen saturation and arterial PaO_2 are completely acceptable for this lesion, and maneuvers to increase the saturations are not likely to be helpful. In fact, by stimulating ductal closure, they may actually be harmful. In a stable patient with no risk factors, there is no indication for starting antibiotics.

Take-Home Message

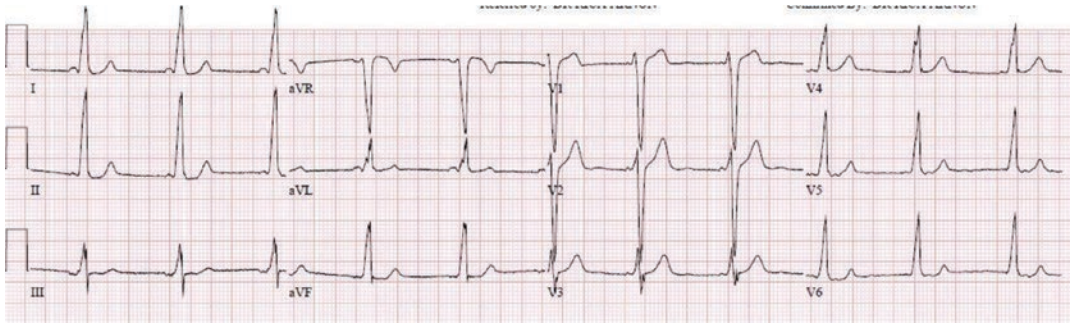
When the diagnosis of a duct-dependent lesion is made prenatally, the immediate postoperative management involves institution of prostaglandin therapy. This measure ensures adequate perfusion of the systemic or pulmonary circulation and prevents the development of acidosis or severe hypoxia.

ABP Content Specification

- Understand the pathophysiology and plan management of acutely symptomatic congenital heart disease.

Question 20

You are evaluating a healthy 16-year-old boy who had a screening electrocardiogram in his pediatrician's office as part of a pre-sport's participation physical. The pediatrician is concerned about abnormalities on the ECG. The patient denies any complaints of chest pain, palpitations, dizziness, or syncope. No family history of sudden cardiac death is noted. His vital parameters are normal for age, and cardiac examination is completely benign. You review the 12-lead electrocardiogram, which is shown as follows.



Your appropriate response with regard to sports participation would be:

A. All forms of physical exertion are permissible because personal or family cardiac history is not a concern.
B. The patient can participate in isotonic but not isometric exercises.
C. A detailed cardiac evaluation, including possibly an electrophysiology study, is necessary before the patient can take part in moderate-to-high intensity competitive sports.
D. The patient can participate in all sports as long as he is taking atenolol.
E. The patient can never participate in competitive sports.

Correct Answer: C. A detailed cardiac evaluation, including possibly an electrophysiology study, is necessary before the patient can take part in moderate-to-high intensity competitive sports.

The athlete in this vignette has evidence of preexcitation (Wolff-Parkinson-White) on the electrocardiogram. This has a prevalence of 0.1–0.3% in the general population and is frequently discovered as an incidental finding. Because these patients have an accessory pathway, atrial impulses can be transmitted to the ventricles with a shorter refractory period between consecutive impulses. Thus, the risk that atrial tachycardia can result in ventricular tachycardia is evident. However, the incidence of sudden cardiac death in an asymptomatic patient is very low (around 0.6%).

The approach to the asymptomatic younger patient with WPW on the ECG is somewhat uncertain. While the incidence of sudden cardiac death is very low, the young age of the patient implies that he might still be at risk for a first episode of supraventricular tachycardia, syncope, or sudden cardiac death. Hence, the recommendation is that such patients be evaluated, including possibly with an electrophysiology study. If the refractory period of the aberrant pathway is low, the pathway should be ablated. Following successful ablation, if there is no recurrence of the preexcitation on the ECG for 2 to 4 weeks, then the patient would be permitted to take part in all competitive sports. However, the use of atenolol alone does not mitigate the risk in these patients, and the patient would still not be permitted to play competitive sports.

Take-Home Message

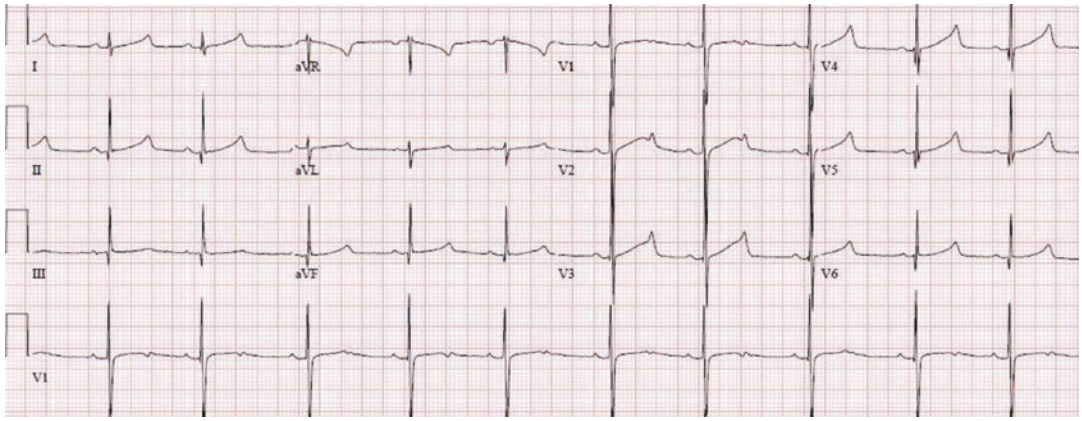
The standards for pre-participation clearance before taking part in competitive sports vary in different countries. While an ECG is not part of the workup in the United States, abnormalities on an ECG warrant evaluation by a cardiologist.

ABP Content Specification

- Recognize ECG abnormalities in children.

Question 21

A 12-year-old girl is brought to the Emergency Department after she fainted at home. She experienced a fast heart beat for a couple of minutes, felt dizzy, and "passed out." She lost consciousness for about 5 minutes, but had no loss of bladder or bowel tone, nor any tonic-clonic movements. By the time the EMT crew came home, she was awake and alert. In the ED, the patient appears well and in no distress. Her vital parameters are as follows: heart rate 70 beats/minute, respiratory rate 14 breaths/minute, and blood pressure 102/70 mm Hg. She is warm and well-perfused. Her pulse is regular. Her heart sounds are normal, and there is no murmur or rub. You review the ECG obtained on her in the ED, which is shown as follows.



At this time, the most appropriate next step in the management of this patient is:

A. Admit the patient for observation and request a cardiology evaluation.
B. Reassure the patient and her parents that her heart rate is within normal limits and discharge her.
C. Obtain intravenous access and give the patient a bolus of 20 ml/kg of normal saline.
D. Refer the patient to a geneticist for heritable arrhythmias.
E. Obtain a neurology consultation for syncope.

Correct Answer: A. Admit the patient for observation and request a cardiology evaluation.

Syncope is a common occurrence in the pediatric population and occurs in 15% of all patients by the end of adolescence. Fortunately, the vast majority of patients have benign vasovagal syncope. Cardiac causes of syncope, though extremely rare, are life-threatening. These would include pump failure as occurs with dilated cardiomyopathy and left ventricular obstructive lesions such as aortic stenosis, pulmonary hypertension, anomalies of coronary arteries, and malignant arrhythmias. While most malignant causes of cardiac syncope tend to manifest with exercise or strenuous activity, malignant arrhythmias may manifest without exertion. The history of preceding palpitations, the absence of postural changes precipitating syncope, and the somewhat prolonged duration of lack of consciousness make an arrhythmia a reasonable etiology for this patient's symptoms. The surface ECG of this patient shows normal sinus rhythm at a rate in the low 70s. However, the corrected QT interval, at 512 milliseconds, is prolonged.

Prolongation of the QTc interval may be secondary to the use of certain medications or to electrolyte abnormalities. Some individuals have congenital long QT syndrome, which is a heritable disorder. Because there is a potential for individuals with prolonged QTc interval to have a malignant polymorphic ventricular tachycardia, an individual with a history of syncope who has a prolonged QTc interval should be observed in a telemetry unit, until an electrophysiology study can be performed.

Torsade may occur in patients with prolonged QT interval during sinus rhythm. It is due to abnormal ventricular repolarization. In stable patients, the treatment of torsade is IV magnesium sulfate. In unstable patients, the treatment is defibrillation.

While the heart rate on the surface ECG is normal, the prolonged QTc warrants further workup, and reassurance of the patient and discharging her is not appropriate. There is no evidence of dehydration on physical examination in this patient, and fluid resuscitation is not called for. Congenital long QT syndrome is a heritable disorder, and genetic evaluation is part of the management of this patient with implications for the family unit. However, this can be done in an elective fashion. The patient's presentation does

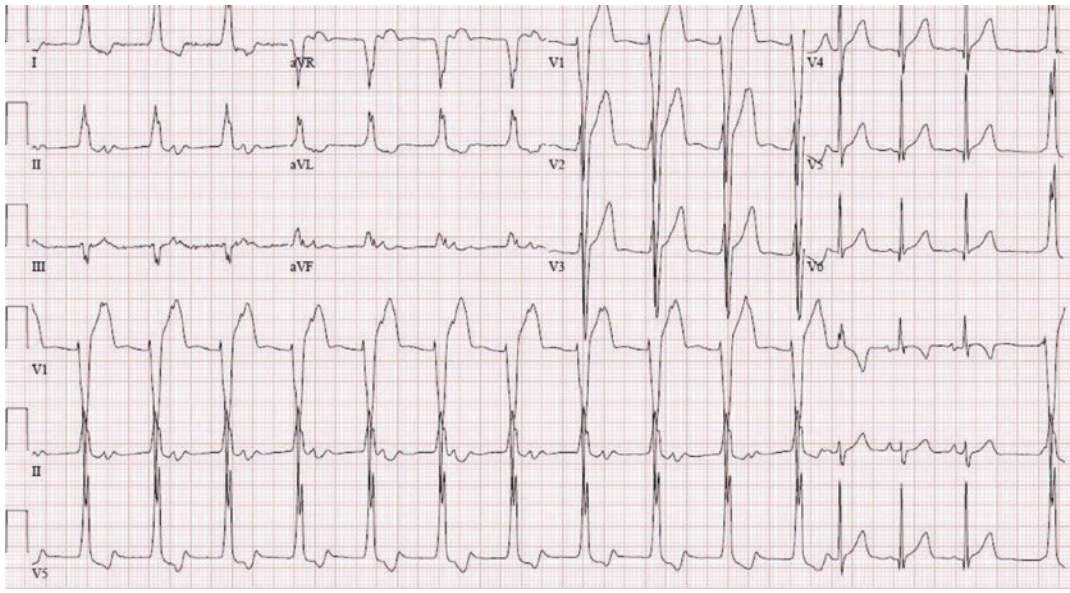
not suggest a neurological cause, and in the scenario where a cardiac cause appears likely, neurological consultation is not required.

Take-Home Message
While most cases of syncope in the pediatric age group are benign, a history of palpitations preceding the episode should prompt a detailed evaluation for a possible arrhythmia. Prolonged QT syndrome may present with ventricular tachycardia and syncope.

ABP Content Specification
- Recognize and plan the management of ECG abnormalities in children.

Question 22

A 6-year-old boy is transferred from a dialysis facility where he presented for his routine dialysis. His nurse noted that his heart rhythm tracing on his cardiac monitor appeared abnormal. On evaluation in the ED, the patient does not appear to be in any distress. His vital parameters are as follows: heart rate 90 beats/minute, respiratory rate 20 breaths/minute, and blood pressure 128/78 mm Hg. He is afebrile. He responds appropriately to your questions. Heart sounds are normal and no heart murmur is noted. Breath sounds are clear. Extremities are warm and well-perfused. The nurse brings over the 12-lead ECG for your review, which is shown as follows.



At this time, the next step in the management of this patient is:

A. Transfer the patient back for his routine dialysis treatment.
B. Electrical cardioversion with 0.5 J/kg of direct current.
C. Admit the patient for telemetry and obtain a complete set of electrolytes in the meantime.
D. Give intravenous potassium at a dose of 0.5 mEq/kg over a half hour.
E. Give oral potassium at dose of 2 mEq/kg/dose.

Correct Answer: C. Admit the patient for telemetry and obtain a complete set of electrolytes in the meantime.

The ECG of the patient in this vignette shows a wide-complex rhythm without a preceding P wave that spontaneously reverts to a sinus rhythm for two beats. This wide-complex rhythm is an

idioventricular rhythm until proven otherwise. Rhythm alterations may be noted in electrolyte abnormalities (commonly of potassium, calcium, and magnesium ions), which are present in patients with end-stage renal disease who have an impaired ability to maintain homeostatic levels of these ions. It is important to note that the patient in this vignette does not have ventricular tachycardia, nor does he have any hemodynamic compromise from his rhythm abnormality. Accordingly, the correct approach would be to assess his electrolyte balance and admit him for telemetry. Sending him back for dialysis without first working him up for his rhythm abnormality is inappropriate. As the patient does not have any hemodynamic compromise from his idioventricular rhythm, pharmacological or electrical intervention is not called for, and empirical therapy with potassium might even be dangerous.

Take-Home Message

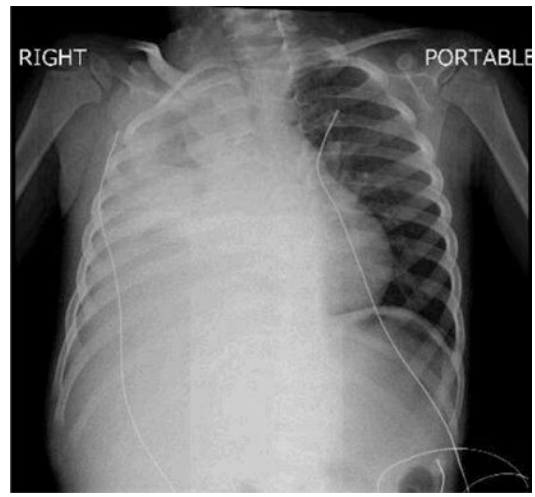
Rhythm abnormalities may be present in patients at risk for dyselectrolytemias. However, in the absence of hemodynamic compromise, they do not warrant emergent treatment, but rather, can be observed.

ABP Content Specification

- Recognize and plan management of acute cardiac dysrhythmias.

Question 23

The parents of a 3-year-old boy bring him to the Emergency Department because they are concerned about his breathing. He underwent surgical closure of an atrial septal defect 10 days ago. His postoperative recovery was uneventful, and he was sent home in 2 days. He appears to have recovered completely, with no history of fever, cough, chest pain, or decreased activity. Over the past 2 days, his parents have noted him to be breathing somewhat faster than before. He has a scheduled appointment with his cardiologist in 2 days. On examination, you notice a well-appearing child in no significant distress. His vital parameters are as follows: temperature 98.6 °F, heart rate 108 beats/minute, respiratory rate 36 breaths/minute, and blood pressure 98/60 mm Hg. Extremities are warm and well-perfused. While he is slightly tachypneic, he is not using accessory muscles of respiration. On auscultation, you do not hear any breath sounds over the right back. Heart sounds are heard, and there is 2/6 ejection systolic murmur over the left sternal border. You obtain a chest radiograph, which is shown as follows.



At this time, the best next step in the management of this patient is:

A. Instruct the parents that it is very important that they keep their follow-up appointment with their cardiologist.
B. Consult the cardiology team and admit the patient for medical management and observation.
C. Arrange for an emergent pleurocentesis.
D. Send the patient home on a regimen of furosemide.
E. Send the patient home on a regimen of steroids.

Correct Answer: B. Consult the cardiology team and admit the patient for medical management and observation.

The patient in this vignette has clinical and radiographic evidence of a large right-sided pleural effusion. This is probably secondary to a post-inflammatory response, which is seen 2 to 5 weeks following cardiac surgery. While the effusion is clearly quite large and occupies most of the right hemithorax, the child in question is not in significant distress. It might be appropriate to admit the child for observation and medical management with diuretics and anti-inflammatory agents (ibuprofen or steroids). If there is no significant change in the fluid volume, or if there is evidence of clinical decompensation, then a pleurocentesis, with placement of an indwelling pigtail catheter, can be performed. Despite the patient's lack of symptoms, the effusion is quite large. Asking the patient to follow-up with the cardiologist or sending him home on medications is not appropriate.

Take-Home Message

Following open-heart surgery, patients are at risk of developing a post-pericardiotomy syndrome which may manifest with a pleural and/or pericardial effusion. It may respond to medical therapy alone and occasionally might require drainage.

ABP Content Specification

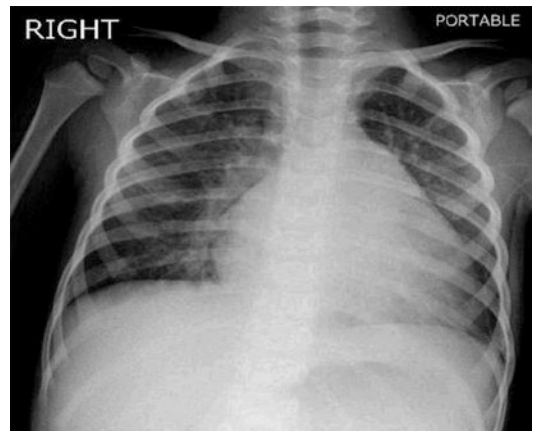
- Recognize the signs, interpret investigations, and plan management of pleural effusion.

Question 24

A 3-year-old girl has been seen in the Emergency Department for the third time in 3 weeks. She started attending preschool 3 weeks ago. She was exposed to a sick contact and developed a fever and runny nose 3 days after starting school. She was evaluated in the ED and diagnosed with a viral upper respiratory tract infection. Ten days later, the parents brought her again for the assessment of a cough. She was diagnosed with reactive airways disease and sent home with a nebulized bronchodilator. Today the parents have brought her with the complaint that the cough remains unresponsive to the medication. The child appears to be breathing heavily and gets tired quite quickly.

Her vital parameters are as follows: afebrile, heart rate 136 beats/minute, respiratory rate 36 breaths/minute, and blood pressure 86/48 mm Hg. Her extremities are warm and well perfused. Faint crackles are heard at the bases of her lung fields. Heart sounds are heard, and there is no murmur. The liver edge is felt 3 cm below the right costal margin.

A chest radiograph is obtained and is shown as follows.



At this time, the next best step in the management of this patient should be:

A. Obtain pediatric cardiology consultation.
B. Give the patient the first dose of prednisone and discharge her on a regimen of albuterol and prednisone.
C. Treat the patient with an antibiotic for pneumonia.
D. Obtain pediatric infectious disease consultation to exclude tuberculosis
E. Consult the pediatric pulmonologist and request a bronchoscopy for a foreign body.

Correct Answer: A. Obtain pediatric cardiology consultation

The patient in this vignette has a chest radiograph with an enlarged cardiac shadow, raising the

possibility of dilated cardiomyopathy or a pericardial effusion in someone with no previous history of heart disease. The prior history of a viral infection and the lack of muffled heart sounds on the physical examination makes myocarditis leading to ventricular dilation a more likely diagnosis. Children, especially neonates, appear to be vulnerable to fulminant viral myocarditis, which can progress to dilated cardiomyopathy.

The diagnosis is easily established by an echocardiogram. Treatment would depend on the clinical manifestations. Those with evidence of low cardiac output would benefit from the use of inotropes and diuretics, whereas those that are well-perfused but present with respiratory symptoms would benefit from the use of diuretics alone.

Patients with reactive airways disease should not have evidence of cardiomegaly or hepatomegaly. There is no evidence of pneumonia or a foreign body on the chest radiograph.

Take-Home Message

Tachypnea in a patient with no past history of asthma, and which is unresponsive to bronchodilators, should warrant additional investigations to rule out another etiology.

ABP Content Specification

- Recognize the signs and interpret investigations of cardiomyopathy.

Suggested Reading

Question 1

Sharoni E, et al. Superior vena cava syndrome following neonatal cardiac surgery. Pediatr Crit Care Med. 2001;2(1):40–3.

Question 2

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Question 3

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Question 4

Maron BJ, Thompson PD, Ackerman MJ, et al. Recommendations and considerations related to preparticipation screening for cardiovascular abnormalities in competitive athletes: 2007 update. Circulation. 2007;115:1643–55.

Question 5

Guidelines for the diagnosis of rheumatic fever. Jones Criteria, 1992 update. Special Writing Group of the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the Council on Cardiovascular Disease in the Young of the American Heart Association. JAMA. 1992;268:2069.

Question 6

Baddour LM, Wilson WR, Bayer AS, et al. Infective endocarditis: diagnosis, antimicrobial therapy, and management of complications: a statement for healthcare professionals from the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, and the Councils on Clinical Cardiology, Stroke, and Cardiovascular Surgery and Anesthesia, American Heart Association: endorsed by the Infectious Diseases Society of America. Circulation. 2005;111:e394.

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Question 10

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Question 11

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Question 12

Yang EH, Shah S, Criley JM. Digitalis toxicity: a fading but crucial complication to recognize. Am J Med. 2012;125:337–43.

Question 13

Beekman RH III. Coarction of the aorta. In: Allen HD, Shaddy RE, Penny DJ, Cetta F, Feltes TF, editors. Moss and Adams' heart disease in infants, children and adolescents: including the fetus and young adult. Philadelphia: Walters Kluwer; 2016.

Question 14

Chan CW. Evaluation of digital clubbing. Aust Fam Physician. 2015;44(3):113–6.

Question 15

Friedman KG, Alexander ME. Chest pain and syncope in children: a practical approach to the diagnosis of cardiac disease. J Pediatr. 2013;163:896.

Question 16

Gupta A, Daggett C, Behera S, et al. Risk factors for persistent pleural effusions after the extra-cardiac Fontan procedure. J Thorac Cardiovasc Surg. 2004;124:1664–9.

Question 17

Massin MM, Bourguignont A, Coremans C, et al. Chest pain in pediatric patients presenting to an emergency department or to a cardiac clinic. Clin Pediatr (Phila). 2004;43:231.

Question 18

Atallah J, Rutledge JM, Dyck JD. Congenitally corrected transposition of the great arteries. In: Allen HD, Shaddy RE, Penny DJ, Cetta F, Feltes TF, editors. Moss and Adams' heart disease in infants, children and adolescents: including the fetus and young adult. Philadelphia: Walters Kluwer; 2016.

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Question 22

Astan R, Akpinar I, Karan A, et al. The effect of hemodialysis on electrocardiographic parameters. Ann Noninvasive Electrocardiol. 2015;20(3):253–7.

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