

Cardiovascular Emergencies in the Pediatric Patient

William A. Woods, MD^{a,*},
Michael A. McCulloch, MD^b

^a*Departments of Emergency Medicine and Pediatrics, University of Virginia Health System,
PO Box 800699, Charlottesville, VA 22908, USA*

^b*Department of Pediatrics, University of Virginia Health System, PO Box 800386,
Charlottesville, VA 22908, USA*

Children with heart disease may present to the emergency department (ED) in many stages of life. They can present with undiagnosed congenital heart disease with their disease undiagnosed, after a temporizing procedure has been performed or after definitive repair. The practicing emergency physician needs to be prepared to make the diagnosis of congenital heart disease and to recognize complications common after a surgical correction of heart disease. Children can also present at any stage of life with syncope or chest pain; therefore, emergency physicians must also be comfortable with the most common types of heart disease associated with these symptoms.

The purpose of this article is to describe the physiology and presentation of undiagnosed congenital heart disease, to describe the complications that can occur after a staged or definitive repair, and to discuss acquired heart disease in children.

Evaluation of the child who has suspected congenital heart disease

The first step in recognizing a previously undiagnosed congenital heart disease is being familiar with the common presentations. In general, patients with significant congenital heart disease present with signs and symptoms of congestive heart failure (CHF), cyanosis, or shock. Infants manifest CHF as feeding difficulty, tachypnea, fussiness, or agitation. Physical examination

* Corresponding author.

E-mail address: waw9h@virginia.edu (W.A. Woods).

may reveal a murmur, tachypnea, diaphoresis, feeding intolerance, or hepatomegaly. In this setting, any murmur, especially if present in diastole, must be addressed. If a murmur is benign, it is imperative the emergency physician seek reassurance from the primary physician or a consultant.

Central cyanosis refers to excessive dissociated hemoglobin and is classically visible in the lips and mucous membranes [1]. Data suggest that 3 to 5 g/dL of dissociated hemoglobin are necessary before central cyanosis becomes visible [2–4]. Peripheral cyanosis, by comparison, is a manifestation of poor peripheral perfusion. In this situation, a decreased flow of oxygenated blood to the periphery exists with an increased oxygen extraction at the end organs. Often the lips and mucous membranes are spared. Thus, peripheral cyanosis can occur without hypoxia. Affected children can present with central and peripheral cyanosis. This combination can be seen when children with hypoplastic left heart syndrome (HLHS) or critical coarctation of the aorta present in shock.

The second step in identifying congenital heart disease is classifying the lesion. Significant congenital heart disease can be broken down into two groups: mixing and obstructive lesions. Mixing lesions can result in a left-to-right shunt (oxygenated blood mixes with deoxygenated blood and exits the heart into the pulmonary system) as in atrial or ventricular septal defects. These patients can present with pulmonary edema, heart failure, inadequate peripheral perfusion, and peripheral cyanosis. Mixing lesions can also result in a right-to-left shunt (deoxygenated blood mixes with oxygenated blood and exits the heart into the systemic circulation) as in a teenager with pulmonary hypertension as a result of an undiagnosed atrial septal defect. Right-to-left lesions may present with central cyanosis and progress to peripheral cyanosis, as seen in children having hypercyanotic episodes (“Tet spells”).

Symptomatic obstructive lesions can be a result of intracardiac obstruction (eg, valvular atresia or stenosis) or extracardiac obstruction caused by atretic vasculature (eg, pulmonary atresia). Obstructive lesions may present after closure of the ductus arteriosus, as in coarctation of the aorta. These patients often demonstrate peripheral cyanosis signs and symptoms of CHF or shock. Severe CHF may result in hypoxia with secondary central cyanosis.

Obstructive lesions that present in the ED are typically caused by closure of the ductus arteriosus. Other forms of obstruction lesions are usually identified in the nursery. Ductal dependent lesions typically become symptomatic within the first two weeks of life; however, reports of children up to 6 weeks old presenting with HLHS or coarctation are not uncommon.

This classification scheme is not perfect. For example, the child who develops HLHS may return home from the nursery with a mixing lesion balanced to keep them asymptomatic. When the ductus arteriosus closes the infant develops an obstruction to peripheral flow. This obstruction results in peripheral cyanosis, yielding an infant in shock. Although not perfect,

thinking of congenital heart disease in these terms help the treating emergency physician manage the patient until able to reach definitive care. No matter how complicated the congenital heart disease lesion, stabilizing treatment depends only on considering mixing and considering obstruction to flow; therefore, further attempts at classifying the lesions are unnecessary. Further description of cardiac lesions are not presented here. The reader is referred to standard texts [5,6] if interested and is urged to consider this classification system to deal with the lesions not listed in the standard texts, the proverbial “scrambled heart.”

One lesion poorly suited for this categorization is transposition of the great vessels. Children with this condition have a mixing lesion until the ductus closes, typically within the first 3 days of life. After closure, children suddenly develop severe central cyanosis due to significantly decreased mixing between the pulmonary and systemic circulations.

The third step in identifying children with undiagnosed congenital heart disease is the diagnostic evaluation in the emergency department. The initial diagnostic evaluation is the attempt at classification done by the history and physical examination. Further workup should be done to verify this suspicion. Blood pressures in all four extremities can identify an obstructive ductal dependent lesion presenting after closure of the ductus arteriosus. Blood pressures higher in the arms than the legs suggest a lesion becoming symptomatic with closure of the ductus arteriosus. Recording oxygen saturations in pre- and postductal extremities also assists in identifying a ductal dependent lesion. If blood pressures are adequate and comparable in all four extremities but oxygen saturations are lower in the legs, then right to left shunting is occurring at the ductus arteriosus after the takeoff of the head and neck vessels. The child, thus, has an open ductus arteriosus with a right to left shunt.

Chest radiography can identify a particularly abnormal heart size or shape. Radiography can also assist with determining the degree of pulmonary blood flow. A child who has a ventricular septal defect with mixing left-to-right may have pulmonary edema present on chest radiograph. Conversely, a child who has developed a ventricular septal defect and pulmonary atresia may have central cyanosis from right-to-left mixing across the defect, and that child may have a paucity of pulmonary vascularity on chest radiograph.

Similar to adults with shortness of breath, in children it may be difficult to determine if hypoxia is caused by a pulmonary or cardiac problem. The hyperoxygenation test may assist the physician in making this distinction. Arterial oxygenation should be greater than 150 mm Hg if a child is allowed to breath 100% oxygen for 10 minutes and there is a respiratory cause for the hypoxia. Values less than 100 mm Hg suggest congenital heart disease with a mixing lesion as the cause for the hypoxia [4].

No diagnostic evaluation of the child who has suspected congenital heart disease is complete without the consideration of alternative diagnoses,

such as dehydration, sepsis, intracranial hemorrhage, pneumonia, and hypoglycemia.

The final step when identifying children who have undiagnosed congenital heart disease is to initiate ED therapy. The aggressiveness of the initial therapy is determined by the classification of the lesion, the tempo of presentation, and the severity of the child's clinical condition. Mixing lesions typically present with gradual onset of symptoms, unless an associated obstructive ductal dependent lesion also exists. Left-to-right mixing lesions may worsen from supplemental oxygenation if pulmonary edema is present. The pulmonary vasculature differs from the systemic vasculature in that oxygen is a vasodilator; therefore, supplemental oxygen may cause a decrease in pulmonary artery pressure with a resultant increase in pulmonary edema and worsening central cyanosis. Alternatively, positive pressure ventilation may improve the shunt fraction. The increase in pulmonary artery pressure with positive pressure ventilation may decrease pulmonary blood flow (which is already excessive) and reduce central cyanosis (improve oxygenation).

Right-to-left lesions may improve with oxygenation. Again, oxygen decreases pulmonary vascular resistance, lessening the right-to-left shunt and improving oxygenation. Supplemental oxygen has a minimal effect on arterial oxygenation unless the shunt fraction alters because the inadequate flow of blood through the pulmonary vasculature enters the left atrium 100% saturated with oxygen. Conversely, positive pressure ventilation may increase pulmonary vascular resistance, worsening the shunt fraction and worsening arterial oxygenation. Intravenous (IV) fluid administration may be beneficial in these children and should be considered.

In the case of a hypercyanotic episode, or "tet spell," a child with the tetralogy of Fallot has an acute worsening of the right-to-left shunt after an increase in the intracardiac obstruction or decrease in systemic vascular resistance. In this case, placing the child in the knee chest position or compressing the abdominal aorta [7] increases systemic vascular resistance and may acutely decrease the right-to-left shunt.

Acute obstructive lesions presenting in the ED typically do so after closure of the ductus arteriosus. Infusion of prostaglandins may open the ductus, relieving the obstruction, and allowing the child to stabilize until they are able to reach definitive care. Complications of prostaglandin infusion include apnea, a mild decrease in systemic vascular resistance and nonpathologic hyperthermia [8,9]. Although unstudied, conventional teaching is that the risk for apnea increases with higher infusion rates. Initial infusion rate is controversial. Some clinicians prefer to start at a higher dose (0.10–0.20 $\mu\text{g}/\text{kg}/\text{min}$) and decrease the dose after the ductus reopens. Other investigators prefer to start at a lower dose (0.05 $\mu\text{g}/\text{kg}/\text{min}$) and increase the dose every 15 to 20 minutes until an effect is achieved. Although there is concern that starting prostaglandins in a child who has sepsis and not congenital heart disease, may worsen resuscitation and

outcome as a result of the decrease in systemic vascular resistance, no data support or refute this concern. Careful administration of a bolus of IV fluids may be beneficial in these children.

Corrected congenital heart disease

ED care of the child who has corrected congenital heart disease has received little attention in the emergency medicine literature; however, care of these children also can follow simple principles, allowing the emergency physician a starting place until consultation can be obtained.

Once a child is diagnosed with congenital heart disease, they may be discharged home with medical management until they grow large enough, which decreases their surgical mortality risk. They may also undergo a temporizing or initial surgical repair. Finally, children undergo definitive repair. They may present to the ED with a wide variation in original anatomy in various stages of surgical repair. Fortunately for the emergency physician, a few principles apply to these repairs.

One common problem across all surgical repairs is stenosis at the extracardiac surgical anastomosis. In this situation, the child resorts to presurgical physiology because whatever repair performed to encourage flow has become stenotic, resisting additional flow. Consider, for example, a child who has had a repair of coarctation of the aorta. If stenosis develops at the anastomosis, the child presents with symptoms of an obstructive lesion. Peripheral cyanosis in the lower extremities with decreased pulses results. Another example is the child who had a Blalock-Taussig shunt placed after diagnosis with tricuspid valve atresia with an associated hypoplastic right ventricle and pulmonary artery stenosis. Blood exits the right atrium directly into the left atrium. A Blalock-Taussig shunt is typically a gortex graft from the right subclavian artery to the right pulmonary artery. This shunt is a temporizing procedure, which creates a left-to-right shunt, allowing adequate mixing until the child grows. If stenosis develops at the anastomosis, the child may develop lessening pulmonary blood flow (which may be visible with comparison chest radiographs) with an increase in central cyanosis. The result is the child's preoperative physiology.

Another common complication in postsurgical patients is arrhythmias. Practically all patients that have undergone surgery for congenital heart disease are at increased risk for sudden death from a presumed ventricular arrhythmia. Those procedures that do not increase the risk for sudden death are an atrial septal defect repair and patent ductus arteriosus ligation. **Box 1** provides a list of common procedures associated with an increased risk for sudden death. No child who has undergone these procedures should be discharged from the ED after presentation with syncope or palpitations unless thoroughly discussed with the child's cardiologist.

Box 1. Corrected congenital heart disease lesions associated with sudden death*High Risk*

Tetralogy of Fallot

Aortic stenosis

Transposition of the great arteries

Coarctation of the aorta

Atrioventricular septal defect

Pulmonary stenosis

Anomalies that undergo the Fontan procedure

Low Risk

Ventricular septal defect

Because of the increased atrial pressures present before and sometimes after surgery, many of these patients are also at increased risk for atrial arrhythmias. Arrhythmias can also arise from the cardiac chamber manipulated during surgery. For example, a suture in the ventricular septal defect repair can be a focus of ventricular arrhythmia.

The final group of complications includes those specific to a particular surgical repair. The Fontan repair is the procedure with the longest list of serious complications. The Fontan (Fig. 1) is typically performed in the case of single ventricle physiology. The most effective ventricle becomes the systemic ventricle, whereas the superior and inferior vena cava drain directly into the right pulmonary artery. Box 2 contains a list of the most common complications of a Fontan procedure presenting after discharge from the hospital. Thromboembolism can occur in the pulmonary or systemic circulation. Thrombotic complications occurred in 25% of patients within 4 years after surgery in one study [10]. Twenty-five percent of these thrombi were in the systemic arterial circulation, whereas 75% were in the pulmonary circulation [10]. Many of the complications arise from the persistent increased central venous pressure that develops to drive pulmonary blood flow [11,12]. If a fenestrated Fontan is performed, a “pop-off valve” exists between the inferior vena cava and the right atrium. Because of the fenestration, it is possible for these children to develop cyanosis with a significant right to left shunt across the fenestration.

The Blalock-Taussig shunt is a temporizing procedure whereby the right subclavian artery is anastomosed to the right main pulmonary artery. In addition to the risks for stenosis at the anastomosis, these shunts may thrombose when a child is dehydrated. Additionally, the ipsilateral vertebral artery may have been intentionally ligated to prevent a basilar steal phenomenon. The long-term risks of this ligation are unknown.

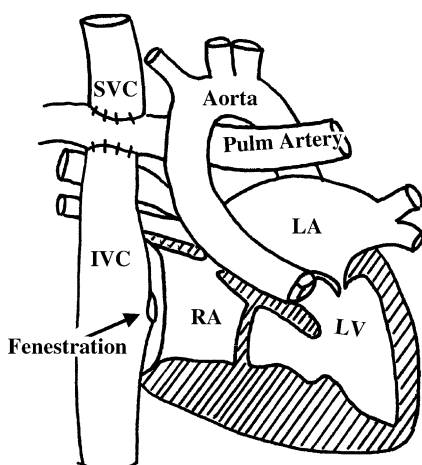


Fig. 1. Fenestrated Fontan in repair of tricuspid atresia with resultant hypoplastic right ventricle. Inferior vena cava and superior vena cava drain into right pulmonary artery. Fenestration into the right atrium from the inferior vena cava.

Surgical repair of coarctation of the aorta carries another set of post-operative risks. One recent series details the complications of 383 patients available for follow-up of 580 patients who had repair. Aneurysmal dilation at the site of the coarctation occurred in 2% of patients. Two patients had massive hemoptysis from an aortobronchial fistula. Intracardiac stenosis requiring surgery occurred in three patients, and one patient had a rupture of a sinus of Valsalva aneurysm [13].

Heart transplantation

Children presenting to the ED after orthotopic heart transplantation have complications that differ from other children who have corrected

Box 2. Complications that may occur after a Fontan procedure

- Thromboembolism
- Thrombosis
- Increased central venous pressure
- Ascites
- Pulmonary effusion
- Pericardial effusion
- Protein-losing enteropathy
- Atrial arrhythmia
- Sudden death

congenital heart disease and require separate attention. These children may present to the ED with an invasive infection, acute/chronic rejection or with posttransplant lymphoproliferative disorder.

Infectious complications in transplant recipients may be caused by viral, bacterial, protozoan, or fungal infections. Most bacterial infections occur within the first month after transplant. Viral infections predominate over the following month. Throughout the life of the transplant recipient, an increasing risk for protozoan or fungal infections exists [14].

In a review of 74 outpatient febrile episodes in 22 pediatric heart transplant patients, 22 of 74 (30%) were considered serious and required inpatient therapy [15]. Three episodes were pneumonia, two were caused by cytomegalovirus (CMV) infections, and two were caused by streptococcal infection. One child in the series died.

No data are available to reassure the emergency physician that clinical presentation or acute laboratory evaluation can exclude the presence of bacterial illness in the transplanted patient who has an acute illness. In a small series, pneumococcal bacteremia occurred in 9 of 80 patients (11%) at 3 to 48 months posttransplant [16]. Of those 9 children, 3 had pneumonia, 2 presented with otitis media, and 1 child had petechiae and purpura. Most of these children, but not all, had a temperature higher than 101°F at presentation.

Pneumocystis carinii pneumonia (PCP) can present with symptom durations from 1 to 2 days to 2 weeks [17]. In this small series, all 10 patients had tachypnea and room air oxygen saturations less than 95%.

CMV is a recognized cause of morbidity in the transplant recipient [18]. CMV infections can present as a mononucleosis-type syndrome, an interstitial pneumonitis, a hepatitis, or as a gastrointestinal disease. The symptoms are typically nonspecific and include headache, myalgias, abdominal pain, and diarrhea. CMV-mononucleosis differs from Epstein-Barr Virus (EBV) mononucleosis because it does not include pharyngitis, tonsillitis, or splenomegaly.

Acute rejection has been defined as the clinical decision to intensify immunosuppression in association with histopathology or dysfunction [19]. The distinction between acute and chronic rejection does not refer to the time of onset after transplantation. Acute rejection is a more fulminant process than chronic rejection. Children may be asymptomatic or present in cardiogenic shock. Common symptoms include fever, myalgias and vomiting. Chest pain may occur, but is uncommon. The electrocardiogram may show a decreased R wave amplitude and an increased QRS duration; however, these findings are not sensitive. Laboratory findings are not helpful. There is no data to suggest that cardiac enzymes are reliably elevated with acute rejection. Elevated hepatic transaminases, although nonspecific for acute rejection, always suggest an invasive complication in the transplant recipient.

Chronic rejection differs microscopically from acute rejection. The clinical manifestation of chronic rejection is accelerated atherosclerosis.

Symptoms are caused by ischemia or infarction and usually include decreased exercise tolerance or malaise. Patients may also present after syncope or sudden death. As with acute rejection, no specific laboratory, or ECG findings exist to secure this diagnosis.

Posttransplant lymphoproliferative disorder (PTLD) is typically a B-cell lymphoma associated with transplant patients [20,21]. The cause is most likely a result of EBV. Presenting symptoms include fever, malaise, a mononucleosis-type syndrome, palpable lymphadenopathy, or mass effect from tumor location. The most common location of the masses are (in descending order): abdomen, thorax, head and neck, and brain [21]. Abdominal PTLD may present with abdominal pain, hepatomegaly, splenomegaly, an abdominal mass, gastrointestinal bleeding, anemia, intussusception, diarrhea, or a protein-wasting enteropathy [21].

In caring for the transplant recipient with an acute illness, the emergency physician should perform several steps. A careful history allows the treating physician to identify a clinical syndrome consistent with a self-limited or more aggressive illness. Noncompliance with the immunosuppressive regimen and PCP prophylaxis increases the risk for acute rejection [22] or acute PCP infection [17]. After a careful physical examination, a chest radiograph and ECG should be obtained in all children who experience an unclear symptom complex. A comparison ECG is necessary because changes including right bundle branch block, left atrial enlargement, and right ventricular hypertrophy are not associated with clinical deterioration [23]. Laboratory studies should include a complete blood count with differential, serum electrolytes, hepatic transaminases, and blood cultures.

The admission recommendations are unclear for children who develop an obvious etiology for their symptoms. The emergency physician should consider admission for any child who experiences a significant fever or fever history. Children with vomiting without a history clearly suggestive of acute viral gastroenteritis are also a concern. Dehydration should be treated more aggressively in transplant recipients because of the nephrotoxicity of their immunosuppressive regimens.

Infective endocarditis

Infective endocarditis refers to a condition in which an organism infects the valves, endocardium, or other cardiac structures. The term acute bacterial endocarditis refers to a fulminant illness whereby death can occur within weeks. Subacute bacterial endocarditis refers to an indolent course evolving over months. The most common bacterial causes of infective endocarditis are *Staphylococcus aureus*, *Streptococcus pyogenes* and *Streptococcus viridans*. The most common symptoms in patients who have infective endocarditis are fever, malaise, chest and abdominal symptoms, and arthralgias. Physical findings may include embolic events, a changing or new murmur, splenomegaly, and petechiae. The classically described findings of

Roth spots, Osler's nodes, Janeway lesions, and splinter hemorrhages are uncommon. The most common laboratory abnormalities in patients who have infective endocarditis are elevated erythrocyte sedimentation rate (ESR), anemia, hematuria, and positive blood cultures.

Children who have congenital heart disease are at an increased risk for bacterial endocarditis. Surgical manipulations of the aortic valve or aorta are the highest risk anomalies. Surgically corrected septal defects or right-sided lesions are at a low risk. The cause of the initial bacteremia leading to endocarditis is frequently unknown. In the most comprehensive series available, a precipitating event could be identified in only 87 of 214 cases of endocarditis in patients who had congenital heart disease [24]. Of these 87, 42 cases were associated with dental procedures and 10 were associated with skin infections. Interestingly, patients received appropriate endocarditis prophylaxis in one half of the cases occurring after dental procedure [24].

The extensive series by Li and Somerville [24] does not list any cases of bacterial endocarditis occurring after traumatic injury. Current recommendations do not suggest endocarditis prophylaxis for cases of traumatic injury [25,26]. Investigators state that prophylaxis is not indicated for urethral catheterization without infection or for endotracheal intubation [25,26]; however, one investigator recommends prophylaxis for urethral catheterization in the face of a urinary tract infection [26].

Endocarditis prophylaxis is indicated for all patients with congenital heart disease. Prophylaxis is not indicated for patients at least 6 months from a surgically repaired ventricular septal defect or patent ductus arteriosus without residual defect. Patients with an isolated atrial septal defect do not require prophylaxis before repair or 6 months after complete repair [25,26]. **Box 3** details the current dosing regimen recommended by the American Heart Association for infective endocarditis prophylaxis.

Arrhythmias or congestive heart failure in children

Not all cardiac causes of acute illness in children are a result of congenital heart disease. Children may present to the ED with syncope, palpitations, chest pain, or shortness of breath. The following section describes the most common pathologic cardiac conditions that may result in ED presentation. Before starting though, it is necessary to have an understanding of the normal age-related progression of the pediatric ECG.

Normal changes in the pediatric ECG

The pediatric ECG is an evolving process, changing on a nearly weekly basis as pulmonary vascular resistance (PVR) decreases, left ventricular (LV) mass increases relative to right ventricular mass (RV), and various other changes occur in the developing cardiovascular system. Though the basic premises are unchanged, physicians must be familiar with these age-

Box 3. Endocarditis prophylaxis recommendations*Dental, oral, respiratory tract, or esophageal procedures*

Give 1 hour before procedure:

Amoxicillin (50 mg/kg with a maximum of 2.0 g) by mouth.

No follow-up dose recommended.

Genitourinary or gastrointestinal procedures in high risk patients

Give within 30 minutes of starting procedure:

Ampicillin (50 mg/kg with a maximum of 2.0 g) and

Gentamicin (1.5 mg/kg not to exceed 120 mg) IV.

Give 6 hours after the procedure:

Ampicillin or amoxicillin (25 mg/kg with a maximum of 1.0 g).

For more information see: Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease. Prevention of bacterial endocarditis: recommendations by the American Heart Association. *JAMA* 1997;277:1794–1801; *Circulation* 1997;96:358–66; *JADA* 1997;128:1142–50.

related variations associated with pediatric ECG to correctly interpret its data. This discussion focuses on this and anatomic variations specific to the pediatric population from birth to 2 months of age to help physicians accurately detect anomalies during this critical period.

Most of the age-related variations demonstrated in the pediatric ECG are because of the changes in the ratio of LV to RV mass. As PVR continues to decrease relative to systemic vascular resistance (SVR), RV dominance rapidly wanes from an LV/RV ratio of 0.8 to 1.5 by 1 month of age. Concurrent with these changes is a gradual decrease in the severity of the right QRS axis deviation. The precordial R/S ratios also change, increasing and decreasing in the left and right precordial leads, respectively. As a point of reference, the newborn ECG may demonstrate a complete reversal of the adult R/S progression, but by 1 month of age, the R/S ratio is typically greater than 1 in V6 and between 1–6 months of age, the R/S ratio in V2 also approaches 1. During this last age range, it is not uncommon to erroneously suspect bilateral ventricular hypertrophy based on the large precordial QRS deflections.

T waves are typically dynamic during the first week of life. At birth the T waves are upright in V1 and V6 with the vector anterior and to the left. Within a few days the vector changes rightward leaving flattened or inverted T waves laterally. By the end of the first week of life, the T wave axis changes posterior and leftward, producing inverted T waves in lead V1 and upright T waves in lead V6. The inverted T wave in V1 may not turn upright until late adolescence.

Supraventricular tachycardia

Pediatric arrhythmias in the first 2 months are rare but can become hemodynamically significant. The most commonly experienced arrhythmia in this age group is supraventricular tachycardia (SVT), generally producing narrow complex heart rates greater than 240 beats per minute. At these rates, P waves (if present) are often buried in the preceding T waves making it difficult to differentiate an atrial from a nodal focus. SVT is not, however, always associated with a heart rate greater than 240. An inappropriately elevated heart rate in an otherwise calm or sleeping infant is SVT until proven otherwise. Any infant found to have an initial episode of SVT should be thoroughly evaluated by a pediatric cardiologist to rule out problems, such as SVT-induced CHF, anatomic anomalies (ie, Ebstein's anomaly), or a reentrant/aberrant pathway as with Wolff-Parkinson-White (WPW) syndrome. The prognosis for infants with SVT is excellent with 60% to 90% of patients experiencing complete remission of symptoms by 6 to 12 months of age.

Wolff-Parkinson-White syndrome

WPW syndrome is associated with ventricular pre-excitation by way of an accessory pathway between the atria and ventricles. These accessory pathways do not demonstrate the obligatory pause as seen in the atrioventricular node. As a result, ventricular pre-excitation is responsible for the short PR interval and delta waves frequently detected on ECG.

WPW typically occurs spontaneously; however, familial clusters have been described. Pre-excitation can be intermittent, making true prevalence in the population difficult to define. WPW frequently coincides with congenital cardiac abnormalities, including Ebstein's anomaly, hypertrophic cardiomyopathy, and atrial septal defects.

The WPW syndrome refers to a patient with symptoms of tachyarrhythmias in the face of ECG evidence of pre-excitation. Three ECG features are described: (1) short PR interval (<0.12 seconds), (2) a prolonged QRS duration (>0.12 seconds), and (3) a delta wave or a slow gradual upslope leading into the QRS complex. The degree of pre-excitation detected on the ECG depends on the relative conduction times between the atrioventricular node and the accessory pathway. Measures to slow conduction through the atrioventricular node may make an accessory pathway more prominent. Described methods include Valsalva or vagal maneuvers and adenosine therapy.

Not all patients with ECG evidence of pre-excitation are symptomatic. Most of those with symptoms are limited to chest discomfort, dizziness, syncope, or shortness of breath [27]. The most common arrhythmias in this population are atrial fibrillation and orthodromic reciprocating tachycardia. Orthodromic reciprocating tachycardia is a type of paroxysmal supraventricular

tricular tachycardia that allows antegrade conduction down the atrioventricular node and retrograde conduction in the accessory pathway. Episodes of the less common antidromic tachycardia place the patient at higher risk for ventricular fibrillation [28].

Although atrial fibrillation is common in patients who have WPW, its occurrence in the presence of rapid conduction across an accessory pathway may predispose a patient to ventricular fibrillation [29,30]. Medications that slow conduction across the atrioventricular node should not be used in patients who develop atrial fibrillation and WPW syndrome. Agents, such as digoxin, calcium channel blockers, beta-blockers, and adenosine, may increase the risk for ventricular fibrillation. Procainamide or amiodarone are drugs of choice for rate control in this situation.

Anomalous coronary artery

Anomalous origin of a coronary artery, although a rare form of congenital heart disease, can cause myocardial ischemia in the infant or syncope in the young adult. Many types of congenital coronary artery abnormalities are described [31]; however, the two most common variations that cause symptoms are discussed here.

The left main coronary artery may arise from the pulmonary artery. This rare lesion occurs in 1 of 300,000 children [32]. Infants with this anomaly present after infarction or with angina. Children with angina may be diagnosed with colic due to inconsolable crying, especially after feeding. In Mahle's series [33], the most common finding in the 11 children identified were cardiomegaly on chest radiography and ischemia on ECG [32]. The classic ECG finding are deep Q waves in leads I and aVL with poor R-wave progression across the precordium [31].

If the left coronary artery originates from within the right coronary sinus or from the right coronary artery, syncope and sudden death may occur in adolescence. Syncope and sudden death are most common if the left coronary artery courses between the pulmonary artery and aorta. Left coronary artery blood flow may be asymptomatic until, classically, flow is impaired by external compression from the aorta and pulmonary artery leading to ischemia, arrhythmia, or sudden death. This occurs during exertion when cardiac output increases causing a mild dilation in diameter of the aortic and pulmonary artery roots. Detection of this lesion requires diagnosis of acute ischemia or clinical suspicion confirmed by echocardiography or catheterization.

Kawasaki Disease

Kawasaki Disease (KD), formerly described as mucocutaneous lymph node syndrome, was originally defined by Kawasaki in a series of 50 cases in 1967 [34]. KD is an acute febrile syndrome of childhood of uncertain

etiology that is also the leading cause of acquired heart disease and myocardial infarction (MI) in children in the United States. Eighty percent of all cases occur in children less than 5 years old. KD is rare in infants less than 4 months old and in adults.

Diagnostic criteria for KD include fever of at least 5 days, and four of the following five findings: (1) nonpurulent bilateral bulbar conjunctivitis, (2) polymorphous exanthema, (3) cervical lymphadenopathy, (4) involvement of the extremities, and (5) changes in the lips and oral cavity (injected pharynx or lips, fissured lips or strawberry tongue). The polymorphous rash is typically truncal without vesicles. Lymph nodes must be at least 1.5 cm in diameter. Extremity involvement includes edema and erythema of the hands and feet followed by periungual desquamation. Other findings in children who have Kawasaki Disease may include extreme irritability, arthritis, gallbladder hydrops, mild hepatitis, uveitis, urethritis, aseptic meningitis, or myocarditis. Laboratory abnormalities may include thrombocytosis (usually in the second and third weeks of illness), elevated sedimentation rate, sterile pyuria, and proteinuria. At its peak, the platelet count may exceed 1,000,000. ECG changes that may be detected during the acute phase include PR prolongation, corrected QT prolongation, and T-wave flattening.

Children may have symptoms of myocarditis or pericarditis during the acute phase; this is typically self-limited. Aneurysms of the coronary arteries may develop during the second and third weeks of illness. Those with aneurysms may suffer acute MI. While occurring in less than 3% of patients, MI typically occurs within 1 year of onset of symptoms and frequently within 3 months of onset of symptoms. MIs may be silent, or the child may present with abdominal pain, vomiting, or crying.

Treatment in the acute phase of Kawasaki Disease in the ED is supportive. Intravenous fluids are typically required because the children have had poor oral intake with increased insensible losses. Aspirin therapy and intravenous gamma-globulin have been shown to decrease complication rates; however, these therapies would not typically be started without appropriate consultation.

Myocarditis

Myocarditis refers to an inflammatory disease of the myocardium that is not caused by acute ischemia. Myocarditis, then, is a clinico-pathologic diagnosis, which does not refer to a specific etiology. While most myocarditis in the United States is viral, the cause can be bacterial, rickettsial, protozoal, parasitic, fungal, autoimmune or pharmacologic [35,36]. Coxsackie B virus, an enterovirus, and adenovirus are the most common viral causes of myocarditis. While newborns are particularly susceptible to coxsackie B myocarditis, all ages are at risk. Presenting symptoms include fever, poor appetite, irritability, sudden death, diaphoresis, pallor, and congestive heart

failure. Older children may not present until dilated cardiomyopathy is already present. Older patients are more likely to present with a gradual onset of congestive heart failure, or atrial or ventricular arrhythmias.

The electrocardiogram may demonstrate sinus tachycardia, low-voltage QRS complexes, and inverted or low-voltage T waves. Wide Q waves and ST-segment changes may also be apparent. The chest radiograph typically shows a dilated heart and pulmonary edema. Treatment is supportive until the child can be transferred to a center whereby aggressive diagnostic evaluation and therapy can be performed.

Hypertrophic cardiomyopathy

Hypertrophic cardiomyopathy, in this section, refers to the idiopathic condition sometimes referred to as idiopathic hypertrophic subaortic stenosis or familial hypertrophic subaortic stenosis. Hypertrophic cardiomyopathy can also be a descriptive diagnosis that includes cardiac conditions secondary to obstructive congenital heart disease (aortic stenosis) or an inborn error of metabolism (glycogen storage disease).

The left ventricular hypertrophy in hypertrophic cardiomyopathy is variably asymmetric and, at the cellular level, myocardial cells are organized in a bizarre fashion. Commonly, this condition is a result of mutations in the cardiac myosin heavy-chain gene. A familial component commonly exists to this disorder.

Clinical manifestations are typically caused by obstruction of flow or arrhythmias. The disorganized contraction of the hypertrophic section of ventricle results in subaortic outflow obstruction during systole. The increased ventricular pressure required to overcome this obstruction results in increased myocardial-wall stress and oxygen demand. Arrhythmias may result from this ischemia or may result from the arrhythmogenic foci of the bizarrely arranged myocardial fibers.

Physical examination of the child who has developed hypertrophic cardiomyopathy may reveal a systolic murmur exacerbated by a Valsalva maneuver or by standing. The murmur is best detected at the left lower sternal boarder or at the apex. A prominent left ventricular lift may exist on palpation. ECG frequently reveals left ventricular hypertrophy and ST-segment and T-wave changes. Chest radiograph is typically normal. Treatment at presentation is limited to supportive therapy for the patient's clinical condition. Consultation is necessary even in children who experience only mild symptoms.

Summary

Although the myriad of congenital heart lesions and their surgical repairs can seem overwhelming, ED evaluation and care of children who experience

these conditions requires the practitioner to understand a few principles. A solid understanding of the physiology of cyanosis helps the clinician determine the appropriate diagnostic studies and interpret the results. Remembering common patterns of postsurgical complications allows the physician to initiate treatment in the child who has surgically corrected congenital heart disease, even if the family cannot relate the surgical history to the physician. Finally, reviewing the common pathologic cardiac causes of CHF and syncope should allow the physician to appropriately exclude a pathologic cause of syncope or palpitations in a child.

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