

The Infant With Undiagnosed Cardiac Disease in the Emergency Department

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Infants who present to the emergency department with previously undiagnosed cardiac disease will often present with nonspecific complaints. A thorough physical exam and appropriate testing will typically lead to the correct diagnosis and treatment. These infants can be divided into 3 groups: those with structural congenital heart disease; those with an arrhythmia; and those with acquired cardiac disease such as myocarditis. This article will provide an overview to the identification of infants with undiagnosed cardiac disease. Clin Ped Emerg Med 6:200-206 © 2005 Published by Elsevier Inc.

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The infant who presents to the emergency department (ED) with previously undiagnosed cardiac disease is an unusual and often difficult diagnostic and management problem for the emergency physician [1]. Children, especially infants who are nonverbal, generally do not present with cardiac-specific symptoms such as chest pain or palpitations. In fact, most infants with a cardiac problem will present with nonspecific complaints such as fussiness or poor feeding. The differential diagnosis for such complaints is vast, whereas undiagnosed cardiac disease is rare. In addition, the presenting signs often suggest other problems that are more commonly seen in infants [2]. Therefore, these children often undergo extensive testing before a cardiac diagnosis is made. However, most of these children will demonstrate signs that can be detected on a thorough physical exam usually allowing the emergency physician to correctly diagnose and treat them.

Infants may present with unrecognized cardiac disease such as structural congenital heart disease (CHD) or a predisposition for an arrhythmia. They can also develop acquired cardiac disease such as myocarditis. This article will review the presentation, diagnosis, and management of these patients.

Undiagnosed Structural CHD

Congenital heart disease occurs in 8 of 1000 live births. Many newborns with CHD are diagnosed in utero or

before leaving the hospital after birth. However, some may be discharged home as their symptoms are not yet apparent or are missed on initial physical exam, especially with the recent trend toward earlier discharge after delivery. Wren et al [3] found that 55% of 1061 children with CHD undergoing a routine neonatal examination had no detectable murmur. Another recent study found that screening pulse oximetry was not reliable in detecting CHD in the newborn nursery [4]. Most cases of CHD present during infancy, but there are still reported cases of CHD diagnosed after infancy [1,5]. There is little published research on the ED presentation of pediatric patients with CHD. A retrospective review by Savitsky et al [1] described 77 patients who presented to an ED over a 5-year period with an acute problem related to their CHD. Only 8 of these patients were previously undiagnosed.

Infants presenting to the ED with undiagnosed structural CHD can be divided into groups based on their presentation, which can be related to their underlying pathophysiology. These 4 groups include those with

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cyanotic CHD and sudden onset of acute or worsening cyanosis; those with left-sided outflow obstruction or ventricular failure who present with shock due to diminished systemic blood flow; those with abnormal coronary arteries who present with myocardial infarction (MI) and shock; and those who present with congestive heart failure (CHF).

Cyanotic CHD

Patients with cyanotic CHD have a structural lesion that causes blood to be shunted from the right to the left side of the heart, bypassing the lungs and oxygenation. The shunt may be through another structural defect such as a ventricular septal defect (VSD) or a patent foramen ovale, through the ductus arteriosus, or by a combination of these mechanisms. The most common lesions that cause cyanosis include pulmonary atresia or stenosis, cyanotic tetralogy of Fallot (TOF), total anomalous pulmonary venous return (TAPVR), transposition of the great arteries, tricuspid atresia, and truncus arteriosus. These children are usually diagnosed at birth or before leaving the newborn nursery but can be discharged from the nursery unrecognized if they have a patent ductus arteriosus (PDA) masking the lesion. In this situation, the ductus arteriosus provides sufficient pulmonary blood flow to maintain a relatively high systemic oxygen saturation, so that hypoxia is not readily apparent.

Eventually, when the ductus arteriosus closes in these patients, pulmonary blood flow decreases, and these children become acutely hypoxic. This generally occurs in the first 2 weeks of life but can happen as late as many weeks after birth. However, cyanosis can be missed on physical exam, especially in children with anemia or dark skin. Pulse oximetry readings may be difficult to obtain in an agitated or distressed infant, especially if the perfusion is compromised because of acidosis. Because cyanosis may not be recognized, the chief complaint may be nonspecific or seemingly unrelated, such as fussiness, vomiting, or shortness of breath. If the cyanosis is unrecognized for a period of time, the child may present in shock or with end organ failure.

When cyanosis is noted, it must first be determined whether it is central (lips, tongue, mucous membranes) or peripheral (circumoral, hands, feet). Peripheral cyanosis or acrocyanosis is common in normal newborns and may be accentuated if the child is in distress or ill. Poor perfusion from shock can also lead to peripheral cyanosis. If the cyanosis is central, a hyperoxia test can help to differentiate cardiac causes from other causes of hypoxia. The hyperoxia test consists of putting the patient in 100% inspired oxygen and measuring the PO₂ from an arterial blood gas (preferably obtained from the right radial artery). If the PO₂ remains less than 150 mm Hg, then the child's hypoxia is more consistent with cardiac shunting than with a pulmonary problem. It should be noted that pulse oximetry is not a reliable substitute for PO₂ in the

hyperoxia test. Another cause of hypoxia that may not respond to the hyperoxia test is methemoglobinemia.

It is important to remember that it is not necessary to diagnose the particular type of cyanotic CHD before initiating therapy as described below. However, once stabilization has begun, diagnostic tests that may be helpful include an electrocardiogram (EKG), chest x-ray, and echocardiogram. Typical EKG and x-ray findings in cyanotic CHD are presented in Table 1.

Emergency department management of these children involves first attending to the airway and breathing. Many of these children will have already been intubated for their hypoxia and respiratory distress before the diagnosis of CHD is made. They may also require management for shock.

To reopen the ductus arteriosus and increase pulmonary blood flow, prostaglandin E₁ (PGE₁) at a dose of 0.05 to 0.1 μm/kg per minute should be initiated as soon as cyanotic CHD is recognized as the cause of the hypoxia. It is not necessary to know what form of cyanotic CHD the child has before initiating PGE₁. An immediate effect on the ductus with concomitant improvement in oxygenation should be seen. The most important side effect of PGE₁ is apnea, occurring in about 12% of patients [6]. For that reason, if the child has not already been intubated, it may be prudent to do so before transport. One study looked at the use of aminophylline in the management of PGE₁-induced apnea in 42 infants with cyanotic CHD and found a decrease in the rate of apnea and need for endotracheal intubation [7]. Hypotension and fever are also side effects of PGE₁ [6].

Table 1 Cyanotic CHD and typical chest x-ray and EKG findings.

CHD lesion	EKG	Chest x-ray
TOGV	RVH	Increased pbf, "egg on string" cardiac silhouette
TAPVR	RVH	Increased pbf, "snowman" or "figure-of-8" cardiac silhouette
Tricuspid atresia	LVH	Decreased pbf
Pulmonary atresia/stenosis	RVH	Decreased pbf
TOF (cyanotic form)	RVH	Decreased pbf, "boot-shaped" heart, 30% with right aortic arch
Truncus arteriosus	RVH or BVH	Increased* or decreased pbf, 30% with right aortic arch

BVH indicates biventricular hypertrophy; LVH, left ventricular hypertrophy; pbf, pulmonary blood flow; RVH, right ventricular hypertrophy; TOGV, D-transposition of the great vessels.

* The more pbf the less cyanosis.

A pediatric cardiologist should be consulted as soon as possible for definitive diagnosis. If necessary, arrangements should be made to transfer the child to a facility that has pediatric critical care, cardiology, and cardiothoracic surgery services. It is important to remember that the risks associated with the transport of these infants are significant, and a specialized transport team with the most experience available should be used [8]. After initial stabilization, infants with ductal-dependent lesions will require intervention to provide a more long-term solution for their abnormality. Surgical or catheter-directed therapy may be useful, depending on the specific anomaly.

Left-Sided Outflow Obstruction or Ventricular Failure and Shock

Other ductal-dependent congenital lesions that present in early infancy (as the ductus closes) include those that depend on a PDA for systemic circulation. These include coarctation of the aorta and aortic stenosis. In these patients, the PDA allows blood to bypass the obstruction and reach the systemic circulation. An infant with a hypoplastic left ventricle will also be dependent on a PDA for adequate systemic circulation. When the ductus closes, these patients have poor systemic perfusion and may present with shock and CHF. They may also, however, present with nonspecific symptoms and signs including lethargy, irritability, and mottling. In patients with coarctation, one may note a decreased blood pressure and oxygen saturation in the lower extremities as compared with the right arm.

Recognition that the shock is due to a ductal-dependent cardiac lesion is essential. These patients are often initially thought to have septic shock and are treated accordingly. Once the need for reopening the ductus arteriosus has been recognized, PGE₁ should be initiated, as with the patients with cyanotic CHD. The dose is 0.05 to 0.1 µg/kg per minute, and immediate improvement in systemic perfusion should be seen. In patients with coarctation of the aorta, a direct effect on the aorta has been postulated to explain a series of cases in which the ductus did not open and yet the patient's condition improved in response to PGE₁ [9]. With an open ductus, the relative flow to the systemic and pulmonary vascular systems depends largely on pulmonary vascular resistance. Interventions that decrease pulmonary vascular resistance should be avoided, as this will divert more flow from the systemic circulation. In some cases, systemic perfusion can be improved by increasing pulmonary vascular resistance through controlled hypoventilation and positive end expiratory pressure.

Patients with impaired systemic perfusion may have significant acidosis and other complications of poor perfusion. This may include multiple organ failure, including neurologic injury. Inotropic support, sodium bicarbonate, and medications to decrease systemic vascular resistance may also be helpful.

A pediatric cardiologist should be consulted as soon as possible for definitive diagnosis. If necessary, arrangements should be made to transfer the child to a facility that has pediatric cardiology and cardiothoracic surgery services. The concerns mentioned above in the discussion on PGE₁ for cyanotic lesions, regarding the side effects of PGE₁ and risks of transport, should be considered. After initial stabilization, these children generally require surgical or catheter-directed therapy to stabilize their CHD. Surgical interventions may be performed in stages for some lesions, such as hypoplastic left heart syndrome. Therapeutic cardiac catheterization may be used for certain critical lesions such as coarctation of the aorta or aortic stenosis.

Anomalous Coronary Arteries and Myocardial Infarction

Myocardial infarction is exceedingly rare in infancy. It is most commonly due to congenitally aberrant coronary arteries, particularly anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA).

In ALCAPA, the left coronary artery arises from the pulmonary artery. Coronary perfusion of the left ventricle ordinarily occurs during diastole. The problem in ALCAPA is that the diastolic pressure in the pulmonary artery is too low to adequately perfuse the left ventricular myocardium. The left coronary system is therefore dependent on collateral circulation from the right coronary artery.

Immediately after birth, the pulmonary vascular resistance is relatively high, so there is adequate coronary perfusion of the myocardium. However, as pulmonary vascular resistance falls in the weeks after delivery, an increasing proportion of the left coronary flow drains retrograde to the pulmonary artery, creating a "steal syndrome," and resulting in hypoperfusion of the left ventricular myocardium. As the pulmonary vascular resistance and pulmonary artery pressure drop, flow from the left coronary artery into the pulmonary artery progressively increases. Symptoms typically begin after the first few weeks of life. Initially, ischemia is intermittent, occurring with exertion; for an infant, this includes feeding and crying.

Symptoms of myocardial ischemia or CHF typically appear between 2 weeks and 6 months after birth, and include recurrent episodes of restlessness, irritability, incessant crying, and dyspnea, often associated with pallor and sweating. These episodes are most frequent during feeding. A further increase in myocardial oxygen demand from stresses such as a viral infection may lead to infarction of the left ventricle. This effect on the myocardium leads to the development of CHF. Infants may present with wheezing and be mistakenly diagnosed as having bronchiolitis. Signs of CHF may be present, including tachypnea, tachycardia, gallop rhythm, cardiomegaly, and hepatomegaly. A murmur of mitral insuffi-

ciency may be heard, the result of infarction of a papillary muscle.

With ALCAPA, a chest radiograph usually shows cardiomegaly with evidence of interstitial pulmonary edema. The EKG classically shows abnormal Q waves in leads I, aVL, and V4 through V6, as well as ST-segment elevation in leads V4 through V6, consistent with an anterolateral infarct [10]. Echocardiography with Doppler color flow mapping currently is the method of choice to confirm the abnormal origin of a coronary artery. Doppler color flow mapping shows blood flowing from the coronary artery into the pulmonary artery. Mitral insufficiency, decreased cardiac function, and regional left ventricular wall motion abnormalities may also be seen. A scoring system using EKG and Doppler echocardiogram findings has been proposed to differentiate ALCAPA from dilated cardiomyopathy [11]. Cardiac catheterization to identify the origin and course of coronary arteries carries a high degree of risk in these patients and is necessary only when diagnosis by echocardiography is not definitive.

The definitive treatment for this anomaly is surgical. Left coronary artery reimplantation into the aorta is the preferred approach. Before surgical repair, supplemental oxygen should be administered to prevent hypoxia. Sedation and analgesia should be used to reduce anginal pain and prevent the tachycardia that increases myocardial oxygen demand and decreases oxygen supply. The patient should be monitored for arrhythmias. Inotropic support, to raise diastolic pressure and coronary artery perfusion, should be used cautiously because it also may increase afterload, myocardial work, and myocardial oxygen demand.

The prognosis for patients after surgery depends to a large extent on the degree and duration of preoperative myocardial insult. Early repair carries a good prognosis [12]. Patients diagnosed beyond infancy can sustain permanent myocardial damage, and a heart transplant may be the only therapeutic option. More than 80% of infants with this anomaly develop signs and symptoms of cardiac damage or failure in infancy, and about 65% to 85% die before 1 year of age, if unrepaired. Those who are diagnosed beyond infancy typically have well-established coronary artery collateral circulation. These individuals may present later in childhood, adolescence, or even in adulthood with angina on effort or with CHF from mitral incompetence.

Other causes of MI in infancy and childhood include other congenital coronary artery anomalies, Kawasaki disease, CHD (postoperative), and hypertrophic cardiomyopathy. Older patients typically present with more classic myocardial ischemia symptoms or arrhythmias, whereas younger patients have less specific complaints, such as respiratory distress, feeding problems, or limited endurance. Initial stabilization focuses on the acute findings, whether ischemia, heart failure, hypotension, or arrhythmia. Long-term therapy is individualized based

on the underlying pathology and is beyond the scope of this discussion.

CHD Presenting With CHF

Ninety percent of cases of CHF in children occur in the first year of life. The vast majority of these are related to CHD. Many types of CHD in infants can cause CHF. These conditions may be divided into groups by the typical age of presentation (Table 2). At birth, volume overload lesions such as severe tricuspid or pulmonary insufficiency and large systemic arteriovenous fistulae are most common. Lesions affecting left ventricular output that are ductal-dependent may also lead to CHF in infancy. Examples include hypoplastic left heart, coarctation of the aorta, aortic stenosis, aortic atresia, and mitral valve atresia. These lesions usually present within the first week of life, when the ductus closes. As discussed above, these children typically have signs of hypoperfusion and shock.

Lesions primarily characterized by left-to-right shunting will cause symptoms as the normal pulmonary vascular resistance falls over the first month of life, with increased shunting from the systemic to the pulmonary circulation. If the resulting shunt is large enough, this may lead to CHF in the first several weeks of life (typically 4-8 weeks of age). Lesions such as VSDs or atrioventricular canal defects are in this category. Compared to the infants with ductal-dependent lesions discussed above, these children will present with a more gradual onset of symptoms, which may also be vague. The chief complaint is often poor feeding or weight loss. They can also present with shortness of breath or irritability. Commonly, they are initially thought to have a respiratory problem such as bronchiolitis or pneumonia.

A chest x-ray may be helpful toward confirming the diagnosis of CHF, showing an enlarged heart and pulmonary edema. Electrocardiogram may reveal ventricular hypertrophy. Definitive diagnosis is usually achieved via echocardiography. These patients with CHF will often require diuresis. Furosemide (0.5 mg/kg orally for patients with mild symptoms; 0.5-1 mg/kg intravenously [IV] for patients with more severe symptoms) should be administered. Pressor therapy as well

Table 2 Presentation of CHF in infancy.

Age	More common lesions presenting with CHF
Newborn	Aortic stenosis, severe pulmonary or tricuspid insufficiency, TOF with absent pulmonary valve syndrome
First week of life	Critical aortic stenosis, hypoplastic left heart, TAPVR, truncus arteriosus
2-8 wk	Acyanotic TOF, atrioventricular canal, coarctation of the aorta, endocardial cushion defect, PDA, VSD
2-6 mo	ALCAPA, PDA, VSD

as angiotensin converting enzyme inhibitors may be indicated. If necessary, arrangements should be made to transfer these patients to a facility with a pediatric cardiologist for further diagnostic evaluation and management.

Arrhythmias

Primary arrhythmias without a previous diagnosis of CHD in infancy are relatively rare, but do sometimes present to the ED. Paroxysmal supraventricular tachycardia (SVT) is the most common symptomatic dysrhythmia in infants and children. Other syndromes that may present with dysrhythmias in infancy include the prolonged QT syndrome.

Supraventricular tachycardia

Recent estimates suggest that SVT occurs in 1 of 250 to 1 of 1000 children [13]. About half of pediatric patients with SVT initially present in infancy [14]. Another spike in incidence occurs in adolescence. In infants, the most common cause of SVT is idiopathic (approximately 50%), most likely secondary to a concealed accessory atrioventricular pathway. Approximately 25% have associated conditions such as infection, fever, or drug exposure, 23% have previously diagnosed CHD, and 22% have Wolff-Parkinson-White (WPW) syndrome. In older children and adolescents, atrioventricular node reentry tachycardia becomes more prevalent [15].

In previously healthy infants, the presentation of SVT is almost always nonspecific and may not obviously suggest a cardiac etiology [14]. Common complaints include fever, fussiness, not feeding well, and pallor. Alternatively, an infant who has been in SVT for a prolonged period or who has other medical problems may present with CHF or shock and require resuscitation.

In newborns and infants with SVT, the heart rate is almost always more than 220 beats per minute. However, in this age group, sinus tachycardia also frequently reaches this rate, and it may sometimes be hard to distinguish between the two in a sick infant. The lack of discernible P waves may help to distinguish between sinus tachycardia and SVT but often is difficult to determine. The lack of beat-to-beat variability is frequently more helpful. (Patients with sinus tachycardia have discernible P waves and beat-to-beat variability.) Widened QRS is rare in pediatric SVT, and such patients should be considered to have ventricular tachycardia unless there is evidence to suggest otherwise.

Regarding treatment, if the child is unstable and IV access is not immediately available, cardioversion (the *Pediatric Advanced Life Support* textbook recommends a dose of 0.5-1 J/kg) should be attempted. If an IV catheter is in place or immediately insertable, adenosine may be used (see below).

If the patient is stable, vagal maneuvers may be attempted. In infants, the only practical way to do this

is by inducing a diving reflex with an ice bag to the face. Reported complications of this maneuver include profound vagal response, retinal detachment if pressure is placed on the eye, and fat necrosis from cold injury to the face [16]. Vagal maneuvers are frequently ineffective in infants and young children.

If the child remains stable and the vagal maneuvers fail, IV access should be obtained. Once access is obtained, adenosine (initial dose = 0.1 mg/kg; maximum = 6 mg) as a rapid IV push followed by a rapid saline flush should be given. If the initial dose is unsuccessful, an increased dose of 0.2 mg/kg (maximum dose = 12 mg) can be tried. Various sources list a range of maximum adenosine doses of 0.2 to 0.4 mg/kg. A multicenter study demonstrated that adenosine successfully terminated SVT in 71 of 98 episodes of presumed SVT in children presenting to 7 pediatric EDs [17].

If adenosine is unsuccessful at terminating the tachycardia, other antiarrhythmics may be added, although none has the rapid action of adenosine. These medications are best used in consultation with a pediatric cardiologist. Of course, if the patient is unstable, DC cardioversion should be considered. Few controlled trials have evaluated the efficacy of individual antiarrhythmic agents in pediatric patients. Most of the information about antiarrhythmic agents has been extrapolated from studies of adults. Digoxin is generally not used because it is ineffective in the prophylaxis of SVT. Moreover, digoxin is not recommended in patients with WPW syndrome because it can precipitate ventricular fibrillation. Because WPW cannot be diagnosed during the tachyarrhythmia, digoxin is discouraged in patients presenting with SVT for the first time. Options include procainamide (10-15 mg/kg over 30-45 min) and amiodarone (5 mg/kg over 20-60 min). Verapamil has been reported to cause cardiovascular collapse in infants and should not be used in this age group. Relative young age and ventricular dysfunction at presentation have been associated with the need for additional medications (refractory SVT).

Many infants who have an episode of SVT do not have a recurrence [18,19]. Roughly 65% of infants with SVT no longer have episodes after their first birthday. However, because they frequently do not present until they are significantly symptomatic, prophylactic therapy often is initiated. Propanolol, amiodarone, and other antiarrhythmic agents have been used safely for long-term treatment of SVT in infants. Less than one third require medication beyond 1 year of age [20]. For older children with WPW or recurring SVT, catheter ablation procedures may eliminate the site of abnormal conduction.

Other Dysrhythmias

Other dysrhythmias are rare in children without previously diagnosed cardiac disease. Congenital prolonged QT syndrome (LQTS) is a disorder of delayed ventricular repolarization characterized by prolongation of the QT

interval. Patients with congenital LQTS are predisposed to ventricular tachycardia, including torsade de pointes. They commonly present between the ages of 9 and 15 years with recurrent episodes of syncope. However, LQTS may also present in infancy, usually as cardiopulmonary arrest that is originally diagnosed as sudden infant death syndrome [21].

Myocarditis

Myocarditis is an inflammatory condition of the myocardium. The most common etiology of myocarditis in infants is viral. Enteroviruses, including coxsackie A and B, and adenoviruses are the most common causes in the US. Although it is the viral infection that triggers acute myocarditis, it is believed to be the patient's own immune response that leads to the myocardial damage [22,23].

The presentation is often nonspecific. There frequently will be a history of a preceding or concurrent viral illness such as an upper respiratory tract infection or gastroenteritis. Infants may initially present with very nonspecific symptoms or signs that are attributed to their viral syndrome. They are often initially misdiagnosed as having bronchiolitis, dehydration, or sepsis. Infants may present with lethargy, irritability, poor feeding, or pallor. In such cases, the underlying heart disease can be missed. They also may present with symptoms more suggestive of heart failure such as diaphoresis or tachypnea. Dysrhythmias are not uncommon in severe cases. Very young infants usually present more suddenly and severely ill. There are case reports of neonatal myocarditis presenting as an acute life-threatening event or neonatal collapse, and reports of enteroviral myocarditis originally misdiagnosed as sudden infant death syndrome [24-28].

A high index of suspicion is needed to differentiate the infant with viral myocarditis from those with a simple viral syndrome, especially during peak viral seasons. Early recognition and treatment may be lifesaving [22]. A chest x-ray may show cardiomegaly, and an EKG may show sinus tachycardia, dysrhythmias, or low QRS voltages. Electrocardiogram patterns mimicking MI have also been reported. An echocardiogram may show an enlarged left ventricle or ventricular wall dysfunction with normal coronary arteries (as distinguished from those with ALCAPA). Initially, it may be difficult to distinguish whether heart failure results from myocarditis, dilated cardiomyopathy, or ischemia secondary to ALCAPA. Cardiac enzymes may be elevated in these patients and may help to distinguish myocarditis from dilated cardiomyopathy [29]. The gold standard for diagnosis is endomyocardial biopsy, but a presumptive diagnosis can often be made from the above studies. Biopsy is associated with some risk, and its benefits must be considered in each case. It is important to remember that the definitive diagnosis will rarely be made in the ED, and patients should be treated based on their presentation.

Initial management should focus on both respiratory and circulatory status. Endotracheal intubation is indicated for those in cardiogenic shock. Inotropic support and afterload reduction, if tolerated, should be initiated. On occasion, PGE₁ may be considered in neonates with left ventricular dysfunction and relatively preserved right ventricular function. Right-to-left flow through the ductus may provide systemic circulation, similar to patients with hypoplastic left heart syndrome. Extracorporeal membrane oxygenation may be considered in those who fail to respond to inotropic support.

High-dose intravenous γ globulin may help improve ventricular function but has not been shown to improve survival in adult patients. Although the benefits of intravenous γ globulin in pediatric myocarditis remain unproven, it continues to be used in some institutions. Immunosuppressive therapy is also of questionable efficacy in patients with viral-induced myocarditis. In general, these therapies should not be initiated in the ED setting without cardiology consultation. After initial stabilization, these patients should be managed in an institution with pediatric critical care facilities and pediatric cardiology specialists. These patients can deteriorate rapidly and may need intensive management during transport; therefore, the most experienced and capable transport team available should be used. The availability of resources such as extracorporeal membrane oxygenation and heart transplantation at the referral facility should also be considered when making transport decisions. Ultimately, some of these patients will require heart transplantation for long-term survival [30].

Summary

Infants presenting to the ED with previously undiagnosed cardiac disease may do so with nonspecific complaints. A thorough physical exam and appropriate testing will generally lead to the correct diagnosis and treatment. Infants with CHD may present with cyanosis or shock due to outflow obstruction, MI, or with CHF. Chest radiograph and EKG may be helpful, but definitive diagnosis is usually made by echocardiogram. Emergency department treatment should be symptomatic and supportive. In infants in whom a "ductal-dependent" lesion is suspected, PGE₁ should be initiated.

The presentation of SVT in infants is usually nonspecific. The heart rate is almost always more than 220 beats per minute but must be differentiated from sinus tachycardia in this age group. If the patient is stable, vagal maneuvers may be attempted. If the child is unstable and IV access is not immediately available, cardioversion should be attempted. If an IV catheter is available, adenosine may be used.

Viral myocarditis in infants typically presents with nonspecific findings. These infants are often initially misdiagnosed as having bronchiolitis, dehydration, or

sepsis. A chest x-ray may show cardiomegaly, and an EKG may show sinus tachycardia, dysrhythmia, or low QRS voltages. An echocardiogram may show an enlarged left ventricle or ventricular wall dysfunction. Initial ED management is supportive.

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