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Emergency Care for Infants and Children with Acute Cardiac Disease

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Children occasionally present to the emergency department with life-threatening congenital or acquired cardiac disease. Presenting symptoms may be nonspecific, and accurate assessment and timely interventions are required to achieve optimal outcomes in this heterogeneous and complex patient population. In this article, we review 4 common scenarios: neonates presenting with ductal-dependent congenital heart disease, infants with tetralogy of Fallot who develop hypercyanotic episodes, children with decompensated congestive heart failure, and those with cardiac tamponade. In each instance, presenting signs and symptoms are discussed, and practical suggestions are offered for the initial diagnostic approach and management.
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Congenital heart disease (CHD) is the most common birth defect, afflicting 32000 newborns (8 per 1000 live births) each year in the United States. Although less common, acquired heart disease may develop at any time throughout childhood and present with life-threatening sequelae. There are dozens of categories of congenital heart defects, and substantial anatomic and pathophysiologic variations exist within each category. Furthermore, the cardiovascular physiology present after surgical or transcatheter intervention for congenital heart defects may be unique, and interventional strategies continue to evolve. The complexity inherent to this patient population is challenging even for the most seasoned practitioners. This article is intended to provide a practical overview of the more common cardiac disorders with which children present to the emergency department in critical condition. The initial diagnostic and therapeutic approach to these patients will be discussed. The topic of acute arrhythmia management in children is beyond the scope of this article, and the reader is referred to recent guidelines from the American Heart Association [1].

Ductal-Dependent CHD

Despite the common use of screening prenatal ultrasound, most patients with CHD are diagnosed postnatally. Neonates with unrecognized ductal-dependent CHD present with cyanosis or shock, or a combination thereof, during the first days of life as the ductus arteriosus closes. Such patients may initially seem healthy and be discharged from the newborn nursery, only to present soon thereafter to the emergency department in extremis. A high index of suspicion for CHD is required because such patients are often misdiagnosed as having more common conditions such as sepsis or pulmonary disease. Ductal-dependent CHD may manifest with nonspecific symptoms including tachypnea, poor feeding, pallor, and/or decreased wakefulness. Findings on physical examination that suggest CHD include abnormal cardiac impulses on palpation of the precordium, pathological heart murmurs, and/or a discrepancy between upper

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and lower extremity pulses or systolic blood pressure (Table 1). In the neonate, rapid shallow respirations suggest heart disease, whereas labored respirations suggest respiratory disease. For specific congenital heart defects, the classic findings present on electrocardiogram (ECG) and chest radiograph (CXR) are summarized in Table 1, but it is important to recognize that variability exists for these test results because of the influence of patient age as well as their ability to reflect the underlying cardiac anatomy. An echocardiogram will establish a definitive diagnosis of CHD and clarify cardiac structural and functional details. Given the extensive anatomic variations for all congenital heart lesions that may have important implications for initial stabilization and timing of interventions, prompt consultation with a pediatric cardiologist is required for neonates with suspected CHD.

Evaluation of the Neonate with Cyanosis

In neonates with CHD, severe cyanosis is present when pulmonary blood flow is inadequate, when mixing is inadequate in the setting of parallel systemic and pulmonary circulations, or when pulmonary venous

Table 1 Classic findings for the most common congenital heart defects.

Lesion	Presentation	Physical Examination	ECG	CXR
RVOTO				
Pulmonary stenosis	Critical PS: cyanosis Mild-mod PS: asymptomatic	SEM, click	RVH	Critical PS: ↓ PBF as ductus closes Mild-mod PS: ± CM
Tetralogy of Fallot with PS	Murmur, cyanosis	SEM	RVH	± CM; ± ↓ PBF with worsening PS; Older infants: "boot-shaped" heart; 25% with right aortic arch
Tricuspid atresia	Variable, cyanosis if restrictive VSD and PS	SEM	LVH	± CM; ± ↓ PBF with worsening PS
LVOTO				
Aortic stenosis	Critical AS: shock Mild-mod AS: asymptomatic	SEM, click	Critical AS: RVH Mild-mod AS: ±LVH	Critical AS: PE Mild-mod AS: ±CM
Coarctation of aorta	Critical CoA: shock Older child: hypertension	↓ Femoral pulses; >10 mm Hg arm-leg BP gradient	Critical CoA: RVH Older child: ±LVH	Critical CoA: PE Older child: ±CM; rib notching
HLHS	Shock, mild cyanosis	Poor perfusion	RVH	↑ PBF as PVR falls
Left-to-right shunts				
Atrial septal defect	Murmur	Fixed split S2	Mild RVH	±CM; mild ↑ PBF
VSD	Murmur; large VSD: CHF as PVR falls	Pansystolic murmur	LVH if ≥ mod. VSD; RVH if large VSD	CM; ↑ PBF as PVR falls
Complete atrioventricular canal	Murmur; CHF as PVR falls	Pansystolic murmur	Left axis deviation	CM; ↑ PBF as PVR falls
Patent ductus arteriosus	Murmur; CHF with large PDA	Continuous murmur	LVH; ±RVH	±CM; ↑ PBF if large PDA
Others				
D-TGA, intact ventricular septum	Cyanosis	Tachypnea with severe cyanosis	Normal or RVH	Normal
D-TGA, VSD	Mild cyanosis, CHF as PVR falls	Pansystolic murmur	BVH	CM; ↑ PBF as PVR falls
Unobstructed TAPVR	Mild cyanosis, CHF as PVR falls	Soft systolic ejection murmur	RVH	CM; ↑ PBF as PVR falls
Obstructed TAPVR	Cyanosis at birth	No murmur	RVH	Normal heart size, pulmonary venous congestion

Note that exceptions to these findings are not uncommon given anatomic heterogenity within each lesion.
± indicates may or may not be present; ↑, increased; ↓, decreased; AS, aortic stenosis; BVH, biventricular hypertrophy; BP, blood pressure; CHF, congestive heart failure; CM, cardiomegaly; CoA, coarctation; D-TGA, D-transposition of the great arteries; HLHS, hypoplastic left heart syndrome; LVH, left ventricular hypertrophy; LVOTO, left ventricular outflow tract obstruction; PBF, pulmonary blood flow; PDA, patent ductus arteriosus; PE, pulmonary edema; PS, pulmonary stenosis; PVR, pulmonary vascular resistance; RVH, right ventricular hypertrophy; RVOTO, right ventricular outflow tract obstruction; SEM, systolic ejection murmur; TAPVR, total anomalous pulmonary venous return; VSD, ventricular septal defect.

egress is obstructed. Neonates with severe right ventricular outflow tract obstruction or pulmonary atresia will develop significant cyanosis upon closure of the ductus arteriosus. These cyanotic neonates have a low ratio of pulmonary (Qp) to systemic (Qs) blood flow (ie, low Qp/Qs) due to right-to-left shunting through the atrial or ventricular septum. Congenital heart defects that may present with cyanosis include severe tetralogy of Fallot, critical pulmonary valve stenosis, pulmonary atresia with intact ventricular septum, severe Ebstein anomaly, and, in selected patients, tricuspid atresia. Neonates with D-transposition of the great arteries with an intact ventricular septum by definition have parallel systemic and pulmonary circulations and often develop marked cyanosis upon closure of the ductus arteriosus. Neonates with obstructed total anomalous pulmonary venous return typically will be cyanotic soon after delivery regardless of ductal patency. Although usually detected during the first 24 hours of life due to the presence of cyanosis or a heart murmur, patients with these lesions occasionally escape detection and are discharged from the normal newborn nursery.

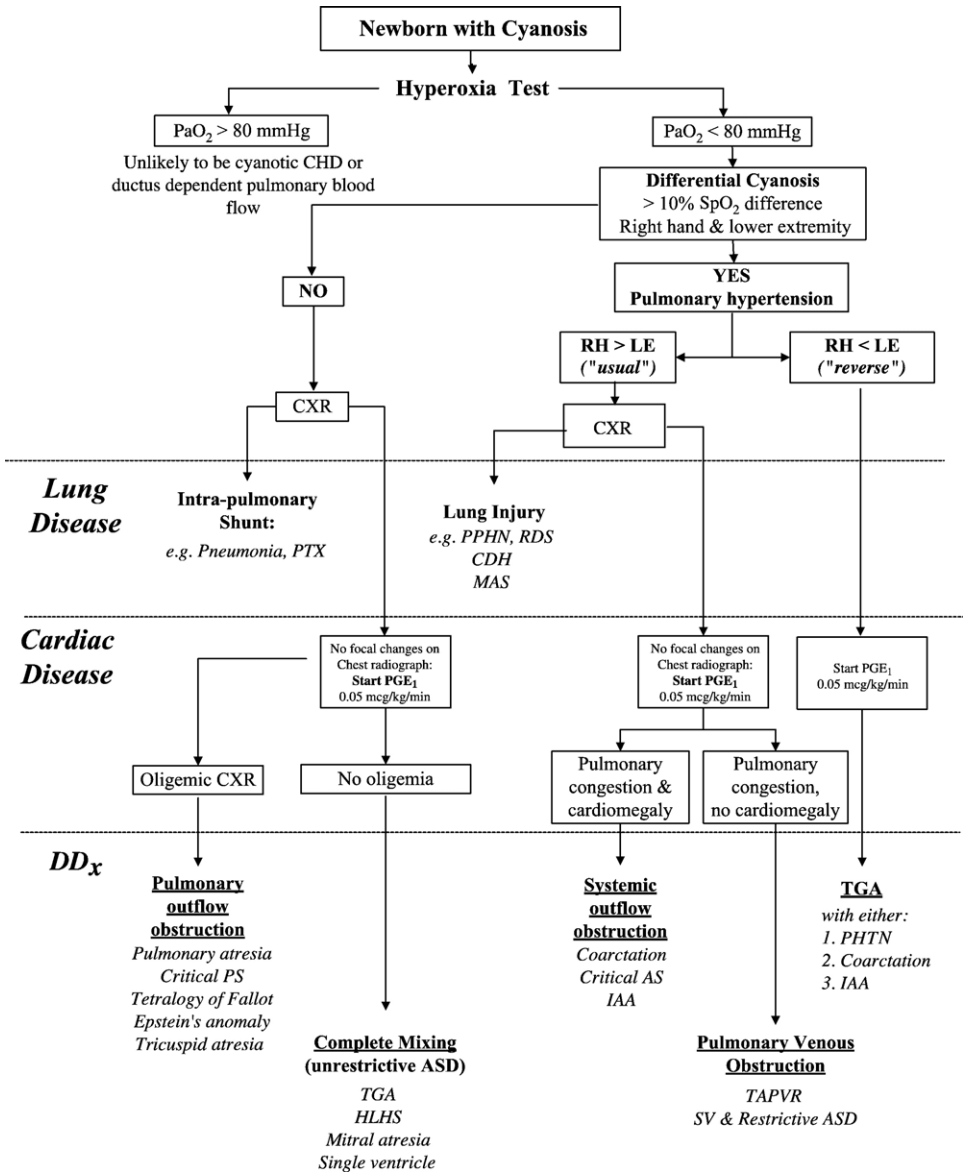

Figure 1 Algorithm for initial assessment of cyanotic newborns. AS indicates aortic stenosis; ASD, atrial septal defect; CDH, congenital diaphragmatic hernia; HLHS, hypoplastic left heart syndrome; IAA, interrupted aortic arch; LE, lower extremity; MAS, meconium aspiration syndrome; PTX, pneumothorax; PPHN, primary pulmonary hypertension; PHTN, pulmonary hypertension; PS, pulmonary stenosis; RDS, respiratory distress syndrome; RH, right hand; SV, single ventricle; TAPVR, total anomalous pulmonary venous return; TGA, D-transposition of the great arteries.

Cyanosis in neonates is almost always due to either pulmonary or cardiac disease. A brief review of the maternal, family, and gestational histories as well as a directed physical examination will allow one to formulate an initial impression that favors either cardiac or pulmonary disease. A CXR, ECG, and hyperoxia test should be obtained in all neonates with unexplained cyanosis. An algorithm similar to that used at Children's Hospital Boston to triage cyanotic newborns is shown in Figure 1. This decision tree begins with the hyperoxia test, during which an initial arterial blood gas is obtained from the right radial artery on room air. A subsequent arterial blood gas is obtained after 10 minutes of breathing 100% FIO_2. The PaO_2 is often between 25 and 40 mm Hg on room air. In general, the PaO_2 will rise on 100% FIO_2 to greater than 80 mm Hg in neonates with pulmonary disease, many of whom will also have hypercarbia ($PaCO_2$ ~45-50 mm Hg). In contrast, the PaO_2 will remain unchanged or only increase slightly in those neonates with cyanotic CHD in whom mild hypocarbia may also be present. For those neonates whose PaO_2 remains less than 80 mm Hg, the presence or absence of differential cyanosis and interpretation of the CXR will allow one to formulate a short list of the most likely diagnoses (Figure 1). The CXR should be inspected for heart size, signs of parenchymal lung disease, increased or decreased pulmonary vascular markings, and sidedness of the aortic arch.

Evaluation of the Neonate with Shock

Neonates with severe left ventricular outflow tract obstruction (LVOTO) include those with critical aortic valve stenosis, hypoplastic left heart syndrome and variants, critical coarctation of the aorta, and interrupted aortic arch. Upon closure of the ductus arteriosus, neonates with these lesions will develop shock with renal insufficiency and metabolic acidosis. A high ratio of pulmonary to systemic blood flow (ie, high Qp/Qs or large left-to-right shunt) develops if a sizable atrial or ventricular septal defect (VSD) is present. The physiologic consequences include elevated left atrial pressure, pulmonary venous congestion with reduced lung compliance, and pulmonary hypertension. Tachypnea develops due to pulmonary venous congestion, due to left-to-right shunting, and as a compensatory response to metabolic acidosis. Neonates with severe LVOTO and a patent ductus arteriosus often present with some degree of cyanosis because of intracardiac mixing, the dependence on the right ventricle (which contains desaturated systemic venous blood) to partially or fully provide systemic perfusion, and pulmonary venous desaturation resulting from pulmonary congestion. Note that classic findings of aortic arch obstruction (diminished or absent femoral pulses or an arm-leg blood pressure gradient) may not be evident when the ductus is patent and may be difficult to detect once the ductus has closed if severe metabolic acidosis and myocardial dysfunction are present. In addition to a directed history, detailed cardiovascular examination, ECG, CXR, and echocardiogram as discussed above, neonates with ductal-dependent systemic blood flow who present with shock also require an assessment of end-organ function. Basic laboratory studies to assess hepatic and renal function are needed in all cases. An abdominal radiograph to assess for necrotizing enterocolitis and a head ultrasound to screen for intracranial pathology should be considered.

Initial Stabilization of Ductal-Dependent CHD

During fetal circulation, endogenous prostaglandins and exposure to low PaO_2 contribute to the maintenance of patency of the ductus arteriosus. Reduced prostaglandin concentration, along with increased PaO_2, contribute to ductal closure after birth. Since the late 1970s, exogenous administration of prostaglandin (prostaglandin E_1 [PGE_1]) has been a reliable therapy to "reopen" or maintain patency of the ductus arteriosus, which allows for resolution of severe cyanosis and shock in neonates with ductal-dependent CHD [2,3]. A PGE_1 infusion will maintain systemic blood flow in neonates with severe LVOTO and pulmonary blood flow in those with severe right ventricular outflow tract obstruction. Prostaglandin E_1 also allows for adequate mixing of systemic and pulmonary blood flow in the setting of parallel anatomic circulation (eg, D-transposition of the great arteries). Finally, the use of PGE_1 allows adequate time for interhospital transport, detailed cardiac evaluation, treatment of noncardiac disorders, and semielective scheduling of most cardiac interventions.

A PGE_1 infusion is indicated for all neonates with known ductal-dependent pulmonary or systemic blood flow. Prostaglandin E_1 is also indicated for those markedly cyanotic newborns whose examination, hyperoxia test, and CXR suggest CHD, and those newborns with suspected LVOTO who present with shock. Prostaglandin E_1 administration should not be delayed in critically ill neonates with suspected ductal-dependent CHD while awaiting a definitive cardiac diagnosis. However, if pediatric cardiology consultation is readily available and severe cyanosis (SaO_2 <80%) and metabolic acidosis (pH < 7.3) are not present, then ideally an echocardiogram should be obtained before initiation of a PGE_1 infusion. Prostaglandin E_1 may be administered through a peripheral intravenous catheter or a central venous line. A dose of 0.05 to 0.1 μg/kg/min may be used to reopen the ductus arteriosus, and a dose of 0.01 μg/kg/min is usually sufficient to maintain ductal patency. Clinical efficacy is usually seen within a few minutes in neonates with cyanosis due to right ventricular outflow obstruction or reduced pulmonary blood flow. Although the duct usually reopens promptly in neonates with LVOTO, shock may require many hours of treatment before

improvement is evident. Apnea is the most troublesome side effect of PGE_1, which may occur less frequently with lower doses [4,5]. Endotracheal intubation may be considered for recurrent apnea, for airway maintenance during interhospital transport, or to aid in the treatment of shock. Other common side effects of PGE_1 include hypotension, fever, and harlequin rash [6]. Uncommonly, cyanosis persists after initiation of PGE_1. For example, neonates with D-transposition of the great vessels and intact ventricular septum may have a restrictive or intact atrial septum that prevents adequate intracardiac mixing of blood between the systemic and pulmonary circulations. Prolonged medical therapy is not warranted in this situation, and an emergent balloon atrial septostomy is indicated to alleviate cyanosis [7,8].

Endotracheal intubation and mechanical ventilation may aid in the resuscitation of selected neonates with ductal-dependent CHD as noted above. In addition to management of PGE_1-induced apnea, positive pressure ventilation may reduce work of breathing and systemic oxygen consumption and may decrease systemic ventricular afterload while improving ventricular wall stress characteristics resulting in improved stroke volume and cardiac output. However, care must be taken with ventilator management to avoid interventions that lower pulmonary vascular resistance in patients with hypoplastic left heart syndrome or interrupted aortic arch. Specifically, excessive supplemental oxygen may be detrimental because it will lower pulmonary vascular resistance and thus encourage available cardiac output to recirculate through the lungs at the expense of end-organ perfusion [9]. Similarly, hyperventilation may lower pulmonary vascular resistance and exacerbate pulmonary overcirculation.

Myocardial function may be compromised in critically ill neonates with ductal-dependent CHD. Volume overload from left-to-right shunting or atrioventricular valve regurgitation, pressure overload due to outflow tract obstruction, cyanosis, and metabolic acidosis all may be contributory. Sodium bicarbonate may be administered to aid in the correction of metabolic acidosis. Inotropic support may be beneficial, usually starting with dopamine at 3 to 5 μg/kg/min. Systemic vasodilation with either milrinone (which has both inotropic and vasodilator properties) or pure vasodilators may be beneficial provided that blood pressure is adequate. Central venous and/or arterial line placement may be necessary in selected cases for administration of medications, for serial blood draws, and for hemodynamic monitoring. Close monitoring of patient temperature is required to avoid the development of hypothermia, which may lead to bradycardia and increased systemic vascular resistance [10]. Hypoglycemia may occur and require treatment. Maintenance of normocalcemia is important given the dependence of the neonatal myocyte on extracellular calcium. Prompt transfer to a congenital heart center is indicated for all neonates with known ductal-dependent pulmonary or systemic blood flow and for all neonates with suspected ductal-dependent CHD if pediatric cardiology consultation and echocardiography are not readily available.

Tetralogy of Fallot and TET Spells

The primary anatomic features of tetralogy of Fallot are an anterior malignment VSD, multilevel right ventricular outflow tract obstruction, an overriding aorta, and right ventricular hypertrophy. The amount of blood that shunts right-to-left through the VSD, and thus the extent of cyanosis, varies with the severity of right ventricular outflow tract obstruction and the prevailing systemic vascular resistance. Pulmonary vascular resistance usually falls soon after birth and has minimal influence on intracardiac shunting. Neonates with tetralogy of Fallot and more severe right ventricular outflow tract obstruction or pulmonary atresia will develop excessive cyanosis upon closure of the ductus arteriosus. Such patients are stabilized with PGE_1 and referred for early surgical intervention. Infants with a minimal degree of obstruction to pulmonary blood flow ("pink TETs") are usually asymptomatic and fairly well oxygenated (SaO_2 >90%) soon after birth. These patients may be discharged from the nursery to await surgical repair at 3 to 6 months of age but are at risk for hypercyanotic episodes, or TET spells, during this period.

TET spells are potentially life-threatening events notable for significant desaturation and irritability that result from an acute decrease in pulmonary blood flow. The precise etiology of these spells has been attributed to some

Table 2 Treatment options for hypercyanotic episodes (TET spells).

Intervention	Effect on Pathophysiology
Comfort patient; minimize noxious stimuli; allow parent to hold	↓ Agitation, ↓ Heart rate
Knee-chest position	↑ SVR
Oxygen	↓ PVR
Fluid bolus	↑ CVP, ↓ Heart rate
Morphine	↓ Agitation
Ketamine	↓ Agitation, ↑ SVR
β-Blocker	↓ Infundibular spasm, ↓ Heart rate
Sodium bicarbonate	↓ Metabolic acidosis
Phenylephrine	↑ SVR
ECMO or CPB for surgical repair	Rescue therapy when above measures fail

CPB, cardiopulmonary bypass; CVP, central venous pressure; ECMO, extracorporeal membrane oxygenation; PVR, pulmonary vascular resistance; SVR, systemic vascular resistance.

combination of spasm of the infundibular conus, an imbalance between the systemic and pulmonary vascular resistances, hypovolemia, and/or tachycardia. A progressive cycle of decreased pulmonary blood flow, increased cyanosis, and worsening metabolic acidosis develops. Although TET spells can occur at any age, the incidence seems to increase after 4 to 6 months of age, which is one of several reasons that complete surgical repair is commonly recommended before this age. TET spells can be triggered by any event that provokes significant patient agitation (eg, placement of intravenous catheters, phlebotomy) or any event that decreases systemic vascular resistance (eg, infection, sedation). Infants with tetralogy of Fallot have a fixed component of obstruction to pulmonary blood flow and thus will normally develop transient desaturation with agitation and feeding due to increased oxygen consumption and cardiac output. Clinical judgment is thus required to differentiate a true TET spell from these more common and generally benign desaturation episodes. The absence of a murmur during a true TET spell, signifying a substantial reduction of blood flow across the right ventricular outflow tract, may be useful in this regard. Treatment strategies are implemented with the goal of decreasing patient agitation and heart rate, repleting intravascular volume, increasing systemic vascular resistance and pulmonary blood flow, and correcting metabolic acidosis (Table 2). Given the likelihood of recurrence, hospitalization and prompt surgical intervention are indicated once an infant has had a TET spell.

Decompensated Congestive Heart Failure

Congestive heart failure (CHF) exists when the heart is unable to provide an output sufficient to meet the metabolic demands of the body. Although CHF is often a chronic condition, acute decompensation may occur when compensatory mechanisms are overwhelmed. Common etiologies of CHF in children are listed in Table 3. Note that the diagnosis of CHF does not imply the presence of myocardial dysfunction. For example, older neonates and young infants with large shunts at the ventricular or great vessel level will develop CHF as pulmonary vascular resistance falls due to an increasing left-to-right shunt. Myocardial function is usually preserved in these patients. In contrast, infants with an anomalous left coronary artery from the pulmonary artery often develop myocardial ischemia and left ventricular dysfunction as pulmonary vascular resistance falls and perfusion pressure to the left coronary artery is compromised.

Clinical symptoms of CHF vary depending on age and underlying cardiac pathophysiology. Neonates and infants may have failure to thrive, feeding difficulties, diaphoresis, and tachypnea. Viral upper respiratory tract infections may be poorly tolerated. Infants with an anomalous left coronary artery from the pulmonary artery may have symptoms of myocardial ischemia, including irritability or crying, which is most prominent with feeding. Older children with failure of the left ventricle may have dyspnea on exertion, exercise intolerance, and/or syncope. Common signs of left-sided decompensated heart failure include rales on lung auscultation and tachycardia. In advanced cases, patients may have evidence of cardiogenic shock (poor perfusion, hypotension), severe pulmonary edema with respiratory failure, and other end-organ insufficiency.

Right-sided heart failure may develop in selected children with CHD, for example, in patients after Tetralogy of Fallot repair or in those with failing Fontan physiology. Increased systemic venous pressure may result in peripheral edema, pleural effusions, exercise intolerance, and

Table 3 Common etiologies of CHF in children.

General category	Pathophysiology	Examples
CHD	Left-to-right shunting (volume overload)	VSD, CAVC, PDA, AP window
	Valvular regurgitation (volume overload)	MR, AR
	Outflow tract obstruction of either ventricle (pressure overload)	AS, PS
	Coronary insufficiency (myocyte dysfunction/death)	ALCAPA
Cardiomyopathies (innate and acquired)	Structural or functional myocyte abnormalities	Mitochondrial disorders, defects in fatty acid metabolism, cardiomyocyte protein abnormalities, toxins, infections (eg, myocarditis), nutritional deficiencies, hypoxemia, and idiopathic processes
Incessant arrhythmias	Myocardial dysfunction, MR	SVT (multiple types) VT (less common)
High output states	Volume overload	Thyrotoxicosis, systemic arteriovenous fistula, severe anemia

ALCAPA indicates anomalous left coronary artery from pulmonary artery; AP, aortopulmonary; AR, aortic regurgitation; AS, aortic stenosis; CAVC, complete atrioventricular canal defect; MR, mitral regurgitation; PDA, patent ductus arteriosus; PS, pulmonic stenosis; SVT, supraventricular tachycardia; VSD, ventricular septal defect; VT, ventricular tachycardia.

hepatic and splanchnic congestion leading to loss of appetite and abdominal pain. Right-sided heart failure manifests with fluid retention, hepatomegaly, and ascites. Signs of both right- and left-sided heart failure are present in some patients.

The physical examination in patients with CHF includes an assessment for jugular venous distension, precordial hyperactivity, respiratory effort, hepatomegaly and ascites, peripheral edema, and abnormal quality or rate of peripheral pulses. Auscultation should focus on the detection of any murmurs, gallops or bruits, or rales on pulmonary examination. Nutritional status should also be assessed. An ECG should be obtained in all patients with decompensated CHF and inspected for abnormalities in rate, rhythm, voltages, ST segments, and T waves. The ECG must be carefully scrutinized for evidence of a reversible diagnosis that may be mistaken for idiopathic dilated cardiomyopathy. For example, patients with an anomalous left coronary artery originating from the pulmonary artery typically have characteristic findings including q waves in the anterior-lateral leads [11]. One must also exclude incessant tachyarrhythmas, such as ectopic atrial tachycardia, that are an often overlooked cause of decompensated CHF in children with structurally normal hearts and in those with underlying CHD [12,13]. A CXR often demonstrates cardiomegaly and increased pulmonary vascular markings. An echocardiogram will provide definitive diagnostic information in some cases and will quantify the severity of myocardial dysfunction, valvular regurgitation, or outflow tract obstruction. In selected cases, additional testing may be indicated, including cardiac catheterization for hemodynamics, angiography, and possibly myocardial biopsy. Additional workup may be required, including further imaging studies (eg, cardiac magnetic resonance imaging), thyroid function tests, or a metabolic evaluation.

The presentation and initial evaluation of patients with possible myocarditis is one etiology of decompensated CHF that warrants special comment. Myocarditis is defined as an inflammatory disease of the myocardium [14]. In North America, viruses usually cause myocarditis, including enteroviruses (eg, coxsackievirus, echovirus) and adenovirus. Less common causes of myocarditis include bacterial infections, toxins (eg, diphtheria), certain drugs (eg, anthracyclines, penicillin, phenytoin, tetracycline), Kawasaki disease, and autoimmune disease (eg, rheumatic fever, systemic lupus erythematous). Myocarditis typically occurs in children without a prior history of cardiac disease, although exceptions certainly exist. The spectrum of presentation for myocarditis ranges from mild myocardial dysfunction in an asymptomatic patient to fulminant myocarditis with cardiogenic shock or sudden death. Patients often report a recent or ongoing flu-like illness, and thus a high index of suspicion is required to avoid a delay in diagnosis. Symptoms may include fatigue, dyspnea, chest pain (due to concurrent pericarditis), or abdominal pain (secondary to hepatic congestion). In patients with myocarditis, the physical examination may be notable for crackles in the lungs, a gallop rhythm, hepatomegaly, and (in older patients) peripheral edema. Fulminant myocarditis manifests with poor perfusion and hypotension that has little response to volume repletion. The ECG often reveals sinus tachycardia with nonspecific ST-segment changes [15]. In fulminant cases, a diffuse low-voltage pattern (<5 mm QRS amplitude in the limb leads) may be present. Arrhythmias or conduction defects may exist, such as primary atrial or ventricular tachyarrhythmias or high grade second- or third-degree heart block. The CXR may reveal pulmonary edema and varying degrees of cardiomegaly. Note that in fulminant myocarditis, the cardiac silhouette may be relatively normal in size despite very poor ventricular function, as there has been insufficient time for the chambers to dilate. An echocardiogram is necessary to assess ventricular function and pericardial effusion. There is growing experience with the diagnostic capabilities of cardiac magnetic resonance imaging in patients with possible myocarditis [16]. Viral direct fluorescent antibody tests, serum antibody titers, and viral cultures may suggest a causative agent. Myocardial biopsy may be useful during the assessment of patients with possible myocarditis, but this procedure is not without risk in critically ill patients, especially infants and small children. Biopsy specimens are examined for severity of lymphocyte infiltrate and myocyte necrosis. The identification of viral DNA in the myocardium by PCR amplification provides strong evidence for a causative agent [17].

Children having undergone heart transplantation may also present with decompensated CHF due to rejection, advanced transplant coronary artery disease, or rarely incessant tachyarrhythmias [18]. Symptoms may be nonspecific, and gastrointestinal complaints are common.

Initial Treatment of Decompensated CHF

Children with decompensated CHF require hospitalization for diagnostic evaluation, initiation of therapy, and assessment of response to treatment. Continuous electrocardiographic monitoring is recommended for all patients during the initial period of hospitalization. Continuous blood pressure monitoring using an arterial line is warranted in patients with cardiogenic shock or in those requiring mechanical ventilation. An internal jugular vein or subclavian venous catheter may be placed such that the tip is in the superior vena cava, where serial mixed venous oxygen saturation ($MVSvO_2$) measurements may be obtained. Continuous measurement of central venous pressure (CVP) as an estimate of preload may also be useful during the initial care of patients with advanced decompensated CHF. In children, pulmonary

artery catheters are infrequently used in hospitalized patients with CHF. However, in difficult cases, a pulmonary artery catheter may provide additional data to assess severity of illness and guide patient management. Blood lactate concentration may provide useful information about the adequacy of tissue oxygenation and thus the state of systemic perfusion before and after hemodynamic interventions.

Treatment of decompensated CHF is directed at optimizing the 4 determinants of cardiac output: preload, contractility, afterload, and heart rate. Simultaneously, maneuvers may be necessary to limit or reduce systemic oxygen consumption until adequate cardiac output and oxygen delivery can be established. Such maneuvers include the use of mechanical ventilation (invasive and noninvasive), sedation and analgesia (with or without muscle relaxation), and avoidance of febrile states among others.

Preload reduction is often indicated to alleviate dyspnea and right-sided congestion and to reduce ventricular wall stress. Treatment is guided by patient symptoms, signs on physical examination, and the presence of pulmonary edema on CXR. If available, serial CVP measurements and knowledge of the patient's "dry" weight are useful to help titrate diuretic therapy. In the acute setting, the first-line diuretic is furosemide, which inhibits sodium and chloride reabsorption in the ascending limb of the loop of Henle and distal renal tubule. The initial dosing is 0.5 to 2 mg/kg, IV, Q 6 to 12 hours depending on the clinical condition and prior exposure to loop diuretics. Spironolactone is a potassium-sparing diuretic that competes with aldosterone for receptor sites in the distal renal tubule, increasing sodium chloride and water excretion while conserving potassium and hydrogen ions. In the hospitalized patient, it is most commonly used in conjunction with furosemide to prevent hypokalemia. When the response to loop diuretics is insufficient, thiazide diuretics may be added. Chlorothiazide, which inhibits sodium reabsorption in the distal renal tubule, may be prescribed at 5 to 10 mg/kg, IV, Q 12 hours. The goal of diuretic therapy is to achieve a euvolemic state, and fluid intake may need to be restricted in selected cases. The response to diuretic therapy is dependent upon the presence of an adequate renal perfusion pressure, which may be estimated by subtracting the CVP from the systemic mean arterial pressure.

Contractility may be augmented in selected children with advanced decompensated CHF by administration of intravenous inotropic agents. Those patients with myocardial systolic dysfunction as the primary cause of CHF stand to benefit the most from an acute trial of inotropic therapy, whereas those with CHF due to left-to-right shunting (eg, complete atrioventricular canal, large VSD) often have preserved myocardial function and thus may not require inotropic support. Because most inotropic drugs raise intracellular calcium levels, are proarrhythmic, and increase myocardial oxygen consumption, they should be initiated only when a clear indication exists for their use and promptly weaned once this indication no longer exists.

Catecholamines activate adrenergic receptors that increase intracellular cyclic adenosine monophosphate (cAMP) concentrations, leading to organ-specific manipulation of intracellular calcium levels and cellular function. Activation of β_1 receptors in cardiac myocytes increases cytosolic calcium levels, resulting in enhanced contractility and increased heart rate. Activation of β_2 receptors in the systemic and pulmonary vascular beds decreases cytosolic calcium levels, resulting in vascular smooth muscle relaxation and vasodilatation, whereas activation of α_1 receptors increases vascular resistance. All catecholamines have a very short half-life, allowing for easy titration. Central venous access is preferred for administration of catecholamine infusions to avoid the risk of extravasation and soft tissue necrosis. Dopamine, an endogenous catecholamine, may be useful in children with cardiogenic shock. Doses of 3 to 10 μg/kg/min stimulate both dopaminergic and β_1-adrenergic receptors leading to increased heart rate and cardiac contractility. Doses of 10 to 20 μg/kg/min primarily stimulate α-adrenergic receptors causing vasoconstriction and increased blood pressure and generally should be avoided in patients with decompensated CHF. Dobutamine, a synthetic catecholamine, increases cardiac index by stimulating β_1 receptors leading to increased intracellular concentration of cAMP. Through its effects on peripheral β_2 receptors, dobutamine may minimally reduce systemic vascular resistance, thus further contributing to increased cardiac output, but it may cause significant tachycardia in some children. Dobutamine may be used to treat decompensated CHF, with a typical starting dose of 5 μg/kg/min. Epinephrine may be useful in children with refractory cardiogenic shock. Low doses (<0.05 μg/kg/min) activate β_1 receptors, thereby augmenting contractility and heart rate, as well as β_2 receptors, which decrease vascular resistance. However, higher doses activate α_1 receptors resulting in increased systemic vascular resistance and should generally be avoided in isolated CHF.

Children who present with decompensated CHF and preserved blood pressure may benefit from intravenous vasodilators, which reduce systemic vascular resistance, arterial blood pressure, ventricular filling pressures, and myocardial wall stress. Vasodilators also alleviate pulmonary edema and dyspnea. Nitroprusside, a nitric oxide donor, may be initiated at 1 μg/kg/min and titrated to 3 to 4 μg/kg/min as needed. Cyanide and thiocyanate toxicity may occur when higher doses are used for more than 3 days. Nesiritide, a recombinant human B-type natriuretic peptide, has vasodilatory, lusitropic, and neurohumoral inhibitory properties in patients with decompensated CHF [19]. Nesiritide binds to dedicated natriuretic peptide guanylate cyclase receptors, which increases intracellular

cyclic guanosine monophosphate, resulting in smooth muscle cell relaxation and modulation of cardiomyocyte calcium levels. Preliminary reports suggest that nesiritide may be efficacious when used in infants and children with CHF [20,21]. After a loading dose of 2 mg/kg, an infusion is started at 0.01 μg/kg/min.

Milrinone, a phosphodiesterase-III inhibitor, prevents the breakdown of cAMP, which ultimately augments myocardial contractility and lusitropy while causing vascular smooth muscle vasodilatation. Milrinone may acutely benefit those children with decompensated CHF, myocardial dysfunction, and preserved blood pressure. Milrinone is devoid of the chronotropic effect of catecholamines and may be administered through a peripheral intravenous catheter. A loading dose of 50 μg /kg may be followed by an infusion of 0.25 to 0.75 μg/kg/min.

Positive pressure ventilation decreases left ventricular wall stress and afterload and reduces work of breathing and oxygen consumption. Infants with decompensated CHF due to left-to-right shunting or shock due to ductal-dependent CHD typically have better perfusion and improved hemodynamics after initiation of mechanical ventilation. Patients with primary myocardial diastolic dysfunction and pulmonary edema also rapidly improve. However, it is critically important to recognize the potential for cardiovascular collapse when patients with decompensated CHF are transitioned from spontaneous breathing to positive pressure ventilation. Patients with myocardial systolic dysfunction as the primary etiology of CHF, as seen with fulminant myocarditis, rejection of a transplanted heart, or advanced dilated cardiomyopathy, are at highest risk for cardiac arrest immediately after endotracheal intubation. In these patient populations, cardiac arrest may be precipitated by the provision of induction agents that diminish the release of endogenous catecholamines. In addition, preload may be compromised by raised intrathoracic pressure. Several management strategies may be useful to minimize the risk of peri-intubation cardiovascular collapse. Preload should be optimized, and volume (crystalloid or colloid) should be immediately available for rapid infusion. If inotropic support seems indicated, it should be started before initiation of induction agents. Although induction agents may blunt endogenous catecholamines as noted above, appropriate sedation and analgesia are nonetheless necessary to prevent further increases in heart rate and systemic vascular resistance that would otherwise occur during laryngoscopy. Careful titration of fentanyl, a narcotic with a stable hemodynamic profile, along with a small dose of benzodiazepine (eg, midazolam) is one commonly used regimen for induction. A paralytic agent will facilitate laryngoscopy and prevent laryngospasm. Despite the above precautions, some patients will still experience cardiac arrest. If a patient's condition allows, endotracheal intubation may be delayed until the patient is in an environment capable of providing rapid-response extracorporeal membrane oxygenation (ECMO) or other forms of rapidly deployable mechanical cardiac support.

Heart rate and rhythm should be optimized in patients with decompensated CHF. The loss of atrioventricular synchrony is often poorly tolerated, and arrhythmias should be treated. Anemia may require treatment with a red blood cell transfusion. Patients with hypoxemia due to pulmonary venous desaturation should receive supplemental oxygen. Fever will increase metabolic demands and should be aggressively treated. Care should also be taken to avoid hypothermia in neonates and young infants, which may increase systemic vascular resistance.

In a child with possible myocarditis, vigilance is required for the development of cardiogenic shock (which may develop rapidly) or arrhythmias. Data from animal models and adult humans do not support the use of corticosteroids in patients with myocarditis. A single retrospective study in children with myocarditis found that treatment with intravenous immunoglobulin (2 g/kg) was associated with improved intermediate term recovery of myocardial function [22]. In patients with myocarditis and refractory cardiogenic shock, high survival rates have been achieved using mechanical cardiac support (eg, ECMO) [23,24]. For this reason, children with acute myocarditis requiring inotropic support should ideally be managed in cardiac centers with ECMO capability.

The initial management of a child with a heart transplant who presents with acute CHF warrants special comment. Diagnostic considerations include rejection, advanced transplant coronary artery disease, or tachyarrhythmias. The transplanted heart is usually denervated, and thus, there is often a blunted chronotropic response given the degree of myocardial dysfunction. Atropine is usually ineffective for increasing the heart rate. In patients with acute rejection, mild symptoms may progress to cardiogenic shock and cardiovascular collapse within a short period. Thus, if rejection is suspected, initiation of pulse-dose corticosteroids should not be delayed while awaiting confirmatory testing by echocardiogram or myocardial biopsy [25].

In young infants with structural heart disease, consideration of the timing of surgical intervention should occur early in the management plan. In infants with a large VSD or complete atrioventricular canal defect, a satisfactory response to a trial of medical therapy may allow surgery to be deferred until 3 to 5 months of age. Symptomatic relief may be obtained using caloric supplementation, diuretics, and oral afterload reduction. In contrast, the risk-benefit ratio for an extended period of medical therapy for infants with D-transposition of the great vessels with VSD, truncus arteriosus, or aortopulmonary window is not favorable, and prompt repair is usually indicated. Urgent repair of an anomalous left coronary artery from the pulmonary artery is indicated at

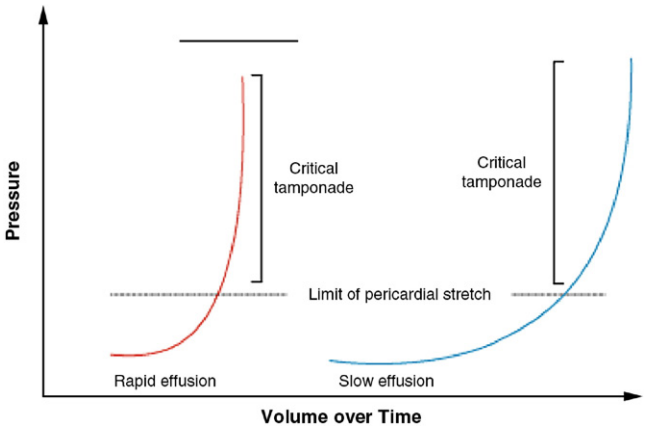

Figure 2 Cardiac tamponade. Pericardial pressure-volume curves are shown in which the volume increases slowly or rapidly over time. In the left-hand panel, rapidly increasing pericardial fluid quickly exceeds the limit of pericardial stretch, causing a steep rise in pericardial pressure. In the right-hand panel, a slower rate of pericardial filling takes longer to reach the pressure required for critical tamponade because there is more time for the pericardium to stretch and for compensatory mechanisms to become activated. Reprinted with permission from Spodick [26].

the time of diagnosis to limit further myocardial injury and optimize the potential for myocardial recovery.

Cardiac Tamponade

Cardiac tamponade may be defined as compression of the heart due to pericardial accumulation of blood, clots, air, fluid, gas, or pus [26]. Tamponade may be caused by a number of diverse diagnoses including acute pericarditis, trauma, or right atrial perforation from a central venous line. Late cardiac perforation due to erosion of percutaneously placed atrial septal defect occluding devices is a recently recognized cause of tamponade [27]. In the developed world, viral agents most commonly cause pericarditis, and concurrent myocarditis may be present. Noninfectious causes of acute pericarditis that may progress to cardiac tamponade include rheumatic fever, connective tissue disorders, Kawasaki disease, radiation, postpericardiotomy syndrome, uremic pericarditis, and malignancy.

In cardiac tamponade, diastolic filling of the heart is impaired, leading to decreased stroke volume and hypotension. Compromise in diastolic filling is exacerbated with cyclic changes in intrathoracic pressure, as occurs during spontaneous (negative pressure) breathing and mechanical (positive pressure) ventilation. Important variables of tamponade physiology include the volume of pericardial fluid, the time over which it accumulates, and available compensatory mechanisms. Although the pericardium can stretch over time, at any given moment, it is inextensible. Thus, a small volume of fluid that accumulates quickly in the pericardial space may cause cardiac tamponade and be poorly tolerated, whereas a large volume of fluid that accumulates slowly over time may cause little hemodynamic embarrassment (Figure 2).

Symptoms of cardiac tamponade are nonspecific and include dyspnea, cough, anorexia, and emesis [28]. The physical examination may be notable for distant heart tones, jugular venous distension, tachycardia, hypotension with a narrow pulse pressure, and poor peripheral perfusion. Pulsus paradoxus is defined as a decrease of 10 mm Hg or greater in systolic blood pressure with inspiration. This variation in blood pressure may be palpable in large arteries. Although pulsus paradoxus is an important diagnostic finding, it is neither sensitive nor specific for cardiac tamponade. Pulsus paradoxus is rarely present in neonates due to rapid respiratory rates and does not typically occur in patients with intracardiac shunts.

The ECG may reveal signs of pericarditis. Diffuse low voltage may be present if a large effusion or severe myocarditis is present. Electrical alternans, defined as the beat-to-beat change in QRS axis due to the heart "swinging" within the effusion, is uncommonly present. The cardiac silhouette on CXR may be normal or only mildly enlarged in children with an acute pericardial effusion. The classic radiographic appearance of a "water-bottle heart," in which the cardiac silhouette is triangular with smoothed out heart borders, is seen with a massive pericardial effusion that has developed over time. Echocardiography will quantify the pericardial effusion and detect the presence of right atrial or right ventricular collapse, and evaluate myocardial function.

A pericardiocentesis is indicated for cardiac tamponade, both for diagnostic and therapeutic purposes. Attention to the patient's cardiopulmonary status throughout the periprocedure period is important. Ideally, one clinician

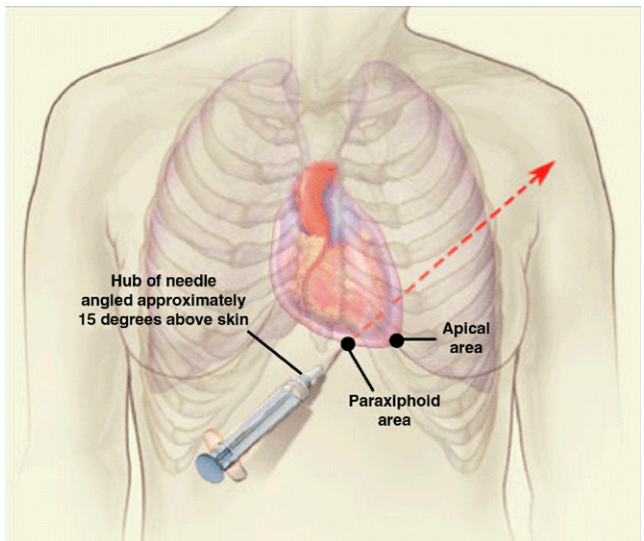

Figure 3 Most common sites of needle insertion for pericardiocentesis. Reprinted with permission from Spodick [26].

is solely responsible for patient monitoring, provision of sedation and analgesia, and management of volume administration. In hypovolemic patients, intravascular volume expansion may temporarily stabilize hemodynamics while preparations are made for pericardiocentesis. In contrast, patients with euvolemia or hypervolemia may develop worsening tamponade with intravascular volume expansion as higher filling pressures enlarge the heart that is confined by pericardial fluid. Positive pressure ventilation (invasive and noninvasive) may also decrease cardiac output, dictating extreme caution when approaching a patient with cardiac tamponade and tenuous respiratory function. Thus, spontaneous breathing, a sedative-analgesic regimen that causes minimal respiratory depression and adequate local analgesia, is desirable for patients with tamponade physiology undergoing pericardiocentesis. The procedure itself can be technically challenging and is associated with serious complications including myocardial perforation, coronary artery injury, pneumothorax, and arrhythmias. Pericardiocentesis is thus ideally performed by an experienced operator. The patient should be positioned in a 30° sitting-up position. Echocardiographic guidance is useful to identify the ideal location for insertion of a 16- or 18-gauge angiocatheter needle into the pericardial space. Using sterile technique, a paraxiphoid approach directing the needle tip toward the left shoulder is commonly used (Figure 3). For prolonged drainage, a guidewire may be inserted through the needle and used to place a pigtail catheter in the pericardial space. Specially designed pericardiocentesis catheters that have a pigtail shape with additional side holes may provide optimal safety and drainage [29].

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