Essentials of Cardiology

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Congenital Heart Disease

Incidence

Estimates of the incidence of congenital heart disease (CHD) range from 0.3% to 1.2% in live neonates.[1] This represents the most common form of congenital pathology and a major cause of mortality during the neonatal period.[2] As medical, interventional, and surgical therapies continue to improve, survival into adulthood has become the expectation for most congenital cardiovascular malformations.[3] At present it is estimated that there are more than a million adults with CHD in the United States, surpassing the number of children similarly affected for the first time in history. A similar trend of an increasing number of adults with CHD, also known as "grown-up congenital heart disease" (or GUCH) patients, has also been reported in Canada and several European countries.[4-8]

Overall, a bicuspid aortic valve (Video Clip 14-1) is the most common cardiac defect, occurring in up to 1% of the population.[9,10] Ventricular septal defects (Video Clip 14-2) represent the next most common congenital lesion,[9,11-15] followed by secundum atrial septal defects (Video Clip 14-3).[9,16] Among the cyanotic conditions, tetralogy of Fallot is the most common, affecting nearly 6% of children with CHD.[17] In the first week of life, however, D-transposition of the great arteries is the most common cause of cardiac cyanosis, because a subset of infants with tetralogy of Fallot will either be acyanotic or mildly desaturated at birth so that their cardiac disease will go undetected until later in life.

Segmental Approach to Diagnosis

A number of classification schemes have been proposed to characterize and categorize the various congenital cardiac malformations.[18-24] The one known as "the segmental approach to the diagnosis of CHD" assumes a sequential, systematic analysis of the three major cardiac segments (atria, ventricles, and great arteries) to characterize the abnormalities in a given patient. The guiding principle of this approach is that specific cardiac chambers and vascular structures have characteristic morphologic properties that determine their identities, rather than their positions within the body.[25] An organized, systematic identification of all cardiac structures or segments and their relationships to each other (connections or alignments between the segments) is carried out to define a given patient's anatomy.[26]

The initial steps to characterize the anomalies and classify a child's cardiovascular disease are to determine the cardiac position within the thorax and the situs of the thoracic and abdominal organs. The position of the heart can be described in terms of its location within the thoracic cavity and the direction of the cardiac apex. For simplicity, the following approach is frequently used. The term *levocardia* indicates that the heart is in the left hemithorax, as is normally the case. *Dextrocardia* specifies that the heart is located in the right hemithorax and *mesocardia* that the heart is displaced rightward but not completely in the right thoracic cavity. It is important to consider that an abnormal location of the heart within the thorax (cardiac malposition) may result from displacement of the heart by adjacent structures or underlying noncardiac malformations (e.g., diaphragmatic hernia, lung hypoplasia, scoliosis). The visceral situs or sidedness of the abdominal organs (liver and stomach) and atrial situs are considered independently. The visceral situs is classified as *solitus* (normal arrangement of viscera; liver on the right, stomach on the left and single spleen on the left), *inversus* (inversion of viscera; liver on the left, stomach on the right), or *ambiguous* (indeterminate visceral position). Abnormal arrangements or sidedness of the abdominal viscera, heart, and lungs suggests a high likelihood of complex cardiovascular pathology. The atrial situs, atrioventricular connections, ventricular looping (referring to the position of the ventricles as a result of the direction of bending of the straight heart tube in early development), ventriculoarterial connections, and relationship between the great vessels are then delineated. Finally, any associated malformations are described, including number and size of septal defects if present, valvar and/or great vessel abnormalities, and so on. Whereas many types of congenital defects fall neatly into this classification scheme, others, such as heterotaxy syndromes (a condition associated with malposition of the heart and abdominal organs) are often more difficult to precisely define.

Physiologic Classification of Defects

The wide spectrum of cardiovascular pathology in the pediatric age group presents a unique challenge to the clinician who does not specialize in the care of these children. Even for those with a focus or interest in cardiovascular disease, the range of structural defects and the varied associated hemodynamic perturbations can be overwhelming.

Although the segmental approach is extremely helpful in characterizing CHD, there are many instances when a physiologic classification system can facilitate understanding of the basic hemodynamic abnormalities common to a group of lesions, congenital or acquired, and assist in patient management.[27,28] Several pathophysiologic classification schemes have been proposed, including some that categorize structural defects into simple versus complex lesions, consider the presence or absence of cyanosis or whether pulmonary blood flow is increased or decreased, and so on.[29-31] The following classification approach groups pediatric heart disease into six broad categories according to the underlying physiology or common features of the pathologies.

Volume Overload Lesions

Volume overload lesions are typically due to left-to-right shunting at the atrial, ventricular, or great artery levels. In general, if the location of the left-to-right shunt is proximal to the mitral valve (e.g., as is the case in atrial septal defects, partial anomalous pulmonary venous return, or unobstructed total anomalous pulmonary venous return), right heart dilation will occur. Lesions distal to the mitral valve (e.g., ventricular septal defect, patent ductus arteriosus, truncus arteriosus) lead to left-sided heart dilation. Children with atrioventricular septal defects (also known as atrioventricular canal defects) also fit into this category. The magnitude of the shunt and resultant pulmonary to systemic blood flow ratio ($\dot{Q}p:\dot{Q}s$) dictates the presence and severity of the symptoms and similarly guides medical and surgical therapies. Diuretic therapy and afterload reduction are beneficial in controlling pulmonary overcirculation and ensuring adequate systemic cardiac output. Transcatheter approaches or surgical interventions may be required to address the primary pathology associated with ventricular volume overload (see Chapter 20).

Obstruction to Systemic Blood Flow

Lesions characterized by systemic outflow tract obstruction include those with ductal-dependent systemic blood flow. These range from critical aortic stenosis, severe coarctation of the aorta, interruption of the aortic arch, and hypoplastic left heart syndrome. Prostaglandin E_1 infusion maintains ductal patency and ensures adequate systemic blood flow until either surgical or transcatheter intervention is performed in the first few days of life to relieve the systemic outflow tract obstruction. Often, inotropic and/or mechanical ventilatory support is necessary. Many of these infants also have significantly increased pulmonary blood flow with a high $\dot{Q}p:\dot{Q}s$ ratio, requiring diuretic therapy and manipulation of the systemic and pulmonary vascular resistances to control blood flow.

Obstruction to Pulmonary Blood Flow

Lesions with pulmonary outflow tract obstruction include those with ductal-dependent pulmonary blood flow. Pulmonary

atresia with intact ventricular septum and critical pulmonary valve stenosis are defects that rely on patency of the ductus arteriosus for pulmonary blood flow. These infants may also require prostaglandin E_1 infusions to manage their cyanosis until the pulmonary outflow tract obstruction is relieved or bypassed.

Parallel Circulation

In the neonate with D-transposition of the great arteries the pulmonary and systemic circulations operate in parallel rather than the normal configuration in series. In this condition, deoxygenated blood from the right ventricle is ejected into the aorta and the left ventricle is in subpulmonary position ejecting oxygenated blood into the lungs. Neonates with this lesion depend on mixing of blood at the atrial, ventricular, or ductal levels. Although an infusion of prostaglandin E_1 can maintain ductal patency, many neonates benefit from a balloon atrial septostomy shortly after birth to create or enlarge an existing restrictive interatrial communication and optimize mixing. This is because mixing at the atrial level is much more effective than either the ventricular or ductal levels.

Single-Ventricle Lesions

This category is the most heterogeneous group, consisting of defects associated with atrioventricular valve atresia (i.e., tricuspid atresia), heterotaxy syndromes, and many others.[32] In some cases, both atria empty into a dominant ventricular chamber (i.e., double-inlet left ventricle); and although a second rudimentary ventricle is typically present, the physiology is that of a single ventricle or univentricular heart. Other cardiac malformations with two distinct ventricles (i.e., unbalanced atrioventricular septal defect) may also be considered in the functional single-ventricle category because of associated defects that may preclude a biventricular repair. A common feature among these lesions is complete mixing of the systemic and pulmonary venous blood at the atrial or ventricular level. Another frequent finding is aortic or pulmonary outflow tract obstruction. These children require careful delineation of their anatomy because each child represents a unique challenge to the practitioner. An important goal in single-ventricle management involves optimization of the balance between the pulmonary and systemic circulations early in life. This relates to the fact that a low pulmonary vascular resistance is an essential prerequisite for later palliative strategies and favorable outcomes. These considerations are also relevant in the anesthetic management of these children during noncardiac surgery.[30,33,34]

Intrinsic Myocardial Disorders

Children with primary cardiomyopathies or other forms of acquired heart disease such as myocarditis are characterized as having heart muscle disease (see later). They frequently have impaired ventricular function, either systolic or diastolic, and benefit from therapies tailored to their particular disease process.

Acquired Heart Disease

Cardiomyopathies

The term *cardiomyopathy* usually refers to diseases of the myocardium associated with cardiac dysfunction.[35,36] These have been classified into primary and secondary forms. The most common types in children are hypertrophic, dilated or congestive, and restrictive cardiomyopathies. Other forms include left ventricular noncompaction[37-39] and arrhythmogenic right ventricular dysplasia.[40] Secondary forms of cardiomyopathies are those associated with neuromuscular disorders such as Duchenne muscular dystrophy, glycogen storage diseases (i.e., Pompe disease), hemochromatosis or iron overload, and mitochondrial disorders. In addition, chemotherapeutic agents such as anthracyclines may result in dilated cardiomyopathies.[41] It is important to understand the basic hemodynamic processes behind the myocardial disease and implications for acute and chronic management.

Hypertrophic cardiomyopathy (HCM) is characterized by ventricular hypertrophy without identifiable hemodynamic etiology resulting in increased myocardial thickness. This is one of the most common forms, accounting for nearly 40% of cardiomyopathies in children.[42-45] The condition represents a heterogenous group of disorders, with the majority of the identified genetic defects exhibiting autosomal dominant inheritance patterns.[46,47] The mutations typically involve genes that encode sarcomeric proteins. This type of cardiac muscle disease is the most common cause of sudden cardiac death (SCD) in athletes, with an overall incidence estimated to be approximately 1% per year, with children and young adults affected most frequently.[48,49] Most children affected by HCM do not have left ventricular outflow tract obstruction (nonobstructive cardiomyopathy). It is unclear if the remaining minority with hypertrophic obstructive cardiomyopathy (HOCM), previously known as idiopathic hypertrophic subaortic stenosis (IHSS), are at an increased risk of SCD as compared with children without obstruction.[44] Hypertrophic cardiomyopathy may also involve the right ventricle.

The diagnosis of HCM begins with a history and physical examination. Most children are identified upon evaluation of a heart murmur, syncope, palpitations, or chest pain. Occasionally, an abnormal electrocardiogram (ECG) leads to referral. An accurate family history is essential. A prominent apical impulse is often present. Auscultation may reveal a systolic ejection outflow murmur that becomes louder with maneuvers that decrease preload or afterload (standing, Valsalva maneuver) or increase contractility. The murmur decreases in intensity with squatting and isometric handgrip. A mitral regurgitant murmur may also be present. An ECG will meet criteria for left ventricular hypertrophy in most children (Fig. 14-1). In some, the ECG findings may be striking (Fig. 14-2). Echocardiography demonstrating a hypertrophied nondilated left ventricle is diagnostic (Video Clip 14-4).[49] In many children, the hypertrophy may be asymmetrical (Video Clip 14-5). Echocardiography is the primary imaging modality for long-term assessment of wall thickness, ventricular dimensions, presence and severity of obstruction, systolic and diastolic function, valve competence, and response to therapy. Other diagnostic approaches such as cardiac catheterization and magnetic resonance imaging (MRI) may add helpful information in some cases.

The care of children with HCM includes maintenance of adequate preload, particularly in those with dynamic obstruction. Diuretics are generally not indicated and will often worsen the hemodynamic state by reducing left ventricular volume and increasing the outflow obstruction. Drugs that augment myocardial contractility (inotropic agents, calcium infusions) are not well tolerated. Patients usually undergo continuous ECG monitoring (Holter recording) and exercise testing for risk stratification.[50] β blockers and calcium-channel blockers are the primary

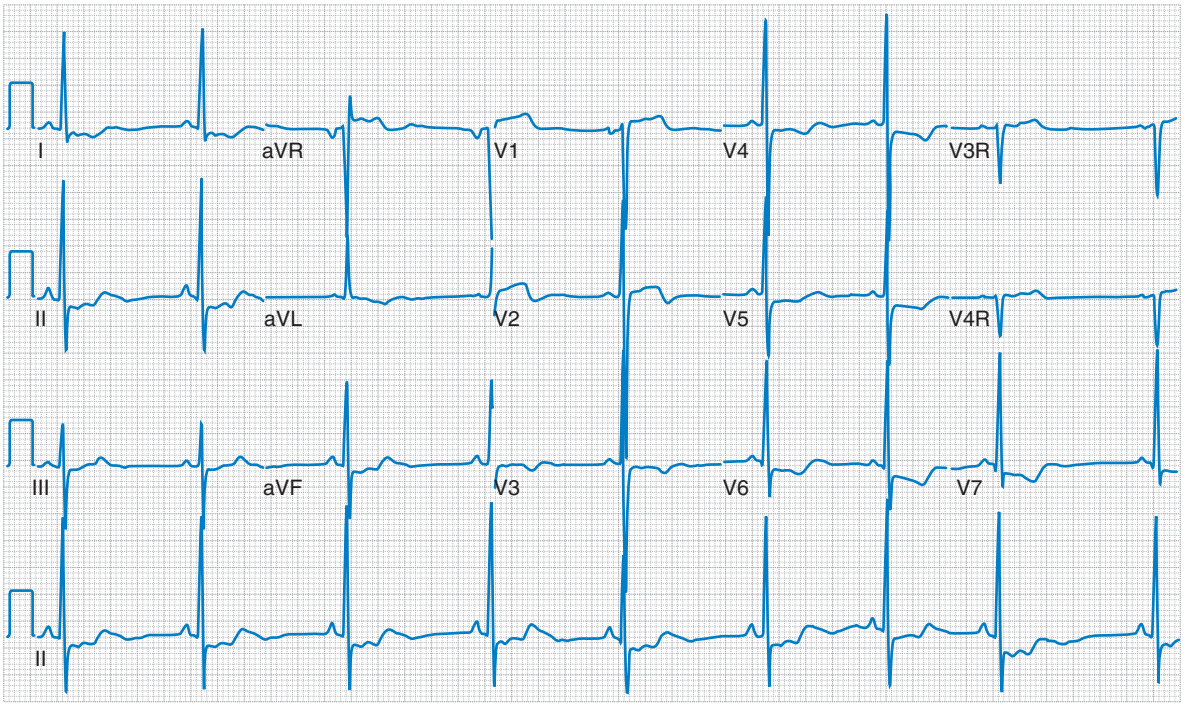


Figure 14-1. ECG in adolescent female with hypertrophic cardiomyopathy demonstrating left ventricular hypertrophy (deep S wave in V_1 and tall R waves over the left precordial leads). There is ST-segment depression and T-wave inversion over the left precordial leads related to repolarization changes associated with left ventricular hypertrophy, also known as a "strain" pattern. Reciprocal ST-segment elevation is noted over the right precordial leads.

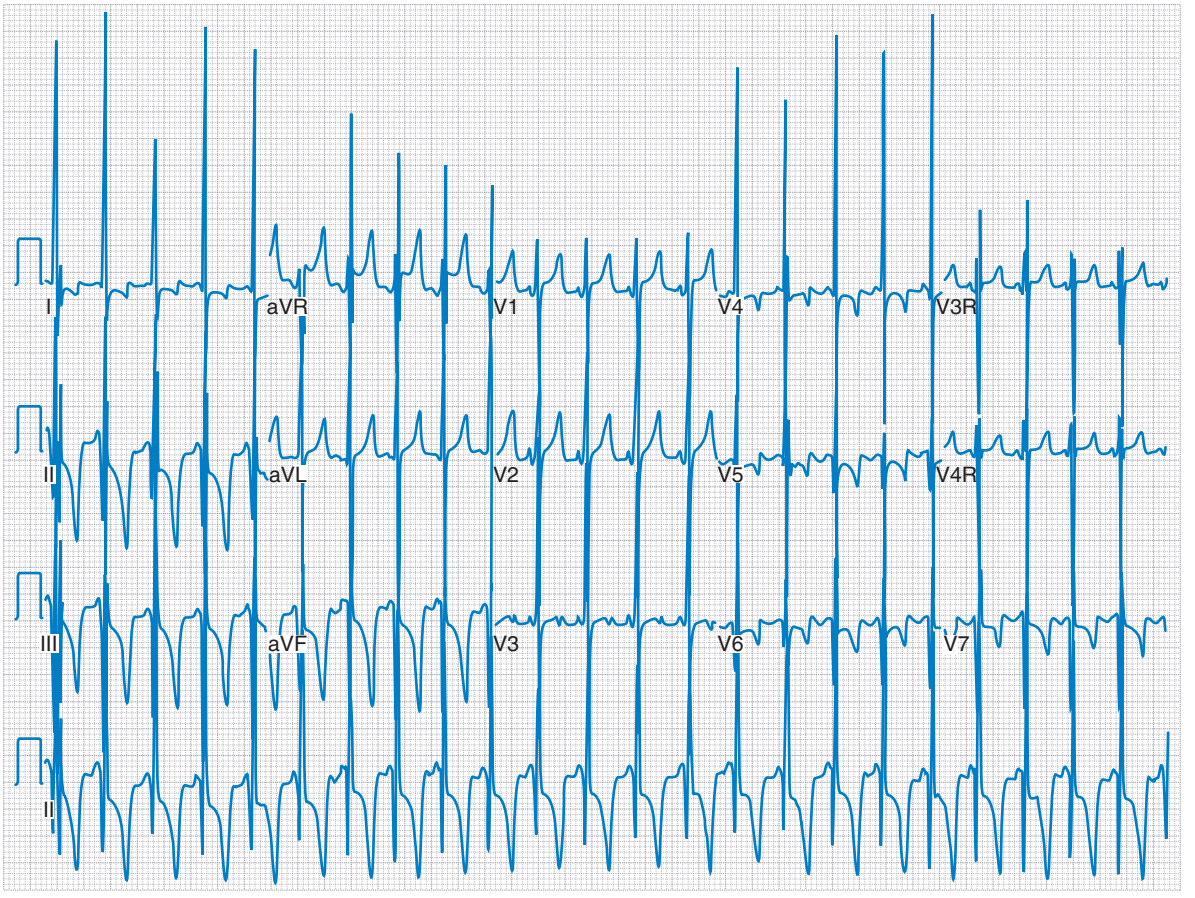


Figure 14-2. Tracing in a patient with Pompe disease, a severe form of hypertrophic cardiomyopathy displaying dramatic right and left ventricular voltages, as well as ST-segment and T-wave abnormalities. The recording is displayed at full standard (10 mm/mV, meaning that the ECG was not reduced in size to fit on the paper) as are all other ECG tracings in this chapter for reference.

agents for outpatient drug therapy.[51] Long-term treatment is individualized and based on the degree of hypertrophy, presence of ventricular ectopy, symptomatology (including syncope and congestive heart failure), family history, and genetic mutation analysis when available. Therapies range widely and include longitudinal observation with medical management of heart failure and arrhythmias, implantation of cardioverter-defibrillators, surgical myotomy/myectomy, transcatheter alcohol septal ablation, and cardiac transplantation. Patients with HCM are usually restricted from participating in competitive sports activities.[50]

Dilated cardiomyopathy (DCM), also known as congestive cardiomyopathy, is characterized by thinning of the left ventricular myocardium, dilation of the ventricular cavity, and impaired systolic function.[52-54] There is a broader range of etiologies than with HCM, ranging from genetic/familial forms with multiple types of inheritance patterns to those caused by infections (adenovirus, coxsackievirus, human immunodeficiency virus[55]), metabolic derangements (thyroid disorders, mitochondrial disorders, carnitine deficiency), toxic exposures (anthracyclines), and degenerative disorders (Becker and Duchenne muscular dystrophy).[56,57] Chronic tachyarrhythmias can also lead to DCM that may or may not improve once the rhythm disturbance is controlled.[58-60]

Most children with DCM present with signs and symptoms of congestive heart failure (tachypnea, tachycardia, gallop rhythm, diminished pulses, hepatomegaly). The chest radiograph typically demonstrates cardiomegaly, pulmonary vascular congestion, and, in some cases, atelectasis (Fig. 14-3). An ECG may identify the likely cause of the cardiac dysfunction in those with cardiomyopathy secondary to rhythm disorders or anomalous origin of the left coronary artery from the pulmonary root. The echocardiogram is essential for diagnosis and characteristically demonstrates a dilated left ventricle and systolic functional impairment (reduced shortening fraction and ejection fraction) (Video Clip 14-6).[61] Therapy in the short term is supportive and aimed at stabilization by controlling congestive heart failure and ventricular dysfunction. Management may include inotropic support (including phosphodiesterase inhibition) and mechanical ventilation. Unlike children with HCM, those with DCM have a volume-loaded, poorly contractile ventricle(s); therefore, gentle diuresis is beneficial. The infusion of large fluid boluses might be poorly tolerated and result in hemodynamic decompensation and cardiovascular collapse. The outcome of children with dilated cardiomyopathy is variable. In most, recovery of left ventricular systolic function occurs; however, others eventually require cardiac transplantation.[62] In a subset of children with severe disease, mechanical circulatory support may be necessary as a bridge to recovery or cardiac transplantation (Fig. 14-4, see Chapter 19).[63-65] Once initial stabilization is achieved, the treatment strategy typically switches to β blockade and afterload reduction with angiotensin-converting enzyme inhibitors.

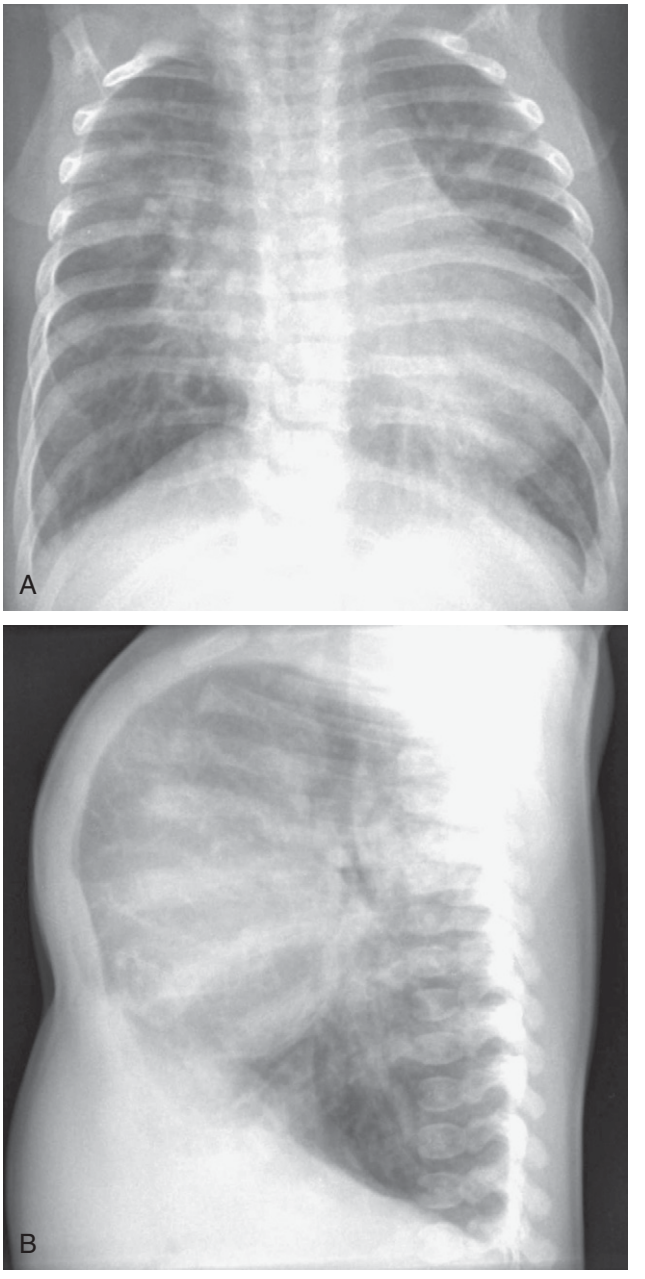


Figure 14-3. Chest radiographs in a young child with dilated cardiomyopathy in the posteroanterior (**A**) and lateral (**B**) projections demonstrating moderate to severe cardiomegaly and pulmonary vascular congestion. The lateral film shows the heart bulging anteriorly against the sternum.

Restrictive cardiomyopathy (RCM) is the least common of the major subsets of cardiomyopathies (5%) and portends a worse prognosis when it presents during childhood (survival <50% at 2 years from diagnosis).[66-71] The disorder is characterized by diastolic dysfunction related to a marked increase in myocardial stiffness resulting in impaired ventricular filling. Most cases are thought to be idiopathic. The presenting symptoms are generally nonspecific and primarily relate to the respiratory system. Occasionally the diagnosis is made following a syncopal or near sudden death event. The physical examination may demonstrate hepatomegaly, peripheral edema, and ascites.

The echocardiographic hallmark of RCM is that of severe atrial dilation (due to elevated atrial pressures) associated with normal- to small-sized ventricles (Video Clip 14-7). The marked diastolic dysfunction leads to increased end-diastolic pressures, left atrial hypertension, and secondary pulmonary hypertension. Children with RCM are prone to thromboembolic

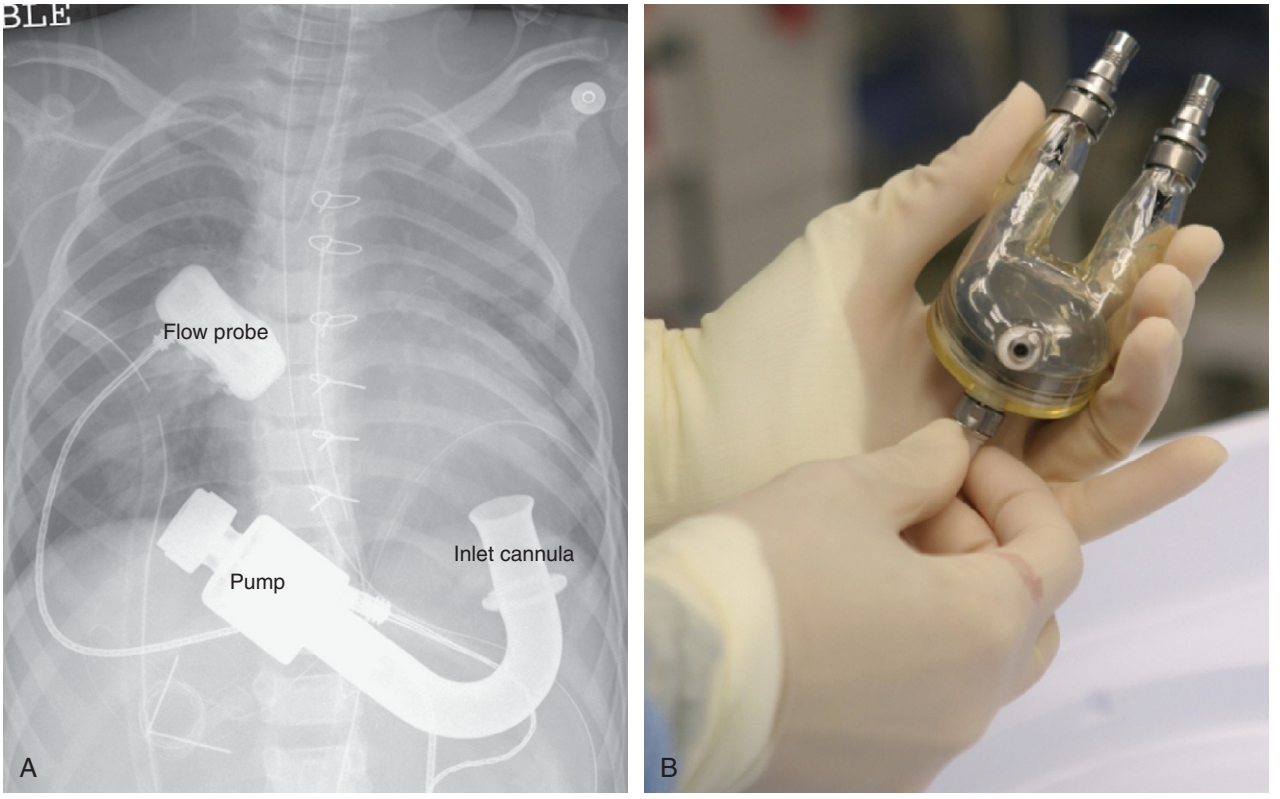


Figure 14-4. Mechanical circulatory assist devices. **A,** Chest radiograph in a child with end-stage dilated cardiomyopathy after placement of a MicroMed/DeBakey Child ventricular assist device for circulatory support while awaiting cardiac transplantation. This miniaturized device includes a titanium pump, inlet cannula (placed in the left ventricle), percutaneous cable, flow probe and outflow graft (placed in the aorta, not radiopaque). **B,** The Berlin Heart ventricular assist device. This device allows for paracorporeal placement and circulatory support in infants and small children. Although not approved by the U.S. Food and Drug Administration, the device has been used under specific guidelines and is available in several European countries.

complications, and anticoagulation therapy is frequently recommended. This is an important consideration during perioperative care as adjustments in the anticoagulation regimen may be necessary. Atrial and ventricular tachyarrhythmias may also occur. Optimal medical treatment is controversial because no specific agents or strategies have been shown to significantly alter outcomes.[71] Similar to children with HCM, diuretics often cause a decrease in the needed preload with detrimental effects on hemodynamics. Inotropic agents are not beneficial because systolic function is typically preserved and the arrhythmogenic properties of inotropic drugs can induce a terminal event. In many centers cardiac transplantation has been effectively utilized.[72,73]

Myocarditis

Myocarditis is defined as inflammation of the myocardium, often associated with necrosis and myocyte degeneration. In the United States it is most often caused by a viral infection. Over the past 20 years the spectrum of viral pathogens causing myocarditis has changed, such that adenovirus, enteroviruses (i.e., coxsackievirus B), and parvovirus have become the most frequent causes of fulminant disease.[74-76] The pathogenesis of myocyte damage in myocardial inflammatory diseases is quite complex.

The overall incidence of myocarditis is not known because it is frequently underdiagnosed, going unrecognized as a nonspecific viral syndrome. A large 10-year population-based study on cardiomyopathy found an annual incidence of 1.24 per 100,000 children younger than 10 years of age, only a fraction of which represented those with myocarditis.[77] The diagnosis is made utilizing clinical history, physical examination, and imaging modalities. A child with new-onset congestive heart failure or ventricular arrhythmias without evidence of structural heart disease should be considered to have myocarditis until proven otherwise. An ECG will typically demonstrate low-voltage QRS complexes with tachycardia, sometimes ventricular in origin. Chest radiography often shows cardiomegaly with pulmonary vascular congestion (Fig. 14-5). Echocardiography reveals dilated ventricles with impaired systolic function, similar to dilated cardiomyopathy, and is useful in the exclusion of alternative diagnoses, such as pericardial effusion or anomalous origin of a coronary artery, which can present in a similar manner. Myocarditis is generally a clinical diagnosis, because definitive

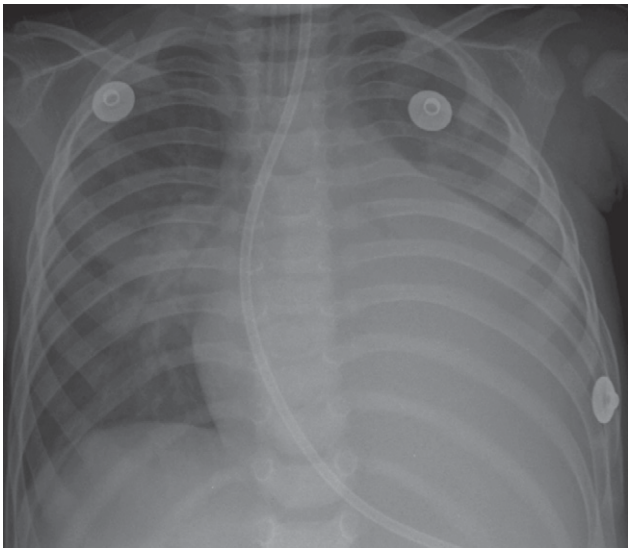


Figure 14-5. Chest radiograph in a child with acute myocarditis. Note the severe cardiomegaly and increased pulmonary vascularity.

confirmation requires the analysis of tissue obtained through myocardial biopsy either in the catheterization laboratory or the operating room (rarely performed).[78]

Many children with myocarditis have subclinical or mild clinical disease, whereas others progress to overt heart failure and/or arrhythmias. Among children with overt heart failure, approximately one third will regain full ventricular function, one third will recover but continue to demonstrate impaired systolic function, and one third will require cardiac transplantation.[79,80] A subset of children, not all of whom initially manifest severe symptoms in the acute period, will progress to develop DCM.

Although no specific therapies have been identified to directly treat the myocardial injury, a variety of strategies have been employed.[81-84] In the past, aggressive inotropic support was used to maintain cardiac output, perfuse vital organs, and prevent metabolic complications. However, increasing inotropy concomitantly increases myocardial oxygen demand and can have detrimental effects. The paradigm has therefore shifted toward diuresis and gentle inotropic support, often with agents such as phosphodiesterase inhibitors (milrinone), to improve myocardial performance without placing a large burden on an already failing heart.[81] Rhythm disturbances should be treated appropriately. Therapy with immune modulation or suppression with intravenous immunoglobulin is the standard of care at many centers.[80,85-87] Mechanical circulatory support may also be required in fulminant disease.[88-93]

Rheumatic Fever and Rheumatic Heart Disease

Acute rheumatic fever and rheumatic heart disease are leading causes of acquired cardiac disease in developing countries, with between 5 and 30 million children and young adults affected worldwide and a mortality rate of 1% to 10%.[94] In the United States, the availability of antibiotics to treat streptococcal pharyngitis has markedly reduced the incidence of this disease but sporadic cases still occur.[95] In children, the peak incidence occurs between 5 and 14 years of age.

Rheumatic fever results from infection by particular strains of group A β-hemolytic *Streptococcus* or *Streptococcus pyogenes.* Both cellular and humoral immune responses to the bacterial antigens cross react with native tissues to result in a multisystemic inflammatory disorder. The incubation period for most strains of group A β-hemolytic *Streptococcus* is typically 3 to 5 days, although some children will present with a more remote history of pharyngitis.

The diagnosis of rheumatic fever is made clinically based on the modified Jones criteria.[96] Major criteria include carditis, polyarthritis, chorea, subcutaneous nodules, and erythema marginatum. Evidence of a prior streptococcal infection along with two major or one major and two minor criteria are required to make the diagnosis in children without prior history of rheumatic fever or echocardiographic evidence of typical valvular involvement. Fever and arthritis are common symptoms. The polyarthritis has a migratory pattern, typically affecting large joints. Cardiac involvement or carditis occurs in nearly 50% of children with their first attack of rheumatic fever. Rheumatic heart disease represents a sequela of the acute process, affecting the mitral and aortic valves most frequently.

Primary prevention of rheumatic fever and rheumatic heart disease begins with prompt recognition and appropriate treatment of the initial streptococcal infection.[97,98] Secondary prevention, with ongoing therapy in individuals with a known history of rheumatic fever, has been shown to be extremely effective at preventing recurrent attacks. Although there is some debate regarding the optimal regimen, intramuscular injections of benzathine penicillin every 3 to 4 weeks appear to be most efficacious.[99]

Finally, in a subset of children with severe cardiac involvement, elective or emergent surgery plays an important role.[100] Valvular disease, rather than global myocarditis, is often the cause of congestive heart failure symptoms; and medical management is, therefore, limited in efficacy. When possible, valve repair rather than replacement is preferred.

Infective Endocarditis

Children with structural or acquired heart disease are at risk for developing infective endocarditis.[101,102] The risk varies from negligible to significant depending on a number of factors but to a great extent is based on the nature of the cardiac condition. The infection results from deposition of bacteria or other pathogens on tissues in areas of abnormal or turbulent blood flow. The diagnosis of endocarditis is made clinically by applying the modified Duke criteria.[103, 104] Major criteria include demonstration of microorganisms and evidence of pathologic lesions. The presentation of the disease may be acute or subacute. New or changing heart murmurs may indicate the development of either regurgitation or obstruction on an affected valve. Among the physical findings in children with endocarditis (part of the minor criteria) are signs of systemic embolization. Splinter hemorrhages (linear streaks under the nail beds), Janeway lesions (painless macules on the hands or feet), Osler nodes (small, painful nodules on the fingers), and Roth spots (retinal hemorrhages with clear centers) may be present. Inflammatory markers, such as erythrocyte sedimentation rate and C-reactive protein levels, are typically elevated, albeit nonspecific. Microscopic hematuria, as a manifestation of renal involvement, is frequently seen.

Acute bacterial endocarditis is caused most commonly by *Staphylococcus aureus.*[105,106] Such individuals typically present with high fevers, chills, myalgias, fatigue, and lethargy and sometimes are critically ill or in shock. Unlike in the adult population in whom right-sided heart endocarditis occurs predominantly in the context of intravenous drug abuse, both left- and right-sided endocarditis can occur in children with CHD.[107] Furthermore, children with indwelling venous catheters have an expanded spectrum of pathogens known to cause acute endocarditis, including coagulase-negative staphylococcal species or other nonbacterial organisms.

Subacute bacterial endocarditis (SBE), on the other hand, often has a more indolent course and presentation. Children generally manifest low-grade fevers, malaise, anemia, and fatigue. Most frequently the *Streptococcus viridans* group or *Enterococcus* species are the underlying pathogen.

Initial evaluation of a child suspected of having bacterial endocarditis includes serial blood cultures drawn from separate sites before initiation of antimicrobial drug therapy. The temporal frequency of these cultures depends on the clinical scenario and stability of the child. It is important to consider that even with excellent blood culture techniques up to 20% of children with evidence of endocarditis are culture negative and require empirical treatment throughout. Transthoracic echocardiography is routinely performed to evaluate for evidence of vegetations or other abnormalities.[108] While visualization of a vegetation establishes the diagnosis, a negative study does not exclude the diagnosis, so that, depending on the clinical index of suspicion, further imaging including transesophageal echocardiography may be necessary (Video Clip 14-8).[109] These imaging modalities are also particularly helpful during follow-up.

Parenteral antibiotics are initiated after blood cultures have been obtained. Broad-spectrum agents are used initially, and once a pathogen has been identified the antibiotic regimen is narrowed.[101] Daily blood cultures are obtained until three consecutive cultures remain sterile. A prolonged course of therapy is required in all children.

In addition to medical therapy, some children require surgical intervention. Failure of medical therapy (inability to clear the bacteremia), abscess formation, refractory heart failure, or serious embolic phenomenon are all indications for surgical intervention. Typically, the procedures involve resection of a vegetation, tissue debridement, and/or repair of consequent cardiac abnormalities. These children should subsequently receive antibiotic prophylaxis for endocarditis prior to at-risk procedures (see later).

A high level of suspicion for endocarditis must be maintained when evaluating a child with known heart disease and either persistent bacteremia (or fungemia) or fever of unknown origin. The same holds true for any child with foreign material in the heart or vascular tissue, such as indwelling central venous catheters, pacemaker/defibrillators, and closure devices.[110]

Endocarditis Prophylaxis

Transient bacteremia may result from disruption of mucous membranes or infected tissue during dental, surgical, or instrumentation procedures, causing seeding of the bloodstream with bacteria. In children with normal intracardiac anatomy, the risk for developing endocarditis from transient bacteremia is extremely low. However, certain cardiac conditions have a predisposition to the acquisition of endocarditis. The recently published guidelines no longer recommend prophylaxis based exclusively on an increased lifetime risk of endocarditis but propose that antibiotic prophylaxis should be restricted to those with the highest risk of an adverse outcome resulting from endocarditis. Patients in this category include those with prosthetic cardiac valves, previous history of endocarditis, certain congenital heart defects/post specific interventions, and cardiac transplant recipients with valvular disease (Table 14-1).[111]

Endocarditis prophylaxis is recommended for these individuals when undergoing dental procedures that involve the gingival tissues or the periapical region of teeth or perforation of the oral mucosa. Although a number of respiratory tract procedures are associated with transient bacteremia, no definitive data demonstrate a cause-and-effect relationship between these procedures and endocarditis. Consideration may be given for high-risk patients undergoing invasive procedures of the respiratory tract that involve incision or biopsy of the mucosa. In contrast to prior guidelines, routine prophylactic administration of antibiotics solely to prevent endocarditis is not recommended for those undergoing genitourinary or gastrointestinal tract procedures. However, for specific clinical scenarios antibiotic prophylaxis may be considered in these patients. Transesophageal echocardiography or routine endoscopy does not merit routine prophylaxis.[112] Prophylaxis is not considered necessary for routine hemodynamic cardiac catheterization with angiography; and although many practitioners administer routine antibiotics when transcatheter placement of an intracardiac device is undertaken, there are presently insufficient data to support this practice.[113]

Table 14-1. Cardiac Conditions Associated with the Highest Risk of Adverse Outcome from Endocarditis

- Prosthetic cardiac valve
- Previous infective endocarditis
- Congenital heart disease (CHD)*
  - Unrepaired cyanotic CHD, including palliative shunts and conduits
  - Completely repaired congenital heart defect with prosthetic material or device, whether placed by surgery or by catheter intervention, during the first 6 months after the procedure†
  - Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device (which inhibit endothelialization)
- Cardiac transplantation recipients who develop cardiac valvulopathy

*Except for the conditions listed above, antibiotic prophylaxis is no longer recommended for any other form of CHD.
†Prophylaxis is recommended because endothelialization of prosthetic material occurs within 6 months after the procedure.
Adapted with permission from Wilson W, Taubert KA, Gewitz M, et al (eds): Prevention of Infective Endocarditis: Guidelines from the American Heart Association. A Guideline from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee, Council on Cardiovascular Disease in the Young, and the Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and the Quality of Care and Outcomes Research Interdisciplinary Working Group. Circulation 2007; 116:1736-1754.

The guidelines are to administer antibiotic prophylaxis 30 to 60 minutes preceding the procedure to achieve adequate tissue levels before the time of potential bacteremia (Table 14-2).[111] The standard prophylactic regimen for children is for oral amoxicillin, 50 mg/kg dose (maximum of 2 g). For those allergic to penicillins or ampicillin the following oral drug choices are available: cephalexin, clindamycin, azithromycin, or clarithromycin. In children unable to take oral medication, options include ampicillin, cefazolin, or ceftriaxone via the intravenous or intramuscular routes. In those allergic to penicillins or ampicillin and unable to take oral medications, cefazolin, ceftriaxone, or clindamycin may be used.

Kawasaki Disease

Kawasaki disease was first described in 1967 as mucocutaneous lymph node syndrome.[114] It represents a fairly common and potentially fatal form of systemic vasculitis.[115-117] The etiology of Kawasaki disease remains unknown.[118] It occurs predominantly in infants and young children, most frequently in those of Asian origin, with the lowest incidence in whites. The disease has a predilection for the coronary arteries and may lead to dilation and aneurysm formation.[119,120]

The diagnosis rests on clinical features, because there are no specific laboratory tests for Kawasaki disease. To meet criteria, a child must have persisting fevers and at least four of the following findings[121,122]:

- Polymorphous exanthem
- Peripheral extremity changes (including erythema, desquamation, and edema of the hands or feet)
- Bilateral nonexudative conjunctivitis
- Cervical lymphadenopathy (often unilateral)
- Oral changes (strawberry tongue; red, dry, or cracked lips)

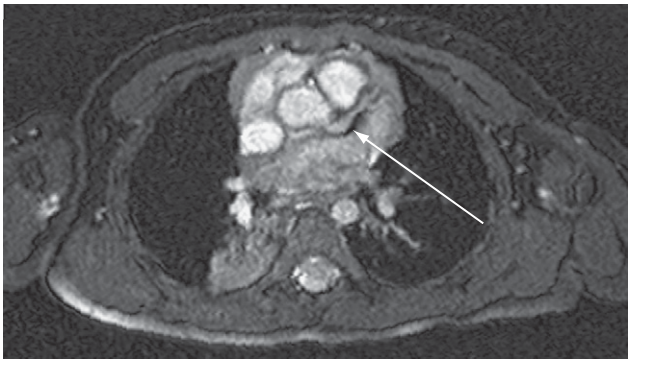


Figure 14-6. MR image in the axial plane at the level of the great vessels in a child with Kawasaki disease demonstrating a large fusiform aneurysm involving the left main coronary artery (*arrow*).

Nonspecific findings may include irritability, hydrops of the gallbladder, sterile pyuria, arthritis, and aseptic meningitis. Acute-phase reactants and thrombocytosis are usually present.

Intravenous gamma globulin is usually administered during the acute phase of the disease, in addition to high doses of aspirin, which are then decreased to lower doses for several months. The presence of coronary artery aneurysms is considered diagnostic of Kawasaki disease (Fig. 14-6). Coronary artery aneurysms or ectasia occur in 15% to 25% of untreated children and less than 5% of those who receive intravenous gamma globulin within the first 10 days of their illness.[122] In children with coronary artery aneurysms low-dose aspirin therapy is continued, in some cases in combination with anticoagulants or

Table 14-2. American Heart Association Guidelines for the Prevention of Infective Endocarditis: Antibiotic Regimens

Situation	Antibiotic	Dose (single dose 30 to 60 min before procedure)	
		*Children***	*Adults*
• Able to take oral medication	Amoxicillin	50 mg/kg	2 g
• Unable to take oral medication	Ampicillin	50 mg/kg IM or IV	2 g IM or IV
	or		
	Cefazolin or ceftriaxone	50 mg/kg IM or IV	1 g IM or IV
• Allergic to penicillins or ampicillin and able to take oral medication	Cephalexin*†	50 mg/kg	2 g
	or		
	Clindamycin	20 mg/kg	600 mg
	or		
	Azithromycin or clarithromycin	15 mg/kg	500 mg
• Allergic to penicillins or ampicillin and unable to take oral medication	Cefazolin or ceftriaxone†	50 mg/kg IM or IV	1 g IM or IV
	or		
	Clindamycin	20 mg/kg IM or IV	600 mg IM or IV

IM, intramuscular; IV, intravenous.
**Total pediatric dose not to exceed adult dose.
*Or other first- or second-generation oral cephalosporin in equivalent pediatric or adult dosage.
†Cephalosporins should not be used in an individual with a history of anaphylaxis, angioedema, or urticaria with penicillins or ampicillin.
Adapted with permission from Wilson W, Taubert KA, Gewitz M, et al (eds): Prevention of Infective Endocarditis: Guidelines from the American Heart Association. A Guideline From the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee, Council on Cardiovascular Disease in the Young, and the Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and the Quality of Care and Outcomes Research Interdisciplinary Working Group. Circulation 2007; 116:1736-1754.

antiplatelet drugs.[123] Myocardial ischemia and infarction, although infrequent, are important potential complications in children with coronary artery involvement.[120,124] Anesthetic care thus requires careful considerations regarding myocardial oxygen demand and supply; on rare occasion coronary revascularization may be necessary.

Cardiac Tumors

In children, cardiac tumors in general are extremely rare, and thus the natural history and optimal treatment strategies are often determined from limited case series.[125-127] Unlike adults, in whom atrial myxomas represent more than 90% of cardiac tumors, in children they tend to be either rhabdomyomas or fibromas.[128] Less common types include hemangiomas, myxomas (Video Clip 14-9), Purkinje cell tumors, and teratomas. Whereas in adults, most tumors are found in the left atrium, cardiac tumors in children occur in all four cardiac chambers. Malignant primary tumors are extremely rare, and data on their outcomes are even more limited.

Rhabdomyomas are the most common primary cardiac tumors in children. They often involve the ventricular septum and left ventricle and are multiple in the majority of cases.[129] Although they are considered benign, children may present with cardiomegaly, congestive heart failure, arrhythmias, or sudden death. The significance of a rhabdomyoma is determined largely by its size and any obstruction it may cause. Many tumors regress over time or completely resolve; thus, surgery is not indicated unless symptoms are present.[130,131] Many children with cardiac rhabdomyomas have associated tuberous sclerosis.[132,133]

Cardiac fibromas are the second most common type of pediatric primary cardiac tumors.[134] They are typically single and involve the ventricular free wall. In a subset of fibromas, the tumor will invade the conduction system causing atrioventricular nodal disease or arrhythmias.[135] Surgery or cardiac transplantation may be required in some cases.[136,137] The tumors may be very large, so that complete surgical resection may result in severely depressed cardiac function. Partial resections have been found to result in an arrest in growth with good outcomes while sparing cardiac function.[131] The primary concern in the perioperative care of children with cardiac tumors relates to the impact of the mass on hemodynamics (i.e., ventricular filling, patency of outflow tracts) and associated abnormalities of cardiac rhythm.[138]

Heart Failure in Children

Definition and Pathophysiology

Over the last several decades heart failure has been a major field of interest and investigation in adult medicine. Although this entity is less common in children, the interest in heart failure has been the subject of various publications,[139] scientific meetings, and the focus of several textbooks.[140,141] It is important to emphasize that pediatric heart failure results from markedly different causes than those reported in adults.[142] Understanding the cellular basis of heart failure in children, compensatory mechanisms, and therapeutic advances in this area are now at the forefront of pediatric medicine.[139-141,143,144] The discussion that follows highlights a few of the key concepts as they relate to anesthetic practice.

The definition of heart failure has evolved over the years. It is considered not only a pump failure but also a circulatory failure involving neurohumoral aspects of the circulation.[145] A number of conditions may ultimately compromise the ability to generate an adequate cardiac output to meet the systemic circulatory demands. It is important to recognize that heart failure does not necessarily imply impairment of ventricular systolic function but that diastolic heart failure is now an increasingly recognized clinical entity.

Etiology and Clinical Features

Causes of heart failure differ with age. In the perinatal period, cardiac dysfunction is typically related to birth asphyxia or sepsis or may represent an early presentation of CHD. The neonate with heart failure typically presents with clinical signs of a low-cardiac output state. Causes include left-sided outflow obstruction (as in aortic stenosis, aortic coarctation, hypoplastic left heart syndrome), severe valve regurgitation (as seen in Ebstein anomaly), or absent pulmonary valve syndrome.

During the first year of life most cases of heart failure are caused by structural heart disease. Other causes of heart failure are cardiomyopathies secondary to inborn errors of metabolism or acute events such as myocarditis. In infants with heart failure, tachypnea, dyspnea, tachycardia, as well as feeding difficulties and failure to thrive are prominent symptoms.[146] The physical examination is characterized by grunting respirations, rales, intercostal retractions, a gallop rhythm, and hepatosplenomegaly. Frequently, a mitral regurgitant murmur is present.

Beyond the first year of life, heart failure is either a consequence of previous surgical interventions, unpalliated/unrepaired cardiovascular disease, cardiomyopathies, myocarditis, or anthracycline therapy for a malignancy. Occasionally, a child may present with severe ventricular systolic impairment related to ongoing myocardial ischemia as a result of a coronary artery anomaly or rarely due to acquired pathologies such as Kawasaki disease. Older children with heart failure exhibit exercise intolerance, fatigue, and growth failure, whereas adolescents have symptoms similar to adults.

Treatment Strategies

Therapy is tailored to the etiology of the cardiac dysfunction and may include supportive care, mechanical ventilation, inotropic support, afterload reduction, prostaglandin E_1 therapy to maintain pulmonary or systemic blood flow, maneuvers to balance the systemic and pulmonary circulations, catheter-based interventions, or surgery.[147-151] Maintaining organ perfusion is the main goal in acute heart failure therapy. The primary therapeutic agents include catecholamines and inodilators. If the cause of the heart failure is not correctable, treatment consists of the standard drug regimen for chronic heart failure (diuretic therapy, β blockade, angiotensin-converting enzyme inhibition).[152,153] Newer agents that have received increasing attention in the management of pediatric heart failure include nesiritide (a recombinant form of human B-type natriuretic peptide)[154-156] and carvedilol (a third-generation β blocker).[157-159] For all ages, nutrition requires particular attention, because the work of breathing causes high caloric demands in the setting of fluid restriction and often inadequate intake; hypercaloric nutrition through an enteral feeding tube may be required.

Syndromes, Associations, and Systemic Disorders: Cardiovascular Disease and Anesthetic Implications

A wide variety of disorders including those resulting from chromosomal abnormalities, single-gene defects, gene deletion syndromes, as well as known associations (nonrandom occurrence of defects), and teratogenic exposure may manifest as cardiovascular disease. The coexistence of frequently associated multiple organ system comorbidities with cardiovascular disease presents a number of challenges to the anesthesia care provider.

Chromosomal Syndromes

Down Syndrome

Down syndrome is the most frequent chromosomal aberration, occurring with a frequency of 1 in 800 living births. The incidence increases sharply with advanced maternal age. In most children, it is the result of trisomy 21 but it may occur from balanced or unbalanced translocations of chromosome 21 or mosaicism. The phenotypes are indistinguishable. In general, these children are typically smaller than normal for age and craniofacial features include microbrachycephaly, short neck, oblique palpebral fissures, epicanthal folds, Brushfield's spots, small low-set ears, macroglossia, and microdontia with fused teeth. Mandibular hypoplasia and a broad flat nose are typical. A narrow nasopharynx with hypertrophic lymphatic tissue (tonsils, adenoids), in combination with generalized hypotonia, frequently leads to sleep apnea. Other conditions that affect these children include mental retardation, cervical spine disorders with vertebral and ligamentous instability, thyroid disease, leukemia, obesity, subglottic stenosis,[160] and gastrointestinal problems.

Airway issues include the potential for upper airway obstruction due to the presence of a prominent tongue, postextubation stridor,[161] and cervical spine injury.[162-164] Vascular access can be difficult. Subjectively, there is an impression that these children have small vessel sizes, vascular hyperreactivity, and fragile tissue consistency and may suffer from more complications after arterial cannulation.

Cardiovascular defects occur in 40% to 50% of children, so it has been recommended that all children undergo screening for CHD in early infancy.[165,166] The most common lesions include atrioventricular septal defects (Video Clip 14-10), ventricular septal defects, tetralogy of Fallot, and patent ductus arteriosus. Pathology after repair of atrioventricular septal defects is most often related to mitral regurgitation, left ventricular outflow tract obstruction, residual intracardiac shunts, and, rarely, mitral stenosis. Bradycardia under anesthesia occurs commonly.[167] Pulmonary hypertension resulting from either the cardiac pathology or from chronic hypoxemia secondary to upper airway obstruction should be considered in the management of these children.[168] Reduced nitric oxide bioavailability has been reported, leading to endothelial cell dysfunction,[169] possibly explaining the increased pulmonary vascular reactivity in these children with an early and higher incidence of pulmonary obstructive disease, even after corrective cardiac surgery.

Trisomy 18

Also known as Edwards syndrome, trisomy 18 is recognized as the second most common chromosomal trisomy (incidence of 1 in 3500 newborns). Most patients exhibit microcephaly, delayed psychomotor development, and mental retardation.[170] Characteristic craniofacial features include micrognathia or retrognathia, microstomia, malformed ears, and microphthalmia.[171] These abnormalities may affect airway management.[172,173] Skeletal anomalies include clenched fingers and severe growth retardation. Neurologic abnormalities are characterized by developmental delay, hypotonia, and central nervous system malformations. The high mortality rate in children with trisomy 18 is usually related to the presence of cardiac and renal problems, feeding difficulties, sepsis, and apnea caused by neurologic abnormalities.

Cardiovascular disease is present in most children and consists primarily of ventricular septal defects and polyvalvular disease.[174,175] Implications for anesthetic care include the high incidence of congestive heart failure and aspiration pneumonia.[173] These children may require interventions to address associated gastrointestinal or genitourinary anomalies.

Trisomy 13

Trisomy 13, an uncommon autosomal trisomy, is also known as Patau syndrome. The incidence of this chromosomal disorder ranges from 1 in 5,000 to 12,000 live births. Major features of this syndrome include cleft lip and palate, holoprosencephaly, polydactyly, rocker-bottom feet, microphthalmia, microcephaly, and severe mental retardation.[176,177] Nearly all children have associated cardiovascular defects that include patent ductus arteriosus, septal defects, valve abnormalities, and dextrocardia.[175] The overall prognosis for children with this syndrome is extremely poor.

Turner Syndrome

Turner syndrome is a genetic disorder (estimated incidence of 1 in 5,000 liveborn female infants) characterized by partial or complete X chromosome monosomy.[178] There is a high degree of spontaneous abortion among affected fetuses. Features of this syndrome include webbed neck, low-set ears, multiple pigmented nevi and micrognathia, lymphedema, short stature, and ovarian failure.[179] Systemic manifestations include cardiac defects (notably aortic coarctation and bicuspid aortic valve), hypertension, hypercholesterolemia, renal anomalies, liver disease, and inflammatory bowel disease. Obesity is common in older children as well as a high incidence of endocrinologic abnormalities such as hypothyroidism and diabetes.[178,180] Renal anomalies occur in up to one third of children.

Gene Deletion Syndromes

Williams Syndrome

Williams syndrome is a congenital disorder with an incidence of 1 in 20,000 live births. The genetic abnormality responsible for this syndrome in the majority of cases is a chromosomal deletion on the long arm of chromosome 7, altering the elastin gene.[181,182] The absence of this gene is detected by fluorescent in-situ hybridization (FISH). Features of Williams syndrome include characteristic elfin facies, outgoing personality, endocrine abnormalities (including hypercalcemia and hypothyroidism), mental retardation, growth deficiency, and altered neurodevelopment. Associated cardiovascular pathology includes valvar and supravalvular aortic stenosis (Fig. 14-7) and coarctation of the aorta.[183,184] The arteriopathy found in these

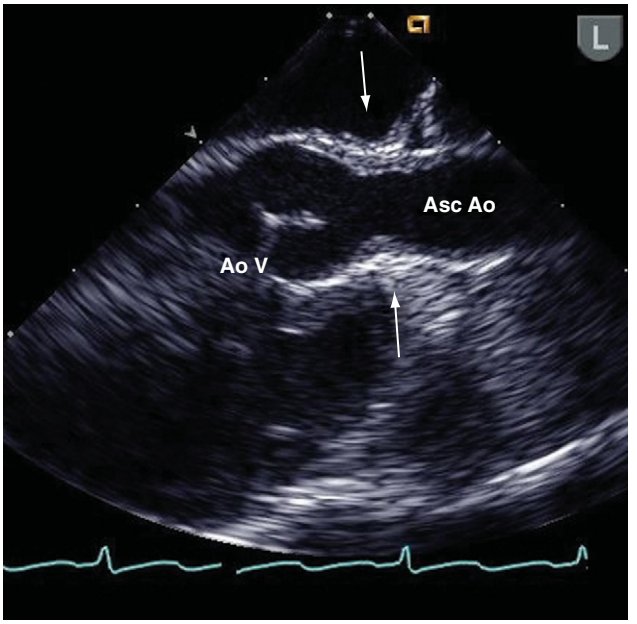


Figure 14-7. Echocardiogram displaying the classic supravalvar aortic narrowing in a patient with Williams syndrome (*arrows*). Ao V, aortic valve; Asc Ao, ascending aorta.

children may also involve the origin of the coronary arteries or other systemic and pulmonary vessels. Diffuse narrowing of the abdominal aorta may occur in association with renal artery stenosis.

Several reports in the literature have described unanticipated events during anesthetic care in these children.[185-187] Two conditions may result in increased anesthetic morbidity and the potential for mortality: coronary artery stenosis leading to myocardial ischemia and severe biventricular outflow tract obstruction. Because the extent of the disease in the affected individual is variable and may have devastating implications, a thorough cardiac evaluation is advisable in all children.[188] On occasion, children may require further evaluation before undergoing anesthetic care.

Children with Williams syndrome may exhibit some degree of muscular weakness; thus, the cautious use and application of muscle relaxants has been suggested.[188] Associated neurodevelopmental delay, attention-deficit disorder, and autistic behavior often requires adequate premedication. A high prevalence of subclinical hypothyroidism has been reported in these children.[189] Renal manifestations include renovascular hypertension, reduced function, and hypercalcemia-induced nephrocalcinosis.

Chromosome 22q11.2 Deletion Syndrome: DiGeorge and Velocardiofacial Syndrome

The 22q11.2 deletion syndrome, with an estimated incidence of approximately 1 in 3,000, encompasses DiGeorge, conotruncal face, and velocardiofacial syndromes. It is also known as CATCH 22 syndrome: an acronym for cardiac defects, abnormal facies, thymic hypoplasia, cleft palate, and hypocalcemia, all of which are commonly present.[190] Cardiac malformations, speech delay, and immunodeficiency are the most common features of the chromosome 22q deletion syndromes. Because no single feature is overwhelmingly associated with the deletion, it should be considered in any child with a conotruncal anomaly, neonatal hypocalcemia, or any of the less common features when seen in association with dysmorphic facial features.

Cardiac anomalies are often described as conotruncal anomalies; however, outflow tract anomalies are also frequently seen.[191] The remainder of the cardiac defects crosses an enormous spectrum, ranging from hypoplastic left ventricle to vascular rings. Only a minority of children with this chromosomal deletion have a normal cardiovascular system. The immune system is affected in a significant number of children. As a consequence of thymic hypoplasia, children typically have diminished T-cell numbers and function. The immunodeficiency requires the use of irradiated blood products and strict aseptic precautions during vascular access. Neurodevelopmental features include primarily speech delay as well as attention-deficit disorders. Psychiatric disorders are well described in these individuals.[192]

Single-Gene Defects

Noonan Syndrome

Noonan syndrome, an autosomal dominant syndrome, occurs with a frequency of 1 in 1,000 to 2,500 livebirths. It is characterized by distinctive dysmorphic features that include neck webbing, low-set ears, chest deformities, hypertelorism, and short stature.[193] Some male children also have cryptorchidism. The diagnosis of Noonan syndrome depends primarily on clinical features.[194] In neonates, the facial features may be less apparent; however, generalized edema and excess nuchal folds may be present as are seen in Turner syndrome. The facial features are more difficult to detect in later adolescence and adulthood.

The disorder is associated with a high incidence of CHD (~50%), with pulmonary valve dysplasia/stenosis being the most common.[195] Hypertrophic cardiomyopathy may develop during the first few years of life (in 10% to 20% of patients).[196] Clinical problems may also include developmental delay and bleeding diathesis (von Willebrand disease, factors XI and XII deficiency, and thrombocytopenia).[197]

Marfan Syndrome

Marfan syndrome is a multisystem disorder with variable expression resulting from a mutation in the fibrillin gene, a connective tissue protein.[198] It is estimated that about 1 in 10,000 individuals in the United States is affected by this syndrome. Clinical manifestations typically involve the cardiovascular, skeletal, and ocular systems.[199,200] Cardiovascular pathology includes mitral valve prolapse and regurgitation, ascending aortic dilation (Fig. 14-8), and main pulmonary artery dilatation. Dilatation of the sinuses of Valsalva is found in 60% to 80% of adults. The risk of aortic dissection rises considerably with increasing aortic size but may occur at any point in the course of the disease.[201] Cardiac arrhythmias may be related to valvular heart disease, cardiomyopathy, and/or congestive heart failure.

β-blocker therapy and aggressive blood pressure control has been the standard of care in patients with aortic root dilation[202] and should be continued perioperatively. Aortic root replacement in Marfan syndrome has been associated with a greater risk of re-dissection and recurrent aneurysm when compared with other patients who have undergone similar interventions.[203] Therefore, it is wise to maintain hemodynamics near baseline values during anesthetic care even in the postoperative period.

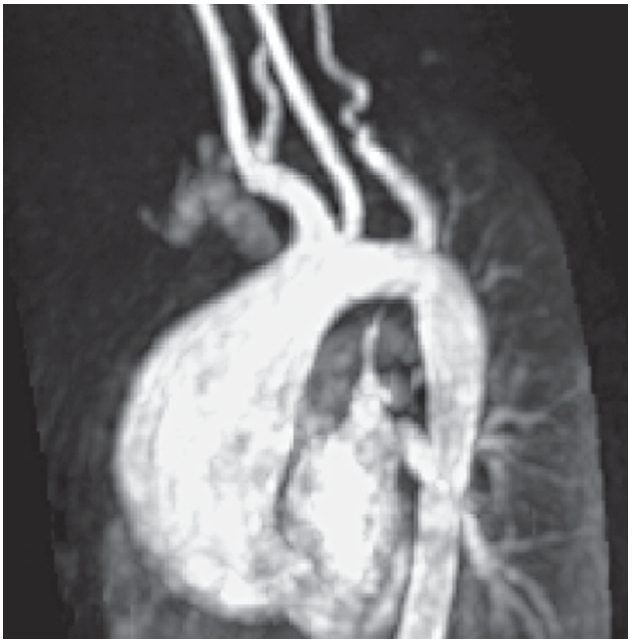


Figure 14-8. Dilated aortic root as displayed by MRI in a patient with Marfan syndrome. There is a marked discrepancy between the diameters of the ascending and descending aorta.

After aortic root surgery some individuals may require chronic anticoagulation therapy. Preoperative hospitalization may be necessary to adjust the anticoagulation regimen in anticipation of surgery. In emergency cases, infusion of coagulation factors and other blood products may be required to offset the anticoagulants. In addition to vascular pathology, children with Marfan syndrome have a predisposition for ventricular dilatation as well as abnormal ventricular function.[204,205]

Several factors can result in pulmonary disease in these children.[206] Chest wall deformities and progressive scoliosis can contribute to restrictive lung disease. In addition, it is thought that the fibrillin defect may affect both lung development and homeostasis impairing pulmonary function. The development of pneumothoraces is relatively common.

Associations

VATER or VACTERL Association

VACTERL association is an acronym given to describe a series of nonrandom anomalies that include vertebral, anal, cardiovascular, tracheoesophageal, renal, and limb defects.[207,208] Up to three fourths of children with VACTERL association have been reported to have CHD. The most common lesions include ventricular septal defects, atrial septal defects, and tetralogy of Fallot. Complex pathology such as truncus arteriosus and transposition of the great arteries occur less frequently.

Vertebral and tracheal anomalies can complicate airway management and regional anesthesia. Approximately 70% of children with VACTERL have vertebral anomalies usually consisting of hypoplastic vertebrae or hemivertebra. These may predispose children to developing scoliosis. Anal atresia or imperforate anus is reported in about 55% of children. These anomalies are usually noted at birth and often require surgery in the first days of life. Esophageal atresia with tracheoesophageal fistula is found in a large number of affected infants. Low birth weight (<1500 g) and associated cardiac pathology have been identified to be independent predictors of mortality in infants undergoing surgery for esophageal atresia/tracheoesophageal fistula (see Chapter 36). The presence of a ductal-dependent cardiac lesion further increases perioperative morbidity and mortality.[209] Limb defects occur in most children and include absent or displaced thumbs, polydactyly, syndactyly, and forearm abnormalities. These may impact vascular access and monitor placement. Renal defects are noted in approximately 50% of children.

CHARGE Association

CHARGE association is characterized by congenital anomalies that include coloboma, heart defects, choanal atresia, retardation of growth and development, genitourinary problems, and ear abnormalities. The association is estimated to occur in approximately 1 in 10,000 to 12,000 live births. The etiology is unknown, but it has been suggested that deficiency in migration of neural crest cells, deficiency of mesodermal formation, or defective interaction between neural crest cells and mesoderm play a part in these defects of blastogenesis.[210,211] Specific genetic abnormalities have also been identified in some individuals.[212,213]

Cardiac defects occur in as many as 50% to 70% of children and commonly include conotruncal and aortic arch anomalies.[214] Retardation of growth and development is usually due to cardiac disease, nutritional problems, or growth hormone deficiency. The developmental delay often is associated with sensory deficits (vision and hearing loss). Most children have some degree of mental retardation. Anesthetic implications, in addition to the cardiac defects, are focused around the airway.[215] A retrospective review of 50 cases reported upper airway abnormalities in 56% of children apart from choanal atresia and cleft lip and palate.[216]

Other Disorders

Tuberous Sclerosis

Tuberous sclerosis is a genetic disease with an autosomal dominant inheritance pattern and an incidence of approximately 1 in 25,000 to 30,000 births.[217] In a relatively large number of children this can be attributed to spontaneous mutations. This systemic disease primarily presents as cutaneous and neurologic symptoms, but cardiac and renal lesions are frequent findings.

The presence of upper airway nodular tumors, fibromas, or papillomas in affected children may interfere with airway management. Cardiac pathology is frequent and includes cardiac rhabdomyoma in 60% of children and coexisting CHD in 33% of cases.[218-220] Cardiac abnormalities with obstruction to flow, heart failure, arrhythmias, conduction defects, or preexcitation may affect the selection of anesthetic agents. Preoperative evaluation in most cases should include an ECG to exclude arrhythmia, conduction defects, or preexcitation.[218] Blood pressure and renal function should also be assessed. Anticonvulsants should be optimized and continued until the morning of surgery. Generally, baseline medical treatment should be resumed as soon as possible because seizures are the most common postoperative complication.[221] The presence of mental retardation may require the administration of agents such as midazolam or ketamine to facilitate parental separation.

Selected Vascular Anomalies and Their Implications for Anesthetic Care

Aberrant Subclavian Arteries

An aberrant or anomalous subclavian artery usually arises from the descending aorta as a separate vessel distal to the "usual" last subclavian artery in a posterior location. In a left aortic arch the aberrant vessel is the right subclavian artery. In this anomaly the arrangement is as follows: the first branch of the left aortic arch is the right carotid artery, followed by the left carotid and left subclavian arteries. The aberrant right subclavian artery, rather than arising proximally from the innominate artery as the first arch branch, originates distal to the last (left) subclavian artery as the fourth branch and courses behind the esophagus toward the right arm. This variant is one of the most common aortic arch anomalies. It is reported to occur in 0.4% to 2% of the general population and may or may not be associated with CHD.[222] There is a high incidence of this anomaly in children with Down syndrome and an association with ventricular septal defects and tetralogy of Fallot among other lesions. In a right aortic arch, the anomalous left subclavian artery originates distal to the origin of the right subclavian artery (Fig. 14-9). This anomaly may be seen in the context of conotruncal malformations. The diagnosis of an aberrant subclavian artery is made by most currently available imaging modalities.

Implications of this anomaly include the following:

- The presence of an aberrant subclavian artery may influence the location of placement of a systemic-to-pulmonary artery shunt.
- This anomaly should be considered in the selection of a site for arterial line placement if the need for transesophageal monitoring is also anticipated during surgery. The aberrant vessel may be compressed along its retroesophageal course by the imaging probe, resulting in inaccurate readings.[223] It may be wise, regardless of the site of arterial line placement, to monitor the arm supplied by the anomalous vessel by pulse oximetry or other methods during esophageal instrumentation.
- On occasion, an aberrant subclavian artery may be part of a vascular ring.
- Rarely, older children with a left aortic arch and aberrant right subclavian artery and without the findings of a complete vascular ring may complain of mild dysphagia (dysphagia lusorium).

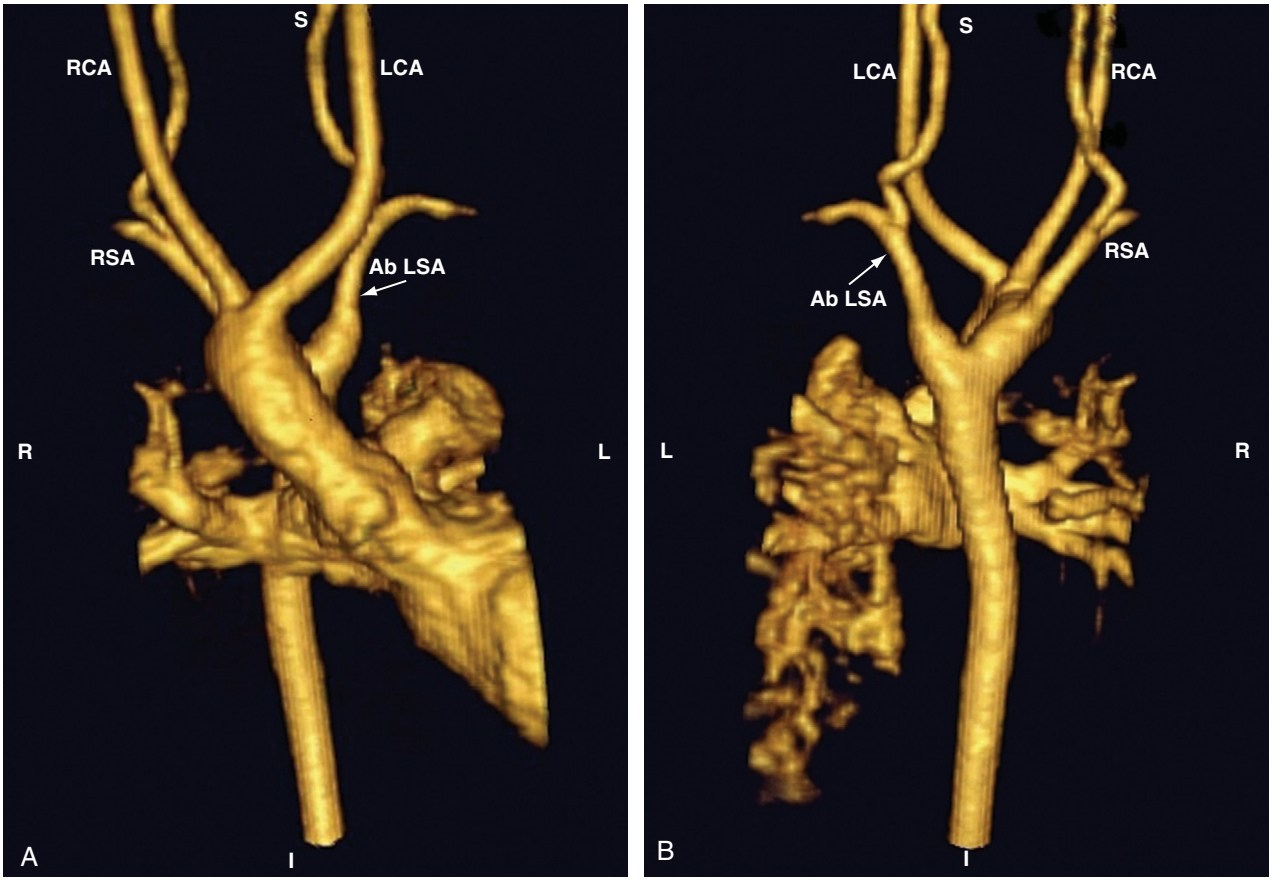


Figure 14-9. The MR images, as visualized anteriorly (**A**) and posteriorly (**B**), demonstrate a right aortic arch with an aberrant left subclavian artery (Ab LSA). The first arch vessel is the left carotid artery (LCA), followed by the right carotid (RCA), and right subclavian artery (RSA). The Ab LSA is the most distal branch originating from the descending aorta and coursing toward the left arm. This vessel may be compressed by a transesophageal echocardiographic probe.

Persistent Left Superior Vena Cava to Coronary Sinus Communication

A persistent left superior vena cava (LSVC) is a form of anomalous systemic venous drainage identified in 4.4% of children with CHD, most frequently those with septal defects.[224] It represents a remnant of the left anterior cardinal vein that typically obliterates during development. If it persists, it remains patent and drains into the right atrium via an enlarged coronary sinus. Bilateral superior vena cavae can be present (Fig. 14-10), or the right superior vena cava might be absent. In the presence of bilateral superior vena cavae the two vascular structures may communicate via an innominate or bridging vein.

Implications of this anomaly include the following:

- In the absence of an innominate vein a catheter placed in the left arm or left internal jugular vein and advanced into the central circulation may rest within the coronary sinus, a potentially undesirable location in a small infant. On chest radiography an unusual course is identified as the catheter courses along the left side of the mediastinum and can be mistaken for intracarotid, intrapleural, or mediastinal locations.
- An LVSC may be of relevance during venous cannulation for cardiopulmonary bypass to ensure adequate venous drainage and optimal operating conditions.
- The presence of an LSVC is important in patients with single-ventricle physiology undergoing palliation involving a cavopulmonary (Glenn) connection(s).
- Association with a dilated coronary sinus. On transesophageal echocardiography it may be confused with other defects, including an ostium primum atrial septal defect (one that lies in the inferior aspect of the atrial septum) and anomalous pulmonary venous return to the coronary sinus.
- On occasion, an LSVC may drain to an unroofed coronary sinus or directly into the left atrium, in which case a right-to-left shunt is present. This may be identified by injection of agitated saline into a left arm or left neck vein while performing an echocardiogram and may be associated with systemic arterial desaturation. This constitutes a risk for paradoxical systemic embolization.
- During cardiac surgery an enlarged coronary sinus may interfere with the administration of retrograde cardioplegia.
- It may confound placement of a pulmonary artery catheter and cardiac output determinations.

Evaluation of the Patient with a Cardiac Murmur

The finding of an incidental murmur during the perioperative period in many cases results in significant distress to the child or family, may trigger additional diagnostic studies including cardiology consultation, and has the potential to delay the scheduled procedure when identified preoperatively. Although cardiac auscultation is a challenging skill that takes many years of practice to master,[225] it is important for the practicing anesthesiologist who cares for children to recognize the main physical findings that may distinguish an innocent cardiac murmur from a pathologic one. In addition, knowledge of several core concepts and red flags can help avoid overlooking potentially important diagnoses.

The majority of normal children, in the range of 90%, will have a murmur at some point in their lives. This is most commonly identified during the neonatal period and early school

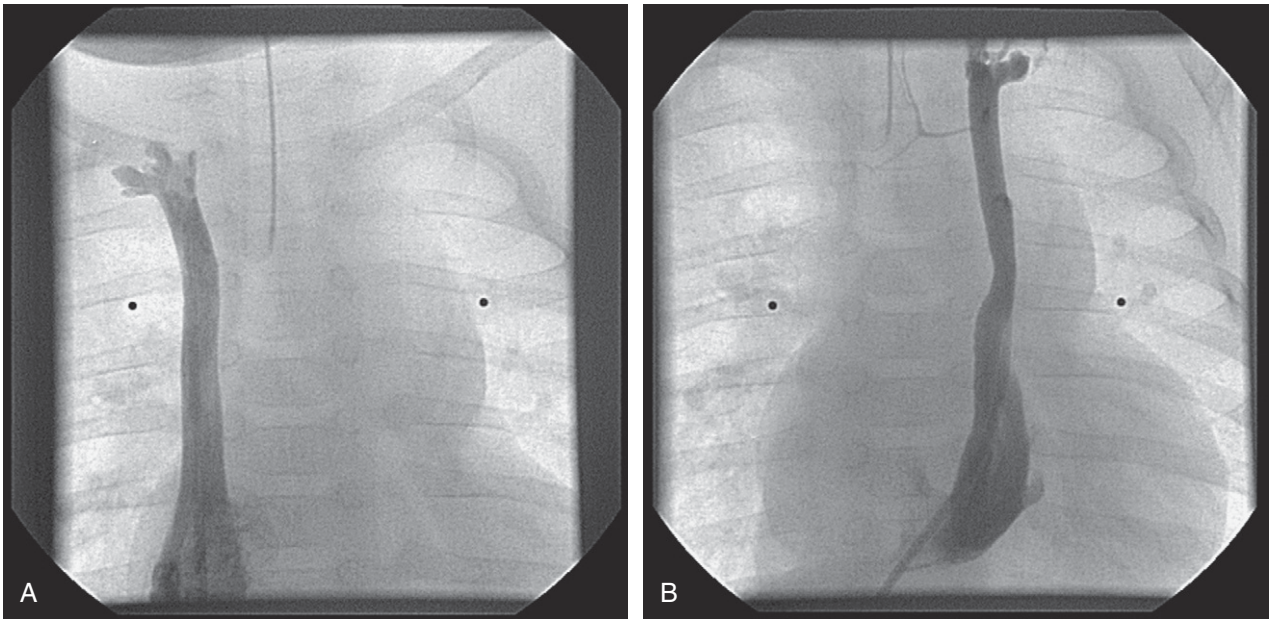


Figure 14-10. Bilateral superior vena cavae. **A,** Angiogram depicting the superior vena cava, normally a right-sided structure, as it drains into the right atrium. **B,** Angiogram in the same child demonstrating drainage of a large left superior vena cava into the coronary sinus. The catheter courses from the inferior vena cava into the right atrium, coronary sinus, and left superior vena cava. Contrast medium into the left superior vena cava demonstrates no innominate (or bridging) vein. A dilated coronary sinus is noted.

years. A great majority of murmurs are functional, otherwise considered innocent in nature, and require no special treatment. This diagnosis is based on physical findings consistent with the benign nature of the specific murmur.

While a complete discussion of cardiac murmur evaluation is beyond the scope of this chapter, it is pertinent to review a few key concepts related to the distinction between innocent versus pathologic murmurs.[226,227] The basic systematic approach when assessing a heart murmur is the same as when evaluating any child's cardiovascular system. Auscultation with both the diaphragm and bell of the stethoscope in the positions of the four primary cardiac valves should occur with the child and environment as quiet as possible. Innocent murmurs of infancy and childhood include pulmonary flow murmur, Still's murmur, physiologic pulmonary branch stenosis, venous hum, and carotid bruit. Innocent murmurs are usually of low intensity (grades I-II of VI) and are associated with a normal cardiovascular examination (e.g., normal precordial activity, first and second heart sounds, peripheral pulses, capillary refill). Innocent murmurs, such as those associated with peripheral pulmonary branch stenosis, right ventricular outflow murmurs, and Still's murmurs, tend to be soft, systolic ejection type and not holosystolic in duration. Physiologic murmurs often resolve by changing the child's hemodynamic state, either with maneuvers such as lying down or sitting up, or temporal changes such as resolution of fever, improvement in anemia, and so on. Diastolic or continuous murmurs are typically abnormal, with the exception of a venous hum. This murmur is thought to be related to turbulent flow of systemic venous return in the jugular veins and superior vena cava and is best heard at the base of the neck. *Murmurs accompanied by a palpable thrill are always pathologic.*

Whenever there is doubt regarding the benign versus the pathologic nature of a murmur, consultation with a pediatric cardiologist is indicated. A chest radiograph and ECG, although thought by some to add minimal value in the initial diagnostic assessment of a cardiac murmur[228,229] and to not be cost effective,[230] may be helpful when considering if further consultation is indicated.

Basic Interpretation of the Pediatric Electrocardiogram

Despite the increasing applications of imaging modalities in the structural and functional assessment of pediatric heart disease, electrocardiography continues to play a significant role in the diagnosis and management of these children. An ECG is considered an integral part of the evaluation of most children with congenital and acquired cardiovascular pathology.

Although the characteristic features of a normal ECG in infants and children were described many decades ago it is surprising that it continues to be one of the most often misinterpreted screening tests in pediatric medicine.[231] This is largely due to the developmental changes that occur in the normal individual as he or she progresses from the neonatal period through childhood, adolescence, and adulthood.[232] Normal values for children of various ages have been established.[233] Indeed, knowledge of normal configurations and values for various ages of children is essential for accurate interpretation.[232-235] Immediately after birth there is a predominance of right ventricular forces represented by tall R waves in the right precordial leads (V_1 and V_2) (Fig. 14-11). Over the first several years, the typical ECG changes to a more familiar left-sided heart

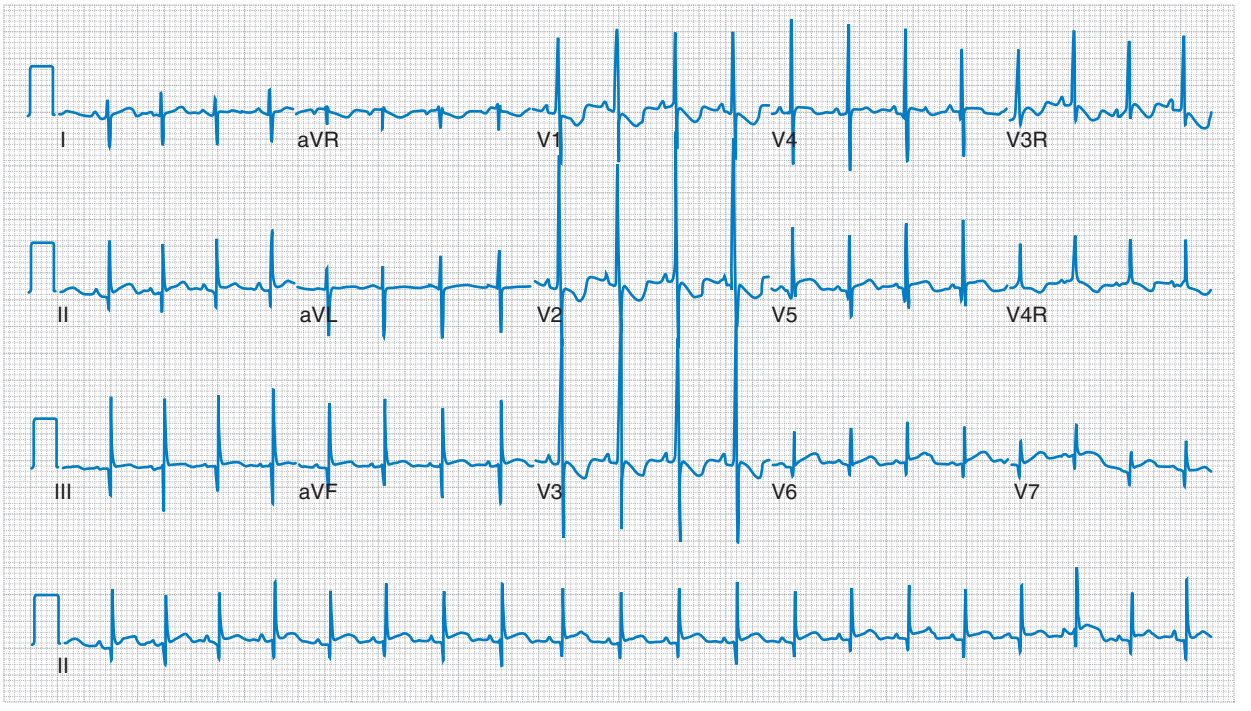


Figure 14-11. Normal ECG in a 2-day-old infant. The normal predominance of right ventricular forces during the neonatal period (tall R waves over the right precordial leads (V_1 and V_2) is evident. The inverted T waves in V_{1-3} are normal for age.

dominant configuration with larger S waves in the right precordial leads and a gradual RS progression with tall R waves in the left precordial leads (V_5 and V_6) (Fig. 14-12). The predominance of right-sided heart forces coupled with the need to evaluate for dextrocardia are the primary reasons that pediatric ECGs should include the V_3R and V_4R leads, which are not routinely obtained in adult studies. These electrodes are placed in the corresponding V_3 and V_4 locations over the right precordium.

Clinical information of relevance in the interpretation of an ECG includes the child's age, gender, suspected or documented diagnosis, and indications for the examination. A number of requirements are essential for accurate interpretation. These include appropriate skin preparation, electrode placement, and an artifact-free recording. A systematic, organized approach to the pediatric ECG is suggested. Determination of the rate and rhythm, with evaluation of the P wave vector and the relationship between each P wave and QRS complex, is the first step. It is important to consider the influences of age, autonomic nervous system, level of physical activity, medications, pain, and temperature on the child's heart rate. The P wave should be upright or positive in leads I and aVF, indicating that the sinus node is the pacemaker of the heart (sinus rhythm, see Fig. 14-12). Normally, the P wave should precede the QRS complex. Next, the QRS electrical axis should be determined. The QRS frontal plane axis is determined by identifying the most isoelectric lead, which will be perpendicular to the direction of ventricular depolarization. Alternatively, the direction of depolarization in leads I and aVF can be examined to roughly approximate the axis. As with all other components of the evaluation discussed here, the physiologic changes that occur with growth are responsible for the change in normal values for the QRS axis based on age. Regardless of age, QRS axes that lie in the northwest quadrant (between 180 and 270 degrees with an S-wave dominant pattern in both I and aVF) are always abnormal and merit further investigation. This is a frequent finding in children with atrioventricular septal defects. Evaluating the T wave, or repolarization axis, is also important, because a difference of greater than 90 degrees between the QRS and T-wave axes can represent strain on the ventricle, which is frequently associated with ventricular hypertrophy (see Fig. 14-1).

Once the evaluation of rhythm and axes is complete, each component of the cardiac cycle as represented on the ECG should be examined. The P wave represents atrial systole; and its morphology, with particular interest in leads II and V_1, can demonstrate either right atrial (P wave amplitude >2.5 mm or 3.0 mm based on age) or left atrial (P wave duration >100 to 120 msec based on age) enlargement (Fig. 14-13). The PR interval represents the time required for passage of an impulse from the sinoatrial node until ventricular depolarization and is largely composed of the atrioventricular nodal delay. A prolonged PR interval, which is age specific, indicates first-degree atrioventricular block. A short PR interval, on the other hand, should alert the practitioner to closely evaluate the QRS duration for signs of pre-excitation (Wolff-Parkinson-White syndrome, Fig. 14-14) although a short PR interval may also reflect a low right atrial pacemaker.

The QRS complex represents ventricular depolarization. The QRS duration should be examined in a lead with a Q wave present (often lead V_5 or V_6) for signs of conduction delay. The importance of age-dependent normal values is obvious here, because the upper limits of normal QRS durations are only 80 msec in neonates. The presence of a wide QRS complex with an RSR′ pattern in V_1 is indicative of right bundle branch block, and a QS pattern in V_1 and a tall notched R wave in V_6 is indicative of left bundle branch block. Other conditions associated

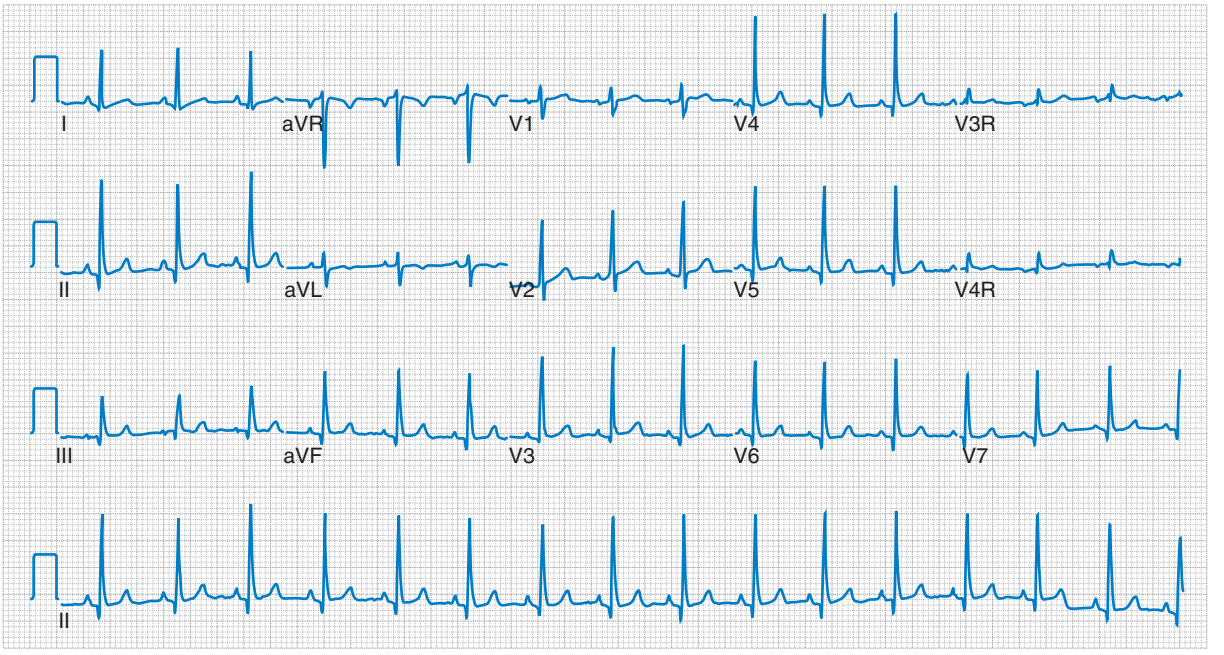


Figure 14-12. Normal ECG in a 10-year-old child. The typical left-sided heart dominant configuration of children this age is noted (gradual RS progression with tall R waves in the left precordial leads [V_5 and V_6]). This is in contrast to the right ventricular dominant pattern seen during infancy and early childhood. The tracing demonstrates normal sinus rhythm, as documented by positive P waves in leads I and aVF.

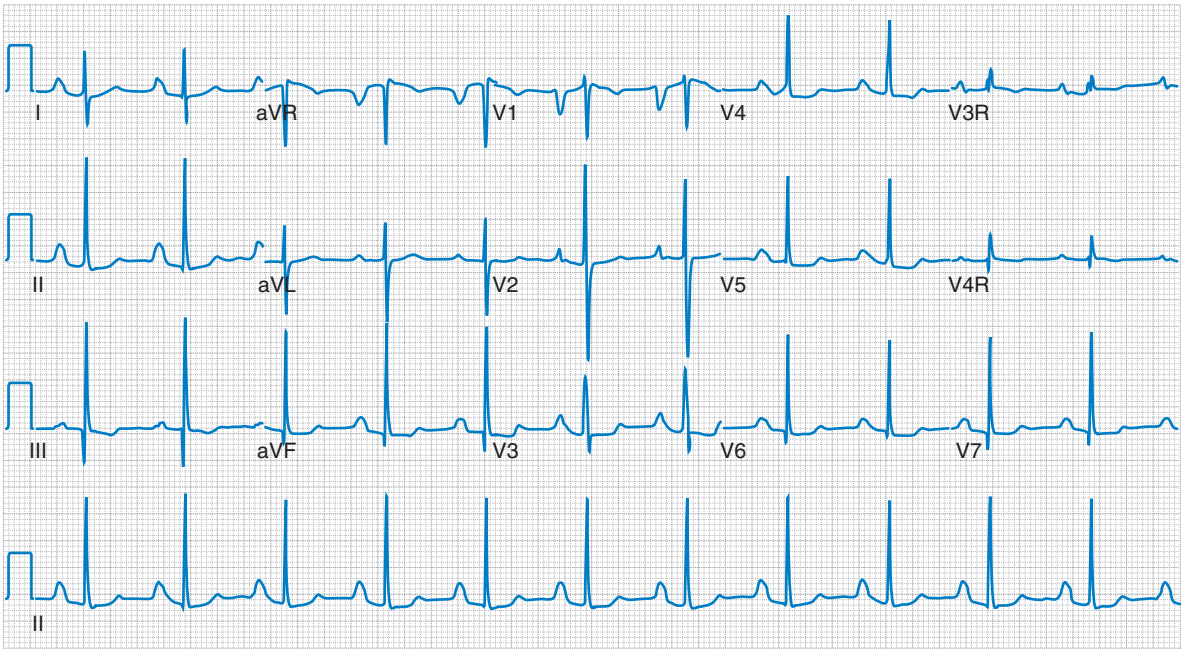


Figure 14-13. Biatrial enlargement. The tracing was obtained in the emergency department setting in a child subsequently found to have restrictive cardiomyopathy. There is evidence of biatrial enlargement as determined by the tall and wide P waves in leads II and V_1, respectively.

with prolongation of the QRS duration include ventricular preexcitation and ventricular pacing.

In addition to the QRS duration, the components of the QRS complex should be examined. Q waves are often present in the lateral and inferior leads, as well as lead aVR, but should be narrow (less than 40 msec) and shallow (age dependent, but generally less than 5 mm deep). Deep or wide Q waves are suggestive of myocardial ischemia and require further evaluation. An additional, relatively rare but crucial, finding occurs in infants with anomalous origin of the left coronary artery arising from the pulmonary artery (ALCAPA). Classically, the ECG in these children demonstrates deep, wide Q waves in leads I and aVL with ST-segment and T-wave changes in the anterior distribution (V_2-V_4) consistent with compromised myocardial

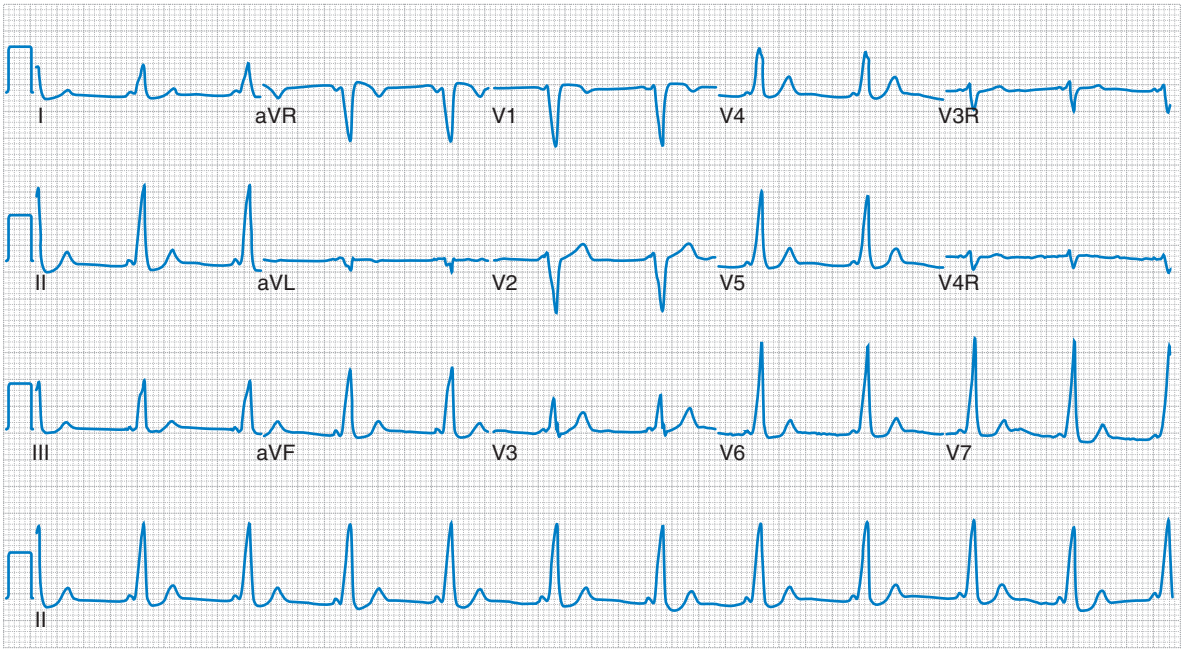


Figure 14-14. The tracing demonstrates the typical ECG features of the Wolff-Parkinson-White syndrome: short PR interval, delta wave, and prolongation of the QRS interval.

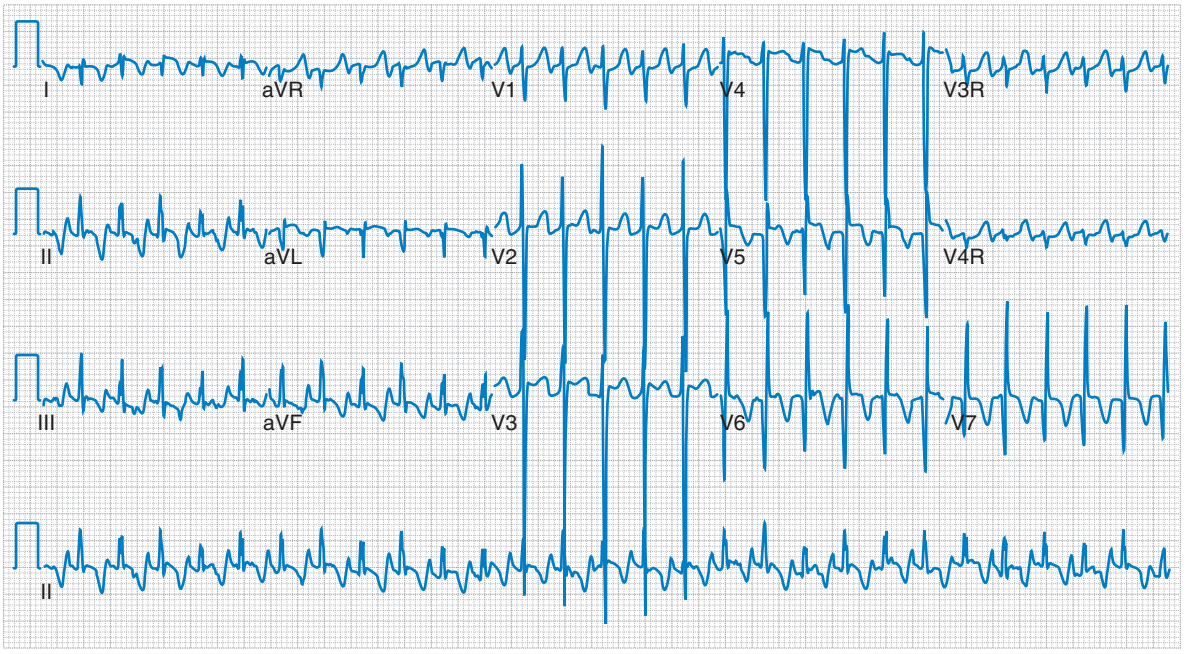


Figure 14-15. ECG tracing obtained in infant with poor ventricular function found to have anomalous origin of the left main coronary artery from the pulmonary root. The presence of Q waves in aVL and the diffuse ST-T wave changes suggestive of ischemia are classic for this anomaly.

blood flow (Fig. 14-15). The QRS amplitudes are also important in assessing both left and right ventricular hypertrophy. Conditions associated with increased QRS voltages that likely require echocardiographic assessment include hypertrophic cardiomyopathy, left ventricular noncompaction, and Pompe disease (see Fig. 14-2).

ST segments should be flat and should not be depressed more than 0.5 mm or elevated more than 1 mm in any lead. The major exception to this rule is when there is gradual upsloping of the ST segment in the mid-precordial leads as seen in early repolarization. T waves represent ventricular repolarization and should all be upright in the precordial leads at birth. Within 1 to 3 days they become inverted, initially in V_1, and eventually in V_2, V_3, and sometimes in V_4. Starting at several years of age the T waves will return to the upright position in the reverse order. In normal adolescents and adults the T wave in lead V_1 may be upright or inverted. The only limb lead that typically displays an inverted T wave is aVR.

One final aspect of the cardiac cycle that must be examined on any ECG is the QT interval—the time from the onset of ventricular depolarization, marked by the onset of the QRS complex, until the completion of repolarization, marked by the end of the T wave. It represents the duration of electrical activation and recovery of the ventricular myocardium and is measured as follows:

$$\text{Corrected QT (QTc)} = \text{measured QT interval/square root of preceding RR interval}$$

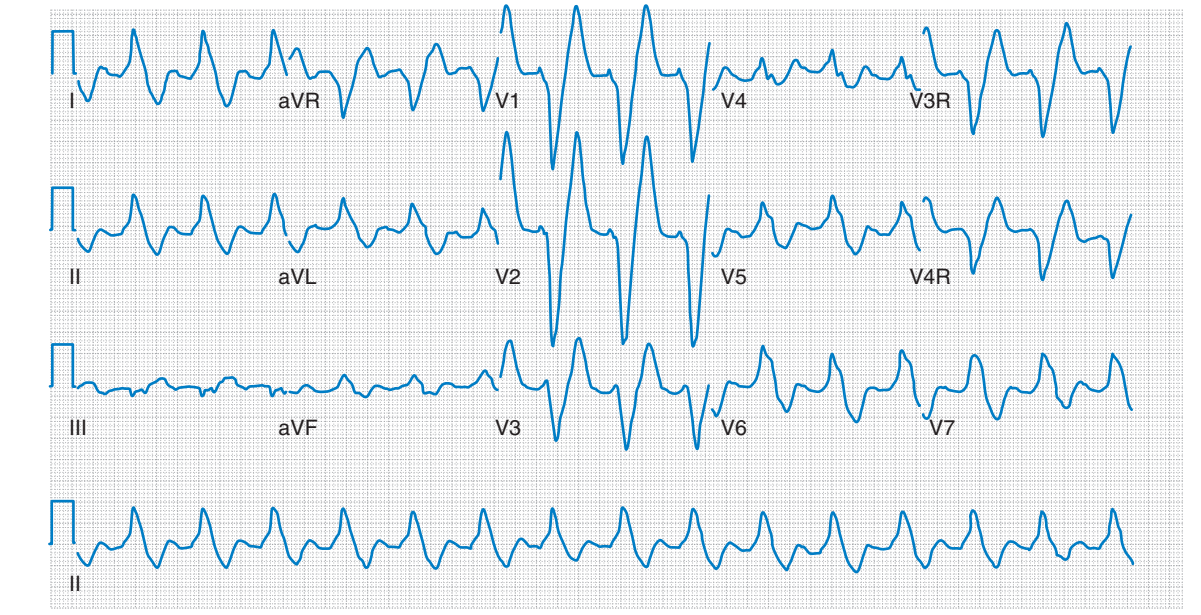
A QTc that exceeds 460 msec is considered abnormal regardless of age. All QTc values that exceed normal values for age merit further investigation. Medications that prolong the QT interval should be avoided until the child has been evaluated by a cardiologist.

Although a detailed organized approach to pediatric ECG interpretation is necessary, there are occasions when particular conditions or circumstances cause global ECG changes that must be quickly recognized. One such case that may occur in the operating room relates to the ECG changes associated with hyperkalemia. As the potassium level rises, the T-wave amplitude will increase. This is followed by widening of the QRS duration (Fig. 14-16) due to an intraventricular conduction delay and, finally, atrioventricular block and arrhythmias, including ventricular tachycardia and fibrillation. Other electrolyte disturbances that may result in characteristic changes on the ECG are as follows:

- Hypokalemia: decreased T-wave amplitude, ST-segment depression, and the presence of U waves
- Hypercalcemia: shortening of the QT interval, sinus rate slowing, and sinoatrial block
- Hypocalcemia: lengthening of the QT interval
- Hypomagnesemia: enhanced effects of hypocalcemia

Essentials of Cardiac Rhythm Interpretation and Acute Arrhythmia Management in Children

Rhythm abnormalities may be identified during the preoperative assessment, in the operating room, or in the postoperative period. The considerations usually revolve around (1) identification of the rhythm disorder; (2) establishing the need for acute therapy; (3) deciding whether to consult a pediatric cardiologist; and (4) conveying pertinent information to the consultant to assist in the characterization of the rhythm disturbance and to establish a management plan.



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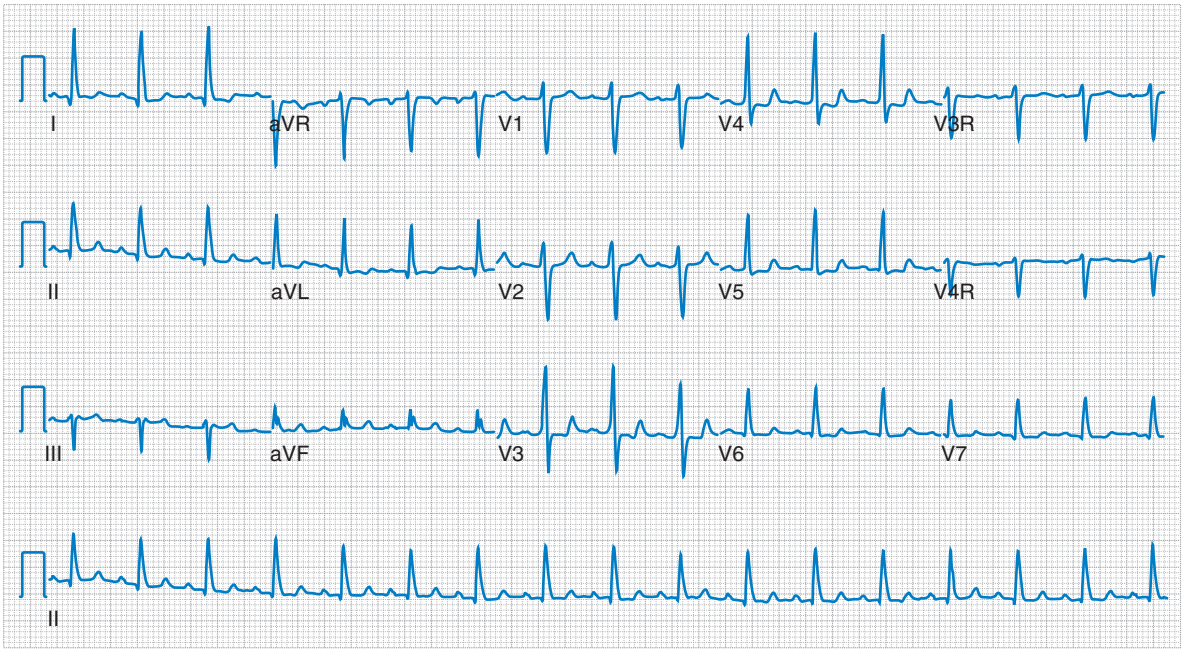

B

Figure 14-16. ECG shows changes that may result from hyperkalemia. **A,** In this 12-year-old male's ECG, there is marked widening of the QRS complexes, associated with peaked T waves. If untreated, this may progress to ventricular fibrillation and asystole. **B,** Tracing in same patient obtained several hours after treatment of electrolyte disturbance demonstrating resolution of the ECG changes.

The following principles should be considered in addressing these issues:

1. Operating room, bedside, or transport monitors and strip recordings facilitate the recognition of rhythm disorders but in most cases are inadequate for definitive diagnosis. A 15-lead surface ECG and rhythm strip should be obtained in all children when feasible.
2. In general, clinicians caring for children should have a basic knowledge of cardiac rhythm interpretation. Although a comprehensive discussion of arrhythmia interpretation is beyond the scope of this chapter, a brief overview of the characteristic features of normal and abnormal cardiac rhythms in the pediatric age group is presented in the section that follows.
3. The need for acute therapy for a rhythm disturbance should be based primarily on the nature of the disorder, urgency of the situation, and the likelihood that this abnormality would or would not be tolerated beyond the immediate short-term

period. The guidelines established by the American Heart Association for Pediatric Advanced Life Support should be followed in all patients.[236] In general, in otherwise healthy children, and in contrast to ventricular arrhythmias, supraventricular tachyarrhythmias are rarely life threatening.

4. The degree of comfort in the characterization and management of pediatric cardiac arrhythmias is likely to be quite variable among anesthesia care providers. For arrhythmias secondary to respiratory compromise, electrolyte imbalance, or metabolic derangements, consultation with a pediatric cardiologist is probably not required. This is also the case for variants or benign rhythm disturbances such as sinus arrhythmia, low atrial rhythms, or occasional premature atrial beats. Consultation is appropriate in most children with known structural heart disease (unoperated, palliated, or those who have undergone definitive interventions) or acquired cardiovascular pathology, in those with a history of a cardiac rhythm disorder under the care of a cardiologist, and in the majority of those with acute arrhythmias, particularly when the initiation of antiarrhythmic drug therapy is anticipated.
5. When consulting a specialist information that may be helpful to communicate includes pertinent details regarding the child's history, clinical diagnosis, nature of the procedure/intervention, relevant laboratory values, description or characterization of the rhythm abnormality, associated hemodynamic parameters, circumstances surrounding the event—including the presence or absence of an intracardiac catheter, review of the pharmacologic agents administered (including anesthetic agents), and other therapies if applicable. The consultant should assist in the characterization of the rhythm disorder, advise as to whether further evaluation is indicated, make recommendations for treatment, and facilitate diagnostic/therapeutic interventions as necessary.

Basic Rhythms

Sinus Rhythm

As discussed in the section on basic electrocardiography, sinus rhythm is characterized by a P wave that precedes every QRS, a QRS that follows every P wave, and an upright P wave in leads I and aVF (see Fig. 14-12).

Sinus Arrhythmia

This represents cyclic changes in the heart rate during breathing. This is a normal finding in healthy children (Fig. 14-17).

Sinus Bradycardia

This is characterized by sinus rhythm with heart rates below normal for age (see Fig. 14-1). Slow heart rates can be observed during sleep or at times of high vagal tone. When there is significant sinus bradycardia, a slow junctional escape rhythm or a slow atrial rhythm originating from an ectopic focus may be present. Certain forms of CHD may be prone to slow heart rhythms (i.e., heterotaxy syndromes).

In the intraoperative setting, particularly on induction of anesthesia, with laryngoscopy, endotracheal intubation, or tracheal suctioning, sinus bradycardia may occur. Sinus bradycardia may also be due to drug administration (i.e., opioids) or increased parasympathetic tone. This type of sinus bradycardia rarely poses significant hemodynamic compromise and, if necessary, can be easily treated with removal of the stimulus, administration of a vagolytic agent (pancuronium), or chronotropic agents such as atropine or epinephrine. Sinus bradycardia may also result from hypoxemia, hypothermia, acidosis, electrolyte imbalance, or increased intracranial pressure. Bradycardia related to hypoxemia should be treated promptly with the administration of supplemental oxygen and appropriate airway management (see Chapter 40). The approach to other secondary forms of sinus bradycardia should focus on addressing the underlying cause. For worrisome low heart rates, particularly in

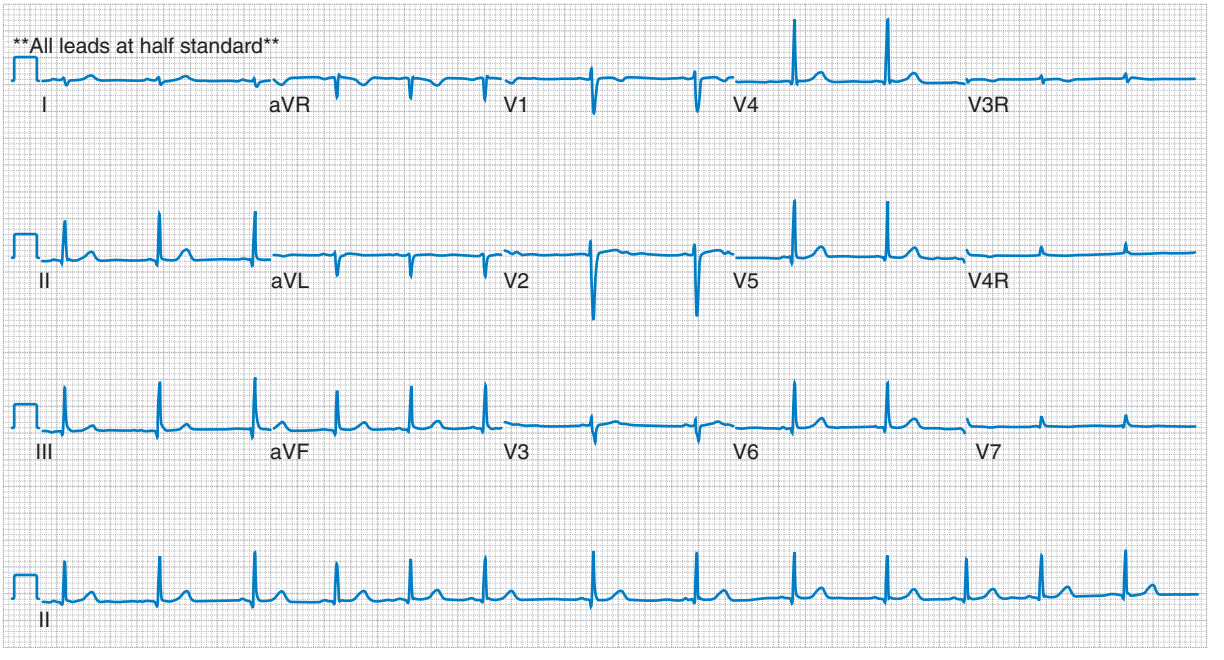

Figure 14-17. Sinus arrhythmia. The rhythm tracing demonstrates the normal heart rate variability with respiration. There is a normal increase in heart rate during inspiration. This is a natural response and is more frequently seen in children.

small infants, or clinical evidence of compromised hemodynamics, pharmacologic therapy (isoproterenol infusion) or temporary pacing should be considered.

Sinus Tachycardia

During sinus tachycardia, sinus rhythm occurs at a heart rate above normal for age (Fig. 14-18). In the perioperative setting this is often the result of surgical stimulation, stress, pain, hypovolemia, anemia, fever, medications (i.e., inotropic agents), or a high catecholamine state. Treatment is directed at the underlying cause. Prolonged periods of sinus tachycardia may impair diastolic filling time, limit ventricular preload, and compromise cardiac output. Children at risk of hemodynamic decompensation include those with significant degrees of ventricular hypertrophy or diastolic dysfunction.

Junctional Rhythm

A junctional rhythm is characterized by QRS complexes of morphology identical to that of sinus rhythm without preceding P waves. This rhythm is slower than the expected sinus rate. When this rhythm completely takes over the pacemaker activity of the heart, retrograde P waves and atrioventricular dissociation may be seen. Junctional rhythms during cardiac surgery are frequently the result of manipulation/dissection in the proximity of the right atrium. The central venous pressure contour typically demonstrates prominent v waves (right atrial pressure wave at the end of systole) due to the loss of AV synchrony (Fig. 14-19). The lack of atrial contribution to ventricular filling may result in decreases in the systemic arterial blood pressure.

Conduction Disorders

Bundle Branch Block

Incomplete right bundle branch block pattern (RSR′ in right precordial leads with near normal QRS duration) occurs in children with right ventricular volume overload (e.g., those with atrial septal defects). Complete right bundle branch block (QRS complex greater than 100 msec for infants, 120 msec for older children) is frequently identified in children after surgical procedures that involve the right ventricular outflow tract. This is characterized by an RSR′ wave pattern in V_1, as well as an inverted T wave, and a wide and deep S (slurred) wave in V_6. Left bundle branch block is an uncommon finding in the pediatric age group that may result from cardiac interventions along the left ventricular outflow tract. Criteria for this conduction disorder include a prominent QS or rS complex in lead V_1 and tall, wide, and often notched R wave in leads I, aVL, and V_6.

Atrioventricular Block

First-Degree Atrioventricular Block

In first-degree atrioventricular (AV) block there is prolongation of the PR interval beyond the normal range for age. Each P wave is followed by a conducted QRS. This may be found in healthy individuals but can also be seen in various disease states. In general, first-degree AV block is a benign condition requiring no specific treatment.

Second-Degree Atrioventricular Block

There are two forms of second-degree AV block: Mobitz type I (Wenckebach) and Mobitz type II. These are characterized by a periodic failure to conduct atrial impulses to the ventricle (P wave without following QRS complex). In type I second-degree AV block there is a progressive lengthening of the PR interval with eventual failure of conduction of the next atrial impulse to the ventricle. The RR intervals concomitantly shorten. The degree of AV block is expressed as the ratio of P waves per QRS complexes (i.e., 2:1, 3:2). This can occur during periods of high vagal tone or in the postoperative setting. It is generally a benign phenomenon that requires no therapy. In the less frequent, type

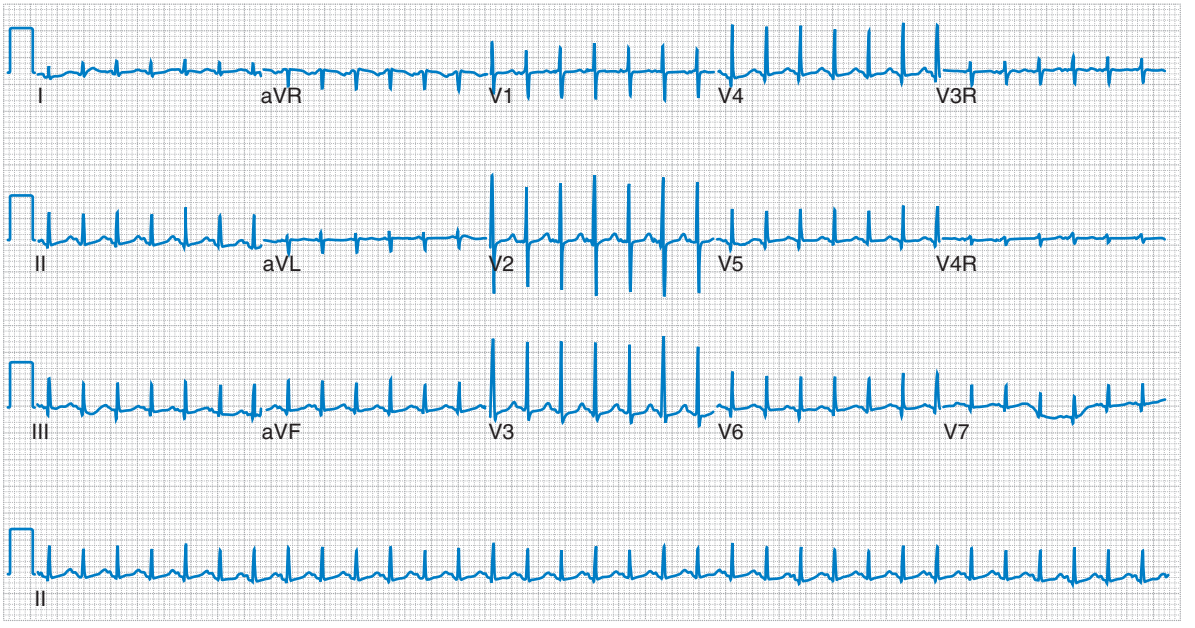

Figure 14-18. ECG in a febrile infant displaying the features of sinus tachycardia (heart rate above normal for age and QRS complexes of normal appearance preceded by P waves that are upright in leads I and aVF).

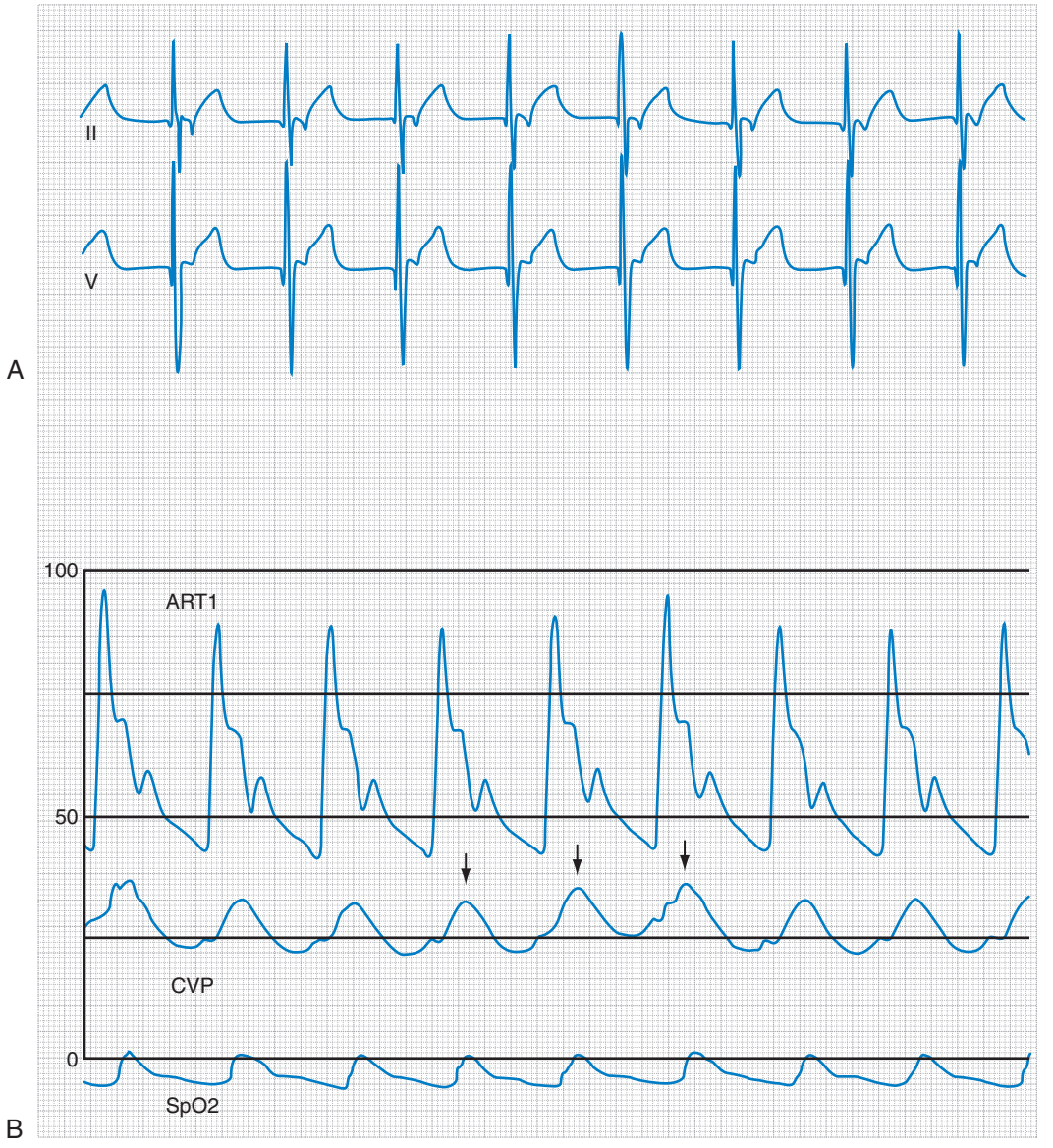

Figure 14-19. (**A**) Intraoperative tracing obtained during cardiac surgery at the time of right atrial dissection demonstrating the features of a junctional rhythm. Retrograde P waves are identified following the QRS complexes. (**B**) The central venous pressure (CVP) tracing demonstrates prominent v waves (*arrows*) related to the loss of atrioventricular synchrony (scale 0-30 mm Hg).

II second-degree AV block there is a relatively constant PR interval before an atrial impulse that fails to conduct. This is considered a more serious conduction disturbance and merits further investigation.

Third-Degree (Complete) Atrioventricular Block

Third-degree AV block is characterized by total failure of conduction of atrial impulses to the ventricle. It can be either congenital or acquired. There is complete AV dissociation, with more atrial than ventricular contractions, and the ventricular rate is usually slow and regular (Fig. 14-20). Temporary pacing may be indicated in the acute setting.

Cardiac Arrhythmias

Supraventricular Arrhythmias

Premature Atrial Contractions or Beats

Isolated premature atrial contractions (PACs) are relatively common in infants and small children. On the ECG the early P waves exhibit a morphology and axis that differ from those in normal sinus rhythm. Premature atrial contractions may be conducted to the ventricles normally, blocked at the AV node, or conduct aberrantly (abnormal QRS morphology). These are usually benign and require no therapy. If a central venous catheter is present, the tip position should be evaluated.

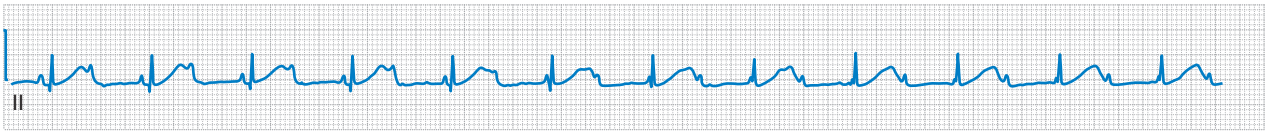

Figure 14-20. Surface ECG tracing demonstrates independent atrial and ventricular activity (atrioventricular dissociation) and failure of any atrial impulses to conduct to the ventricles. These are features of complete atrioventricular block.

Supraventricular Tachycardia

Supraventricular tachycardia (SVT) is the most common significant arrhythmia in infants and children.[237,238] It is characterized by a regular tachyarrhythmia (tachycardia heart rate is age dependent but typically >230 beats per minute in children) with a narrow or "usual" complex QRS morphology. Supraventricular tachycardia can occur in structurally normal hearts as well as in various forms of CHD. "Usual" complex implies that the QRS morphology in tachycardia is similar to that in normal sinus rhythm (Fig. 14-21). On occasion, widening of the QRS in SVT may be secondary to bundle branch block or related to the tachycardia mechanism (SVT with aberrancy). A wide QRS complex may make the distinction between supraventricular and ventricular tachycardia difficult.

There are two general types of SVT: automatic and reentrant. These can be differentiated by evaluating characteristics of the tachycardia, usually assisted by the input from a specialist. The evaluation of a tachyarrhythmia should include a surface 15-lead ECG and continuous rhythm strip to document onset and termination; and, if a medication such as adenosine has been administered, a recording of the response to the drug or pacing maneuvers should also be obtained. The management of SVT depends on the clinical status of the child, type of tachycardia, and precise electrophysiologic mechanism. General management principles include the following:

- Determination of hemodynamic stability. In the presence of hemodynamic instability synchronized direct current cardioversion (0.5-1.0 J/kg) should be performed.[236]
- Antiarrhythmic therapy is based primarily on the clinical condition and suspected tachycardia mechanism. Vagal maneuvers may be considered but should not delay treatment. Adenosine is the drug of choice in the acute setting for diagnosis and termination of most supraventricular tachycardias.[239,240] β blockers are most often used for chronic therapy.
- Others measures include treatment of fever if present, sedation, correction of electrolyte disturbance, decreasing or withdrawing medications associated with sympathetic stimulation (i.e., inotropic agents) or with vagolytic properties (i.e., pancuronium).
- In addition to pharmacologic therapy, atrial pacing or cardioversion may be required.

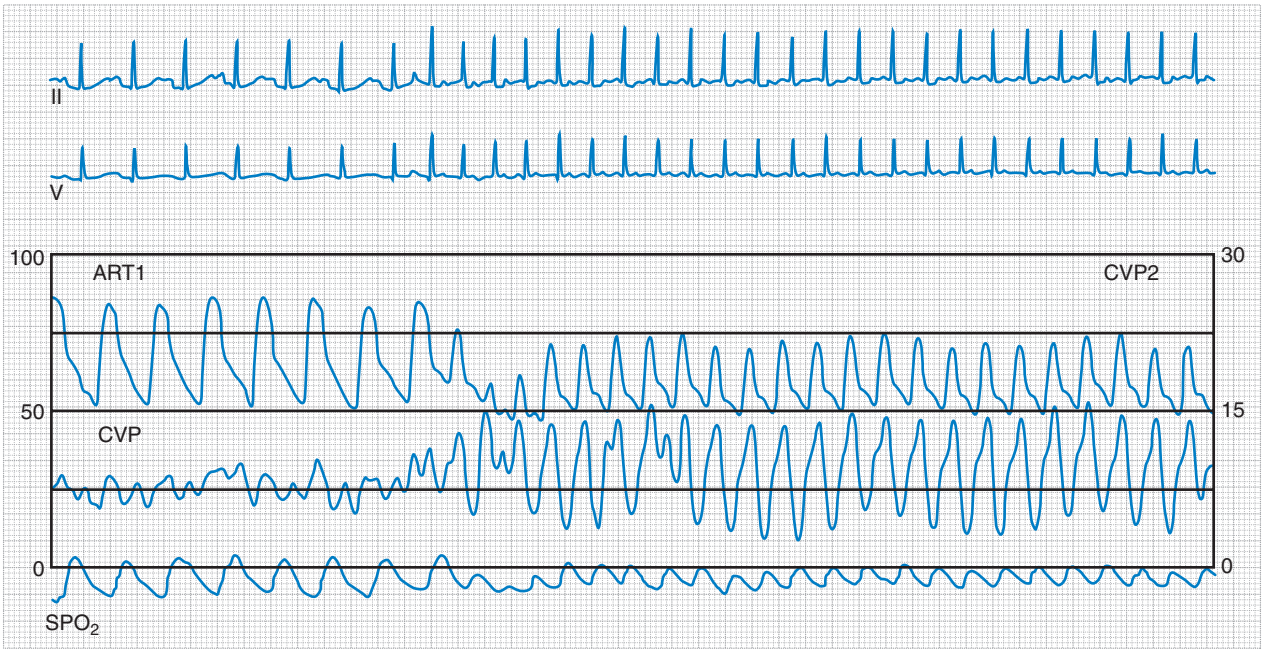

Figure 14-21. The initial portion of the tracing demonstrates normal sinus rhythm. A premature atrial beat initiates a narrow complex tachycardia (QRS morphology same as in sinus rhythm). The supraventricular tachycardia is associated with hemodynamic changes (note the decrease in the systemic arterial pressure, ART 1, scale 0-100 mm Hg and the increase in the central venous pressure, CVP, scale 0-30 mm Hg; oxygen saturation, Spo_2).

Ventricular Arrhythmias

Premature Ventricular Contractions or Beats

Premature ventricular contractions (PVCs) are characterized by (1) prematurity of the QRS complex, (2) a QRS morphology that differs from that in sinus rhythm, (3) usually a prolongation of the QRS duration for age, (4) abnormalities of the ST segment and T wave, and (5) premature ventricular activity not preceded by a premature atrial beat. PVCs of a single QRS morphology (uniform), without associated symptoms, and in children with structurally normal hearts are generally considered benign. An ECG during sinus rhythm should allow for careful measurement of the QT interval. Further investigation and consultation is warranted in the presence of PVCs of multiple morphologies (multiform), if these occur with moderate frequency or in succession (couplets or runs), and are associated with symptoms or a structurally abnormal heart.

Ventricular ectopy in the perioperative period may be the result of profound hypoxemia, electrolyte disturbances, or metabolic derangements. Other causes include the use of recreational drugs, myocardial injury, poor hemodynamics, and prior cardiac surgical intervention.

Ventricular Tachycardia

Ventricular tachycardia (VT) is relatively uncommon in children. It is defined as three or more consecutive ventricular beats occurring at a rate greater than 120 beats per minute (Fig. 14-22). The QRS morphology in VT is different than that in sinus rhythm, and the QRS duration is typically prolonged for age. ECG features that support this diagnosis include (1) AV dissociation, (2) intermittent fusion (QRS complex of intermediate morphology between two other distinct QRS morphologies), (3) QRS morphology of VT similar to that of single PVCs, and (4) tachycardia rate in children usually below 250 beats per minute.

Acute onset of VT in pediatric patients may be due to hypoxia, acidosis, electrolyte imbalance, or metabolic problems. Ventricular tachycardia may also occur in the context of depressed myocardial function, poor hemodynamics, prior surgical interventions, cardiomyopathies, myocardial tumors, acute injury (inflammation, trauma), and prolonged QT syndromes.[241,242] Among patients with CHD and ventricular arrhythmias, those at higher risk include older children after tetralogy of Fallot repair.

Long QT Syndrome

Long QT syndrome (Fig. 14-23) is an electrical cardiac disturbance that can predispose children to arrhythmias that include torsades de pointes ventricular tachycardia (Fig. 14-24), ventricular fibrillation, and bradyarrhythmias resulting in syncope, cardiac arrest, or sudden death.[243] Congenital and acquired forms have been described. The congenital varieties (i.e., Romano-Ward, Jervell and Lange-Nielsen syndromes) are likely the result of a genetic defect in the sodium or potassium channels responsible for maintaining electrical homeostasis in the heart.[244] Diagnostic criteria proposed for the long QT syndrome include ECG findings, clinical history (deafness, syncope), and family history.[245] A frequent feature is prolongation of the QTc on the resting ECG .

An important consideration in the care of children with long QT syndrome is ensuring adequate β-adrenergic blockade pre-

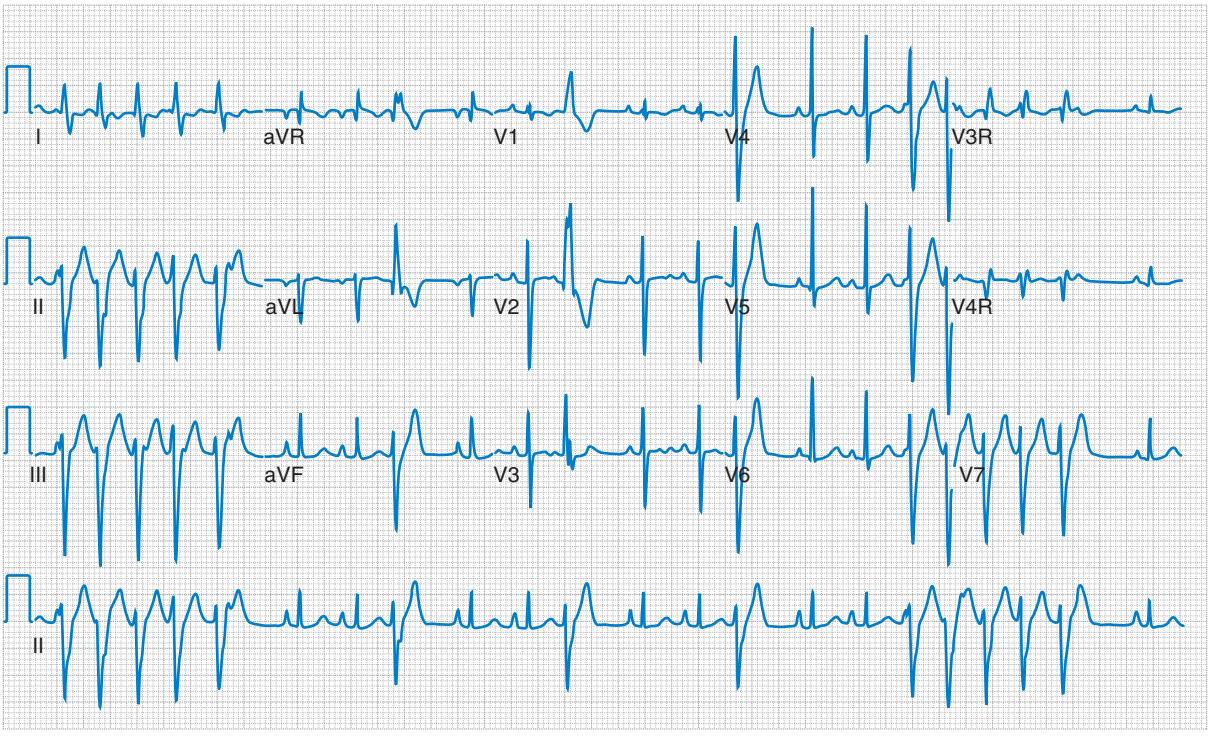

Figure 14-22. The tracing demonstrates frequent uniform premature ventricular beats and episodes of nonsustained monomorphic ventricular tachycardia.

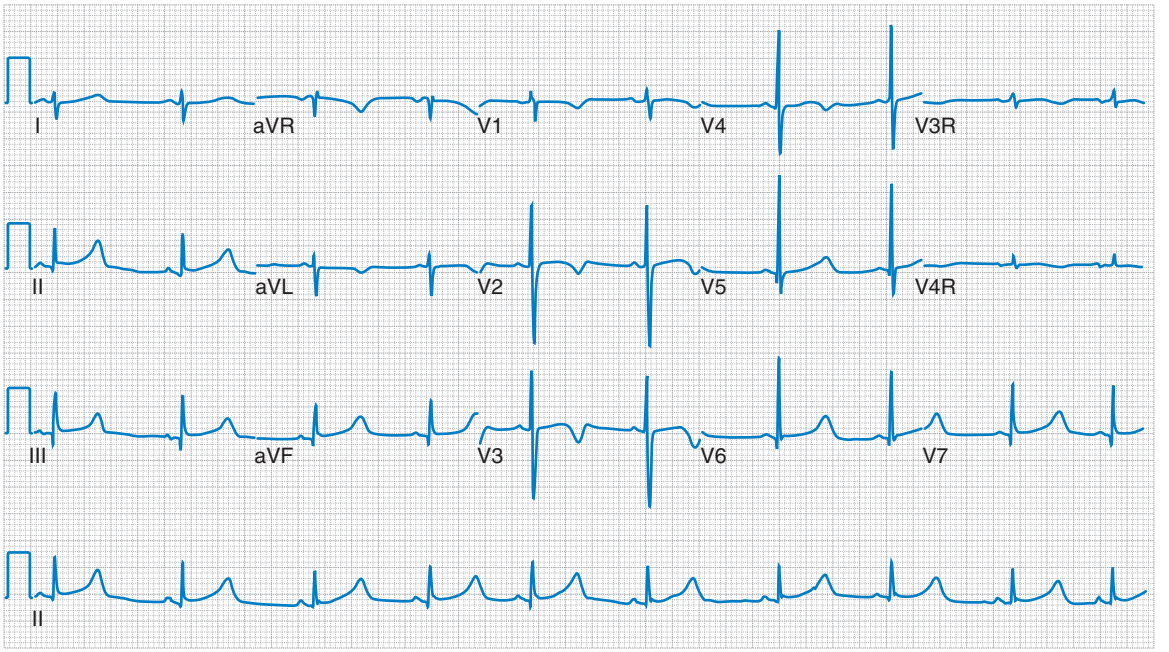

Figure 14-23. Long QT syndrome. Note the appearance of a prolonged QT interval on the ECG.

operatively and minimizing adrenergic stimulation.[243] In view of the fact that the evidence for risk is variable among the many drugs associated with torsades de pointes, the drugs have been divided into several groups (these are listed and updated on www.qtdrugs.org). Intraoperative arrhythmias can be treated with additional doses of β blockers. Magnesium and/or amiodarone may be required in some cases. Bradyarrhythmias can be managed by pacing. Conditions and drugs associated with prolongation of the QT interval should be avoided if possible. Despite the fact that several drugs routinely administered during anesthetic care (intravenous medications and inhalational agents) increase the QT interval, in most cases these drugs are given without untoward effects. This supports the concept that other factors, such as the effect of a drug on the dispersion of myocardial repolarization, may be more predictive of arrhythmogenic potential rather than prolongation of the QT interval per se. The care of these patients should be planned in combination with a specialist.

Prolongation of the QT interval may also result from electrolyte derangements (hypokalemia, hypocalcemia, hypomagnesemia), drug therapy (antibiotics, antiarrhythmic agents, antipsychotic drugs, cisapride), and neurologic or endocrine abnormalities. Therapy in this setting should focus on correction of the underlying cause.

Considerations in the management of VT are as follows:

1. Although some atypical forms of supraventricular tachyarrhythmias may mimic VT, a wide QRS tachycardia should always be considered to be of ventricular origin.
2. The initial approach in the care of a child with an acute ventricular rhythm disturbance is the prompt evaluation of clinical status and hemodynamic stability. Sustained ventricular arrhythmias are generally poorly tolerated and require immediate attention. In the unstable child, cardiopulmonary resuscitation should be instituted while preparing for cardioversion. Expert consultation is advisable when advanced drug therapy is contemplated. Drugs that may be considered include lidocaine, amiodarone or procainamide.[236] The latter two should not routinely be administered concurrently due to QT prolongation.
3. Electrical cardioversion in torsades de pointes should be performed only if the arrhythmia is sustained. Magnesium sulfate is considered a first-line drug. Procainamide is contraindicated due to prolongation of the QT interval.
4. In addition to β blockade, some congenital forms of the long QT syndrome may require implantation of a cardioverter-defibrillator.

Ventricular Fibrillation

Ventricular fibrillation (VF) is an uncommon arrhythmia in children. It is characterized by chaotic, asynchronous ventricular activity that fails to generate an adequate cardiac output. The ECG in VF demonstrates low-amplitude irregular deflections without identifiable QRS complexes. A loose ECG electrode may mimic these surface ECG features; thus, immediate clinical

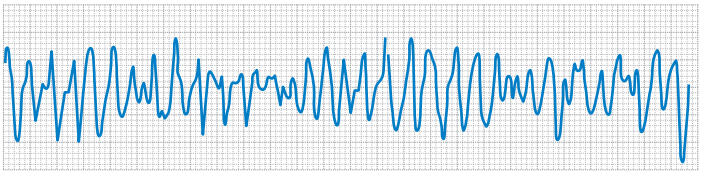

Figure 14-24. The typical positive and negative oscillation of QRS complexes that characterize torsades de pointes ventricular tachycardia are shown.

assessment should be performed and adequate pad contact assured when VF is suspected.

Considerations in the management of VF are as follows:

1. This is a lethal arrhythmia if untreated.
2. Immediate defibrillation (initial dose 2 J/kg) is the definitive therapy. If this is unsuccessful, the energy dose should be doubled (4 J/kg) and repeated. Pediatric paddles (2.2 cm diameter) are generally recommended for children weighing less than 10 kg. Adult paddles (8-9 mm diameter) are suggested for children weighing more than 10 kg to reduce impedance and maximize current flow.
3. Adequate airway control and chest compressions should be rapidly instituted while preparing for defibrillation or between shocks if several defibrillation attempts are needed. Resuscitative drugs should strongly be considered without delaying defibrillation.

Pacemaker Therapy in the Pediatric Age Group

Pacemaker Nomenclature

The pacemaker nomenclature was revised by the North American Society of Pacing and Electrophysiology and the British Pacing and Electrophysiology Group in 2000,[246] as follows (Table 14-3):

- The first letter refers to the chamber(s) paced (A = atrium, V = ventricle, D = dual or both, O = none)
- The second to the chamber(s) sensed (A = atrium, V = ventricle, D = dual or both, O = none)
- The third to the pacemaker's response to sensing (I = inhibited, T = triggered, D = dual response, O = none)
- The fourth to rate modulation (R = rate modulation, O = none)
- The fifth to multisite pacing (A = atrium, V = ventricle, D = dual or both, O = none)

Permanent Cardiac Pacing

Indications

Guidelines for permanent pacing in children, adolescents, and patients with CHD were updated in 2002.[247] Indications include symptomatic sinus bradycardia, bradycardia-tachycardia syndromes, congenital complete AV block, and advanced second- or third-degree AV block.[247,248]

Perioperative Considerations

Device interrogation should be part of the complete preoperative evaluation in all patients with implanted pacemaker systems scheduled for anesthetic care.[249,250] Consultation with a specialist to obtain information regarding unit type, settings, date of and indications for implantation, and underlying rhythm is highly recommended. If unit information is not readily available a chest radiograph provides identification of the device, as indicated by a radiopaque marker. Alternatively, the major pacemaker manufacturers can be contacted on an around-the-clock basis as they maintain computerized records of all implanted devices that can be readily accessed. Results of a recent 15-lead ECG should be reviewed. Reprogramming may be required before the planned procedure to avoid potential problems with pacemaker malfunction related to electrocautery. Electrocautery is one of the most common potential sources of electromagnetic interference in patients with implanted cardiac devices. Recommendations for the perioperative management of these children include (1) the use of bipolar cautery versus a unipolar configuration if possible; (2) avoidance of cauterization near the generator, and (3) positioning of the indifferent plate for electrocautery away from the pacemaker so that the device is not

Table 14-3. NASPE/BPEG Generic Pacemaker Code*

Position Category	I Chamber(s) Paced	II Chamber(s) Sensed	III Response to Sensed Event	IV Rate Modulation	V Multisite Pacing
	A = Atrium V = Ventricle D = Dual (A & V) O = None	A = Atrium V = Ventricle D = Dual (A & V) O = None	I = Inhibited T = Triggered D = Dual (I & T) response restricted to dual chamber devices O = None	R = Rate modulation O = none	A = Atrium V = Ventricle D = Dual (A & V) O = None

*Revised 2000.
Most Common Pacing Modes:
The pacemaker mode, specified by a code, describes the mode in which the pacemaker is operating.
Single-Chamber Pacing:
AAI: atrial demand pacing (atrial pacing and sensing, inhibited on sensed beat)
AAIR: atrial demand pacing (atrial pacing and sensing, inhibited on sensed beat), rate responsiveness
VVI: ventricular demand pacing (ventricular pacing and sensing, inhibited on sensed beat)
VVIR: ventricular demand pacing (ventricular pacing and sensing, inhibited on sensed beat), rate responsiveness
Asynchronous Pacing (No Sensing):
AOO: fixed rate atrial pacing
VOO: fixed rate ventricular pacing
DOO: fixed rate AV pacing
Dual-Chamber Pacing:
DDD: paces and senses both chambers
DDDR: paces and senses both chambers, sensor-driven rate responsiveness

between the electrocautery electrodes. Devices such as the harmonic scalpel and battery operated hot wire hand-held cautery units do not interfere with implanted cardiac devices. Rate-responsive features should be deactivated in the majority of cases. Chronotropic drugs and alternate pacing modalities (transcutaneous, transvenous, epicardial) should be readily available in the event of pacemaker malfunction and compromising underlying rate. Although insertion of a transvenous pacing system has been advised in children with complete AV block undergoing pacemaker implantation, a 10-year review concluded that there is no benefit to routine preoperative temporary pacing in these children.[251] Capture thresholds can be affected by pharmacologic agents, and this should be considered if pacing is required in the child receiving antiarrhythmic drug therapy. Perioperative conditions may also influence pacing thresholds. A magnet should be accessible to allow for asynchronous pacing if required.[252,253] Most generators respond to magnet application by pacing at a fixed rate asynchronously (AOO, VOO, or DOO). A potential issue is that the specific magnet rate, as determined by the manufacturer for the particular device, may differ from the desirable or optimal pacing rate. Thus, the use of a magnet should not be considered a substitute for preoperative pacemaker interrogation/programming. In addition to perioperative ECG monitoring, additional modalities that confirm pulse generation during pacing (esophageal stethoscope for assessment of heart sounds, pulse oximetry, invasive arterial blood pressure monitoring) are strongly encouraged. After the procedure is completed, the device should be tested and reprogrammed.

Transcutaneous Pacing

Several devices that combine defibrillation/cardioversion capabilities and external pacing features are currently available. In children, emergency transthoracic pacing may be considered as a temporizing measure in those with symptomatic bradycardia.[254] Transcutaneous pacing has not been found to be effective in the treatment of asystole in children.[255] Pacing electrode size should be selected according to patient size (usually patients weighing less than 15 kg require smaller adhesive pads). Device settings typically include pacing rate and power output. Sedation may be necessary to tolerate soft tissue discomfort. Prolonged periods of transcutaneous pacing may result in local cutaneous injury. In addition to monitoring for pacemaker capture by ECG, ongoing clinical assessment of the adequacy of cardiac output should be undertaken.

Implantable Cardioverter-Defibrillators

The primary goal of an implantable cardioverter-defibrillator (ICD) is the prevention of sudden death in children at high risk.[247] Although sudden cardiac death is an uncommon occurrence in children, certain patients may be at particular high risk. Individuals with arrhythmogenic right ventricular dysplasia, long QT syndrome, and HCM and those with a history of near-death events may be considered suitable candidates for pacemaker/defibrillator implantation.[248,256] Potential candidates also include those with operated CHD and a history of malignant arrhythmias.[257] At the present time, the experience in the pediatric age group with these devices has been limited and additional data are required to further refine guidelines for use, address safety concerns, and evaluate long-term issues specific to children.[247,257-261] The anesthetic implications of implanted units relate primarily to the potential for surgical electromagnetic interference (electrocautery).[262,263] Perioperative consultation with a cardiologist/electrophysiologist is, therefore, essential in the care of these children. In many cases the devices may need to be adjusted or deactivated before surgery. Careful evaluation and device reprogramming is advisable at the conclusion of the surgical intervention.

Diagnostic Modalities in Pediatric Cardiology

Chest Radiography

The standard posteroanterior and lateral chest radiographs provide a number of clues regarding a child's underlying cardiovascular anatomy; however, plain radiographs in general are an insensitive screening tool for cardiac disease.[264] Children with numerous types of significant CHD may have initially normal-appearing radiographs; alternatively, an infant with a poor inspiratory effort and the presence of a large thymus may give the appearance of cardiomegaly and actually have normal intracardiac anatomy (Fig. 14-25).

Interpretation of a chest radiograph begins with identification of the patient's name and ensuring that the right-left orientation of the radiograph is appropriate. All lines and tubes (including endotracheal tube position if applicable and sternal wires from prior surgeries as well as coils, devices, and stents from prior catheterization procedures) should be followed to verify their location, course, and likely site of termination. The bones and soft tissues should be inspected for evidence of sternal wires, fractures, vertebral anomalies, or wide intercostal spaces, suggesting a prior thoracotomy. Sidedness, including the location of the gastric bubble, liver, and position and orientation of the cardiac mass, should be noted. The lung parenchyma should be examined for evidence of focal consolidation, such as pneumonia or atelectasis, as well as the pulmonary vascular markings. Pulmonary overcirculation or undercirculation helps

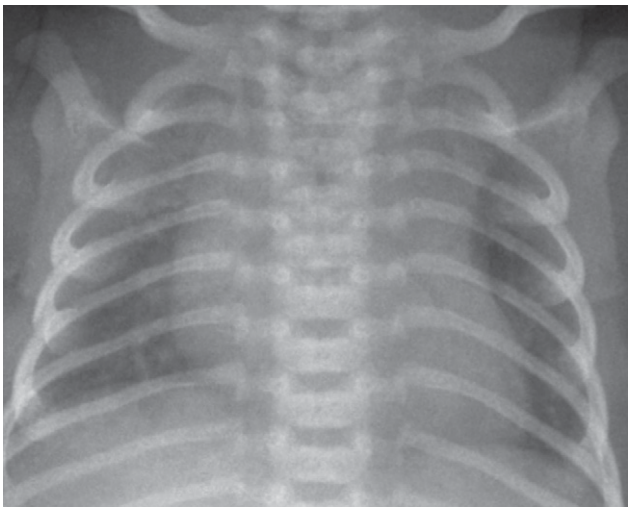

Figure 14-25. Chest radiograph in a neonate with apneic episodes undergoing evaluation to exclude potential cardiac etiology demonstrates the appearance of a poor inspiratory effort and a large cardiothymic silhouette, making the interpretation difficult. No evidence of cardiac pathology was identified in this infant.

guide the differential diagnosis (see earlier discussion on physiologic classification of CHD).

Last, the cardiac silhouette and great vessels should be assessed. In younger children the thymus may obscure the superior portions of the cardiac shadow. Careful inspection of the cardiac silhouette includes an assessment of overall size and evidence of individual chamber/vessel dilation (e.g., rightward cardiac dilation on the posteroanterior film indicating right atrial enlargement or posterior enlargement on the lateral film suggesting left atrial dilation). Prominence of the main pulmonary artery segment if present should be noted because it provides further evidence of the degree of pulmonary overcirculation in left-to-right shunt lesions. The tracheal indentation can usually be seen and is useful in determining aortic arch sidedness, although in a young child with a prominent thymus this can be difficult to assess.

Perhaps more useful than an individual radiograph as a diagnostic tool, however, are serial chest films used to monitor a child's cardiovascular status over time. In a young child with a volume overload lesion such as a large septal defect, the child's physical examination and growth parameters coupled with the degree of cardiomegaly and pulmonary overcirculation are more helpful than other more advanced imaging techniques. Initiation and titration of pharmacologic therapy, as well as the timing of surgery, may also be guided by a plain chest radiograph.

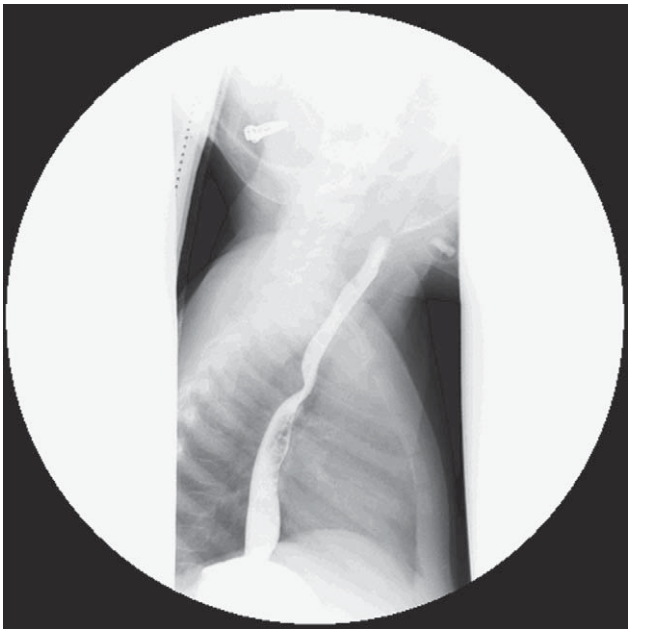

Figure 14-26. Lateral view of a barium swallow in a child with respiratory symptoms demonstrating a filling defect posteriorly in the midesophagus, consistent with an aberrant subclavian artery.

Barium Swallow

The current applications and uses of barium swallow studies in the diagnosis of CHD are limited. To a large extent this modality has been replaced by MRI and chest tomography.[265-267] In some cases, however, a barium esophagogram is used as an initial screening tool when there is concern for the presence of a vascular ring, as in a child with medically refractory gastroesophageal reflux, recurrent pneumonia, or stridor.[268,269] Double aortic arches represent one of the most common types of vascular ring in young children, but other types such as a right aortic arch with an aberrant left subclavian artery and a left-sided ligamentum arteriosus may also cause symptoms. The indentation pattern in the barium column is consistent with the specific vascular anomaly (Fig. 14-26).

Echocardiography

Echocardiography is the diagnostic modality of choice for the initial evaluation and serial assessment in most types of pediatric heart disease. Using a variety of probes, ultrasound waves are used to acquire real time images of cardiovascular structures. Various echocardiographic modalities are available, including transthoracic,[270] transesophageal,[271-274] fetal,[275,276] epicardial,[277] intracardiac imaging,[278] and intravascular ultrasound.[279] Each of these play important roles in the diagnostic evaluation and management of children with suspected or confirmed cardiovascular disease.

Advantages of echocardiography include its noninvasive nature, provision of excellent temporal and spatial resolution, generation of portable real-time images, cost-effectiveness, and ease of use. As with any type of ultrasound, these waves are transmitted well through homogeneous tissues and fluid but poorly through air and bone. In children with significant pulmonary disease, air-filled spaces (pneumothorax) and significant scar tissue image quality may be suboptimal. Another limitation of echocardiography relates to limited acoustic windows in certain patient groups, such as those who have undergone multiple cardiothoracic procedures, older individuals, or children with a significant amount of soft tissue/body fat. For this reason, cardiovascular MRI is being increasingly used for noninvasive imaging in certain subsets of children (see later). Additional challenges of echocardiography include the need to obtain serial two-dimensional (2D) tomographic images by sweeping the transducer scan in multiple planes to mentally translate these images into three-dimensional (3D) structures, and the need for expertise in the interpretation. Despite these limitations, echocardiography remains the primary diagnostic imaging modality in most children and many medical and surgical management strategies are primarily based on the echocardiographic findings.

A standard transthoracic echocardiogram consists of a two-dimensional (2D) examination, M-mode imaging, and Doppler evaluation (color flow, pulsed-wave, continuous-wave modalities). Two-dimensional imaging provides structural assessment of the heart and adjacent vasculature. Cross-sectional images are obtained from a number of windows that allow for excellent anatomic detail in multiple planes (Video Clip 14-11). For the transthoracic approach a typical examination includes the parasternal, apical, subcostal, and suprasternal notch windows. In most cases, this is adequate for a detailed segmental evaluation of the cardiac anatomy as described earlier. M-mode echocardiography allows for a one-dimensional imaging of the heart with excellent temporal resolution (Fig. 14-27). It is otherwise known as an "ice pick" view of the heart in real time. Unfortunately, this technique only provides limited anatomic information, and as such, is primarily used in the assessment of ventricular dimensions and function.

Color flow Doppler techniques allow for evaluation of both directionality and velocity of blood flow. In addition to detecting

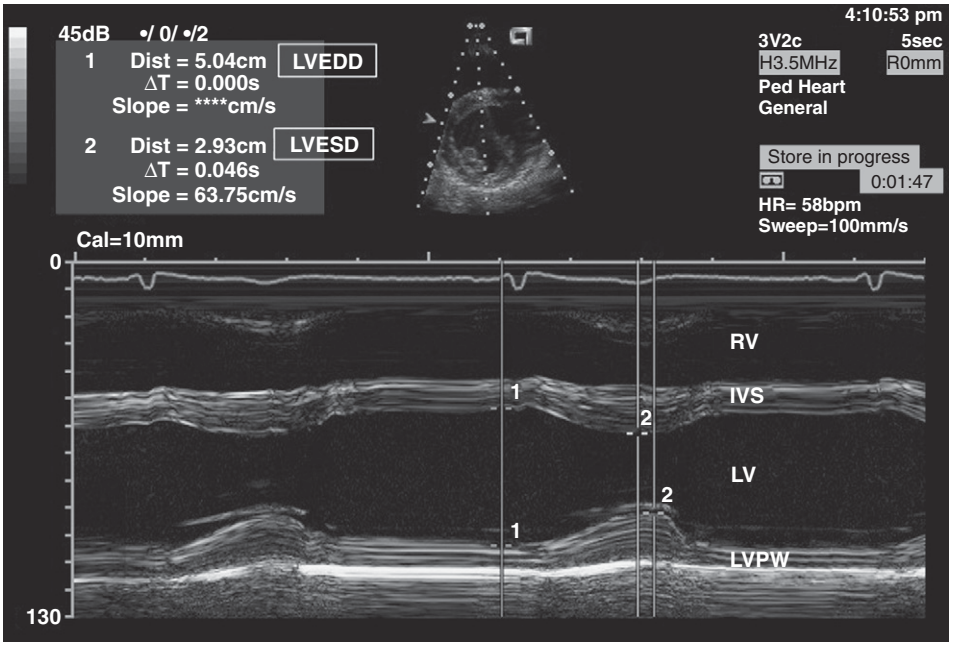

Figure 14-27. M-mode echocardiography allows for determination of left ventricular dimensions and calculation of shortening fraction. RV, right ventricle; IVS, interventricular septum; LV, left ventricle; LVPW, left ventricular posterior wall; LVEDD, left ventricular end-diastolic dimension; LVESD, left ventricular end-systolic dimension.

flow across cardiac valves and great vessels, color flow imaging allows detection of subtle lesions such as small septal defects that can be difficult to identify by standard 2D imaging alone. Traditionally, flow toward the transducer is displayed in red whereas flow away is represented as blue. Turbulent blood flow is associated with increased Doppler velocities and can be readily identified as a mosaic of colors, typically of greenish tint (Video Clip 14-12).

Pulsed- and continuous-wave Doppler represent spectral modalities that complement the color flow data and provide quantitative information. Pulsed-wave interrogation is advantageous in localizing specific sites of stenosis or turbulence but is limited in the magnitude of velocities it can detect. Continuous-wave Doppler, on the other hand, allows for quantification of much higher velocities (see Video Clip 14-12). By using velocities obtained with pulsed- and continuous-wave Doppler, estimates of pressures within various cardiac chambers are possible by applying the simplified Bernoulli equation, which states that the difference in pressure between two locations is approximately four times the square of the velocity of the jet of flow between them:

$$\text{Pressure gradient (in mm Hg)} = 4 \times v^2$$

The applications of 3D echocardiography have been increasingly investigated in the diagnosis of CHD.[280-282] An examination can provide clear and useful volumetric assessments when the images are adequate. A significant advantage of this imaging approach is that it is able to display cardiovascular structures and their interrelationships in detail, in many cases facilitating the understanding of pathologic conditions over 2D imaging. It allows for an enhanced perspective of the margins and geometry of abnormalities, such as septal defects and valvar anomalies. 3D echocardiography may also be particularly useful when interventions are planned. Technologic advances in a number of areas should allow for improved image quality and shorter acquisition times.

Practical Concepts Regarding the Interpretation of an Echocardiographic Report

Measurements of Cardiac Chambers and Vessel Dimensions

A number of measurements are routinely performed during an echocardiographic examination. These generally include ventricular dimensions such as left ventricular end-diastolic (LVED) and end-systolic (LVES) dimensions, thickness of the interventricular septum and left ventricular posterior wall, as well as measurements of valve sizes and great artery dimensions. To determine whether these are appropriate for the child being examined, the measurements are related to values obtained in normal children matched for body-surface area. In many centers the reports include determinations of Z scores to indicate how many standard deviations the observations are from mean values in a comparative population.

Assessment of Ventricular Function

A number of echocardiographic techniques are able to provide information regarding ventricular performance. Two of the most commonly reported indices of ventricular systolic function involve measurements of the extent of shortening, namely, shortening fraction and ejection fraction.

Shortening fraction (SF) represents the percent of change in left ventricular diameter during the cardiac cycle. This is calculated using the following equation:

$$\text{SF (\%)} = \text{LVED dimension} - \text{LVES dimension}/\text{LVED dimension}$$

Values range from 28% to 44%, with a normal mean value of 36%. This index, however, is dependent on ventricular preload and afterload.

Ejection fraction (EF) is the fraction of blood ejected by the ventricle (stroke volume) relative to its end-diastolic volume. In other words, it represents the percentage of blood ejected from the left ventricle with each heart beat. Ejection fraction is derived by volumetric analysis of the left ventricle by means of the following equation:

$$EF\ (\%) = LVEDV - LVESV/LVEDV$$

where LVEDV = left ventricular end-diastolic volume and LVESV = left ventricular end-systolic volume

Normal values vary in the literature but in most studies range between 56% and 78%. A low ejection fraction is generally associated with systolic functional impairment, but cardiac dysfunction may occur in the presence of a normal ejection fraction. Such may be the case of a child with diastolic heart failure.

It is important to also consider that although these functional indices are routinely and easily obtained they have significant limitations. The estimation of ejection fraction is based on geometric assumptions for the elliptical left ventricle. These assumptions thus may not be applicable to a systemic right ventricle or other types of ventricular geometries.[283] There has therefore been an escalating interest in newer echocardiographic approaches and imaging modalities that may provide more sensitive and comprehensive information regarding ventricular performance, even in the absence of clinical disease. Some of the newer techniques being used in the assessment of ventricular function include (1) myocardial performance index (MPI), also known as the Tei index, which combines systolic and diastolic intervals to assess global ventricular function[284-286]; (2) Doppler tissue imaging (DTI) to evaluate intramural myocardial velocities[287]; and (3) strain and strain rate imaging to quantitate the rate of segmental myocardial deformation.[288] Although values in normal children have been established for all of these imaging modalities and alterations in the presence of pathologic conditions have been described,[289,290] additional studies documenting their clinical applications in specific types of cardiovascular pathology are needed.

Estimation of Pressures

When the peak velocity of a tricuspid regurgitant jet is reported, these data can be used to estimate right ventricular systolic pressure and thus pulmonary artery systolic pressure, in the absence of pulmonary stenosis or outflow obstruction (Video Clip 14-13). If, for example, a peak regurgitant velocity of 3 m/sec is recorded across the tricuspid valve, using the simplified Bernoulli equation as discussed earlier, the pressure gradient or difference between the right atrial and right ventricular systolic pressures can be estimated to be $4 \times 3^2 = 36$ mm Hg. If a normal right atrial pressure is assumed (4 to 6 mm Hg), this would predict a right ventricular systolic pressure of approximately 40 mm Hg. Similarly, if the peak or maximal flow velocity across a ventricular septal defect is measured at 4.5 msec, this predicts a pressure gradient of $4 \times 4.5^2 = 81$ mm Hg between the ventricles, implying that the defect is pressure restrictive and the right ventricular and pulmonary artery systolic pressures are relatively low.

Evaluation of Gradients

Estimation of a peak instantaneous gradient is the most clinically useful method for quantifying the severity of obstructions across semilunar valves and outflow tracts. It is derived by application of the simplified Bernoulli equation. These estimates when obtained across the pulmonary valve tend to have a higher correlation with catheterization peak-to-peak gradients than those measured across the aortic valve, where mean gradients (obtained by automated integration of the velocities under a spectral Doppler tracing) are found to have a higher correlation.[291] The mean gradient rather than a peak gradient as determined by Doppler echocardiography is considered a better indicator of the severity of the obstruction across atrioventricular valves and other low-flow venous pathways.

Evaluation of Regurgitant Lesions

Although a number of echocardiographic parameters have been investigated that may facilitate the evaluation of the severity of regurgitant lesions, in most pediatric cardiac centers this remains largely a qualitative assessment. The severity of the pathology is usually characterized as mild, moderate, severe, or combinations thereof when there is overlap among these categories. Serial echocardiographic assessment and comparative data are clinically more meaningful than an isolated report.

Magnetic Resonance Imaging

Cardiovascular MRI/angiography has emerged in recent years as a complementary technology to other traditional imaging modalities (Video Clips 14-14 and 14-15). Benefits have been reported in the assessment of complex pathology,[292,294-298] delineation of systemic and pulmonary vascular anomalies,[297,298] evaluation of global and regional ventricular function,[299] assessment of myocardial viability,[300] and characterization of pulmonary blood supply in children with structural alterations of the pulmonary vascular tree.[301] Particularly interesting applications that may further expand the utility of MRI in cardiovascular medicine include the quantification of left-to-right shunts[302-304] and measurement of blood oxygen saturation.[305] Furthermore, MRI has been found to be of benefit for guiding interventions in pediatric heart disease.[306-308] Most common indications in infants include assessment of complex cardiovascular malformations, delineation of vascular structures (aortic arch, systemic and pulmonary venous drainage), evaluation of possible airway compression, and characterization of cardiac tumors. Despite the significant technical challenges associated with the technique in children, this imaging approach plays an important diagnostic role. In adults with CHD, particularly in those with poor acoustic windows, this is frequently the imaging modality of choice. This may also be the case in children with palliated or repaired CHD.

Although the temporal resolution of MRI is inferior to echocardiography, new sequences and techniques allow for real-time acquisition similar to that of fluoroscopy. Spatial resolution continues to improve as well, particularly with the use of more powerful magnets. An important aspect in the acquisition of MRI data with high spatial resolution is the use of both cardiac and respiratory gating to allow sampling during only specific portions of the cardiac and respiratory cycles. Slow heart rates and low respiratory rates facilitate this process. MRI, in contrast to CT, does not involve radiation exposure, making it preferable for serial examinations that many young children with cardio-

vascular pathology require. However, MRI examinations are associated with the need for multiple anesthetics and their inherent risks.

Because of the nature of the magnetic fields generated in MRI, the presence of several types of metal, including pacemakers, ICDs, cerebrovascular clips/coils, or recently implanted intracardiac or intravascular coils and devices, are considered contraindications. Some artificial devices such as vascular clips, intravascular stents, and atrial septal defect occluder devices are typically made of titanium, allowing for sequences to be acquired that minimize the artifact produced by the foreign material. Stainless steel, on the other hand, as is found in coils used for occlusion of collateral vessels or a patent ductus arteriosus, generates significant artifacts within the study.

An additional limitation of MRI is the need for patient immobility during long examinations for optimal image quality. In small children this requirement usually necessitates the use of deep sedation/general anesthesia.[309-311] In infants with complex, and often cyanotic, CHD, specialists skilled in the care of children with cardiovascular pathology are often asked to provide care during the procedures. The severity of the cardiovascular disease may add to the challenges presented to the anesthesia care provider in a remote location.[312,313] The lengthy nature of the studies and the significant time requirements to perform postprocessing of the images cause MRI to be much more time intensive for the interpreting physician than other noninvasive imaging modalities.

A significant advantage to this technique is avoidance of harmful radiation inherent to traditional catheterization techniques. As MRI technology improves, with faster scans, increasing availability, and decreasing cost, it will continue to play an increasing role in the diagnosis and longitudinal follow-up of congenital and acquired pediatric heart disease.

Computed Tomography

Cardiac computed tomography (CT), with or without ECG gating, has also become an option among the battery of imaging modalities available to the pediatric cardiologist (Fig. 14-28).[292,296,314,315] The major advantage of CT over MRI is the very rapid scan times, such that, in most children, sedation is minimal or not necessary. With multislice CT detectors, a complete CT of the thorax can be performed in less than 10 seconds; and as new 64-slice detectors become increasingly available, the time required for the study will continue to shorten. A significant drawback of CT is the significant radiation burden, although typically estimated to be similar or slightly higher than a diagnostic cardiac catheterization, and the likely need for iodinated contrast agents with their concomitant risks.

Cardiac CT is not as accurate as MRI for delineation of intracardiac anatomy, but it provides excellent spatial resolution and information on extracardiac vasculature. CT has been reported to be of benefit in the evaluation of aortic arch anomalies and vascular rings, as well as in defining the systemic and pulmonary venous returns. In adult patients, multislice CT provides excellent information on coronary arteries and the presence of atherosclerotic disease; but in infants and children with smaller vessels and more rapid heart rates, these images are more difficult to obtain.

Cardiac Catheterization and Angiography

Cardiac catheterization involves the invasive measurement of intracardiac/vascular pressures and blood oxygen saturation coupled with angiography to assess cardiac anatomy and hemodynamics (Figs. 14-29 and 14-30). Before the era of 2D echocardiography, cardiac catheterization was frequently used for diagnostic purposes. At the present time, with the advances in noninvasive imaging, diagnostic procedures represent a relatively small proportion of these studies. Current indications for cardiac catheterization at most centers include (1) the assessment of physiologic parameters such as pressure and resistance data, (2) anatomic definition when other diagnostic modalities are inadequate, (3) need for electrophysiologic testing/treatment, and (4) when interventions are anticipated.

The majority of catheterizations in the current era involve interventions, ranging from endomyocardial biopsies, to angioplasties and stenting of stenotic vessels, dilation of valves, and conduits, to occlusion techniques for both native defects such as a patent ductus arteriosus, septal defects, or fistulous connections and surgically created defects such as Fontan fenestrations (Fig. 14-31, Video Clip 14-16, see Chapter 20). In some cases,

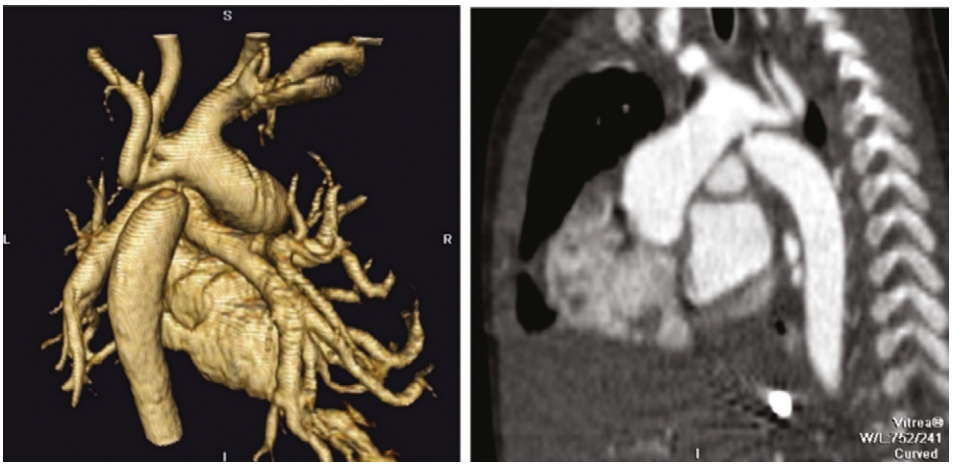

Figure 14-28. These two CT images display the details of the aortic arch anatomy in an infant with severe aortic arch obstruction.

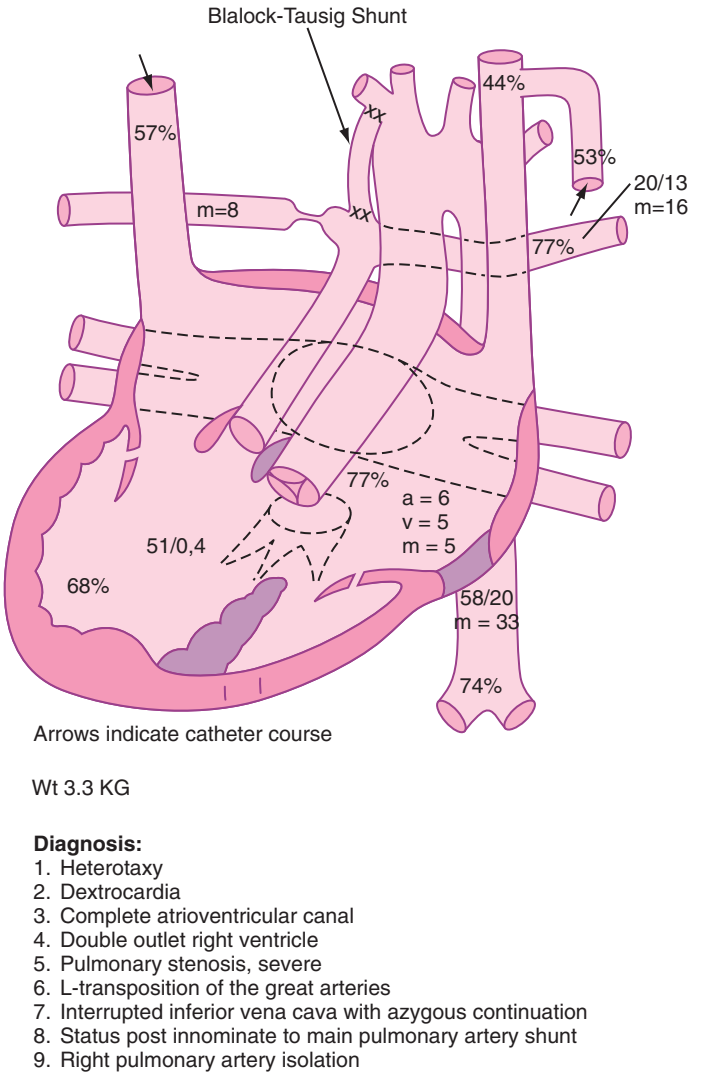

Figure 14-29. Cardiac catheterization. Diagram illustrating the structural abnormalities in a child with complex cardiovascular pathology. Data routinely obtained at cardiac catheterization are depicted, including oxygen saturation determinations, pressure measurements, and hemodynamic calculations. These types of pictorial representations are extremely helpful when caring for children with complex defects.

such as might be the case of critically ill neonates with complex heart disease, catheter-based interventions such as balloon atrial septostomy and other procedures can be lifesaving.

In most children, access to the central circulation is accomplished percutaneously via the femoral approach. Those with occluded femoral veins or with cavopulmonary connections may require venous access through an internal jugular vein. In general, most examinations involve hemodynamic evaluation with recording of pressure data through catheters positioned at various sites of interest. Oxygen saturation data are obtained by reflectance oximetry or blood gas measurement from various cardiac chambers and vessels. It is important to recognize that in contrast to the oxygen saturation calculations derived from a blood gas analysis, reflectance oximetry assessments are actually measured values. This allows for the determination of oxygen content (total amount of hemoglobin in the blood) and, when combined with values of oxygen consumption, for the assessment of blood flows and other calculations (i.e., shunts).[316] Additional data that may be obtained include pressure gradients, cardiac output measurements, and parameters to derive vascular resistances and valve areas.

Fluoroscopy and cineangiography are essential components of most cardiac catheterizations studies. Of the two, cineangiography accounts for the majority of the radiation exposure during the procedure as images are recorded during the injection of contrast material typically at 15 or 30 frames per second.[317] Most angiograms are obtained during biplane imaging by positioning the equipment to obtain optimal views that allow for delineation of the pathology in question (axial angiography) (Video Clip 14-17).[318,319]

Although cardiac catheterization has evolved over the years providing improved safety, it is an invasive procedure involving a number of risks and potential for morbidity and mortality, albeit relatively small. Risks associated with cardiac catheteriza-

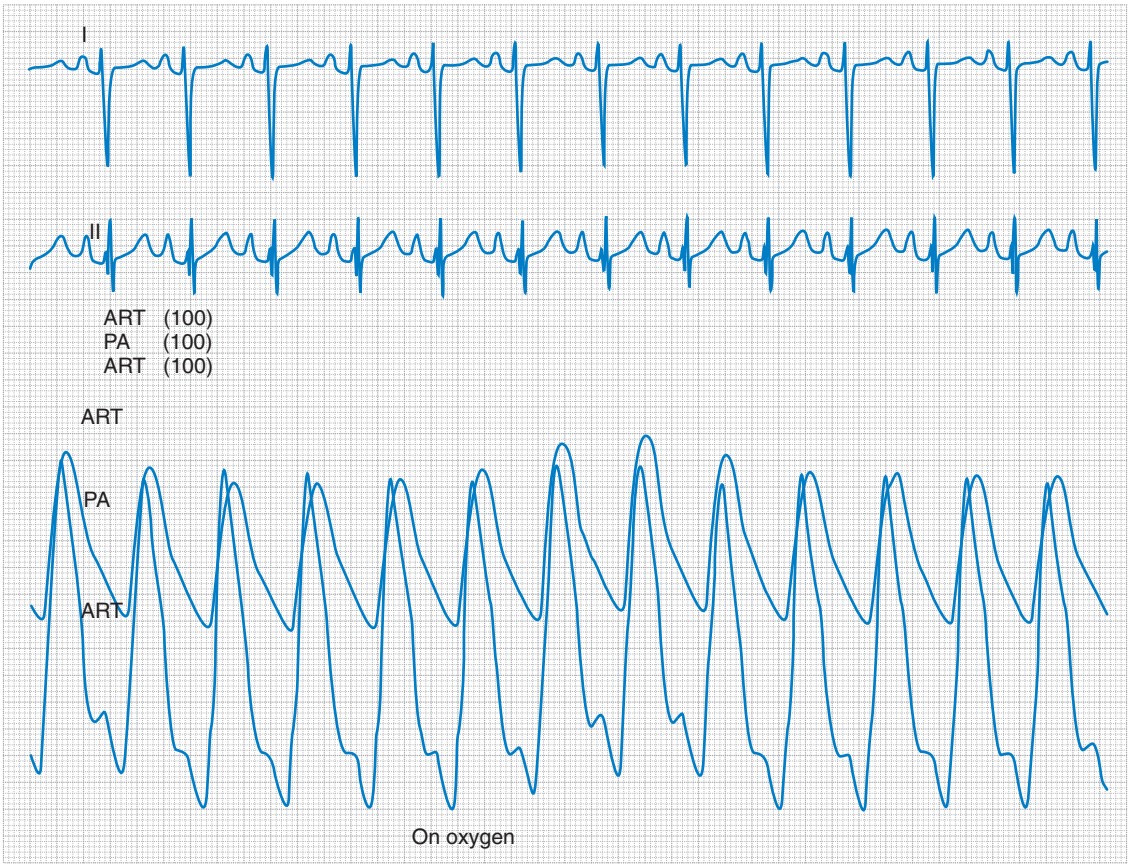

Figure 14-30. Cardiac catheterization pressure recording displays a typical hemodynamic tracing (scales are the same for both pressures). Note that the pulmonary artery systolic pressures are at systemic levels in this child with multiple left-sided obstructions. ART, systemic arterial pressure; PA, pulmonary artery pressure.

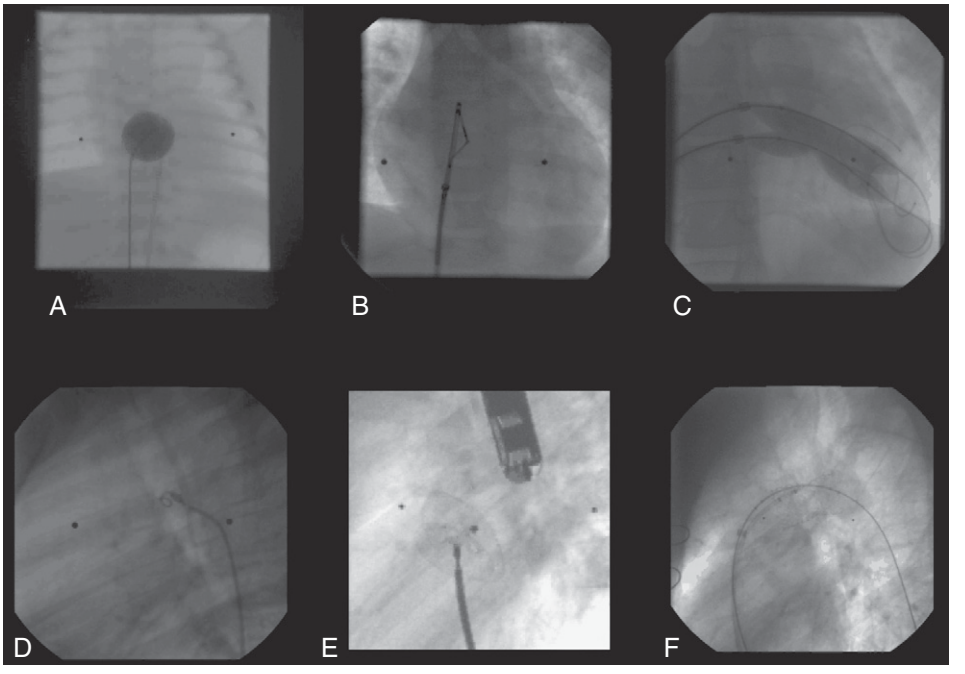

Figure 14-31. Catheter-based procedures. *Upper panel:* (**A**), balloon atrial septostomy; (**B**), blade atrial septostomy; (**C**), mitral balloon valvuloplasty. *Lower panel:* (**D**), placement of ductal occluder device; (**E**), transcatheter closure of secundum atrial septal defect; (**F**), pulmonary artery dilation with stent placement.

tion include excessive blood loss, vascular complications,[320,321] infection, arrhythmias, vascular/cardiac perforation,[322] systemic air embolization, myocardial ischemia, and those associated with the administration of contrast agents. These are more likely to occur in infants and small children.[323] Although transient increases in body temperature may occur after a study, the incidence of endocarditis is extremely rare. Interventional catheterizations, by the nature of the procedures, are associated with a greater rate of complications. However, as transcatheter interventions become safer and more effective, an increasing number of children may obviate the need for surgery, often undergoing procedures on an outpatient basis.[324] New techniques continue to evolve in this field, such as the percutaneous placement of valves,[325] strategies that combine cardiac catheterization and surgical intervention, so-called hybrid procedures,[326-328] and catheter-based interventions during fetal life.[329]

Practical Concepts Regarding the Interpretation of a Cardiac Catheterization Report

Pressure Data

Atrial pressure tracings are characterized by several waves (a, c, and v waves) and descents (x and y). The right atrial pressure is typically "a" wave dominant. The mean right atrial pressure is normally less than 5 mm Hg. In the presence of significant tricuspid valve regurgitation or a junctional rhythm, the "v" wave becomes the dominant wave. The left atrial pressure tracing in contrast to the right atrium, displays "v" wave dominance, which is accentuated during mitral regurgitation. The mean left atrial pressure rarely exceeds 8 mm Hg. The reported values for atrial pressures correspond to the "a" and "v" waves and mean pressures. Ventricular pressures are recorded and reported during systole, at end systole, and at end diastole. For the right ventricle the systolic pressure is normally in the 25- to 30-mm Hg range, with end-diastolic pressure of 5 to 7 mm Hg. The systolic pressure in the left ventricle normally increases with age and should equal the systolic arterial pressure; the end-diastolic pressure is typically less than 10 mm Hg. The pulmonary artery pressure is reported in terms of systolic, diastolic, and mean pressures. The systolic pulmonary artery pressure in a normal child should be equal to the right ventricular systolic pressure, and the mean pulmonary artery pressure should not exceed 20 mm Hg. The pulmonary artery wedge pressure is obtained by advancing a catheter into a distal vessel until this is occluded, reflecting the left atrial pressure. The aortic pressure and contour of the tracing varies depending of the site of interrogation. Typically, there is an increase in the systolic pressure as the catheter navigates toward the peripheral circulation. This phenomenon is known as "pulse wave amplification."

Pressure gradients represent the differences between two distinct sites and can be measured in a number of ways (mean gradient and peak gradient). It is important to consider that a number of factors may affect the determination of pressure gradients. The flow across the lesion is significantly influenced by the severity of the obstruction and the ventricular function.

Shunt Calculations

Shunts are characterized in terms of their direction (left-to-right, right-to-left, bidirectional) and magnitude. Left-to-right shunts can be quantified based on the ratio of the pulmonary ($\dot{Q}p$) to systemic ($\dot{Q}s$) blood flow ratio as follows:

$$\dot{Q}p/\dot{Q}s = \frac{(Sao_2 - MVo_2)}{(PVo_2 - PAo_2)}$$

where Sao_2 = systemic arterial O_2 saturation, MVo_2 = mixed venous O_2 saturation, PVo_2 = pulmonary venous O_2 saturation, and PAo_2 = pulmonary arterial O_2 saturation.

A $\dot{Q}p:\dot{Q}s$ ratio that exceeds 3:1 is considered a significant shunt, although smaller ratios may be associated with significant symptomatology.

Cardiac Output Determinations

The volume of blood ejected by the heart into the systemic circulation, or cardiac output, can be derived in several ways. Thermodilution measurements use saline as an indicator to measure pulmonary blood flow. In the absence of intracardiac shunts this is equivalent to the systemic blood flow or cardiac output (expressed as liters per minute). In the Fick method, oxygen is used as an indicator and cardiac output is obtained by the application of the following formula:

$$\dot{Q}s\ (L/min) = Vo_2\ (L/min)/Sao_2\ content - MVo_2\ content$$

where Vo_2 = oxygen consumption (assumed or measured), Sao_2 = systemic arterial O_2 content, MVo_2 = mixed venous O_2 content, and O_2 content = O_2 saturation × (1.36 × 10 × hemoglobin concentration).

Vascular Resistances

Resistance represents the change in pressure in the systemic or pulmonary circulation with respect to flow. This is expressed as mm Hg/L/min (Wood units) and is usually normalized for body surface area.

The systemic (SVR) and pulmonary vascular resistance (PVR) are derived as follows:

SVR = (aortic mean pressure − right atrial mean pressure)/$\dot{Q}s$

PVR = (pulmonary artery mean pressure − pulmonary capillary wedge pressure or left atrial pressure)/$\dot{Q}p$

Considerations in the Perioperative Care of Children with Cardiovascular Disease

General Issues

The anesthetic care of children with heart disease[330] is challenged by the following factors:

- The remarkable spectrum of disease, from structural defects to acquired pathology
- The wide range of congenital lesions and their underlying physiologic consequences
- The numerous surgical procedures in CHD (Table 14-4) and their hemodynamic implications
- The fact that many parents are unaware of the full extent or details of their child's lesion or abnormalities

At the same time, to optimally care for these patients the following objectives should be met:

- Familiarity with the cardiovascular pathology
- Understanding of the physiologic abnormalities and available therapies

Table 14-4. Surgical Procedures for Congenital Heart Disease

Procedure	Description	Goal/Result
Arterial switch (Jatene) operation	Arterial trunks transected above the level of the semilunar valves, relocated to their appropriate respective ventricles, coronary arteries reimplanted into the neoaortic root	Establishes the normal ventricular-arterial connection (right ventricle to pulmonary artery and left ventricle to aorta)
Atrioventriculoseptal defect (atrioventricular canal) repair	Patch closure of atrial and ventricular communications, reconstruction of atrioventricular valves, closure of cleft in left-sided atrioventricular valve	Eliminates the intracardiac shunt
Blalock-Taussig shunt	Subclavian artery to pulmonary artery communication. Modified implies placement of a graft.	Allows for or increases pulmonary blood flow
Central shunt, Waterston shunt, Pott shunt	Creation of communication between systemic and pulmonary circulations	Allows for or increases pulmonary blood flow
Closure of septal defects	Patch or primary closure of communications at the atrial or ventricular levels	Eliminates the intracardiac shunt
Coarctation repair	Relief of aortic arch obstruction (various approaches)	Establishes patency across the aortic arch
Damus-Kaye-Stansel	End-to-side anastomosis of main pulmonary artery onto the aorta. Necessitates reestablishing pulmonary blood flow via an alternative route (graft from a systemic artery into the pulmonary artery or a right ventricular to pulmonary artery conduit)	Allows for unobstructed systemic outflow in the context of single ventricle associated with obstruction to aortic flow or other settings
Division/ligation of patent ductus arteriosus	Obliteration of the ductus arteriosus	Eliminates shunting at the level of the great arteries
Fontan procedure	Connection that allows for inferior vena cava blood to drain into pulmonary circulation	Separates the pulmonary and systemic circulations in patients with single ventricle physiology. Usually final step in the single-ventricle palliation pathway.
Glenn anastomosis (cavopulmonary connection)	Superior vena cava to pulmonary artery direct anastomosis (bidirectional implies flow from superior vena cava into both pulmonary arteries)	Provides for pulmonary blood flow while unloading the single ventricle. May be first or intermediate step in the single-ventricle palliation pathway.
Konno-Rastan procedure (aortoventriculoplasty)	Enlargement of the left ventricular outflow tract and aortic annulus. The defect created in the ventricular septum to enlarge the outflow tract is repaired with a large patch.	Alleviates subvalvar and valvar aortic obstruction. When the aortic root is replaced by an autologous pulmonary root this is referred to as a Ross-Konno. Alternatively, cryopreserved homograft tissue may be used in the form of an extended root replacement.
Norwood procedure (stage I palliation)	Involves aortic reconstruction, an atrial septectomy, and placement of a systemic-to-pulmonary artery shunt	Addresses systemic outflow tract obstruction by allowing the right ventricle to eject into a reconstructed aorta. The atrial septectomy provides for unobstructed drainage of the pulmonary venous return into the right atrium. The systemic-to-pulmonary artery shunt supplies the pulmonary blood flow.
Pulmonary artery banding	Constrictive band placed around main pulmonary artery	Limits excessive pulmonary blood flow
Rastelli operation	Creation of an intracardiac tunnel that allows for left ventricular output into the aorta while closing a ventricular septal defect and placement of a right ventricular conduit to pulmonary artery	Allows for the left ventricle to eject solely into the aorta at the same time as it abolishes the intracardiac shunting at the ventricular level and provides for unobstructed pulmonary blood flow. The procedure results in separation of the pulmonary and systemic circulations.

Table 14-4 (continued). Surgical Procedures for Congenital Heart Disease

Procedure	Description	Goal/Result
Sano modification of the Norwood procedure	Placement of graft between the right ventricle and main pulmonary artery as an alternative to a modified Blalock-Taussig shunt in the Norwood operation	Provides for pulmonary blood flow
Senning/Mustard procedure (atrial switch)	Intra-atrial baffle procedure	Allows for pulmonary venous blood to be rerouted through the tricuspid valve into the right ventricle (as the systemic chamber that ejects into the aorta). At the same time the systemic venous return is channeled across the mitral valve into the left ventricle, which pumps into the main pulmonary artery.
Tetralogy of Fallot repair	Closure of ventricular septal defect and relief of right ventricular outflow tract obstruction	Eliminates intracardiac shunting at the ventricular level (cyanosis) and addresses the right ventricular outflow tract obstruction (frequently at several levels)
Truncus arteriosus repair	Closure of the ventricular septal defect and establishment of right ventricular to pulmonary artery continuity (usually via a homograft)	Abolishes the intracardiac shunting and restores the normal connection between the ventricles and great arteries
Valvectomy	Valve excision	Relieves valvar obstruction
Valvotomy	Opening of stenotic valve	Relieves valvar obstruction
Valve replacement	Placement of bioprosthetic or mechanical valve	Addresses valvar pathology (obstruction and/or regurgitation)
Valvuloplasty	Valve repair	Relieves valvar regurgitation and/or stenosis

- Recognition of compensatory mechanisms, signs of limited reserve, and potential perioperative risks
- Ability to identify the potential impact of the scheduled intervention/surgical procedure on the patient's underlying condition and anticipate how this will be tolerated

The combination of the daunting challenges and difficult objectives can be intimidating even to the most experienced clinician. Thus, when caring for children with cardiovascular disease, an interdisciplinary approach is desirable and highly recommended, allowing for the formulation and execution of optimal management plans. If available, consultation with the child's cardiologist or primary care physician should include inquiries about the details of the child's disease, overall clinical status, past and current medical treatment, prior surgical interventions, and presence of residual pathology. The interaction between members of the perioperative team should allow the opportunity for an exchange of information, discussion of concerns, and recommendations that may facilitate patient care as well as the development of comprehensive care plans.[331] This is particularly important in the management of children with complex pathologic processes.

A complete medical history and focused examination is essential during the preoperative assessment. In addition to providing the opportunity for evaluation of the child's disease processes, overall clinical status, and functional reserve, this allows for appraisal of issues that may affect anesthetic management (limited vascular access, difficult airway, gastroesophageal reflux, manipulations to manage pulmonary and systemic blood flow and pressures). Available diagnostic studies should be reviewed (e.g., ECG, chest radiograph, echocardiogram, Holter monitor, cardiac catheterization). On occasion, depending on the nature of the procedure, complexity of the disease, and potential impact on perioperative outcome, additional evaluation and/or diagnostic studies may be warranted. In many cases, the anesthesiologist plays a major role in the determination of whether the available information is adequate.

A fundamental goal in the preoperative evaluation is the identification of children who are at increased risk because of cardiac and pulmonary limitations imposed by their cardiovascular disease. After the preoperative visit, the anesthesiologist caring for a child with CHD should have an adequate understanding of the pathophysiology of the cardiac defect and implications of any previous interventions. Abnormal indices that should raise potential concerns include hypoxemia (Spo_2 less than 75%), pulmonary to systemic blood flow ratio ($\dot{Q}p:\dot{Q}s$) exceeding 3:1, outflow tract gradients over 50 mm Hg, pulmonary hypertension (mean pulmonary artery pressure above 30 mm Hg), increased pulmonary vascular resistance index (>2 Woods units · m^2) or polycythemia (hematocrit >60%). In addition, the following clinical states may place children at significant risk for severe cardiopulmonary decompensation during anesthesia and surgery: history of recent congestive heart failure, uncontrolled arrhythmias, severe ventricular dysfunction, unexplained syncope, substantial exercise intolerance, or any condition associated with significant functional cardiac or pulmonary impairment.

Clinical Condition and Status of Prior Repair

Children with CHD may present for anesthetic care before catheter-based or surgical interventions for their cardiovascular disease or after palliation or definitive procedures. It is important to recognize that true "corrective procedures" are those that

result in a normal life expectancy and normal cardiovascular reserve.[332] They generally require no further medical or surgical treatment. As such, only a few procedures fulfill these criteria: ligation/division/occlusion of a patent ductus arteriosus and closure of an isolated secundum atrial septal defect. Other interventions or surgical procedures may result in repair or "correction," however not necessarily in normal hemodynamics or life expectancy. In fact, some assume limitation in cardiovascular reserve and the need for close follow-up, further medical management, and potential or additional surgical therapies. In other cases, as in children with palliated CHD, the circulation may still be abnormal. These individuals have been reported to be at a greater risk of adverse perioperative events.[333-335]

The effect or impact to the heart and other systems during previous procedures also requires careful consideration. A number of problems may remain or develop after surgical intervention. These include residual shunts, valvar stenoses or outflow tract obstruction, valvar regurgitation, pulmonary hypertension, arrhythmias, and ventricular dysfunction. Children who require a detailed appraisal of perioperative risks include those with residual significant pathology; suspected or known pulmonary hypertension, single-ventricle physiology (including patients post Norwood procedures, Glenn operations, or Fontan palliations), and after conduit placement or cardiac transplantation (see Chapters 15, 16, and 21).

Summary

Caring for children with heart disease is a major aspect of pediatric anesthesia practice. The spectrum of pathology in the pediatric age group includes a wide range of structural defects as well as varied acquired diseases. The ability to provide optimal care is dependent on an understanding of the basic pathophysiology of the lesions; familiarity with the commonly used diagnostic modalities and their clinical applications; and medical and surgical treatment options available to affected individuals. In this chapter we have presented basic concepts in cardiology that may enhance the overall knowledge of the practicing anesthesiologist in pediatric cardiovascular disease.

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