Emergency Presentation Of Congenital Heart Disease In Children

It’s January, the middle of “RSV season,” when a 2-week-old infant is brought to the emergency department by his mother because he was “breathing fast all night.” His birth history is unremarkable, and his mother claims that he had been doing well until yesterday — breastfeeding, urinating, and stooling regularly. In triage, physical examination is remarkable for an irritable child with a respiratory rate (RR) of 60 breaths per minute, a heart rate (HR) of 170 beats per minute, right upper extremity blood pressure of 70/50 mmHg, axillary temperature of 99°F, and oxygen saturation as determined by pulse oximetry of 85%. Skin and mucous membranes appear dry. Extremities are cool with a capillary refill time of 4 seconds. Nasal flaring and moderate intercostal retractions are noted, and expiratory wheezing is heard bilaterally. No murmur is noted, and the liver edge is 3 cm below the right costal margin. Supplemental oxygen is administered by a facemask and SpO₂ increases to 90%. The initial concerns are RSV bronchiolitis or sepsis.

An inhaled albuterol treatment is started, a capillary blood gas is obtained, intravenous access is promptly accomplished, and he receives two 20 mL/kg fluid boluses with no improvement. In fact, vital signs have deteriorated to RR 60, HR 180, BP 62/38, SpO₂ 86%, and crackles are auscultated on repeat examination of lungs. Extremities remain cool and mottled, and liver is now 5 cm below costal margin. A chest x-ray shows a normal sized heart, prominent pulmonary vascular markings, and mild right ventricular hypertrophy. The pediatrician identifies a heart murmur.

A chest x-ray shows a normal sized heart, prominent pulmonary vascular markings, and mild right ventricular hypertrophy. The pediatrician identifies a heart murmur.

CME Objectives

Upon completing this article, you should be able to:
1. Describe the physiology of patients with congenital heart disease likely to present to the emergency department.
2. Identify and evaluate the signs and symptoms increasing the likelihood of a diagnosis of congenital heart disease.
3. Practice the fundamental principles of resuscitation and management of patients with suspected or known congenital heart disease.
4. Identify and evaluate the unique anatomical and physiological considerations in patients who have undergone palliation procedures for single ventricle lesions.

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bilateral hyperinflation. Arterial blood gas results are reported as follows: pH 7.26, pCO₂ 20 torr, and pO₂ 60 torr. At this point, one of the residents asks, “Could this patient have congenital heart disease?”

Though the field of antenatal ultrasound and echocardiography has advanced considerably in the last decade, many children with congenital heart disease (CHD) are not diagnosed before birth and do not manifest symptoms until after discharge from the newborn nursery.²⁻⁷ Frequently, when symptoms develop, they are first brought to the attention of an emergency medicine physician. These infants and children are particularly challenging patients, often appearing markedly distressed with non-specific signs and symptoms that resemble more common pediatric diagnoses such as sepsis, respiratory infection, or reactive airway disease. Unfortunately, therapy for these diagnoses may worsen the clinical course of a child with heart disease, emphasizing the need for a high index of suspicion in order to promptly identify patients with CHD.

This issue of Pediatric Emergency Medicine Practice addresses the evaluation and management of pediatric patients with CHD in the context of the evidence available from the medical literature. A comprehensive understanding of the pathophysiology as it relates to clinical manifestations is crucial for appropriate triage and treatment. Patients with known uncorrected CHD and those who have undergone palliation procedures are increasingly encountered in emergency departments for illnesses that may or may not be related to their pre-existing conditions.

Critical Appraisal Of The Literature

A literature review was performed with Ovid MEDLINE and PubMed searches for articles on CHD published from 1965-2007. Guidelines for pediatric patients with acute myocarditis have recently been published, though no such guidelines are available for patients with CHD.⁵⁻¹³ For more information on the guidelines for acute myocarditis in pediatric patients, see last month’s issue of Pediatric Emergency Medicine Practice. Many studies have been published on the epidemiology of CHD, but research on the evaluation and diagnosis of patients presenting with an acute illness is sparse. The available studies, combined with the pathophysiologic considerations involving perinatal and infantile hemodynamic changes, should provide an important framework for emergency physicians taking care of such children.

Epidemiology And Etiology

The incidence of CHD requiring specialized cardiac care is estimated to be 500-600 per 100,000 live births.¹⁴ Of these patients, approximately 170 per 100,000 live births have critical CHD, defined as those that are ductal-dependent or require surgical or interventional attention in the first month of life.¹⁵ Antenatal detection of CHD has improved over the past decade, though recent studies suggest that greater than 60% remain undiagnosed.⁵⁻⁶ If antenatal diagnosis is not made, most patients with CHD appear normal at birth. In a prospective study of 7204 newborns, senior house officers performed routine neonatal examinations after birth and suspected heart lesions in 46 patients, of which 25 were found to have CHD by a cardiologist. Another 31 infants with normal initial examinations presented with symptoms later during their first year of life and were diagnosed as having CHD.¹⁷ Clinical experience is similar to these studies. Murmurs on physical examination are often considered innocent and many children are not referred to a cardiologist. Conversely, of all infants with murmurs referred to a cardiologist, only 44% were diagnosed as having cardiac malformations.¹⁷

Several studies have investigated the use of pulse-oximetry as a screening tool for CHD in asymptomatic infants, and while abnormal values have been found to be very specific for CHD, sensitivity is poor and many cases are still missed.¹⁶⁻²² In one study, any neonate with either an abnormal pulse-oximetry value or a murmur was referred for echocardiography. This approach was 77% sensitive for detection of CHD. Therefore, even if antenatal ultrasound, neonatal examination, and pulse-oximetry are implemented for all neonates, patients with critical CHD will continue to be discharged from the nursery without a diagnosis and become symptomatic later, requiring the attention of the emergency physician.

Most cardiovascular emergencies due to CHD in previously undiagnosed children occur in the first year of life. In a systematic review of 3 large studies on infants with CHD who were not diagnosed antenatally (8651 total patients), the timing of presentation was bimodal, such that ductal-dependent defects primarily presented at less than 1 week of age and left-to-right shunting lesions presented between 1 and 6 months of age (Table 1).²³ This
bimodal distribution can be explained physiologically. The ductus arteriosus (DA) connects the pulmonary artery to the descending aorta (Figure 1). In the fetal circulation, the DA allows for pulmonary arterial blood to be shunted away from the lungs to the descending aorta, going to the placenta for oxygenation. Under normal circumstances, functional closure of the DA, which begins at the pulmonary artery and proceeds to the aorta, occurs within 12-15 hours following birth. Exposure of the lungs to higher concentrations of oxygen and decreased circulating levels of prostaglandin E₂, nitric oxide, and adenosine lead to DA constriction. In addition, the intima of the DA thickens with migrating smooth muscle and endothelial cells, further narrowing the lumen. During the ensuing 1-3 weeks, the smooth muscle becomes fibrotic, leaving the DA permanently closed as the ligamentum arteriosum. There are 2 types of ductal-dependent defects. Obstructive lesions of the right side of the heart (e.g., tricuspid atresia, pulmonary atresia) require an open DA to provide adequate pulmonary blood flow. Obstructive lesions of the left side of the heart (e.g., hypoplastic left ventricle, coarctation of the aorta) require the DA to maintain adequate systemic blood flow. As the DA begins to close, obstructive right heart lesions will develop cyanosis and obstructive left heart lesions will develop hypoperfusion, shock, and pulmonary edema. Table 1 lists the diagnoses that fall into each category. In patients with cyanotic lesions, DA constriction may be impeded as a result of the decreased arterial PO₂. In patients with left-sided obstructive lesions, pulmonary arterial pressures will be higher than normal, leading to

<table>
<thead>
<tr>
<th>Age</th>
<th>Mechanism</th>
<th>Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Newborn (less than 1 month)</td>
<td>Lesions requiring DA for pulmonary blood flow</td>
<td>Tricuspid atresia, Pulmonary atresia w/intact ventricular septum, Tetralogy of Fallot with pulmonary atresia, Tetralogy of Fallot with severe pulmonary stenosis, Double outlet right ventricle with severe pulmonary stenosis, Critical pulmonary stenosis, D-transposition of the great arteries, Ebstein’s anomaly</td>
</tr>
<tr>
<td></td>
<td>Lesions requiring DA for systemic blood flow</td>
<td>Critical coarctation of the aorta (CoA), Critical aortic stenosis, Aortic atresia, Interrupted aortic arch, Double outlet right ventricle with aortic stenosis</td>
</tr>
<tr>
<td></td>
<td>Pulmonary venous obstruction</td>
<td>Total anomalous pulmonary venous return (TAPVR)</td>
</tr>
<tr>
<td></td>
<td>Increased arteriovenous pressure difference</td>
<td>Arteriovenous fistula (e.g., hepatic, aneurysm of vein of Galen)</td>
</tr>
<tr>
<td></td>
<td>Excessive left-to-right shunting</td>
<td>Truncus arteriosus, Aortopulmonary window</td>
</tr>
<tr>
<td>Young infant (2-6 months)</td>
<td>Decreasing pulmonary vascular resistance</td>
<td>Ventricular septal defects, Atrioventricular canal defects, Large patent DA, Unobstructed TAPVR</td>
</tr>
<tr>
<td></td>
<td>Decrease pulmonary arterial pressure</td>
<td>Anomalous left coronary artery from the pulmonary artery (ALCAPA)</td>
</tr>
<tr>
<td>At any age</td>
<td>Acquired heart disease</td>
<td>Myocarditis, Idiopathic cardiomyopathy, Pericarditis, Cardiac tumor</td>
</tr>
<tr>
<td></td>
<td>Arrhythmia</td>
<td>Supraventricular tachycardia, Complete heart block</td>
</tr>
</tbody>
</table>
increased pressure and flow through the DA. Consequently, patients with both types of ductal-dependent lesions have physiologic alterations that may lead to delayed closure of the DA. This allows them to be asymptomatic for a variable period of time after birth only to present to an emergency department following discharge from the hospital. A study of 56 infants who had CHD on autopsy revealed that patients with cyanotic CHD became symptomatic early and many died prior to being discharged from the nursery, while all patients who died from left-sided obstructive lesions became distressed after discharge from the hospital. Specifically, the most likely life-threatening defects presenting after hospital discharge were interrupted aortic arch, hypoplastic left heart, and critical coarctation of the aorta. These data have led to the assertion by many experts that cardiovascular collapse in the first month of life is a left-sided obstructive lesion until proven otherwise.

The pathophysiology of large intra-cardiac left-to-right shunts is related to a gradual change in pulmonary vascular resistance (PVR) and right ventricular compliance. In the fetus, PVR is supra-systemic, and consequently, most blood ejected by the right ventricle is shunted across the DA to the descending aorta on its way to the lower body and the placenta. Following birth, with the onset of ventilation and inhalation of oxygen, PVR declines to approximately 50% of systemic vascular resistance (SVR) by 3 days of life. In patients without CHD, PVR will continue to fall as the thick medial muscle layer of the pulmonary arteries gradually involutes, approximating 15% of SVR (similar to adult values) within 6 weeks to 3 months of life. In patients with PVR-dependent lesions, such as ventricular septal defect, aortopulmonary window, and truncus arteriosus, left-to-right shunting begins when PVR falls after birth, causing increased pulmonary blood flow. This increased shear stress on the pulmonary endothelium delays the normal involution of muscle in the pulmonary vasculature, leading to a more gradual decrease in PVR. As PVR declines, left-to-right shunting increases, resulting in an increase in left ventricular end-diastolic volume (preload). In addition, right ventricular compliance increases, further promoting left-to-right shunting. Eventually, volume overload and symptoms of congestive heart failure (e.g., tachypnea, increased work of breathing, tachycardia) appear, typically leading to symptomatic presentation at 2-3 months of age.

Contemporaneous with the fall in PVR and increase in right ventricular compliance, the physiologic anemia of infancy occurs, typically reaching a nadir at 3 months of age. Moreover, a substantial amount of the hemoglobin in young infants is fetal hemoglobin, shifting the oxyhemoglobin dissociation curve to the left, decreasing oxygen delivery to the tissues. These factors, while easily compensated by increasing cardiac output in patients with normal cardiac anatomy and function, will further contribute to the development of congestive heart failure in patients with large left-to-right shunts.

Though total anomalous pulmonary venous return (TAPVR) and truncus arteriosus are classically described as cyanotic lesions, neither lesion is ductal-dependent. In TAPVR, a left-to-right shunt is present as some or all of the pulmonary veins drain into the right atrium, coronary sinus, or into a vertical vein that will traverse via a supra- or infra-cardiac pathway to drain into a vena cava (Figure 2). An obligate right-to-left shunt is present, most often at the atrial level, to allow for systemic cardiac output. More importantly, the pulmonary veins may be obstructed, either by compression from a bronchus or stenosis of the vertical vein. Severe obstruction will lead to marked pulmonary edema, pulmonary hypertension, and cyanosis immediately after birth. This is a surgical emergency. More commonly, mild-to-moderate obstruction is present, and as PVR declines and right ventricular compliance increases, more pulmonary venous blood draining into the
right atrium will enter the right ventricle, leading to increased pulmonary blood flow. As pulmonary blood flow increases, the moderate obstruction becomes manifest, causing the development of pulmonary edema and eventual right ventricular failure. Infants typically present in the first month of life, with the age of presentation being inversely related to the severity of obstruction. Late-presenting patients with TAPVR (greater than 1 month of age) have relatively low degree of obstruction.

Figure 2. Supracardiac TAPVR

In truncus arteriosus, blood from both ventricles traverses a single trun-
cal valve and then mixes at the level of the great arteries. The propor-
tion of blood entering the pulmonary versus systemic circulation depends on the relative resistances of the pulmonary and systemic vascular beds. Here, the pulmonary artery (PA) trunk is shown arising from the Aorta (Ao).

Patients with truncus arteriosus have a common truncal valve and artery that supply both the pulmonary and systemic circulation (Figure 3). Mixing occurs at the ventricular level. Since PVR is lower than systemic vascular resistance, the majority of blood flows preferentially to the pulmonary circulation. In order to maintain adequate cardiac output, left ventricular end-diastolic volume increases. Congestive heart failure ensues when the increase in left ventricular end-diastolic volume is excessive. With both TAPVR and truncus arteriosus, large amounts of pulmonary blood flow relative to systemic blood flow can maintain adequate oxygen-hemoglobin saturation, and cyanosis is often not apparent. Many asymptomatic neonates with abnormally low SpO₂ (but not low enough to cause cyanosis) detected by routine screening had one of these two lesions.¹⁹,²⁰

Though not a left-to-right shunting lesion, patients with anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) will also develop symptoms as a result of decreasing PVR. In ALCAPA, the left coronary artery is perfused with blood from the pulmonary artery. Coronary perfusion pressure to the left coronary artery is compromised when PVR and pulmonary artery pressure fall, leading to myocardial ischemia and infarction. Symptoms of congestive heart failure then develop. These patients classically present in early infancy, from 1–3 months of life, when left coronary artery perfusion becomes inadequate. Some may present later in childhood or adulthood if extensive collaterals develop between the right and left coronary arteries. The decreased PO₂ in the anomalous left coronary artery and the physiologic anemia of infancy will further decrease O₂ delivery to the myocardium.

Atrial septal defects do not typically present in infancy because the amount of left-to-right shunting is small. These patients present in late childhood, adolescence, or adulthood with exercise intolerance, fatigue, arrhythmia, or an embolic event. The diagnosis can be made during early childhood on a routine physical examination when a suspicious murmur is auscultated (i.e., systolic ejection murmur at left second intercostal space due to increased right ventricular output, along with fixed splitting of the second heart sound), or when a child is evaluated for poor weight gain or frequent upper respiratory infections.
The diagnosis of CHD should be considered in any previously healthy infant presenting with respiratory distress and/or shock. Table 1 serves as a reference tool for cardiac diagnoses based on the age of the patient at presentation and the pathophysiology of the symptoms. These guidelines, however, are not absolute. Some children with aortic stenosis, coarctation of the aorta, tetralogy of Fallot, and double outlet right ventricle with pulmonary stenosis may initially have mild obstruction and may not become symptomatic during DA closure. Instead, symptoms develop later in infancy when the obstruction progresses. On the other hand, lesions with marked pulmonary overcirculation from left-to-right shunting, such as aortopulmonary window and truncus arteriosus, will develop congestive heart failure shortly after birth, possibly during the first week of life.

Acquired heart disease such as myocarditis, cardiomyopathy, and arrhythmias — namely supraventricular tachycardia — can present similarly to a patient with CHD. Fulminant viral myocarditis, characterized by lymphocytic infiltration of the myocardium with myocyte necrosis, warrants special attention, as some studies report mortality rates of 25-35%. Coxsackievirus was originally thought to be the most common culprit, but recent data have identified adenovirus as being responsible for 55-60% of the cases. Coxsackievirus was implicated in 30-35% of the cases; parvovirus B19, influenza, herpes simplex, Epstein-Barr, and cytomegalovirus account for the remaining 5-15%. Children have a short viral prodrome followed by dramatic symptomatology such as cardiogenic shock, heart block, ventricular arrhythmias, or death.

Table 2 provides a list of non-cardiogenic diagnoses that share the presenting clinical features of a child with heart disease. While these non-cardiogenic etiologies are more common, failure to maintain a high index of suspicion for CHD or acquired heart disease in the initial work-up could lead to inappropriate therapeutic interventions and/or delay in transport to a surgical center, leading to significant morbidity or death.

### Table 2. Non-Cardiogenic Etiologies In The Differential Diagnosis Of CHD

<table>
<thead>
<tr>
<th>Respiratory</th>
<th>Bronchiolitis</th>
<th>Pneumonia</th>
<th>Spontaneous pneumothorax</th>
<th>Laryngomalacia</th>
<th>Pulmonary hemangioma</th>
<th>Cystic adenomatoid malformation</th>
<th>Reactive airways disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemodynamic</td>
<td>Sepsis</td>
<td>Anaphylaxis</td>
<td></td>
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<tr>
<td>Gastrointestinal</td>
<td>Gastroesophageal reflux</td>
<td>Tracheoesophageal fistula</td>
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<td></td>
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<tr>
<td>Other</td>
<td>Non-accidental trauma</td>
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<tr>
<td>Toxic/Metabolic</td>
<td>Methemoglobinemia</td>
<td>Toxic ingestion</td>
<td>Congenital adrenal hyperplasia</td>
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</table>

Prehospital Care

Most patients with undiagnosed CHD will not be recognized as such by paramedics or emergency medical technicians. Careful attention to the fundamentals of pediatric advanced life support (e.g., airway, breathing, and circulation) should, however, provide a certain degree of stabilization to many of these patients prior to arrival to the emergency department. Infants with severe tachypnea, cyanosis, and/or shock should receive manual bag-mask ventilation or endotracheal intubation, regardless of the etiology. Once the airway is secure, manual ventilations must not be overly aggressive, as frequent large breaths will compromise venous return and could exacerbate pulmonary hypertension in patients with undiagnosed CHD. Patients with undiagnosed CHD who present with shock may inadvertently receive overzealous intravenous fluid resuscitation. While patients with sepsis and hypovolemic shock will respond favorably to vigorous fluid resuscitation, patients with CHD, particularly left-sided obstructive lesions or left-to-right shunts, could deteriorate. Emergency medical personnel must be aware of this possibility and consider discontinuing fluid boluses if respiratory status and cyanosis worsen with no improvement in perfusion or hemodynamics.

If a neonate less than 1 month of age with circulatory failure is en-route to the emergency department, CHD with ductal-dependent systemic blood flow (Table 1) should be suspected and prostaglandin E1 (PGE1) should be ordered from the hospital pharmacy during transport to be administered immediately upon arrival to the emergency department. Also, since oxygen is a strong stimulus for DA constriction, emergency medical personnel should be instructed to minimize supplemental inspired oxygen by accepting pulse oximetry readings between 80 and 90%.24,26
For patients with known CHD, whether corrected, palliated, or uncorrected, signs or symptoms of respiratory or cardiovascular compromise should prompt expeditious transfer to a tertiary care facility with pediatric cardiovascular surgeons. Airway, breathing, and circulation must be assessed prior to transfer, and similar precautions with ventilation and fluid administration should be followed.

**ED Evaluation**

A careful history and physical examination of an infant with tachypnea, cyanosis, or shock may reveal subtle clues that should raise suspicion for CHD. While the sudden onset of shock in a neonate less than 1 month of age is ductal-dependent CHD until proven otherwise, these patients more commonly present with less obvious symptoms. In one study of 120 infants with left-sided obstructive disease, 81 presented with tachypnea and a history of poor feeding. Further questioning of parents of these children, as well as children with non-ductal-dependent heart disease (Table 1), will likely reveal a history of feeding difficulties (e.g., small quantities ingested at each feed, prolonged feeding times, diaphoresis during feeding) and poor weight gain since birth, in contrast to patients with acquired heart disease, SVT, or other non-cardiogenic diseases (Table 2). This association of CHD and failure to thrive has been appreciated for decades. In 1962, a large survey of 890 patients with CHD was published showing approximately half to be below the 16th percentile for height and weight and 27% to be below the 3rd percentile for height and weight. Inadequate caloric intake, malabsorption, delayed gastric emptying, and increased metabolism all play a role in growth failure. A more recent review of 89 patients with severe CHD ages 1–45 months found 65% to be below the 5th percentile for weight and 41% to be below the 5th percentile for both weight and height. Subgroup analysis revealed that patients with cyanotic CHD will more often appear stunted (decreased weight and height) while patients with acyanotic CHD will more often appear wasted (decreased weight but with a normal height). Moreover, patients in both categories with pulmonary hypertension will demonstrate a greater degree of malnutrition. A detailed feeding history, as well as weight and length measurements, should be obtained whenever an infant presents to the emergency department with suspected CHD.

The initial vital signs for a patient with CHD in the emergency department will commonly reveal tachycardia, hypotension, tachypnea, and the absence of fever, all of which can also be found in many non-cardiac illnesses (Table 2). These findings are generally not sensitive or specific indicators of CHD. Historical, physical, and laboratory findings were compared in patients with left-sided obstructive cardiac disease to patients with sepsis or meningitis in order to differentiate infants with undiagnosed CHD from a non-cardiogenic etiology. It was noted that patients with infection more often had fever and irritability while patients with left-sided obstructive cardiac disease had higher respiratory rates. More specifically, 1 patient out of 47 with CHD had a documented temperature greater than 38.3°C (100.4°F) while fever was present in 20 of 38 patients with infection. Therefore, the absence of fever in an infant with tachypnea and/or shock should raise suspicions for CHD, though it should not preclude an investigation for infectious etiologies.

A comparison of right upper extremity blood pressure to lower extremity blood pressure is a readily available and cost-effective tool that aids in the diagnosis of left-sided obstructive CHD. Coarctation of the aorta is the most common left-sided obstructive lesion presenting in infancy. The site of the coarcta-
tion in infants presenting in distress is most often pre-
ductal, situated between the left subclavian artery and the DA. If the DA is widely patent, blood pres-
sure in the upper and lower extremities will be rela-
tively similar. As DA closure begins, the upper extremity blood pressure increases, lower extremity blood pressure decreases, and femoral pulses become diminished or absent. In a study of 74 patients less than 6 months of age with coarctation of the aorta, all patients (n = 58) greater than 4 days of age had either upper extremity systolic hypertension or diminished femoral pulses on presentation. More importantly, these findings are often present before the onset of severe tachypnea and/or shock. While studies demonstrate a considerable degree of variability between upper and lower extremity blood pressures in normal individuals, a systolic blood pressure gradient of greater than 8 mmHg is often deemed significant, especially in the setting of other signs and symptoms such as diminished femoral pulses or tachypnea. Emergency physicians should obtain blood pressures using an appropriately-sized cuff (i.e., bladder length greater than or equal to 80% and cuff width greater than or equal to 40% of limb circum-
ference) from the right arm and either leg in all
infants presenting in distress, and they should carefully compare brachial and femoral pulses.33

After assessment of vital signs, the SpO2 in the right upper extremity and one lower extremity should also be obtained in all patients with suspected CHD. Cyanosis may not be apparent despite abnormally low arterial oxygen saturation. Central cyanosis is visible when approximately 5 g/dL of reduced hemoglobin is present in capillary blood; this correlates to about 2.5-3.5 g/dL of reduced hemoglobin in arterial blood.34 If a neonate presents with a serum hemoglobin of 12 g/dL, the SpO2 would have to drop below 80% before cyanosis would be visible. Consequently, one should not be reassured if a baby who presents in distress is “pink” in color until the SpO2 is measured.

Patients with ductal-dependent pulmonary blood flow (Table 1) classically develop oxygen desaturation as the DA closes, and they may be otherwise asymptomatic for some time prior to closure of the DA. While patients with primary pulmonary disease or myocarditis will typically develop marked tachypnea and increased work of breathing prior to oxygen desaturation, patients with ductal-dependent CHD can have oxygen desaturation out of proportion to the degree of tachypnea. As metabolic acidosis develops from persistent hypoxemia, work of breathing worsens. Patients with ductal-dependent systemic blood flow may also have oxygen desaturation before any other symptoms appear. This finding occurs as some of the desaturated systemic venous blood ejected by the right ventricle is shunted across the DA from the pulmonary artery into the aorta to maintain systemic perfusion. In patients with hypoplastic left heart or aortic stenosis, blood shunted through the DA flows both retrograde though the aortic arch and antegrade through the descending aorta, leading to oxygen desaturation in both the upper and lower extremities. Patients with coarctation of the aorta and interrupted aortic arch, where the obstruction is most often distal to the left subclavian artery, have desaturated blood flow only to the lower extremities. In a study of newborns with known CHD, of the 13 with left-sided obstructive heart disease, 9 had normal SpO2 in the upper extremity with an abnormally low SpO2 in the lower extremity, and 3 had abnormal SpO2 measurements in both extremities.35 In another study examining patients with both ductal-dependent pulmonary and systemic blood flow, 65 of 66 neonates had either SpO2 less than 95% in both upper and lower extremities or a difference of greater than or equal to 4% between the two.36 Therefore, SpO2 measurements in the right upper extremity and one lower extremity are crucial in any neonate presenting with respiratory distress or shock.

Patients with unobstructed TAPVR and truncus arteriosus more commonly present with symptoms of congestive heart failure rather than cyanosis. Since mixing is predominantly left-to-right shunting, SpO2 is only mildly decreased, and cyanosis may not be noticeable. Large ventricular septal defects, atrioventricular canal defects, aortopulmonary windows, ALCAPA, myocarditis, and cardiomyopathy also present with congestive heart failure. The physical examination findings of congestive heart failure in children are different from those classically described in adults. Along with the signs and symptoms of left heart failure such as tachypnea and pulmonary edema on chest radiograph (CXR), children will also develop signs of right heart failure such as facial edema and hepatomegaly.37 Ross et al compared 22 patients with CHD and congestive heart failure to 19 patients with CHD without congestive heart failure.38 Diagnoses in the patients with congestive heart failure included left-to-right shunt lesions, cardiomyopathy, and ALCAPA. The most sensitive and specific findings for moderate-to-severe congestive heart failure were determined to be a history of less than 3 ounces of formula per feed or greater than 40 minutes per feeding, respiratory rate greater than 60 breaths per minute, and liver edge greater than 2.5 cm below the right costal margin. Patients with sepsis or primary pulmonary disease will also be tachypneic, but a history of poor feeding for several days to weeks preceding presentation and the presence of marked hepatomegaly are not commonly observed.

During the initial physical examination, a thorough cardiac evaluation must be performed. The cardiac examination may appear normal in some patients with CHD. Confounding the task, many patients without CHD who present to the emergency department in distress may have an innocent murmur or a flow murmur related to anemia and/or fever. No data exist comparing murmurs in distressed patients with CHD to distressed patients without CHD. One study examined 222 asymptomatic patients who were referred to cardiology clinic for evaluation of a murmur. From the 73 (33%) diagnosed with CHD, 6 cardinal signs were identified that were independent predictors of the presence of

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**Table 1**

Classical Signs of Congestive Heart Failure

<table>
<thead>
<tr>
<th>Sign</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tachypnea</td>
<td>Rate greater than 60 breaths per minute</td>
</tr>
<tr>
<td>Palpitations</td>
<td>Greater than 40 minutes per feeding</td>
</tr>
<tr>
<td>Hepatomegaly</td>
<td>Liver edge greater than 2.5 cm below the right costal margin</td>
</tr>
<tr>
<td>Facial Edema</td>
<td>Face becomes swollen</td>
</tr>
<tr>
<td>Clubbing</td>
<td>Fingernails develop into a club shape</td>
</tr>
<tr>
<td>Cyanosis</td>
<td>Oxygen saturation less than 90%</td>
</tr>
<tr>
<td>Congestion</td>
<td>Edema in the extremities</td>
</tr>
<tr>
<td>Right Heart Failure</td>
<td>Facial edema, hepatomegaly, tachypnea</td>
</tr>
</tbody>
</table>

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These 6 cardinal signs are: pansystolic murmur, murmur intensity greater than or equal to grade 3, point of maximal intensity at the upper left sternal border, harsh quality of the murmur, early mid-systolic click, and abnormal second heart sound. If a patient is critically ill, many of these findings are present or, in certain situations, intensified. Although any murmur should raise suspicion for CHD in an infant who presents with tachypnea and/or shock, the presence of 1 of these 6 cardinal signs should further heighten the suspicion for CHD and lead to an immediate pediatric cardiology consultation.

The key aspects of the history and physical examination are summarized in Table 3. If none of these findings are detected, CHD still cannot be definitively excluded and further diagnostic studies are warranted.

### Diagnostic Studies

The diagnostic evaluation of any infant with tachypnea, cyanosis, and/or shock should include an arterial blood gas, CXR, and electrocardiogram (ECG). Other laboratory studies such as a complete blood count, serum electrolytes, hepatic transaminases and function studies, arterial lactate, and coagulation tests are important indicators of end-organ function in any patient with severe respiratory distress and/or shock. An elevated white blood cell count in and of itself should not steer the physician away from a diagnosis of CHD. Arterial blood gas analysis in many children with CHD will show metabolic acidosis from hypoperfusion, often with concomitant respiratory alkalosis from hyperventilation, but again, similar derangements can be seen in sepsis or bronchiolitis. If the patient is breathing a high concentration of oxygen, the arterial blood gas could provide more helpful information. In 1970, Lees theorized that the change in PaO₂ in an arterial blood gas obtained after administering 100% oxygen to a cyanotic infant with CHD will be less than predicted due to the presence of intracardiac mixing and right-to-left shunting. Despite this assertion, failure to increase PaO₂ is not diagnostic for CHD, as many patients with severe primary lung disease may have a similar result. However, if an infant with cyanosis is endotracheally intubated and provided both 100% oxygen and positive end-expiratory pressure with minimal change in PaO₂, cyanotic CHD is more likely than primary pulmonary disease. It should be remembered that oxygen is a potent stimulus for DA constriction, so 100% oxygen must be used with caution in infants with suspected ductal-dependant CHD. If an infant begins to deteriorate hemodynamically and/or if SpO₂ decreases, further diagnostic studies and possible intervention are needed.

### Table 3. History And Physical Examination Findings That Should Raise Suspiion For CHD In An Infant Or Child With Tachypnea, Cyanosis, And/Or Shock

<table>
<thead>
<tr>
<th>History of poor feeding for several days to weeks preceding illness</th>
<th>Less than 3 ounces or greater than 40 minutes per feed or poor weight gain</th>
</tr>
</thead>
<tbody>
<tr>
<td>Absence of fever in history or in ED</td>
<td>Temperature less than 38.3°C (100.4°F)</td>
</tr>
<tr>
<td>Upper extremity hypertension or decreased lower extremity blood pressure as compared to upper extremity</td>
<td>Should be apparent by 5 days of age</td>
</tr>
<tr>
<td>Oxygen desaturation out of proportion of what would be expected based on physical examination or differential between upper and lower extremity</td>
<td>Less than 95% in both extremities or greater than 4% difference between upper and lower extremity</td>
</tr>
<tr>
<td>Abnormal femoral pulses</td>
<td>Absent or diminished relative to the brachial pulses</td>
</tr>
<tr>
<td>Signs of congestive heart failure</td>
<td>Facial edema, hepatomegaly (especially greater than 2.5 cm below costal margin), pulmonary edema</td>
</tr>
<tr>
<td>Abnormal cardiac examination, especially 1 of the 6 cardinal signs</td>
<td>Pansystolic murmur</td>
</tr>
<tr>
<td></td>
<td>Intensity of the murmur greater than or equal to grade 3</td>
</tr>
<tr>
<td></td>
<td>Point of maximal intensity at the upper left sternal border</td>
</tr>
<tr>
<td></td>
<td>Harsh quality of murmur</td>
</tr>
<tr>
<td></td>
<td>Early mid-systolic click</td>
</tr>
<tr>
<td></td>
<td>Abnormal second heart sound</td>
</tr>
</tbody>
</table>
es after initiation of supplemental oxygen, the FiO₂ should be weaned.

Plasma B-type natriuretic peptide (BNP) and its biologically inactive counterpart amino-terminal pro-BNP (N-BNP) are produced by enzymatic processing of the pro-hormone pro-BNP, which is secreted by the cardiac ventricles in response to increased wall tension. Patients with congestive heart failure from volume overload (e.g., left-to-right shunting), pressure overload (e.g., critical aortic stenosis following DA closure), or ventricular dysfunction (e.g., myocarditis, ALCAPA) may have elevated levels of these hormones upon presentation. Two recent studies have demonstrated the utility of BNP and N-BNP in differentiating between cardiac and pulmonary disease in infants and children with respiratory distress. In one study of pediatric patients presenting with respiratory symptoms, the 23 patients with congestive heart failure from congenital or acquired heart disease had a mean BNP serum concentration of 693 ± 502 pg/mL, whereas the 26 patients with lung disease had a mean BNP serum concentration of 45.2 ± 64 pg/mL. The authors reported that a BNP level of 40 pg/mL was 84% accurate in differentiating CHF from pulmonary disease. In a similar study, the median N-BNP level in 17 infants with acute heart failure from congenital or acquired heart disease was 18,452 pg/mL, in contrast to a median N-BNP level of 311 pg/mL in 18 patients with lung disease. While further larger studies are needed to confirm these results, most experts would agree that markedly elevated serum BNP and N-BNP levels in infants and children with respiratory distress suggest heart disease, while normal BNP and N-BNP levels make a non-cardiogenic diagnosis more likely.

If congestive heart failure is apparent, a serum cardiac troponin T should be obtained. Acute viral myocarditis typically presents with elevated cardiac troponin T, likely due to infiltration of cardiac myocytes and necrosis. In a study of 43 pediatric patients admitted with primary myocardial dysfunction from either acute viral myocarditis, idiopathic chronic dilated cardiomyopathy, or moderate to large ventricular septal defect, a cardiac troponin T greater than 0.052 ng/mL could be used to make the diagnosis of acute viral myocarditis. Due to the high mortality rate in children, differentiation of acute viral myocarditis from CHD and cardiomyopathy is crucial, since prompt initiation of immunotherapy can improve outcome.

Many studies have demonstrated minimal clinical utility of chest radiographs in the evaluation of a child referred to a pediatric cardiologist with an asymptomatic murmur. However, for infants presenting in distress to the emergency department where pediatric echocardiography is not readily available, the chest radiograph may provide valuable information. The cardiothoracic index (CTI) is calculated by measuring the maximal transverse diameter of the heart in the anteroposterior view and dividing it by the maximal width of the thoracic cavity. A CTI greater than 0.6 (Figure 4) is a marker of cardiac enlargement and should be determined in all patients. Infants without heart disease rarely have evidence of cardiac enlargement, while the CTI in patients with CHD and acquired heart disease is variable. Moreover, an enlarged thymic shadow commonly seen in neonates may falsely make the heart appear enlarged on anteroposterior view of the chest. Nevertheless, if the CTI is greater than 0.6 in an infant with respiratory distress and/or shock, congenital or acquired heart disease must be suspected and a pediatric cardiology consult is warranted. If the CTI is less than 0.6, however, CHD or acquired heart disease cannot be excluded.

Pulmonary vascularity should also be assessed when reviewing the CXR. In normal patients, pulmonary vessels can be seen branching to about two thirds of the distance between the pulmonary hila and the pleural surfaces of the lungs. In patients with left-to-right shunting lesions, left-ventricular

![Figure 4. Cardiomegaly CXR](image)
outflow tract obstruction, and poor myocardial function, branching vessels may be noted farther out toward the pleura. In contrast, patients with right ventricular outflow obstruction have decreased pulmonary vascular markings. Table 4 summarizes the defects and their typical pulmonary vascularity.

Other unique radiographic features mentioned throughout the literature may be present in certain types of CHD. In some infants with tetralogy of Fallot, the main pulmonary artery segment is flat or concave on CXR and the cardiac apex is upturned due to right ventricular hypertrophy, producing the classic coeur en sabot (boot-shape) appearance (Figure 5).55 Neonates with complete transposition of the great arteries (D-TGA) have a concave main pulmonary artery segment with a narrow superior mediastinum caused by superimposition of the aorta and pulmonary arteries on anteroposterior view.55 This finding, coupled with the convex appearance of the ascending aorta and right heart border, contribute to what has historically been described as the “egg-on-a-string” appearance (Figure 6). Due to enlargement of a vertical vein and the superior vena cava, CXR of a patient with TAPVR may exhibit a “snowman” or “figure of eight” sign (Figure 7).55 An important caveat to all of these diagnostic clues is that while these findings may be helpful when they are apparent, many patients with these lesions will have a normal appearing CXR. In fact, the presence of the classic findings is the exception and not the rule.55,56

A 12-lead ECG can be normal in an infant with CHD. In many cases of CHD it is abnormal, and in a

![Figure 5. Tetralogy of Fallot CXR](image1)

Chest x-ray of a 1-month-old infant with tetralogy of Fallot. The classic coeur en sabot (boot shape) appearance can be seen, signifying the upturned cardiac apex resulting from right ventricular hypertrophy.

![Figure 6. Complete Transposition Of The Great Arteries](image2)

Chest x-ray of a 1-day-old infant with d-transposition of the great arteries. The narrow superior mediastinum caused by superimposition of the aorta and the pulmonary artery, coupled with the convex appearance of the ascending aorta and right heart border, is referred to as the “egg-on-a-string.”

<table>
<thead>
<tr>
<th>Table 4. Pulmonary Vascularity In CHD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Increased pulmonary vascularity</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
</tr>
<tr>
<td>Atrioventricular canal defect</td>
</tr>
<tr>
<td>Large DA or AP window defect</td>
</tr>
<tr>
<td>Truncus arteriosus</td>
</tr>
<tr>
<td>TAPVR</td>
</tr>
<tr>
<td>D-transposition of the great arteries</td>
</tr>
<tr>
<td>Critical coarctation of the aorta</td>
</tr>
<tr>
<td>Critical aortic stenosis</td>
</tr>
<tr>
<td>Aortic atresia</td>
</tr>
<tr>
<td>Interrupted aortic arch</td>
</tr>
<tr>
<td>Double outlet right ventricle</td>
</tr>
<tr>
<td>with aortic stenosis</td>
</tr>
<tr>
<td>Hypoplastic left heart</td>
</tr>
<tr>
<td>Cor triatriatum</td>
</tr>
<tr>
<td>ALCAPA</td>
</tr>
<tr>
<td>Acute viral myocarditis</td>
</tr>
<tr>
<td>Cardiomyopathy</td>
</tr>
<tr>
<td>Decreased pulmonary vascularity</td>
</tr>
<tr>
<td>Tricuspid atresia</td>
</tr>
<tr>
<td>Pulmonary atresia w/intact ventricular septum</td>
</tr>
<tr>
<td>Tetralogy of Fallot with pulmonary atresia</td>
</tr>
<tr>
<td>Tetralogy of Fallot with severe pulmonary stenosis</td>
</tr>
<tr>
<td>Double outlet RV with severe pulmonary stenosis</td>
</tr>
<tr>
<td>Critical pulmonary stenosis</td>
</tr>
<tr>
<td>Ebstein’s anomaly</td>
</tr>
</tbody>
</table>
few situations it may be diagnostic. Several studies that examined the ability of pediatric emergency department physicians to accurately interpret ECGs demonstrated poor concordance with pediatric cardiologists.57-59 A systematic approach may improve pediatric ECG interpretation skills. The rate should first be determined, followed by characterization of the rhythm and axis. Many infants with uncorrected CHD will present with sinus tachycardia, often related to congestive heart failure from excessive left-to-right shunting, obstruction, or poor myocardial function. Sinus rhythm is present if a P-wave precedes each QRS complex and the P-wave axis is normal, denoted by upright P-waves in leads I, II, and aVF.60 P-wave morphology should also be examined, since P-waves taller than 2 mV (2 small boxes) in infants suggests right atrial enlargement (RAE) (Figure 8), and P-wave duration greater than 0.08 sec (2 small boxes) indicates left atrial enlargement (LAE).61 RAE is common in patients with tricuspid atresia, pulmonary stenosis, TAPVR, and Ebstein’s anomaly, while bilateral atrial enlargement is common in patients with atrioventricular canal defects. Patients with atrial enlargement, especially Ebstein’s anomaly, are at risk for atrial arrhythmias.

Once sinus rhythm is confirmed, the QRS axis should be determined. Abnormalities of the QRS axis are often the only abnormalities apparent on ECG in infants with CHD. Under normal circumstances, neonates have right ventricular dominance and right QRS axis deviation. The presence of severe right QRS axis deviation (negative QRS complex in leads I and aVF) or left QRS deviation (positive QRS complex in leads I and negative QRS complex in leads aVF) is abnormal in an infant less than 1 year of age and should prompt consultation by a pediatric cardiologist (Figure 8). Left-to-right shunting lesions (LV volume overload) and tricuspid atresia (diminutive RV) will have left-axis deviation, whereas lesions with severe pulmonary stenosis can have severe right axis deviation due to right ventricular hypertrophy.61

The P-R, QRS, and QTc interval should be calculated. It is important to note that the upper limits of normal for these intervals are lower in infants as compared to adults. For example, in neonates, the P-R interval ranges from 0.08-0.15 seconds and the upper limit of normal for the QTc is 0.44 seconds, whereas in adults, the P-R interval ranges from 0.12-0.20 seconds and the upper limit of normal for the QTc is 0.49 seconds.61 These intervals are typically normal in patients with CHD, though a prolonged P-R interval is common in patients with an atrioventricular canal defect (Figure 8), and most patients with Ebstein’s anomaly have a right bundle branch block.

All leads should be scrutinized for ST-elevations, T-wave inversions, or abnormal Q-waves (i.e., width greater than 0.35 ms).62 Patients with ALCAPA will classically have abnormal Q-waves in the lateral leads (I, aVL, V4-V6) due to myocardial infarction and may have ST-depressions in the inferior leads (II, III, aVF) due to ongoing myocardial ischemia (Figure 9). Patients with myocarditis may also exhibit ischemic changes on ECG. In a study of 45 patients with acute viral myocarditis, 31 of 45 had flattened or inverted T-waves, and 10 of 45 had ST elevations.63 Myocarditis can also induce SVT, premature ventric-
ular complexes, ventricular tachycardia, ventricular fibrillation, or complete heart block.10

In summary, the arterial blood gas, laboratory analyses, CXR, and ECG may not be diagnostic in an infant with CHD, but the constellation of data, along with the history and physical examination, should lead an astute emergency department physician to consult a pediatric cardiologist and initiate the appropriate management.

Treatment

Morbidity and mortality from surgical correction of CHD have declined significantly, consequent to improved diagnostic tools, surgical techniques, and postoperative care.64 The American Heart Association reports an overall mortality risk of 4.7% after all types of cardiac surgery in the pediatric population.65 However, morbidity remains significant with the incidence of neurologic sequelae as high as 25% in children who have required cardiac surgery.66 Although this morbidity may be attributed to the effects of surgery and cardiopulmonary bypass, a growing body of evidence indicates that decreased preoperative cerebral oxygen saturation and metabolic acidosis may be additional important contributing factors.66-71 Systemic hypoperfusion, shock, and multi-organ damage negatively impact survival. Patients with early diagnosis and appropriate management have improved survival, likely due to a more stable preoperative status.4, 24-27 Thus, appropriate preoperative resuscitation and management are critical to reduce morbidity and mortality in neonatal and pediatric patients who present with CHD.1-2, 67, 76-77

Optimal management of a child with known or suspected CHD is achieved by stabilizing the airway, establishing vascular access, supporting the circulation, correcting metabolic derangements, and in patients with ductal-dependent lesions, maintaining the patency of the DA. Prostaglandin E1 infusion is critical to maintain the patency of the DA in patients with ductal-dependent lesions. In addition, sepsis and abnormalities of other organ systems (such as seizures or pneumonia) should be addressed and treated accordingly. The severity of the child’s presentation and the rapidity of the deterioration should dictate the urgency of the resuscitation. The principles discussed in the following sections have been summarized in the Clinical Pathway on page 17.

Stabilizing The Airway

The neonate or pediatric patient with respiratory distress, cyanosis, or circulatory collapse will require a stable airway to maintain adequate oxygenation and ventilation, as well as to minimize unexpected and sudden deterioration. Although patients with CHD may require emergent endotracheal intubation, several important pathophysiologic concepts must be considered in these patients. PVR, though lower than antenatal levels, will be elevated in patients with CHD as compared to normal children and adults. Perhaps more importantly, increased pulmonary blood flow leads to delayed maturation of the pulmonary vascular bed, such that the typical involution of smooth muscle in the arterioles is delayed.24 As a result, the pulmonary vascular bed is more reactive, especially to catecholamine surges, putting these patients at risk for pulmonary hypertensive episodes. In preparation for endotracheal intubation and mechanical ventilation, sedation and neuromuscular blockade are necessary to prevent adverse effects from noxious stimuli. Ketamine, which increases pulmonary arterial pressure and thus could precipitate a pulmonary hypertensive crisis, should be avoided in this situation. Other medications (such as morphine, benzodiazepines, barbiturates, and propofol) all have the potential to cause systemic vasodilation and reduce aortic diastolic pressure, thereby potentially inducing myocardial ischemia. Use of such drugs should be closely monitored. Etomidate (0.3-0.5 mg/kg/dose), a carboxylated imidazole hypnotic drug, has a more favorable hemodynamic profile.78 It has alpha2-adrenergic receptor activity, which likely blunts the hypertensive effects typically seen with the suppression of endogenous catecholamines by anesthetic agents.
Adrenal suppression has been reported with prolonged infusions of etomidate, but a single dose of etomidate is considered safe by most, but not all investigators.79-82 Fentanyl (1-2 mcg/kg/dose), which does not trigger histamine release as morphine does, can also be used.83 Neuromuscular blockade can be accomplished using rocuronium (1 mg/kg/dose), which has a quick onset of action (1-2 minutes), or vecuronium (0.1 mg/kg/dose), which is less expensive than rocuronium but with a relatively longer onset of action (2-4 minutes). After intubation, a nasogastric or orogastric tube should be inserted to remove intra-gastric air. Even small amounts of air in the abdomen may impact ventilation and perfusion. Once the airway is secure, appropriate oxygenation and ventilation must be established. This can be best achieved with a thorough understanding of the unique physiology of these patients.

Utilizing supplemental oxygen is a fundamental principle in pediatric resuscitation. When neonates present with profound cyanosis or shock, careful attention must be directed at the clinical response to supplemental oxygen as well as other therapies.84 Neonates with ductal-dependent lesions, especially those with left-sided obstructive lesions, may decompensate further when breathing air with a high FiO2. In addition to stimulating constriction of the DA, oxygen causes pulmonary vasodilatation, which may worsen pulmonary edema and respiratory distress.24 Neonates with cyanosis, tachypnea, and/or shock should be given supplemental oxygen, but when the response to this therapy is counter to what is anticipated, CHD should be suspected and supplemental inspired oxygen should be minimized by accepting pulse oximetry readings between 80 and 90%.26

Patients with tachypnea and congestive heart failure due to left-to-right shunting (Table 1) may be hypoxic due to pulmonary edema or concomitant respiratory illness. Supplemental oxygen will be beneficial for these children. If patients are not hypoxic, dilatation of the pulmonary vasculature by supplemental oxygen may increase left-to-right shunting and worsen congestive symptoms. Ultimately, emergency physicians should have a low threshold to intubate these patients and provide positive pressure ventilation. Increased work of breathing due to pulmonary edema with poor lung compliance generates greater negative intrathoracic pressure. This translates to increased transmural pressures (increased LV afterload) as well as increased systemic venous return (RV preload) in an already overloaded heart. Infants with left-to-right shunting lesions have excess pulmonary blood flow and would benefit from strategies to curtail shunting. Positive pressure ventilation increases PVR (RV afterload) and reduces LV afterload, minimizing intracardiac shunting and improving systemic blood flow. Positive pressure ventilation also decreases systemic venous return (RV preload), which may benefit a patient with excessive pulmonary blood flow. Other ventilatory strategies to reduce pulmonary blood flow include mild hyperventilation and minimizing supplemental oxygen.85,86

Positive pressure ventilation is not without undesirable consequences. Impairment of systemic venous return can be potentially harmful, decreasing cardiac output. Systemic vasodilatation from anesthetic agents as well as hypovolemia can further aggravate the compromised systemic venous return. In small infants, an exaggerated vagal response from endotracheal intubation and mechanical ventilation will further debilitate cardiovascular function. Anticipatory management is useful to avoid the adverse hemodynamic consequences associated with intubation and mechanical ventilation. Volume resuscitation, careful selection of anesthetic agents, positive inotropes, and anticholinergic agents such as atropine (0.01 mg/kg/dose IV, minimum 0.1 mg/dose) are useful adjuncts.

### Establishing Vascular Access

A child presenting with shock, cyanosis, and/or tachypnea will require immediate vascular access. Central venous access for central venous pressure monitoring, infusion of resuscitation fluids, and inotropes may also be required. When available, the ideal central venous access in the neonate is an umbilical vein catheter. The distal tip of umbilical venous lines should be above the diaphragm at the 9th or 10th thoracic vertebrae. A low-lying umbilical venous line placed approximately 2 cm into the umbilical vein is an alternative to the supra-diaphragmatic position; it is treated as a peripheral line and is useful in emergency situations.

Radiographic evidence of line positioning is imperative, since the incidence of misplaced catheters is greater than 20%.87 Complications such as blood loss, air embolism, vessel perforation, and infection can occur with either the high or low umbilical catheters. The umbilical venous catheter can also be used to monitor response to therapy by following trends in mixed venous O2 saturation. Interestingly,
infrcardiac TAPVR has been diagnosed by detecting unusually high mixed venous O₂ saturation through blood samples from an umbilical venous catheter.88

Emergency departments may not be equipped with the necessary umbilical catheters, may not have a clinician with the expertise to place an umbilical catheter, or depending on the age at the time of presentation, may not be able to access the umbilical vein. An intraosseous (IO) needle is an acceptable alternative to umbilical catheters and peripheral intravenous lines. IO lines provide reliable and expedient administration of fluids and medications in the setting of shock.89-92 Emergency drugs and fluids can be administered intraosseously at the same rates and dosages as when given intravenously; the IO route is comparable to peripheral venous rather than central venous access. A more important benefit may be the location of IO placement during resuscitation. Chest compressions may preclude the physician from placing an umbilical or other central catheter, whereas an IO site in the tibia would be more feasible.95 Complications include extravasation of fluid and periosteal infiltration potentially accompanied by compartment syndrome, osteomyelitis, localized cellulitis, abscess formation, skin necrosis, tibial fractures, and fat and/or bone marrow microemboli.89,92 The risk of complications is low, but as soon as intravenous access is established, the intraosseous needle should be removed.

When umbilical venous catheterization is unsuccessful or not feasible, central venous catheterization can be obtained using the femoral, subclavian, or internal jugular vein approach. In a 5-year retrospective study of central venous catheter placement in a pediatric emergency department, the femoral vein (83%) was the most common insertion site, but the subclavian (10%) and internal jugular veins (6%) were accessed as well. The complication rate was low and there were no life- or limb-threatening sequelae.93

Maintaining The Patency Of The Ductus Arteriosus

PGE₁ has been shown to be effective at reopening and temporarily maintaining patency of the DA in neonates with ductal-dependent lesions.94-97 There have also been case reports of PGE₁ increasing the caliber of the coarctation site and improving hemodynamic status in infants with coarctation of the aorta.98 In patients with ductal-dependent lesions, PGE₁ infusion is a life-saving therapy.

The decision to initiate PGE₁ should be based on clinical findings and a high index of suspicion for the diagnosis of ductal-dependent lesions. Awaiting definitive diagnosis via echocardiography by a pediatric cardiologist could waste valuable time, during which the patient may further decompensate. Danford et al published a study of the application of information theory to determine clinically relevant variables in the decision to start a PGE₁ infusion. Their analysis revealed that outcome is improved by early PGE₁ treatment in any cyanotic newborn with a murmur or poor pulses regardless of how ill they appear and in any critically ill newborn who has either cyanosis or poor pulses.99

Since PGE₁ is metabolized and extracted rapidly in the lungs, it must be administered as a continuous infusion. There are multiple methods of initiating a PGE₁ infusion.100 Some clinicians prefer to start a higher dose (0.1-0.2 mcg/kg/min) to ensure reopening of the DA. The dose is then titrated downward every 15-20 minutes to minimize the adverse side effects. Dosages as low as 0.005–0.01 mcg/kg/min have been reported to be as effective as traditionally higher doses of 0.02–0.1 mcg/kg/min.101 Several studies have reported dose-related risk of injurious side effects; however, it is controversial whether complications are significantly diminished with a reduction in the infusion rates.101,102,103 Alternatively, a lower dose may be started and titrated upward every 15-20 minutes until the DA is reopened. A typical starting infusion rate is 0.05–0.1 mcg/kg/min, but doses as high as 0.4 mcg/kg/min may be required to achieve the desired clinical response. Neonates with cyanotic ductal-dependent lesions responded within 30 minutes while acyanotic neonates with left heart obstruction required up to 3 hours (range: 15 minutes – 11 hours) for maximal effect.97 The PGE₁ infusion rate may be titrated based on clinical improvement in perfusion, arterial blood pH, arterial blood pressure, pulse oximetry, and urine output. DA closure will occur within 1-2 hours after discontinuing the PGE₁ infusion.94

Although there are no absolute contraindications to initiating a PGE₁ infusion, not all neonates with a cyanotic cardiac lesion will respond favorably. In neonates with obstructed TAPVR, re-opening the DA may increase pulmonary blood flow and thereby exacerbate pulmonary venous congestion. In a neonate with D-TGA, PGE₁ may achieve increased mixing across the DA, but a restrictive atrial and/or ventricular septal defect may make the admixture of
blood across the DA inadequate to achieve sufficient oxygen delivery to the end organs. An emergent atrial septostomy is the necessary life-saving measure in such patients.

The rate of complications associated with PGE$_1$ infusions is reported to be approximately 50%.$^{95, 101, 103}$ More common side effects are minor, including feeding intolerance, fever, tachypnea, thrombocytopenia and jitteriness. However, life-threatening side effects such as necrotizing enterocolitis, respiratory depression, hypotension, bradycardia, convulsions, and intraventricular hemorrhage have been reported as well.$^{94, 95, 101, 103}$ Many of the side effects of PGE$_1$ imitate sepsis, especially when the constellation of symptoms includes fever, hypotension, and respiratory depression. Confounding the suspicion for sepsis, a small retrospective study reported that PGE$_1$ was temporally associated with leukocytosis.$^{104}$ Because sepsis must always be strongly suspected in any ill-appearing neonate presenting to the emergency department in distress, a complete diagnostic evaluation for infection should be performed and antibiotic therapy should be initiated concomitant with the evaluation for CHD. Additionally, fluid resuscitation with an isotonic solution of 10–30 mL/kg should be administered if significant hypotension is encountered during the initiation of PGE$_1$.

Emergency department physicians must be aware that after the DA is reopened, the pulmonary and systemic circulations are in competition and need to be balanced as best as possible while the patient is awaiting transfer to a pediatric intensive care unit.$^{106}$ Excessive blood flow to the lungs results in pulmonary edema and may lead to inadequate systemic tissue perfusion, organ dysfunction, and shock. Specifically, the coronary arteries, which are dependent on diastolic blood flow for perfusion, are compromised when blood flow through the DA is preferentially diverted to the lungs during diastole. In contrast, decreased pulmonary blood flow will lead to unacceptable degree of cyanosis or compromised systemic perfusion.

In single ventricle lesions, pulmonary and systemic circulations are dependent on pressure generated by the same chamber. In such situations, systemic arterial and mixed venous oxygen saturations are reliable measures of the flow between the two circulations.$^{105}$ The goal of systemic arterial oxygen saturation of 80-90% mentioned previously is based on Fick’s Principle, where total consumption of oxygen is equal to the product of the blood flow and the arterial-venous oxygen concentration difference.$^{104}$ Further manipulation of this principle results in the simplified Qp:Qs ratio, which compares blood flow through the pulmonary vasculature (Qp) to blood flow through the systemic circulation (Qs) (Equation 1). The ideal ratio is present when the circulations are perfectly balanced, similar to an anatomically normal heart, where the pulmonary and systemic circulations are in series. In the emergency department, Qp:Qs can only be measured crudely based on several assumptions. First, in single ventricle lesions, pulmonary arterial oxygen saturation (SpaO$_2$) should be equivalent to systemic arterial oxygen saturation (SaO$_2$). This assumption can be made based on the fact that all venous return (systemic and pulmonary) to the heart mixes at the atrial level then enters the single ventricle where it is distributed to both the aorta and pulmonary artery with the assistance of the DA. Secondly, pulmonary venous oxygen saturation (SpvO$_2$) can be assumed to be 100% in the absence of significant lung disease. Lastly, while superior vena cava (SVC) oxygen saturation (which represents mixed venous blood prior to any intracardiac mixing) is the accepted method of measuring SvO$_2$, this value is not typically available to emergency room physicians.$^{107}$ Therefore, one must assume that the systemic O$_2$ extraction is constant such that SvO$_2$ varies with the initial aortic oxygen saturation – i.e., the difference between aortic and mixed venous oxygen saturation remains 25%. Using these assumptions, Qp:Qs can be estimated (Table 5). It becomes evident that as the aortic oxygen saturation increases, the

<table>
<thead>
<tr>
<th>O$_2$ saturation</th>
<th>SaO$_2$ - SvO$_2$</th>
<th>SpvO$_2$ - SpaO$_2$</th>
<th>Qp/Qs</th>
<th>Qp:Qs</th>
</tr>
</thead>
<tbody>
<tr>
<td>75%</td>
<td>75 - 50</td>
<td>100 - 75</td>
<td>25/25</td>
<td>1</td>
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<tr>
<td>85%</td>
<td>85 - 60</td>
<td>100 - 85</td>
<td>25/15</td>
<td>1.7</td>
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<tr>
<td>95%</td>
<td>95 - 70</td>
<td>100 - 95</td>
<td>25/5</td>
<td>5</td>
</tr>
</tbody>
</table>

### Equation 1. Simplified Qp:Qs Ratio

$$\frac{Qp}{Qs} = \frac{(SaO_2 - SvO_2)}{(SpvO_2 - SpaO_2)}$$

- Qp: pulmonary blood flow
- Qs: systemic blood flow
- SaO$_2$: aortic saturation
- SvO$_2$: mixed venous saturation
- SpvO$_2$: pulmonary venous saturation
- SpaO$_2$: pulmonary artery saturation

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**Pediatric Emergency Medicine Practice**

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Clinical Pathway: Management Of An Infant < 1 Month Old With Tachypnea, Cyanosis, Or Shock

Cyanosis or poor perfusion

History & Physical
Secure airway
Establish vascular access
Start PGE1 (0.05 mcg/kg/min)
Maintain SpO2 80-90%
Order chest x-ray & ECG
Blood, urine, CSF cultures
IV antibiotics
Pediatric cardiology consultation

YES

NO

Tachypnea

History & Physical
Vitals signs (including BP and SpO2 measurements in upper right and one lower extremity)
Assess airway & breathing; consider endotracheal intubation if work of breathing is excessive
Chest x-ray
ECG

Fever ( > 38.3°C) (100.4°F)

Blood, urine, CSF cultures
IV antibiotics
Cardiac disease not ruled out.

YES

Decreased lower extremity BP as compared to upper extremity and/or Δ SpO2 of > 3% between upper and lower extremity

Aortic coarctation likely; order PGE1 infusion and obtain pediatric cardiology consultation.

NO

Signs of CHF - facial edema, hepatomegaly (< 2.5 cm below costal margin)

Start dobutamine or dopamine infusion at 5 mcg/kg/min

If improvement in heart rate and perfusion, continue fluid resuscitation.
If no improvement, start dobutamine or dopamine at 5 mcg/kg/min.

YES

NO

Give 10 cc/kg 0.9% saline bolus

Abnormal cardiac examination
pan-systolic murmur
intensity > grade 3
PMI at upper left sternal border
harsh quality
early mid-systolic click
abnormal second heart sound

Cardiomegaly on CXR or abnormal ECG (left axis deviation, RA enlargement, S-T segment changes, abnormal Q-waves)

Pediatric cardiology consultation; myocarditis possible. Send blood for troponin T and prepare for possibility of rapid deterioration.

NO

NO

Pediatric cardiology consultation; myocarditis possible. Send blood for troponin T and prepare for possibility of rapid deterioration.

This clinical pathway is intended to supplement, rather than substitute for, professional judgment and may be changed depending upon a patient’s individual needs. Failure to comply with this pathway does not represent a breach of the standard of care.

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Qp:Qs increases disproportionately. Aortic oxygen saturation of 95% represents a pulmonary flow 5 times the volume of systemic flow. **Figures 10 and 11** illustrate this point graphically.

In children with single ventricle lesions, optimal end organ function and improved survival rates correlate with Qp:Qs between 1 and 2, which corresponds to an arterial oxygen saturation equal to 75-88%. Maintenance of aortic oxygen saturation within this range can be achieved with manipulation of the PVR and SVR. In cyanotic patients, supplemental inspired oxygen and alkalosis will decrease the PVR, promoting pulmonary blood flow. Lung volume can have a profound effect on PVR. Low lung volumes due to atelectasis and high lung volumes due to hyperinflation will increase PVR and must be avoided. This important physiology underscores the need for careful endotracheal intubation in these patients. Right mainstem intubation or overzealous manual positive pressure breaths, while potentially detrimental to any patient, can have devastating consequences in patients with single ventricle physiology. Patients with single ventricle lesions and aortic oxygen saturation greater than 90% likely have low PVR, high pulmonary blood flow, and compromised systemic perfusion. Decreasing supplemental inspired oxygen and mild hypoventilation (pH=7.35) can increase the PVR, directing blood flow to the systemic circulation. Fio2 may have to be decreased to 21%. The risk of pneumothorax and ventilator-induced lung injury precludes the use of hyperinflation to increase PVR. Adequate volume resuscitation and pharmacotherapy, most notably the phosphodiesterase III inhibitor milrinone (0.5 mcg/kg/min) or the vasodilator nitroprusside (0.5-4 mcg/kg/min) via continuous infusion, can decrease SVR and promote systemic blood flow. Lastly, pain and agitation can cause dramatic changes in PVR and SVR and complicate mechanical ventilation. A continuous infusion of fentanyl at 1-3 mcg/kg/min can be useful, providing pain control and a sedative effect without significant hemodynamic derangement.

**Supporting The Circulation**

The ultimate goal during management of an infant or child with tachypnea, cyanosis, and shock is to decrease metabolic demands while increasing oxygen delivery and perfusion. A target mean arterial blood pressure of 40 mmHg to optimize perfusion is a general guideline in a neonate; graphs of age, height, and gender-dependent blood pressures are available to direct therapy in the pediatric population. Patients with normal blood pressure, however, can still be in shock, since systemic arterial pressure is often not reflective of tissue blood flow. Physical examination findings (capillary refill time, quality of the distal pulses, warmth of the extremities, urine output, and mental status) and laboratory markers of tissue perfusion (lactate and base deficit) are useful adjuncts to direct therapy during the shock state.

If shock persists in patients with ductal-dependent lesions after the initiation of PGE1, further fluid resuscitation and/or inotropic support must be considered. If pulmonary edema and hepatomegaly are present, further increase in preload will likely not be helpful and could be detrimental. Furthermore, if the clinical status is worsening during volume adminis-
tration, the fluid bolus should be discontinued and inotropic support should be initiated. In some pediatric centers, bedside limited echocardiography by the emergency physician (BLEEP) has been used to estimate left ventricular function and the inferior vena cava volume, which is an indirect measure of preload. A study of 31 critically-ill children (age range: 23 days to 16 years) demonstrated good correlation between measurements of these two parameters performed by emergency physicians as compared to pediatric echocardiographers. Normal IVC size, extrapolated from adult data, has been reported to be 9.2 ± 2.4 mm/m², and it collapses more than 50% during quiet inspiration. Patients with congestive heart failure may have poor LV function (e.g., myocarditis, cardiomyopathy, ALCAPA), and the IVC will likely be enlarged with little respiratory variation. If this information can be obtained quickly in the emergency department, excessive fluid administration can be avoided while inotropic support is appropriately initiated and titrated.

At low doses (3-5 mcg/kg/min), dopamine is a good beta₁-agonist that increases heart rate, contractility, and blood pressure. It also stimulates beta₂-adrenergic receptors that produce vasodilatation, as well as alpha₁-adrenergic receptors that conversely produce vasoconstriction, with likely no net effect. At higher doses (5-20 mcg/kg/min) there is significantly more beta₁ as well as alpha₁ stimulation and, consequently, tachycardia, increased myocardial oxygen consumption, and ischemic vasoconstriction. At doses higher than 5 mcg/kg/min, dopamine should be preferably infused through a central venous catheter to prevent local tissue damage in the event of extravasation from a peripheral intravenous catheter.

If the patient is still unstable despite a dopamine infusion of 10 mcg/kg/min, an epinephrine infusion should be initiated. At low doses of 0.01-0.05 mcg/kg/min, epinephrine stimulates beta₁-adrenergic receptors and increases heart rate, contractility, and blood pressure. At doses of 0.05-0.1 mcg/kg/min, peripheral beta₂-adrenergic receptors are also activated; this decreases SVR (hence, diastolic blood pressure) and further increases heart rate, cardiac output, and systolic blood pressure. Infusion rates between 0.1 and 1 mcg/kg/min stimulate alpha₁-receptors with resulting constriction of renal and cutaneous arterioles. However, this is balanced by the net increase in cardiac output and stimulation of beta₂-receptors; end organ perfusion is likely to increase. Doses higher than 1 mcg/kg/min further activate alpha₁-receptors and markedly increase afterload, consequently decreasing cardiac output and end organ perfusion. Potential adverse effects include tachycardia, hypertension, myocardial ischemia, dysrhythmias, hypokalemia (due to beta₂-stimulation), and severe vasospasm. Because local infiltration can cause severe tissue damage, epinephrine should be infused through a central venous catheter.

Infants with congestive heart failure from known or suspected left-to-right shunting lesions presenting with a stable blood pressure may benefit from diuretics to mitigate the congestion and improve myocardial function by generating a more favorable end-diastolic pressure. Conversely, if hypotension and acidosis are present, the emergency physician must determine if hypovolemia or myocardial failure is responsible. The presence of marked hepatomegaly and edema will suggest the latter. If hypovolemia is suspected, careful fluid resuscitation is recommended. Prudent clinicians will conservatively manage these patients with multiple 10 mL/kg aliquots and start inotropic therapy with dopamine to prevent fluid overload. Frequent clinical assessments after each fluid bolus are necessary in all such patients to judge the therapeutic response.

Metabolic derangements should also be corrected to optimize tissue perfusion and maximize the benefits of inotropic support. Metabolic acidosis occurs secondary to poor tissue perfusion rather than hypoxemia. Severe tissue hypoxia from inadequate oxygen delivery leads to significant anaerobic metabolism with production of lactic acid. Tissue hypoxia results in metabolic acidosis, which increases PVR and reduces myocardial contractility, leading to a decrease in cardiac output. Cautious administration of sodium bicarbonate (1–2 mEq/kg) may ameliorate metabolic acidosis and can be used as a temporizing measure while attempts are made to restore adequate perfusion. While data suggest that sodium bicarbonate has been shown to reduce afterload and increase myocardial contractility in neonates, deleterious side effects have been reported. It is associated with intracerebral hemorrhage when administered rapidly and undiluted. Furthermore, sodium bicarbonate is metabolized to CO₂ and water, so if adequate pulmonary blood flow and ventilation are not present, the neonate may not be able to eliminate CO₂ effectively; thus, acidosis will be worsened.

Other important therapeutic considerations include prevention of ionized hypocalcemia and hypoglycemia, maintenance of normothermia, and
minimization of unnecessary oxygen requirements with sedation and neuromuscular blockade. Catecholamines act via calcium-dependent mechanisms and thus, calcium replacement will benefit the overall cardiac output. When ionized calcium levels are less than 1.10, patients can be treated with calcium gluconate at 100 mg/kg/dose or calcium chloride at 10 mEq/kg/dose. The increased clinical stress from the cardiovascular abnormalities (i.e., shock and/or respiratory distress) can rapidly deplete the infant’s glycogen stores. Hypothermia and hypothermia both place excess metabolic demands on a fragile myocardium, so strict temperature control to achieve normothermia should be attempted.

**Special Circumstances**

**Tetralogy Of Fallot**

Classic tetralogy of Fallot consists of a malaligned ventricular septal defect (VSD) with an overriding aorta, contributing to right ventricular outflow tract (infundibulum) obstruction (RVOT) and consequent right ventricular hypertrophy. Depending on the location and degree of the obstruction, patients may have baseline cyanosis or normal saturation with “spells” of cyanosis due to infundibular spasm (Figures 12 and 13). Many infants with severe pulmonary stenosis or pulmonary atresia will develop cyanosis after the DA has closed, necessitating the surgical placement of a shunt from the pulmonary artery to the systemic circulation. These patients characteristically have baseline systemic O₂ saturations in the range of 75-85% prior to definitive surgical correction. Some other patients will not develop cyanosis even after DA closure. These “pink tets” can be monitored as outpatients for a few months prior to surgery. During this time, they are at risk for developing “hypercyanotic tet spells,” usually occurring during agitation and crying. Hypercyanotic spells are due to excessive production of endogenous catecholamines which causes infundibular spasm, decreasing blood flow through shock. The presence of a differential in either or both is an important diagnostic clue for the presence of CHD.

- The most sensitive and specific findings for moderate- to-severe congestive heart failure in infants with CHD have been determined to be: a history of less than 3 ounces of formula per feed or greater than 40 minutes per feeding, respiratory rate greater than 60 breaths per minute, and liver edge greater than 2.5 cm below costal margin.
- Many patients with CHD will have increased pulmonary reactivity and/or pulmonary hypertension. Efforts should be made to limit stimuli of pulmonary vasoconstriction, such as agitation, hypoxemia, acidosis, and pulmonary hyperinflation. Careful attention must be paid to the clinical response to therapeutic maneuvers in patients with CHD. If a ductal-dependent CHD is suspected or a patient is presenting after stage I palliation of a single ventricle lesion, supplemental oxygen should be titrated to achieve SpO₂ of 80-85%. If hypovolemia is suspected, careful fluid resuscitation is recommended. Prudent clinicians will conservatively manage with multiple 10 mL/kg aliquots and start inotropic therapy with dopamine to prevent fluid overload.
- Because sepsis is strongly suspected in any neonate presenting to the emergency department in distress, a complete diagnostic evaluation for infection should be performed, and antibiotic therapy should be initiated concomitant with the evaluation for CHD.
- Patients with known uncorrected CHD, palliated single ventricle lesions, and cardiac transplants pose unique challenges in the emergency department setting. The emergency physician must be familiar with these issues in order to provide optimal treatment.

**Key Points**

- Despite recent advances in antenatal ultrasound, the majority of patients with CHD are diagnosed after delivery.
- Functional closure of the DA may be delayed in children with CHD. As a result, patients with ductal-dependent lesions will become symptomatic after discharge from the neonatal nursery.
- The timing of presentation of CHD is bimodal. Patients with ductal-dependent defects will present in the first month of life, while patients with predominantly left-to-right shunting lesions will present between 2-6 months of age.
- A ductal-dependent CHD should be considered in any neonate less than 1 month of age who presents with sudden cardiovascular collapse. These lesions have the highest incidence of mortality prior to surgical correction.
- The decision to initiate PGE₁ should be based on clinical findings and a high index of suspicion for the diagnosis of ductal-dependent lesions. Awaiting definitive diagnosis via echocardiography by a pediatric cardiologist could take valuable time, during which the patient may further decompensate.
- The diagnosis of CHD should be considered in any infant less than 1 year of age who presents with cyanosis, tachypnea, or shock. Suspicion should be enhanced by history of poor feeding and poor weight gain, absence of fever, auscultation of a murmur, hepatomegaly, cardiomegaly on chest x-ray (cardioto- racic index greater than 0.6), or an abnormal ECG, especially the presence of left-axis deviation, atrial enlargement, or ischemic changes.
- Upper and lower extremity blood pressure and SpO₂ measurements should be performed in all neonates less than 1 month of age with cyanosis, tachypnea, or
Tetralogy of Fallot consists of a VSD with an overriding aorta (Ao), pulmonary artery (PA) outflow tract obstruction, and consequent right ventricular hypertrophy. The above diagram shows dynamic subvalvar - infundibular (Inf) obstruction. Depending on the location and degree of obstruction, some patients may have baseline cyanosis, while others will have normal saturation with spells of cyanosis from infundibular spasm.

During hypercyanotic spells, there is decreased blood flow through the pulmonary outflow tract and increased right to left shunting across the VSD. This leads to severe cyanosis and tachycardia, which decreases right ventricular diastolic filling time and reduces the driving pressure for pulmonary blood flow. These patients have symptoms of hyperpnea, tachypnea, and increasing cyanosis, which occasionally progress to syncope, seizures, and death.

Treatment strategies for “tet spells” focus on the promotion of pulmonary blood flow by lowering endogenous production of catecholamines and raising RV filling pressure and SVR. Patients should be comforted by their parents and keep their knees pushed to the chest to increase SVR. Supplemental oxygen should be provided. Intravenous fluid boluses (10 mL/kg) should be given to increase right ventricular preload. If cyanosis persists, sedation should be given, which decreases circulating catecholamines, blunts the hyperpneic drive, and relaxes the dynamic infundibulum, relieving much of the outflow tract obstruction. Morphine 0.05-0.1 mg/kg/dose has historically been used as the first line agent. If the patient is still agitated and cyanotic despite morphine, endotracheal intubation should be performed and more potent sedatives should be employed. Ketamine (1 mg/kg/dose) provides sedation and increases SVR, which can further promote pulmonary blood flow through the stenotic RVOT. A phenylephrine infusion (10-20 mcg/kg bolus followed by 2-5 mcg/kg/min continuous infusion) can be administered to increase SVR and systemic blood pressure, thereby improving systemic oxygenation. Beta-blockers (esmolol 500 mcg/kg bolus followed by 50-300 mcg/kg/min continuous infusion) can be used to decrease RV outflow contractility and possibly increase SVR by blocking peripheral beta receptors. Table 6 summarizes this management.

**Table 6. Management Of "Tet Spell" – C.O.I.N.S.I.P.E.S.**

<table>
<thead>
<tr>
<th>Comfort</th>
<th>Have parents hold child, place knees to chest</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oxygen</td>
<td>Provide oxygen via nasal cannula or face mask</td>
</tr>
<tr>
<td>IV Fluids</td>
<td>Give 0.9% normal saline as 10 cc/kg boluses</td>
</tr>
<tr>
<td>Narcotics</td>
<td>Morphine 0.05-0.1 mg/kg IV</td>
</tr>
<tr>
<td>Sedation</td>
<td>Ketamine 1-2 mg/kg IV</td>
</tr>
<tr>
<td>Intubate</td>
<td>Ketamine 1 mg/kg, rocuronium 1.2 mg/kg; after intubation, continue to give morphine 0.1 mg/kg IV Q° or ketamine 1 mg/kg IV Q°</td>
</tr>
<tr>
<td>Phenytoinphrine</td>
<td>Phenylephrine 10-20 mcg/kg/dose IV, followed by continuous infusion 2-5 mcg/kg/min</td>
</tr>
<tr>
<td>Esmolol</td>
<td>Esmolol 500 mcg/kg load over 1 min followed by continuous infusion at 50-300 mcg/kg/min</td>
</tr>
<tr>
<td>Surgery</td>
<td>If still cyanotic at this point, emergent surgery is needed</td>
</tr>
</tbody>
</table>
Unrepaired hypoplastic left heart syndrome is universally lethal with 95% mortality in the first month and 100% by 4 months. Surgical palliation for hypoplastic left heart syndrome was established in 1983. Since then, mortality has significantly improved from 5-year survival rates of 30% in the mid-1980s to a current survival rate of 70-80%. As surgical techniques and post-operative care have progressed, the periods between the three palliation stages are becoming a critical time for morbidity and mortality. Special consideration must be given to the patient with known single ventricle physiology in the various stages of repair who presents to the emergency department.

The three-stage palliation begins in the first week of life with the Norwood procedure. The main pulmonary artery trunk is transected from the branch pulmonary arteries and used to reconstruct the hypoplastic aortic arch and create a neo-aorta, which converts the right ventricle into the systemic pump. To prevent pulmonary venous hypertension, the atrial septal defect (ASD) is enlarged to eliminate restriction to flow from the pulmonary veins across the ASD and into the right atrium. Finally, to provide a source of pulmonary blood flow, a shunt from the subclavian artery to the undivided branch pulmonary arteries is created. This is known as the Blalock-Taussig shunt (BTS). This parallel circulation is extremely fragile. It is dependent on preload and intolerant of increases in afterload. The interval between the Norwood procedure and the next surgical stage of repair (i.e., Glenn or hemi-Fontan procedures) remains a vulnerable period for potential catastrophic events with an inter-stage mortality of approximately 15% and as high as 30%. Multiple etiologies of mortality include myocardial ischemia from coronary artery impairment, neo-aortic obstruction, restrictive ASD, shunt stenosis or thrombosis, right heart dysfunction, arrhythmias, intolerance to dehydration and respiratory infections, and pulmonary and systemic circulation imbalance with ensuing organ failure or hypoxemia.

If the patient presents to the ED during this period with SpO2 less than 75%, it is reasonable to provide supplemental O2 and fluid resuscitation. It is important to identify the cause of the SpO2 desaturation. Pulmonary venous desaturation (pulmonary edema or infiltrate), decreased pulmonary blood flow (clotted or outgrown BT shunt, increased PVR), or mixed venous desaturation (anemia, hypovolemia, or RV failure) all lead to arterial hypoxemia. Although it is valuable to assess the BTS by echocardiogram, a CXR may provide useful clues to the etiology of the patient’s desaturation. Comparing the patient’s current CXR with the most recent CXR is extremely important. Increased pulmonary vascular markings would suggest pulmonary venous desaturation whereas decreased pulmonary vascular markings might suggest inadequate pulmonary blood flow. In either circumstance, endotracheal intubation with mechanical ventilatory support and supplemental O2 to maintain SpO2 saturation between 75 and 85% is imperative.

The next stage of palliation occurs at 4-6 months of age. The Glenn procedure initiates the separation of the pulmonary and systemic circulation, thereby decreasing the volume load to the right ventricle. The first step in separating the pulmonary and systemic circulation is to remove the BTS and directly route the SVC blood flow to the PA. The SVC is transected from the right atrium and attached directly to the undivided branch pulmonary arteries. This is known as the Blalock-Taussig shunt (BTS) (Figures 14 and 15). The RV is now pumping two circuits in parallel, pulmonary blood flow and systemic blood flow. Similar to unrepaired patients with ductal-dependent lesions on PGE1, these flows must be balanced, with an optimal systemic oxygen saturation of 75-85% reflecting a Qp:Qs of 1. This parallel circulation is extremely fragile. It is dependent on preload and intolerant of increases in afterload. The right atrium receives systemic return from the IVC, so mixing still occurs and SaO2 values are normally in the range of 80-90%. The RV remains the pump for the systemic circulation, but it no longer supports pulmonary blood flow since the BTS has been eliminated. Pulmonary blood flow is now passive and dependent on the pressure gradient between the systemic venous system and the common atrium. These patients are not at risk for pulmonary overcirculation, but they are rather sensitive to elevations in PVR and pulmonary arterial pressure. As a result, a common reason for patients with superior cavopulmonary anastomoses to present to the emergency room after discharge to home following surgery is cyanosis. Factors that decrease systemic venous pressures (namely hypovolemia) or increase PVR (such as atelectasis or pneumonia) are not well tolerated and can result in marked cyanosis.
1. “If the patient had CHD, the diagnosis would have been made in utero or prior to leaving the newborn nursery.”
Recent studies estimate that greater than 60% of CHD cases are not diagnosed on antenatal ultrasound. Most neonates with CHD are clinically asymptomatic immediately after birth. Even when abnormalities such as a murmur are detected on physical examination, many will not be directly referred to a pediatric cardiologist.

2. “The patient is greater than 1 week old, so the diagnosis cannot be a ductal-dependent CHD. The DA should have closed days ago.”
Patients with ductal-dependent cardiac disease, especially ductal-dependent systemic blood flow, may have persistence of DA patency beyond 24 hours of life as a result of increased DA blood flow. Cardiovascular collapse in a child less than 1 month of age should be considered as a CHD with ductal-dependent systemic blood flow until proven otherwise.

3. “The infant is febrile. We should evaluate and treat for sepsis. CHD is not likely.”
While sepsis is more common and should be suspected in any neonate presenting to the emergency department with tachypnea, cyanosis, or shock, a diagnosis of CHD should not be excluded based solely on the presence of fever. In fact, the presentation of a patient with CHD may be precipitated by an infectious process.

4. “The blood pressure and SpO₂ obtained in triage are normal. I’m reassured.”
Infants presenting to the emergency room in distress should have blood pressure and SpO₂ measurements performed on the right upper extremity and one lower extremity. A discrepancy between the two extremities is suggestive of CHD.

5. “No one can hear a murmur on this infant. Therefore, CHD is not likely.”
In many patients with CHD, especially those presenting with marked tachycardia, a murmur will not be present or will be difficult to auscultate. The presence of a murmur should raise suspicions for CHD, but the absence of a murmur does not exclude the diagnosis.

6. “The chest x-ray and electrocardiogram are normal. Therefore, the heart must be fine.”
Similar to a murmur, the presence of cardiomegaly or ECG changes increases the likelihood of cardiac disease, but the absence of either or both does not exclude the diagnosis, especially if the physical examination is abnormal.

7. “The infant is hypoxic – give 100% O₂ stat!”
Infants with ductal-dependent CHD or single ventricle physiology (e.g., following the Norwood procedure) may become worse in a hyperoxic environment. Oxygen stimulates DA constriction and promotes pulmonary blood flow, sometimes at the expense of systemic blood flow. In these patients, SpO₂ should be maintained between 80 and 85%.

8. “The infant’s blood pressure is low; I shouldn’t give sedation prior to endotracheal intubation. I’ll just give a muscle relaxant.”
Many patients with CHD will have increased pulmonary reactivity or pulmonary hypertension. Noxious stimuli may precipitate a crisis. Etomidate and fentanyl have a limited effect on hemodynamics and one or both should be used with muscle relaxation prior to endotracheal intubation.

9. “The baby boy has a history of CHD, but the mother says that the problem has been corrected. He must have normal anatomy now.”
Although a thorough history in patients with known CHD must be obtained in any emergency situation, a firm grasp of the anatomy and physiology is necessary for appropriate management. The patient’s primary cardiologist must be contacted promptly if historical details are lacking.

10. “PGE₁ has a lot of side effects. I will wait to start an infusion until I am sure of the diagnosis.”
The common problems associated with PGE₁ infusion can be managed with respiratory and cardiovascular support. The detrimental effects of a delay in the institution of PGE₁ in an infant with ductal-dependent CHD, however, can be devastating. If a ductal-dependent CHD is suspected, a PGE₁ (0.05 mcg/kg/min) infusion must be immediately instituted and can be life-saving.
Emergency physicians should have a low threshold for endotracheal intubation in patients with lung disease in the presence of a cavopulmonary anastomoses. Once intubated, mean airway pressures should be tailored to decrease atelectasis without hyperinflation. High FiO2 or even inhaled nitric oxide therapy may be needed to decrease PVR and improve pulmonary blood flow. Furthermore, since the majority of SVC blood derives from the cerebral circulation, increasing cerebral blood flow may enhance the pulmonary circulation and improve oxygen delivery to end organs. Cerebral blood flow is exquisitely sensitive to PaCO2. Hyperventilation causes respiratory alkalosis and subsequent cerebral vessel vasoconstriction; consequently, cerebral blood flow is decreased. Conversely, hypoventilation will cause respiratory acidosis, decrease cerebral vascular resistance, and increase cerebral blood flow. Maintaining respiratory acidosis favorably affects cerebral blood flow and pulmonary blood return. On the other hand, concerns may arise over the increase in PVR associated with acidosis. There is evidence that despite this effect, hypoventilation leading to mild hypercapnea improves systemic oxygenation after the Glenn procedure by enhancing cerebral blood flow and venous return to the lungs, whereas hyperventilation results in decreased systemic and cerebral oxygenation. Apparently, the effect on cerebral blood flow dominates the effect on PVR. Hence, the optimal ventilatory strategy for patients with a superior cavopulmonary anastomoses should employ mild respiratory acidosis, maintaining the systemic arterial pH between 7.30 and 7.35.

Elevated pulmonary venous pressures resulting from atrioventricular valve regurgitation or single ventricular dysfunction will also decrease the gradient for pulmonary blood flow. This etiology should be suspected in patients with signs of poor perfusion in the absence of pulmonary disease or dehydration. In this setting, a milrinone infusion (0.5 mcg/kg/min) could be helpful. Its inhibition of phosphodiesterase III decreases the breakdown of cyclic AMP, which provides inotropic support as well as afterload reduction via improved diastolic relaxation and systemic vasodilatation.

Emergency physicians must also be aware of the problems posed by passive pulmonary blood flow in the setting of cardiopulmonary resuscitation. When a patient with a superior cavopulmonary anastomosis presents in cardiac arrest, chest compressions will assist perfusion of the systemic circulation but will be detrimental to oxygenation. In fact, the positive pressure applied to the chest will result in impaired flow through the pulmonary circulation. Fluid resuscitation is vital to augment systemic venous pressures to overcome this obstacle and provide adequate ventricular preload. Compressions must be performed while allowing complete chest recoil. If possible, brief periods of negative intra-thoracic pressure (active compression-decompression CPR techniques) may improve oxygenation through restora-
Palliation Of Single Ventricle Lesions – Stage 3

The final stage of palliation, the Fontan procedure, is scheduled at around 1½ to 3 years of age. The IVC blood flow is now also re-directed to the main pulmonary artery. This eliminates the mixing of oxygenated and deoxygenated blood in the common atrium and further decreases the work required by the right ventricle. The systemic and pulmonary circulations are now completely separated (total cavopulmonary anastomosis) creating blood flow in series (Figures 17 and 18). Ideally, minimal mixing should occur and SaO2 should be nearly 100%. In some patients, a small communication, known as a fenestration, will be left between the Fontan circuit and the common atrium. This modification is typically performed in patients at risk for elevated PVR in the post-operative period. If PVR becomes acutely elevated, blood passing through the Fontan circuit will shunt into the common atrium maintaining adequate systemic venous preload and cardiac output. In these situations, SaO2 will be decreased (85-95%) due to mixing of deoxygenated blood from the Fontan conduit into the common atrium, but “blue” blood flow is better tolerated than no blood flow. Hypoxia, acidosis, and atelectasis can all elevate PVR, precipitating shunting across the fenestration.

As in the Glenn procedure, pulmonary blood flow is passive, so increases in PVR or decreases in systemic venous pressures can result in impaired systemic perfusion or cyanosis. In patients with non-fenestrated Fontan procedure, dehydration and increases in PVR will limit ventricular preload and compromise systemic perfusion. In patients with fenestrated Fontan procedure, worsening cyanosis due to shunting across the fenestration will be more common. Intravascular volume expansion, supplemental O2, mechanical ventilation, and inhaled nitric oxide will all be helpful. If mechanical ventilatory support must be instituted for pulmonary disease or atelectasis, a strategy of low rate, short inspiratory time, and only enough positive end expiratory pressure as needed to correct the atelectasis is employed. The benefits of mild respiratory acidosis seen following Glenn surgery are not apparent in the Fontan circulation, likely because pulmonary blood flow has a lesser degree of dependence on cerebral circulation. Alkalosis to minimize PVR is more prudent.

Patients with Fontan physiology have an additional complication not seen after the Glenn procedure. They frequently develop protein losing enteropathy and subsequent pleural and pericardial effusions, ascites, as well as body edema. The etiology is unclear, although the increase in systemic venous pressures is likely a contributing factor. These patients will require volume resuscitation to maintain adequate intravascular volume and must be considered for drainage of pericardial, pleural, and abdominal fluid if the accumulation is impeding cardiopulmonary function.

Cardiac arrest also poses significant problems unique to the patient with total cavopulmonary anastomosis physiology. As with superior cavopulmonary anastomoses, chest compressions increase intrathoracic pressure and impair flow through the pulmonary vascular bed. In total cavopulmonary anastomoses, since cardiac output is usually completely dependent upon pulmonary venous return, systemic perfusion is compromised. Abdominal compressions alternating with chest compressions (interposed abdominal compressions) should be considered, exploiting the new IVC connection to the pulmonary vascular bed. Additionally, negative extrathoracic pressure (active compression-decompression CPR techniques) has been shown to improve pulmonary blood flow during cardiac arrest and may become critical in management of patients after cavopulmonary anastomoses.

Despite the separation of the pulmonary and systemic circulation and unloading of the systemic veno-
tricle, the risk of cardiac failure and death increases over time. The 5-year and 10-year survival rates after Fontan palliation are 86% and 74%, respectively. Arrhythmias (10-40%) and thromboembolic events from passive flow through the Fontan circuit (3-20%) contribute to increased morbidity and mortality after total cavopulmonary anastomosis.

Heart Transplantation

Pediatric heart transplantation is becoming more prevalent for acquired heart disease (e.g., myocarditis, cardiomyopathy) and some forms of CHD. As 5-year survival rates improve, so does the likelihood that a transplant patient will present to the emergency department. Patients with graft rejection and infection in association with immune suppression can present initially with subtle symptomatology and quickly progress to devastating outcomes, including cardiopulmonary arrest. Nearly 30% of post-transplant patients who presented to the emergency department required admission. Communication with the patient’s transplant team or primary care physicians will help guide evaluation and initial therapy. Emergency physicians must be able recognize the clues of graft rejection and infection in order to provide the primary team with as much information as possible.

A 7-year retrospective analysis at a major pediatric heart transplant center revealed that rejection was the diagnosis in only 9% of post heart transplant patients presenting to the emergency department, yet this diagnosis provoked the most apprehension in physicians. Symptoms associated with rejection may be mild and often mimic infection. Patients can have irritability, malaise, poor feeding, nausea, and changes in sleep patterns. The most common finding is tachycardia, although fever and signs of CHF, including tachypnea, rales, pallor, murmur and hepatomegaly, may be present. Symptoms of angina are rare because the transplant heart is denervated. More commonly, patients with post transplant coronary

<table>
<thead>
<tr>
<th>History &amp; Physical Examination</th>
<th>Fever, poor feeding, irritability, lethargy, palpitations, decreased urine output, tachypnea, wheezing, retractions, rales, dyspnea, grunting, bradycardia, gallop, murmur, organomegaly, pallor, poor capillary refill, cool extremities</th>
</tr>
</thead>
<tbody>
<tr>
<td>CXR</td>
<td>Changes from prior CXR with respect to heart size, pulmonary edema, effusions</td>
</tr>
<tr>
<td>ECG</td>
<td>Changes from prior ECGs with respect to QRS voltages, QRS axis, conduction pattern, arrhythmia</td>
</tr>
<tr>
<td>Echocardiogram</td>
<td>Changes from prior echocardiograms with respect to pericardial effusion, ventricular function, LV thickening, mitral valve insufficiency</td>
</tr>
</tbody>
</table>
artery disease from chronic rejection will present with syncope. Table 7 contains the common signs and symptoms of acute cardiac graft rejection.\(^{136}\)

More than one half of the post transplant patients presenting to the emergency department are diagnosed with infection.\(^{136}\) Although many are minor and can be managed similar to the general population, opportunistic infections, especially CMV and PJP, are common and can be serious. Presumptive treatment with gancyclovir and trimethoprim-sulfamethoxazole, respectively, should be initiated when these diagnoses are considered. Gastrointestinal symptoms are also a common complaint; most are infection-related, but possible immunosuppressive medication toxicity and medication interactions should be evaluated.

### Controversies And Cutting Edge

Extracorporeal membrane oxygenation (ECMO) has been deployed successfully in the ED as a resuscitative tool in certain pediatric cardiac arrest situations.\(^{137}\) Because acceptable neurologic outcome depends on the duration of arrest and hypoxemia, this excludes most pediatric arrests, which are generally secondary to respiratory failure. However, the institution of ECMO in the emergency department during cardiac arrest, also referred to as extracorporeal cardiopulmonary resuscitation (ECPR), may be effective in a select group of patients with witnessed arrest of cardiac etiology, as in ALCAPA, myocarditis, or cardiomyopathy. In a review of 64 patients who required ECMO after a witnessed arrest, survival to discharge was 49% in events of children with heart disease compared to 9.5% in children with other medical conditions.\(^{138}\) At another institution, a review of 103 cardiac patients managed with ECMO over 9 years showed an improvement in survival from 43% to 65% during the first and second half of the study, respectively.\(^{139}\) ECMO can be utilized to stabilize a patient in the ED until a long-term ventricular assist device can be implanted.

The Berlin Heart, a neonatal and pediatric sized ventricular assist device introduced in the 1990s, had an initial mortality rate of 100%. With changes in indications for support, adjustment in cannulation techniques, advances in anticoagulation therapy and monitoring, along with improved ICU management, survival rates are now comparable to the adult experience, which is approximately 70%.\(^{140}\) The device has been successfully explanted in patients with severe but reversible cardiac disease, such as ALCAPA and myocarditis, without subsequent need for a heart transplant.

### Disposition

After initial stabilization of any patient with CHD, communication and coordination between the transporting and accepting institutions are imperative. Expedient transport is not advantageous if the accepting institution does not have the required resources for management or if the resources are not readily available when the patient arrives. Likewise, the transport is futile if the patient deteriorates en route and the transporting team does not have the requisite experience to manage a critically-ill infant with CHD. Two-way inter-hospital transport of pediatric cardiac patients with an experienced pediatric cardiac transport team resulted in decreased incidence of acidosis and hypothermia compared with one-way transport.\(^{141}\) Neonates with ductal-dependent lesions who are stabilized with low dose prostaglandin (less than 0.015 mcg/kg/min) may not require endotracheal intubation and mechanical ventilation for safe transport,\(^{101}\) however, a transport team for self-ventilating patients requires a practitioner who is trained in neonatal intubation and understands the physiologic consequences of mechanical ventilation in the event the need for mechanical ventilatory support arises.\(^{142}\)

While awaiting transfer, maintenance of stabilization of the patient will minimize deterioration in transit. Ideally, a dedicated emergency physician must be committed to the patient for continuous cardiorespiratory monitoring, management of ventilation, institution of further volume and/or inotropic support as needed, control of arrhythmias, and maintenance of the DA in order to optimize clinical status until the transport team has arrived.\(^{136}\)

### Summary

Emergency physicians, especially those working in centers without pediatric sub-specialty support, must have a firm understanding of the fundamentals of CHD. Many infants with CHD will continue to be routinely discharged from nurseries without a diagnosis of CHD. Many others with a diagnosis will have distinctive symptomatology due to their pre- and post-operative anatomy and physiology. A high index of suspicion and a systematic approach utiliz-
ing traditional methods of diagnosis (e.g., history, vital signs, pulse oximetry, physical examination, chest x-ray, and electrocardiogram) should direct appropriate consultation and initial management. Emergency physicians must also remember that some resuscitation measures that are almost universally employed in critically-ill patients, such as supplemental oxygen and fluid expansion, may be harmful in these patients in certain types of CHD.

Case Conclusion

The patient presented in the opening vignette was ultimately diagnosed with critical aortic stenosis. Presentation to the emergency department occurred following delayed closure of the DA. The abrupt development of symptoms and presence of hepatomegaly provided early clues to the diagnosis, but the patient's clinical deterioration following treatment with inhaled albuterol and aggressive intravenous fluid boluses increased the likelihood of CHD. The patient was subsequently endotracheally intubated, administered PGE1 via continuous infusion, and transferred to a tertiary care facility for surgical correction.

References

Evidence-based medicine requires a critical appraisal of the literature based upon study methodology and number of subjects. Not all references are equally robust. The findings of a large, prospective, randomized, and blinded trial should carry more weight than a case report.

To help the reader judge the strength of each reference, pertinent information about the study, such as the type of study and the number of patients in the study, will be included in bold type following the reference, where available. In addition, the most informative references cited in this paper, as determined by the authors, will be noted by an asterisk (*) next to the number of the reference.


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CME Questions

1. Which of the following factors contributes to delayed closure of the DA in infants with CHD, relative to normal neonates?
   a. The presence of fetal hemoglobin
   b. Inspiration of higher concentrations of oxygen
   c. Decreased pulmonary vascular resistance
   d. The physiologic anemia of infancy
   e. Increased flow through the DA

2. Which of the following lesions has ductal-dependent systemic blood flow?
   a. Truncus arteriosus
   b. Critical coarctation of the aorta
   c. Ebstein’s anomaly
   d. Total anomalous pulmonary venous return
   e. Double outlet right ventricle with pulmonary stenosis

3. Which of the following factors contributes to the development of congestive heart failure in a 2-month-old with a large ventricular septal defect?
   a. Physiologic polycythemia of infancy
   b. Decreasing left-to-right shunting
   c. Increasing pulmonary vascular resistance
   d. Increasing right ventricular compliance
   e. The absence of fetal hemoglobin

4. Which of the following factors has the greatest effect on the timing of presentation for infants with TAPVR?
   a. Amount of left-to-right shunting
   b. Severity of pulmonary venous obstruction
   c. Decrease in pulmonary vascular resistance
   d. DA closure
   e. Physiologic anemia of infancy

5. Which of the following murmurs qualities describes 1 of the 6 cardinal signs for the presence of CHD?
   a. Grade II systolic ejection murmur
   b. Point of maximal intensity at the lower left sternal border
   c. Harsh quality of the murmur
   d. Abnormal first heart sound
   e. Diastolic rumble

6. Which of the following is most distinguishing for the diagnosis of congenital heart disease in a 1-month-old infant in respiratory distress?
   a. Elevated arterial lactate
   b. Right axis deviation on electrocardiogram
   c. Left axis deviation on electrocardiogram
   d. Tachycardia
   e. Hypotension

7. When stabilizing the airway in a neonate or pediatric patient with respiratory distress, profound cyanosis, and circulatory collapse, which of the following drug regimens is optimal?
   a. Fentanyl and rocuronium
   b. Morphine, versed, and vecuronium
   c. Etomidate, ketamine, and rocuronium
   d. Pentobarbital and rocuronium
   e. Rocuronium only

8. With respect to supplemental oxygen in a neonate or infant with known or suspected congenital heart disease, which of the following is TRUE?
   a. Supplemental oxygen should not be administered to an infant with congenital heart defect since it constricts the ductus arteriosus.
   b. Administer supplemental oxygen to maintain optimal $O_2$ saturation at 93-97% in order to provide adequate $O_2$ delivery to the tissues.
   c. Careful attention must be paid to the clinical response when supplemental oxygen is administered since it may further compromise systemic circulation.
   d. Supplemental $O_2$ will assist in maintaining the patency of the ductus arteriosus and should be administered in as high a concentration as possible.
   e. Supplemental oxygen will vasoconstrict the pulmonary vasculature and halt the progression of pulmonary edema.

9. A 2-month-old female infant with unrepaired tetralogy of Fallot presents to the emergency department. Despite multiple fluid boluses, endotracheal intubation with a fraction of inspired oxygen of 100%, and sedation with morphine and ketamine, she remains cyanotic. What is the next step in the management of this patient?
   a. Phenylephrine
   b. Dopamine
   c. Epinephrine
   d. Prostaglandin E1
   e. Furosemide

10. Which of the following palliation stages for hypoplastic left heart syndrome relies on passive pulmonary blood flow, complicating cardiopulmonary resuscitation?
    a. First stage Norwood (arch reconstruction, BTS)
    b. Second stage Glenn (superior cavopulmonary anastomosis)
    c. Third stage Fontan (total cavopulmonary anastomosis)
    d. a, b, and c
    e. b and c

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